Recurrence of a Simultaneous Tumor of the Parotid Gland and Scalp Skin Malignant Fibrous Histiocytoma

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Abstract: Malignant fibrous histiocytoma (MFH) is one of the most common soft-tissue sarcomas and generally arises in elderly patients. Most often, MFH occurs in the extremities and the retroperitoneum; MFH of the head and neck region is only reported in 3% of all cases. The distant metastasis appears in one third of all cases, and those cases mainly involve the lung, regional lymph nodes, the liver, or bones. The main treatment of MFH of the head and neck is radical excision of the tumor with modified or radical neck dissection. Recurrences of MFH are commonly seen and identified within the first 2 years of treatment. We report a case of simultaneous MFH tumors of the parotid gland and of the scalp skin in an elderly male patient. The patient underwent a wide surgical excision of the lesion and referred to the oncology department for further treatment. However, 1 month after chemoradiotherapy had commenced, there was a recurrence of the tumor over the skin on the right parotid region.

Key Words: Malignant fibrous histiocytoma, parotid gland, simultaneous tumor, tumor recurrence

Malignant fibrous histiocytoma (MFH) is one of the most common soft-tissue sarcomas and generally arises in elderly patients. Most often, MFH occurs in the extremities and the retroperitoneum; MFH of the head and neck region is only reported in 3% of all cases.[1] Incidences of MFH affecting the parotid gland are exceedingly rare,[2] with only 19 recorded cases. The distant metastasis appears in one third of all cases, and those cases mainly involve the lung, regional lymph nodes, the liver, or bones.[1] A simultaneous MFH tumor in both the parotid gland and the scalp skin has not yet been reported.

The main treatment of MFH of the head and neck is radical excision of the tumor with modified or radical neck dissection. Recurrences of MFH are commonly seen and identified within the first 2 years of treatment. Although no difference in the outcomes of MFH variants has been reported, superficial lesions recur more frequently than deep lesions.[3]

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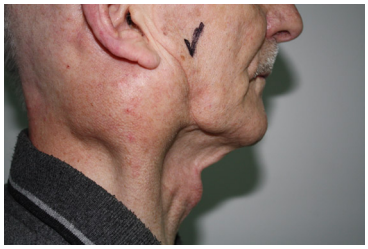

FIGURE 1. Preoperative view of the lesion on the right parotid region.

The following is a case of simultaneous MFH tumors of the parotid gland and of the scalp skin in an elderly male patient. The lesion recurred very early and was located on the right parotid region.

A 76-year-old man presented with a firm, painless, semimobile, and rapidly growing mass in the right parotid region. There was no apparent skin involvement, and the mass measured approximately 6 × 5 cm in size (Fig. 1). In addition, there was a nodular ulcerated lesion on the scalp skin for 2 months that measured 1 × 1.5 cm in size, was irregular in size, and was brownish. Ultrasonographic evaluation of the lesion revealed the existence of a lesion measuring 6 × 5 cm on the tail of the right parotid gland with mixed echogenicity, necrotic areas, and a morphology resembling a malignant mass. The magnetic resonance imaging with contrast enhancing revealed a nodular lesion in the right parotid gland. There was invasion to the surrounding structures, including the sternocleidomastoid muscle. There was no invasion to neighboring vascular structures (Fig. 2). Cytologic examination of the parotid gland mass using a fine-needle aspiration biopsy revealed atypical cells that were consistent with high-grade neoplasm.

The patient underwent total parotidectomy with unilateral right-sided radical neck dissection. The overlying skin was tumor negative. There was no injury to the facial nerve. Otherwise, the lesion on the scalp was excised totally with approximately 2 cm negative margins. Skin defect on the scalp was primarily reconstructed. The histopathologic study of the primary neck lesion revealed the pleomorphic MFH. The scalp lesion was diagnosed as a metastasis of the primary tumor. These highly cellular tumors have prominent fibrous stromata that, in some examples, show a storiform growth pattern. Tumor cells are usually spindly with elongated, tapering nuclei that have a fibroblastic or myofibroblastic appearance (Fig. 3). Immunohistochemical staining revealed positive reaction for vimentin, S-100, and CD68 in the tumor cells but was negative for pancreatin, CD3, CD20, and CD38 representing MFH.

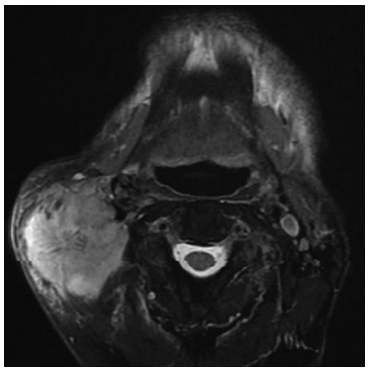

FIGURE 2. A magnetic resonance image with contrast enhancement representing a solid mass in the right parotid gland.

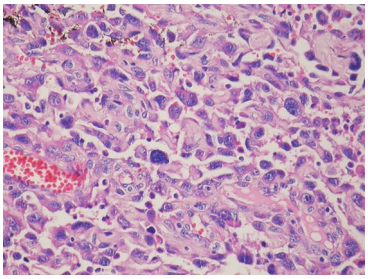

FIGURE 3. Histopathologic evaluation of the tumor showing fibroblastic differentiation with spindle-shaped cells in a storiform pattern (hematoxylin-eosin, original magnification ×400).

The patient was referred to the oncology department for further treatment. However, 1 month after chemoradiotherapy had commenced, there was a recurrence of the tumor on the right parotid region (Fig. 4).

DISCUSSION

Malignant fibrous histiocytoma is described as an undifferentiated high-grade pleomorphic sarcoma.[4] The extremities, trunk, and retroperitoneum are the regions most involved in MFH. It has also been described in the visceral, tracheal, and genital organs.[4] Head and neck involvement has been reported in 10% of the cases, with the majority occurring in the sinonasal tract.[1] Location in the parotid region is not common. Therefore, MFH is not usually considered in the differential diagnosis of parotid tumors, and MFH of the parotid region is frequently mistaken for pleomorphic adenoma. The definitive diagnosis is established by histochemical studies.[5] The present case is a unique case report of simultaneous MFH involving both the parotid gland and the scalp skin. These tumors arise from mesenchymal tissues including cervical fascia and skeletal muscles.[4] Thus, we assume that the MFH of the parotid gland had arisen from the fascia surrounding the parotid gland in our patient. It was also thought that the MFH of the scalp skin was a metastatic lesion. Despite its rare incidence, atypical rapid relapse of the tumor was observed postoperatively in our case 1 month after starting chemoradiotherapy over the skin of the primary-tumor site. This feature has not yet been reported in the literature, although frequent relapse of the tumor is known.

Histopathologic examination of MFH is often misdiagnosed as malignant melanoma, anaplastic lymphoma, pleomorphic leiomyosarcoma, pleomorphic liposarcoma, malignant peripheral nerve sheath tumor, soft-tissue osteosarcoma, anaplastic carcinoma, gliosarcoma, or malignant gliomas.[2] The distinction from MFH typically requires clinical and radiographic correlation with cytopathology.[5]

The primary treatment of MFH of the head and neck is the radical excision of the lesion with radical dissection of the regional lymph nodes.[3] Malignant fibrous histiocytoma is an aggressive tumor; therefore, after surgical excision, postoperative chemoradiotherapy is advised to reduce local recurrence because these tumors recur very commonly. Superficial tumors have a higher recurrence rate than deep tumors.[3] It was reported that the risk of disease progression has been shown to increase when the adjuvant radiotherapy is not used.[3] However, prognosis of MFH differs according to location, histology, and extension of the lesion. The reported 5-year survival rates range from 39% to 64%.[6,7] Some studies describe that the MFHs of the head and neck have a worse survival rate.[3] This is potentially due to high-grade tumors and anatomic location that preclude negative margins.[3] However, in our case, local recurrence was detected on the right upper jugulodigastric region just after starting chemoradiotherapy.

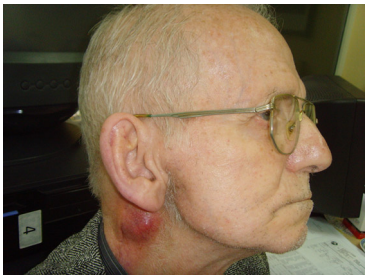

FIGURE 4. Recurrence of the lesion on the right parotid region.

In conclusion, MFHs are highly aggressive mesenchymal tissue tumors and tend to recur very frequently, even after wide resection of tumor and chemoradiotherapy.

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Brucella melitensis Infection Within Warthin Tumor of the Parotid Gland

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Abstract: Brucellosis is a zoonotic systemic infectious disease, and multiorgan involvement is commonly seen, but involvement of the

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neck is a rare presentation of brucellosis. Granulomatous infections of the parotid gland are extremely rare. Warthin tumor is a well-known benign neoplasm of the salivary glands. In this report, we describe a Warthin tumor associated with *Brucella melitensis* in the same parotid gland.

Key Words: Brucellosis, salivary gland, parotid, neck

Brucellosis is a zoonotic systemic infectious disease, and multiorgan involvement is commonly seen.[1,2] However, involvement of the neck is a rare presentation of brucellosis. Warthin tumor (papillary cystadenoma lymphomatosum) is a well-known benign lesion of the salivary glands with epithelial and lymphoid components. It is the second most common tumor of the parotid gland.[3] In this report, we describe a Warthin tumor associated with *Brucella melitensis* in the same parotid gland. According to our knowledge, this is the first documented case with Brucellosis within the parotid gland.

A 52-year-old man was admitted to our otorhinolaryngology department with a complaint of a right preauricular mass that had been present for 9 months. Fine-needle aspiration biopsy of the mass was performed at another hospital, and it was reported as an "inflammatory process." Although the medical therapy (amoxicillin plus clavulanic acid) was given for 2 weeks, the mass did not reduce in size. There was no history of trauma, infection, or skin lesion of the head and neck. His medical history was negative for systemic symptoms or complaints such as fever, chills, or night sweats. On physical examination, there was a smooth-surface, hard mass, 3 × 2 cm in size, on the angle of the right mandible. Other systems of the body were normal. Blood leukocyte count was 6400/mL, erythrocyte sedimentation rate was 7 mm/h, procalcitonin was less than 0.020 ng/mL (reference range, <0.5 ng/mL), serum glucose level was 343 mg/dL, and C-reactive protein was 0.7 mg/L. Other laboratory investigations revealed normal results. Vital signs were stabile.

The computed tomographic scan of the neck revealed an oval semisolid lesion measuring 3 × 2.5 cm, with a well-defined border, just inferior to the right parotid gland. It had both cystic and solid components (Fig. 1).

A superficial parotidectomy was performed under general anesthesia. On gross examination, the lesion was gray, and there was a purulent secretion in the center of the lesion. It was sent for culture. The histopathologic diagnosis of the surgical specimen was reported as "Warthin tumor and epithelioid granulomas without caseation necrosis consistent with chronic granulomatous inflammation." *Brucella melitensis* was isolated in the culture, and medical treatment (rifampicin 600 mg/d and doxycycline 200 mg/d for 6 weeks) was given after surgical excision. The patient is still under our close follow-up and no evidence of recurrence 10 months after therapy.

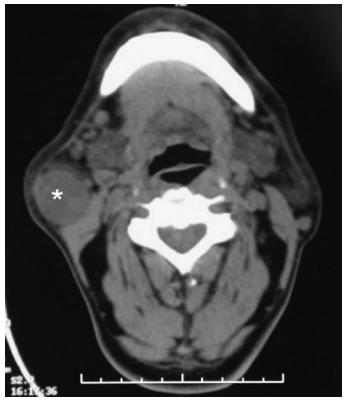

FIGURE 1. A computed tomographic scan shows that an oval semisolid lesion with a well-defined border (asterix) is just inferior to the right parotid gland. It had both cystic and solid components.

DISCUSSION

The parotid gland lesion may occur because of various diseases such as sialolithiasis, inflammatory diseases, and benign or malignant tumor. Parotid gland tumors with granulomatous infections, such as tuberculosis, are reported to be extremely rare in the literature.[3,4] We are reporting the first case in the literature of an isolated parotid infection (Warthin tumor) with *B. melitensis* with no associated symptoms or signs of systemic brucellosis.

Various bacteria have been reported to be involved in the parotid inflammation process; the most common being *Staphylococcus aureus* and anaerobic bacteria in adults.[4] *Streptococcus pneumoniae*, *Haemophilus influenzae*, *Proteus mirabilis*, *Mycobacterium tuberculosis*, and *Candida* species have been reported to be among the most common pathogens in parotid abscesses, but granulomatous infections were extremely rare.[4–6]

The pathophysiology of abscess formation by microorganisms in parotid neoplasms is unclear, and several mechanisms likely contribute. Ductal ectasis, primary parenchymal involvement, or infection of the intraparotid or periparotid lymph nodes can result in abscess formation.[3,4] Infection of the parotid gland can arise from ascending infection via the Stensen duct by the bacteria from the oral cavity or bacteriemia.[6–8] Tan and Goh[9] report on 2 cases of parotid abscess that developed within a preexisting Warthin tumor but did not identify the causative organism. Their case series comprises 15 patients, and they found that diabetes mellitus is a significant comorbid factor, with 6 patients having diabetes. Such microvascular compromise may partly explain the susceptibility of patients with diabetes to parotid abscess.[9] The added compression caused by a neoplasm may lead to salivary stasis and subsequent infection, or erosion of ductal elements and translocation of microorganisms. The lymphoid elements within a Warthin tumor may provide a site of tropism resembling a normal lymphoid tissue where organisms, such as *Bartonella henselae* and *M. tuberculosis*, frequently suppurate.[6,7] The cystic nature of Warthin tumor may predispose to abscess formation by providing a preexisting fluid-filled cavity for infection.[6,8] Our patients had uncontrolled diabetes and occupational exposure. These conditions may have been contributed to the development of infection with *B. melitensis* within Warthin tumor.

In developing countries where the prevalence of brucellosis is high, various clinical manifestations can be seen. Typically, the organism is ingested and enters the bloodstream via the gastrointestinal tract. Once within the bloodstream, the organism quickly becomes an intracellular pathogen contained within circulating polymorphonuclear cells and macrophages.[10] The main transmission way of the agent is the consumption of infected milk and milk products. However, transmission may occur through mucous membranes (eyes, nose, gastrointestinal system, and genitourinary system) and via cuts, abrasions, inhalation, or, in the case of veterinarians and microbiologists, parenteral injection.[1,10] Our patient was a veterinarian.

The most common form of brucellosis is the systemic form, characterized by an undulating fever, diffuse musculoskeletal complaints, and various complications.[1,2] Focal brucellosis, on the other

hand, is much less common, and neck involvement is a rare presentation of brucellosis. There are some extremely rare cases reported in head and neck region such as the thyroid gland, cervical lymph nodes, branchial cyst, and neck abscess.[11–13] Rameh et al[11] reported a case of an isolated branchial cleft cyst infection with *B. melitensis* with no associated symptoms or signs of systemic brucellosis. Nadler et al[12] reported a 54-year-old woman with suppurative lymphadenitis accompanied by erythema, pain, fever, and pus drainage. *Brucella suis* was detected as a causative agent.[12] Varona et al[13] reported a 42-year-old man with an isolated cervical lymphadenopathy due to *B. melitensis* without systemic signs and symptoms. In addition, previous studies reported 2 cases of cervical lymphadenitis due to *Brucella* species in Turkey.[14,15]

Our case presented a slowly growing mass at the parotid gland, and we suspected for parotid gland tumor. Therefore, this patient underwent superficial parotidectomy. Our reported case is the first case of focal *Brucella* infection that presented as a parotid gland tumor with no associated systemic manifestations. The management of Warthin tumor of the parotid gland is superficial parotidectomy. However, infection with *B. melitensis* is present in this case's surgical excision coupled with medical treatment.

To the best of our knowledge, this is the first report of Warthin tumor associated with *Brucella* genus in the parotid gland. It is proposed that, in the differential diagnosis of a parotid gland mass, brucellosis should be kept in mind in the endemic area.

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Solitary Myxoid Neurofibroma of the Soft Palate

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Abstract: The neurofibroma is a benign tumor of neural origin, not frequently located in the oral cavity, and, especially, extremely rare in the soft palate. When it occurs solitarily, it is a spontaneous tumor without any internal manifestations. We report a case of an isolated neurofibroma of the soft palate not associated with von Recklinghausen disease.

Key Words: Neurofibroma, palate, soft

The neurofibroma is a benign neoplasm originated from the peripheral nerve sheath. The subtypes of neurofibroma are localized, plexiform, and diffuse. Localized neurofibroma is solitary, polypoid or nodular, and mostly cutaneous and is usually not associated with neurofibromatosis. It is circumscribed but is an nonencapsulated lesion with varied appearance.[1] In oral peripheral nerve sheath tumors, the frequency identified for neurofibroma was 0.2%.[2]

To our best knowledge, the described case of solitary myxoid neurofibroma of the soft palate is a second report in medical literatures.

CLINICAL REPORT

A 44-year-old man presented with gradually enlarging painless soft mass on the right side of the soft palate for 3 years. Clinical examination revealed a circumscribed mass (approximately 3 × 2.5 cm) on submucosal lesion of the right soft palate (Fig. 1). Dermatologic examination did not reveal any café au lait spots, cutaneous neurofibromas, and axillary or inguinal freckling. He has no family history of neurofibromatosis. Computed tomography demonstrated a well-circumscribed mass in the right soft palate (Fig. 2).

Under nasotracheal intubation, the mass was entirely excised. The tumor mass was well circumscribed, easily dissected without

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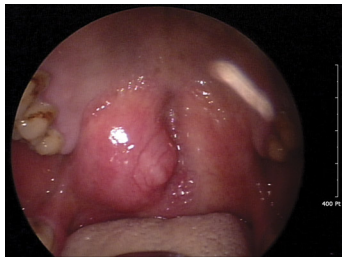

FIGURE 1. Photograph of the oral cavity shows a large mass in the right soft palate.

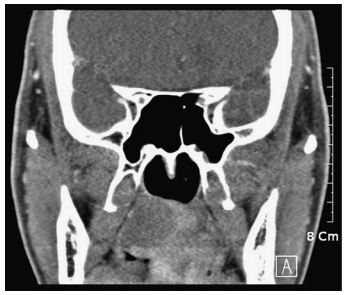

FIGURE 2. Coronal-sectioned computed tomographic scan showing an ill-defined, round 3 × 2-cm mass in the right soft palate.

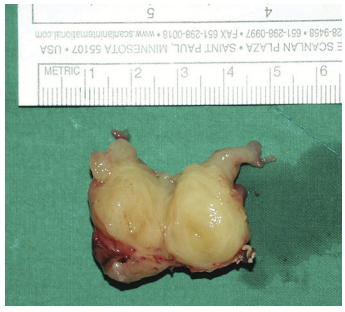

FIGURE 3. Cross-sectioned specimen finding shows myxoid glistening yellowish cut surface.

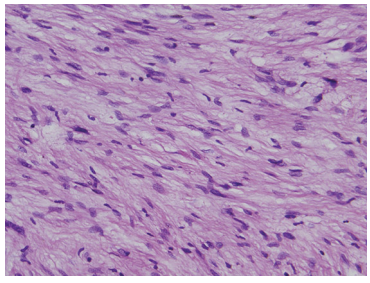

FIGURE 4. Higher magnification shows that tumor is composed of interlacing bundles of elongated cells having wavy nuclei (hematoxylin-eosin, original magnification ×400).

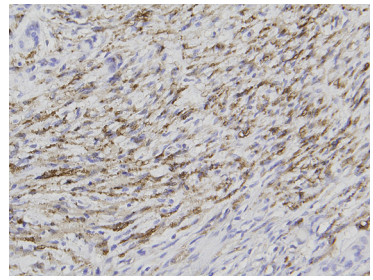

FIGURE 5. Microscopic findings and immunohistochemical staining. The tumor cells are positive signals for S-100 protein (original magnification ×400).

adhesion. The excised mass was 3 × 3 × 2.5 cm in size, ovoid shaped, and nonencapsulated, and the cut surface was greasy and yellow (Fig. 3). Histologic examination showed the proliferation of spindle-shaped cellular elements immersed in a myxoid stroma of wavy fibrillar materials (Fig. 4). The immunohistochemistry highlighted a positive result for S-100 (Fig. 5), indicative of neural origin, confirming the diagnosis of neurofibroma. The region healed without complication.

DISCUSSION

Benign peripheral nerve sheath tumors of neural origin are traumatic neuroma, neurofibroma, neurilemoma, and palisaded encapsulated neuroma. Neurofibromas are benign, heterogenous peripheral nerve sheath tumors that arise from the connective tissue of peripheral nerve sheaths, especially the endoneurium.[3]

To our knowledge, an isolated neurofibroma arising from the soft palate without any other manifestations of von Recklinghausen disease was reported only in 1 case.[4]

Salla et al[2] reported that oral peripheral nerve sheath tumors among oral pathologic specimens represented 0.2%, solitary neurofibroma was extremely rare, and there was no case of soft palate origin.

Neurofibroma is a circumscribed but nonencapsulated tumor consisting of a mixture of Schwann cells, perineural cells, and endoneurial fibroblasts.[5] Only 4% of the neurofibromas were encapsulated or well delineated.[6]

In the solitary neurofibroma, the most common sites were the tongue, the palate, the buccal mucosa, and the floor of the mouth.[7] It was observed that these lesions affected adults and children, related no sex dominance.[8]

Two histologic aspects allow to distinguish a plexiform neurofibroma from a myxoid neurofibroma. The first of these is less common and can more frequently be associated with the von Recklinghausen disease and with multiple endocrine neoplasia, whereas the second one is more frequently highlighted as a solitary finding.[9]

Fumo et al[10] recently proposed a classification of 3 ultrastructural subtypes of neurofibromas, based on the presence, in percentage, of Schwann cells, perineural and endoneurial fibroblasts, and intermediate cells. Because of the S-100, CD34, and epithelial membrane antigen immunocolorings, it is possible, in fact, to establish the cellular lineages present within the lesion and make a differential diagnosis between the various types of tumor of the peripheral nerves.

A distinction between neurofibroma and neurilemoma is imperative of the possibility of malignant transformation. In patients with neurofibromatosis type 1, the transformation of a neurofibroma into a malignant peripheral nerve sheath tumors has been observed in 2% to 5% of cases.[3] Pain, change in texture, rapid increase in size, and neurologic deficits are indicators for the

development of a malignant peripheral nerve sheath tumor within a preexisting neurofibroma.[11]

The treatment choice for palatal tumors is to remove them by excising the portion of the palate to which it is attached; this applies whether the tumor is situated in the hard or soft palate.[4] After complete excision, recurrence is rare.

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A Huge Midline Premaxillary Cyst as a Late Complication of Maxillary Surgery

Joohwan Kim, MD, Inn Chul Nam, MD, Seong Hyun Yun, MD, Jin Hee Cho, MD, PhD

Abstract: A median palatal cyst is an uncommon nonodontogenic cyst, and patients usually present with a painless swelling or the sensation of a mass. The mass is typically a well-defined fixed swelling along the midline. The mass can cause slight elevation of the nasal floor or swelling and drainage from the hard palate. Surgical resection is usually recommended as a definite treatment.

We treated a 30-year-old man with a premaxillary mass with nasal obstruction. He had undergone surgery on both the maxilla and the mandible to correct malocclusion 10 years earlier. A physical examination revealed elevated mucosa of the nasal floor, resulting in near-total obstruction of the nasal cavity, and the gingival mucosa over the upper incisors was also swollen. Preoperative computed tomographic scan demonstrated a midline nonenhancing round cystic lesion in the premaxillary area. Surgical excision was performed via a sublabial approach under general anesthesia, and his recovery after surgery was uneventful.

Key Words: Nasopalatine duct cyst, nasal septum, nasal cavity

A midline palatal cyst is a rare condition, located in the premaxillary area. Patients usually present with a painless swelling or the sensation of a mass in this area. Sometimes, a midline palatal cyst is reported in patients who have had surgery involving the midfacial structures.[1] The proliferation of epithelial remnants is suggested as the etiology, and some investigators classify this as a nasopalatine duct cyst.

We treated a 30-year-old man with a premaxillary mass with nasal obstruction. There are only a few reports of huge midline palatal cysts arising from a surgical defect after maxillary surgery. Here, we describe the clinical, radiologic, and histopathologic findings and treatment method. Our institutional review board approved disclosure of this case.

CLINICAL REPORT

A 30-year-old man presented with a painful swelling in the premaxillary area and bilateral nasal stuffiness. The mass was not painful initially, but discomfort increased gradually during 2 months. He had undergone surgery on both the maxilla and the mandible to correct malocclusion 10 years earlier, and the postoperative course was unremarkable. The patient first noticed the premaxillary swelling after a bout of the flu 5 months earlier and was told he had a right upper gingival abscess. The symptoms subsided after spontaneous rupture and medications, including antibiotics.

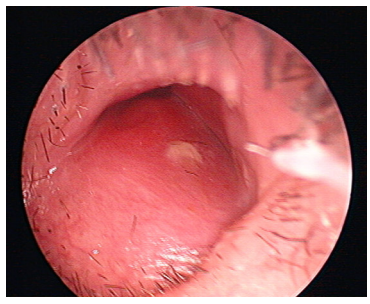

FIGURE 1. Endoscopic view of the left nasal cavity. The nasal floor mucosa was elevated, resulting in near-total obstruction of the nasal cavity. No discoloration of the mucosa or necrotic change was found.

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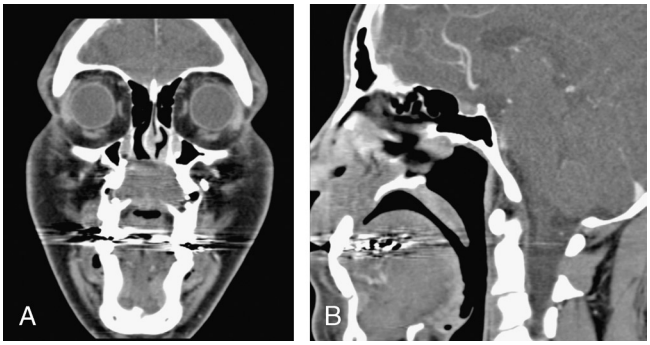

FIGURE 2. Preoperative computed tomographic scans in the coronal (A) and sagittal (B) planes show a lesion approximately 4 cm in diameter, occupying more than half of the anterior hard palate and extending into the nasal cavity, where there is some absorption of the nasal septum.

Anterior rhinoscopy and rigid endoscopy revealed an elevated mucosa of the nasal floor, resulting in near-total obstruction of the nasal cavity (Fig. 1). The upper lip and nasal philtrum were elevated slightly and displaced anteriorly. The swollen premaxillary skin was tender and painful. There was no abnormality in the oral cavity, and the hard palate was not swollen. The gingival mucosa over the upper incisor was also swollen and tender, but there was no color change.

Preoperative computed tomographic scan demonstrated a midline nonenhancing round cystic lesion in the premaxillary area (Fig. 2). The lesion extended bilaterally along the inferomedial wall of both maxillary sinuses and posteriorly to the end of the hard palate. The bone of the anterior nasal floor, including the anterior nasal spine, had been absorbed completely, as had the anterior maxillary cortex and palatal bone. Coronal images showed a round 4 × 4-cm lesion extending superiorly to the nasal septum and bulging inferiorly toward the oral cavity. Sagittal images revealed that some of the bony and cartilaginous components of the nasal septum were missing because of the superior extension of the cystic mass.

On the basis of these findings, we made a diagnosis of a postoperative premaxillary cyst and excised it via a sublabial approach under general anesthesia. At surgery, 2 microplates and screws were found to be attached to both lateral sides of the cystic mass. They were partially fixed to the maxilla and were removed during surgery. The cyst wall was readily dissected from the nasolabial tissue and resected completely without rupture. The cystic fluid was a thick yellowish serous liquid, and the specimen had a thin wall. The histopathologic findings showed a cystic lesion composed of an elongated strip of fibrous connective tissue lined by epithelium, which varied from squamous to cuboidal (Fig. 3). His postoperative recovery was uneventful, and there was no facial deformity. The patient has been monitored with no evidence of recurrence for 2 years.

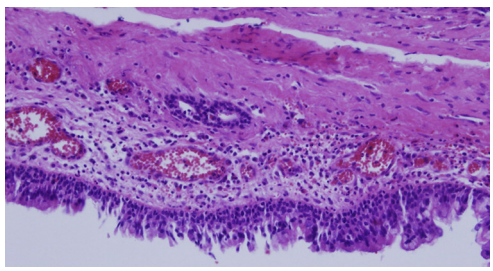

FIGURE 3. Histopathologically, the cyst wall consisted of an elongated strip of fibrous connective tissue lined by multiple layers of cuboidal and squamous epithelium (hematoxylin and eosin stain; original magnification, ×100).

DISCUSSION

A median palatal cyst is an uncommon nonodontogenic cyst located posterior to the anterior papilla of the hard palate.[2] The proximity of the lesion to the incisive canals, extension of borders, nondiagnostic radiographic errors, and ambiguous terminology in the literature make this entity somewhat challenging to diagnose clinically and histologically.[3] Several theories concerning the etiology of these cysts have been proposed. Many authors classify this cyst as a nasopalatine duct cyst because of the occasional presence of epithelial remnants from the 2 embryonic nasopalatine ducts.[4] Another theory suggests that these cysts are derived from odontogenic cell rests and the proliferation of epithelial remnants during movement of the maxillary arch during mastication, leading to synostosis.[5]

Nasopalatine duct cysts are most often detected in patients between the fourth and sixth decades of life. The cysts often present as asymptomatic swelling of the palate, but can present with painful swelling or drainage. Surgical enucleation is generally the recommended treatment. Radiographically, the cyst appears as a well-defined round-to-ovoid radiolucency between the roots of the central incisors. The pathologic findings usually range from squamous epithelium alone or combined with respiratory epithelium.[6] Because this cystic mass originated from the proliferation of epithelial remnants of the nasopalatine canal, a thorough dental examination should be carried out after a procedure such as a root canal filling of the anterior incisor. A nasolabial cyst is a common cyst in the premaxillary area; this cyst is usually positioned to one side, but a larger one can be confused with a midline mass. Pathologically, nasolabial cysts are lined with nonkeratinized epithelium with a fibrous connective tissue lamina propria and characterized by a thin epithelial lining, which may be pseudostratified or squamous in character.[7]

The initial presentation of a midline palatal cyst is typically a painless well-defined fixed swelling along the midline. It is often first recognized by either a dentist or primary physician and the patient is subsequently referred for further evaluation and management. The mass typically causes slight elevation of the nasal floor and needle aspiration of the lesion yields a yellowish-brownish fluid. Associated pain is thought to be secondary to concomitant infection. Surgical resection is usually recommended to remove a cystic mass in the premaxillary area. Marsupialization of the cyst via an intranasal approach is also an accepted treatment, even for larger cysts, and this reduces the postoperative complications, such as a persistent fistula.[6] A huge midline palatal cyst sometimes results in a bone defect of the nasal septum, as in our case. A literature review indicates that reconstruction of the anatomy is not needed in most cases.[3,8,9] We are reminded that the cartilaginous septum plays a more important role in the nasal shape and position than the bony septum.[10] If the defect includes the anterior nasal spine or a large portion of the cartilaginous septum or anterior nasal spine, the patient should be warned about the possibility of an external nasal deformity and close observation is needed.

Our patient had a rare case of a midline premaxillary mass apparently arising from a postoperative defect as a late complication. In this case, the midline palatal cyst was suspected of developing from trapped remnants of epithelium after the surgery to correct malocclusion 10 years earlier. For unknown reasons, the epithelial remnants may become activated and begin proliferating, resulting in a fluid-containing cystic structure. The bony defects that were created as the result of the previous surgery may have contributed to the enlargement of the cyst and the additional bony absorption may have resulted from pressure necrosis. Less likely, a preexisting

cystic mass, such as a nasopalatine duct cyst, happened to grow, and the bony defects provided space for it to enlarge. Patients who have had surgery on the maxilla are at risk for developing such cysts. Clinical, radiologic, and histologic evidence should be gathered before reaching a diagnosis, and awareness of this condition will help with its diagnosis and treatment.

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Bilateral Respiratory Epithelial Adenomatoid Hamartoma of the Olfactory Cleft Penetrating Into the Endocranium

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Abstract: Respiratory epithelial adenomatoid hamartomas (REAHs) of the nose and paranasal sinuses are relatively rare. These tumors usually do not extend over the boundaries of the nose and sinuses. The authors presented a 65-year-old man experiencing progressive hyposmia, followed by intermittent stubborn headache. The symptoms lasted for almost 2 years and were getting worse very slowly. Fiber-endoscopy showed relatively discrete polypoid tissue occupying the olfactory cleft bilaterally. The computed tomography and magnetic resonance imaging suggested the possible lack of the cribriform plate and the unity and uniformity of the tissues located both in the endocranium and high in the nasal cavity. The clinical picture resembled very much a esthesineuroblastoma.

The patient underwent endoscopic sinus surgery under the general hypotensive anesthesia. Frozen sections during the surgery showed REAH. The entire tumor was removed in a piece meal way, including both olfactory bulbs because they were involved within the pathologic tissue as well.

This case showed that REAH could also be a locally aggressive process, penetrating even into the endocranium.

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Key Words: Respiratory epithelial adenomatoid hamartoma, olfactory cleft, endocranial penetration, functional endoscopic sinus surgery

Respiratory epithelial adenomatoid hamartoma (REAH) is a benign lesion characterized by the glandular proliferation originating from the surface of the respiratory epithelium. The term "hamartoma" was first used by Albrecht[1] in 1904 for a tumor-like, but primarily nonneoplastic, malformation or inborn errors of tissue development.

Hamartomas can occur wherever in the body, originating from the epithelium, seromucous glands, fibrous stroma, or vessels.[2] They are commonly seen in the lung, kidney, liver, spleen, and intestine. Fifteen years ago, they were still believed to be extremely rare in the upper aerodigestive tract. Then, Wenig and Heffner[3] described for the first time a prominent glandular proliferation in the nose lined by ciliated respiratory epithelium and originating from the surface epithelium. The authors named it respiratory epithelial adenomatoid hamartoma.

A huge number of REAH located within the nasal cavity have been published by Wenig and Heffner.[3] The most common localization was the nasal septum, particularly its posterior aspect, combined with the lesions along the lateral nasal wall, middle meatus, or inferior turbinate.

Lima and collaborators[4] were the first to highlight the importance of being aware of hamartomas located in the olfactory cleft mimicking nasal polyposis.

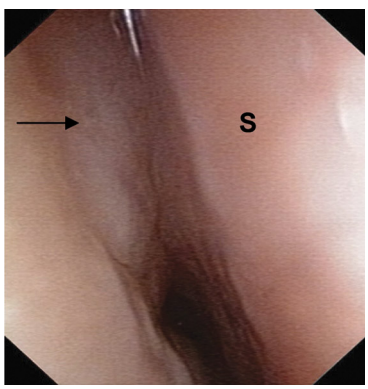

FIGURE 1. Endoscopic view of the ceiling of the right nasal cavity. A grayish area (black arrow) suggests suspected tissue of the right olfactory cleft. S indicates septum.

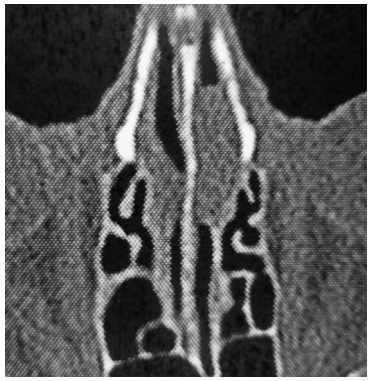

FIGURE 2. Axial CT scan indicating an unusual tissue mass in the olfactory cleft region bilaterally, particularly on the right.

Two basic hypotheses have been presented regarding the etiology of REAH. The first one suggests its congenital nature. The second one suggests that the REAH is related to chronic sinusitis, that is, to a prolonged proinflammatory environment.[2,5,6] The findings of REAH associated with nasal polyposis support this statement.[2,5,7]

The authors describe a case of bilateral REAH of the olfactory cleft, which has penetrated into the endocranial space, thus mimicking both clinically and radiologically an esthesineuroblastoma. The clinical and pathologic features of this lesion in our patient are discussed.

CLINICAL REPORT

A 65-year-old man presented with progressive hyposmia, followed by the intermittent stubborn headache. The symptoms lasted for almost 2 years and were getting worse very slowly. Antibiotic and antiallergic treatments applied by the general practitioner failed. The patient also had a history of previous neurosurgical treatment because of the left trigeminal neuralgia with anesthesia of the middle face.

Anterior rhinoscopy showed almost a normal finding, but fiberendoscopy revealed a relatively discrete polypoid tissue occupying the olfactory cleft bilaterally (Fig. 1). Axial projections of the computed tomography (CT) revealed a mass in both olfactory clefts (Fig. 2), whereas coronals magnetic resonance imaging (MRI) resembled very much the cases of esthesineuroblastoma: the suspected formation extended from the endocranium to the nasal cavity, almost vanishing the bone of the cribriform plate (Fig. 3). The

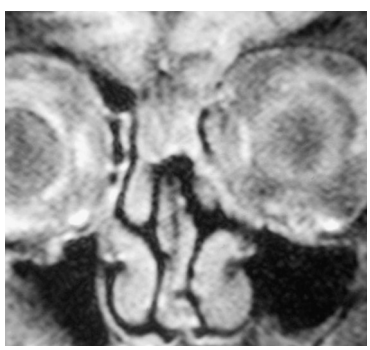

FIGURE 3. A coronal MRI projection suggesting the possible lack of the cribriform plate and the continued propagation of the tissue, located primarily in the endocranium, into the nasal cavity.

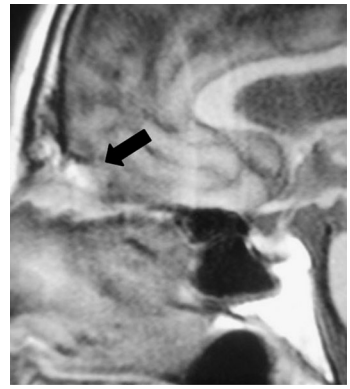

FIGURE 4. Sagittal MRI. Black arrow indicates a tumefaction, which seems to be related to the brain rather than to the nose.

MRI also suggested the same at Figure 4 a propagation of the process from the brain to the nose.

The patient underwent endonasal endoscopic removal of the tumefaction under general hypotensive anesthesia. The endonasal part was the first to be removed. It was sent for an immediate frozen section pathohistologic analysis. The result was REAH. In the meantime, no bony boundary toward the endocranium was identified. It was obvious that the tumor was penetrating the endocranium having vanished the cribriform plate. The entire tumor was removed in a piece meal way including both olfactory bulbs because they were involved within the pathologic tissue as well. At this stage of the procedure, the surgical situation resembled very much a typical finding during the endoscopic endonasal removal of the esthesineuroblastoma. We nicely presented also the anterior surface of the frontal lobe of the brain and endocranial surface of the posterior table of the frontal sinus.

After the removal, the defect of the anterior skull base was closed by means of underlay application of the fascia lata, fibrin glue, and resorbable package. A lumbar drainage was applied for 72 hours.

Despite very extensive surgery, the patient was doing well very quickly after the intervention and did not experience any of possible neurologic disorders that could occur after such a destructive procedure at the skull base. The final pathohistologic result confirmed the diagnosis of REAH. The patient is doing well for more than 1 year after surgery. He does not experience headache any longer but, expectedly, does not have any olfactory abilities. The control CT scans showed no presence of the tumor.

DISCUSSION

Respiratory epithelial adenomatoid hamartoma predominantly occur in adult patients, with male predominance and median age at the sixth decade,[3,4,8] which is in accordance with the age of our patient. No relationship to any specific etiologic agent or correlation with tobacco use or alcohol abuse could be established in these patients.[2,7] The presenting symptoms usually are vague and nonspecific, often including nasal obstruction, congestion, stubborn headache, rhinorrhea, nose bleeds, and hyposmia or even anosmia.[4,6]

The leading symptom in our patient was exactly progressive, rapidly developing hyposmia and a stubborn headache.

In general, hamartomas have no malignant potential but nor do they have any tendency to regress spontaneously.[5,9] Our patient is the best example for that: his hamartoma expanded over the cribriform plate, bony boundary into the endocranium!

The olfactory cleft localization of REAH seems to be more frequent than it was thought before.[4,5] Therefore, it is mandatory to think about this entity in all cases of diffuse nasal polyposis when, because of polypous tissue masses, it is not possible to distinguish where does the suspected mass come from: between the middle turbinate and the nasal septum or from the ostiomeatal complex proper. This is the main point to discover the real background of the disease. Moreover, misinterpretation of the REAH as chronic sinus inflammation and diffuse polyposis may result in an inappropriate treatment.

In our case, the preoperative situation strongly suggested esthesineuroblastoma, not polyposis at all. Because of the opacity between middle turbinate and nasal septum, we thought also about REAH in particular because we knew that Athre and Ducic[9] found that REAH could be a locally aggressive process! They represent the patient with frontal sinus hamartoma with expansion to the posterior right orbital wall. The patient was cured with a frontal craniotomy and cranialization. This is exactly why we decided to remove the entire tumor, including both olfactory bulbs.

Regardless of the localization, the treatment of choice for hamartomas is complete local excision of the tumefaction. Conservative surgical resection seems to be curative, with no recurrences reported in variable follow-up periods.[4,9–12] In comparison with this, our surgical treatment was really a radical one.

CONCLUSIONS

Hamartomas of the nasal cavity and paranasal sinuses are rare. These lesions are characterized by an abnormal overgrowth of tissue elements indigenous to a particular area of the body. Hamartomas do not have unlimited growth potential nor do they have metastatic potential. Extensive progression of the disease to involve the orbit and intracranial vault is exceedingly rare but possible.

Conservative surgical resection of hamartomas is curative, and the literature describes no instances of recurrent, persistent, progressive, or metastatic disease.

Our case denies some literature statements regarding the progressiveness of this tumefaction because it was found to penetrate from the olfactory clefts to the endocranium. To our knowledge, our case is the first one reported with extension of the olfactory cleft REAH to the endocranium.

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Transient Cardiac Failure Due to Takotsubo Cardiomyopathy After Surgical Reduction of Nasal Fracture

Matteo Brucoli, MD, Francesco Arcuri, MD, Mariangela Giarda, MD, Arnaldo Benech, MD, PhD

Introduction: Takotsubo syndrome, also known as ampulla cardiomyopathy, broken heart syndrome, idiopathic apical ballooning syndrome, and stress-induced myocardial stunning, has been first described by Japanese authors in 1996 and subsequently specified in 2001; it derives from the resemblance between the ancient round-bottomed, narrow-necked Japanese fishing pots used to trap octopus in Asia and the end-systolic appearance of the left ventricle on ventriculography.

Clinical Report: We introduce the case of a woman who was involved in a traffic car crash and, subsequently, was admitted to the Maxillo-Facial Unit of the Novara Major Hospital with a diagnosis of nasal fracture. She underwent general anesthesia for the reduction of the fracture; after surgery, she developed acute chest pain, elevated cardiac biomarkers, ischemic electrocardiogram changes, and transient akinesis of the left ventricle without significant epicardial coronary artery disease. A diagnosis of takotsubo syndrome was made.

Conclusions: This syndrome, which presents the same clinical features of a ventricular failure, is probably underdiagnosed, but after the introduction of sophisticated cardiac imaging and coronary intervention, more cases are identified and an unnecessary thrombolytic therapy can be spared. This reversible condition, which

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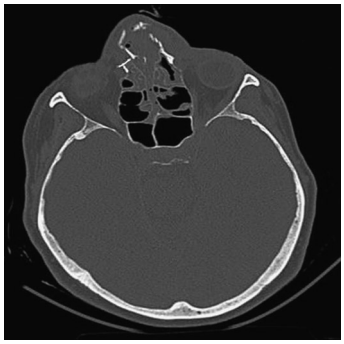


FIGURE 1. Axial computed tomographic scan demonstrating the nasal fracture.

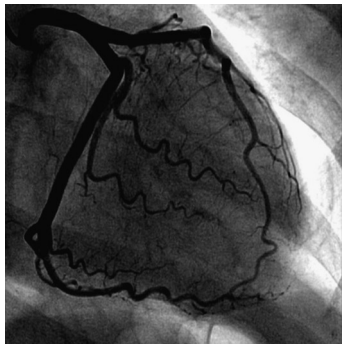


FIGURE 3. Diagnostic coronary angiogram demonstrating normal epicardial coronary arteries.

is, to our knowledge, never mentioned after a craniomaxillofacial surgical procedure, should be considered in the diagnostic algorithm for all patients presenting with acute onset of chest pain, elevated cardiac biomarkers, and ischemic changes on the electrocardiogram after a general anesthesia.

Key Words: General anesthesia, takotsubo syndrome, nasal fracture

Takotsubo syndrome, also known as ampulla cardiomyopathy, broken heart syndrome, idiopathic apical ballooning syndrome, and stress-induced myocardial stunning, has been first described by Japanese authors[1] in 1996 and subsequently specified in 2001[2]; although most cases have been initially described in this country,[3,4] nowadays there are reports from other parts of the world.[5,6]

The name of this syndrome (*tako* means octopus and *tsubo* means pot) derives from the resemblance between the ancient round-bottomed, narrow-necked Japanese fishing pots used to trap octopus in Asia and the end-systolic appearance of the left ventricle on ventriculography.

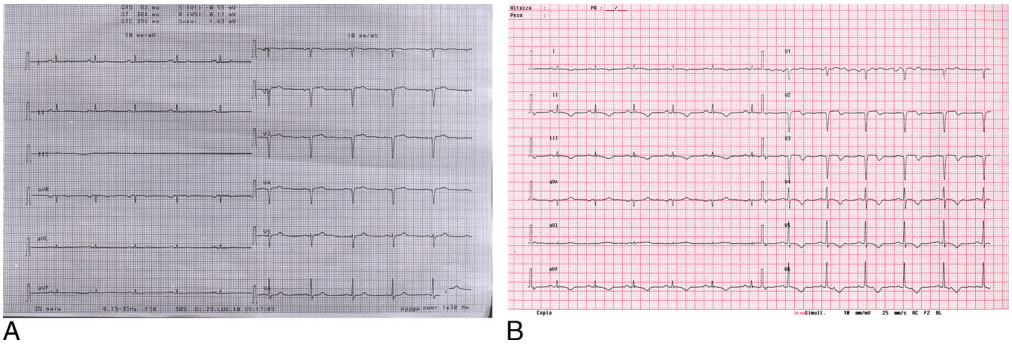
This condition is characterized by a combination of the following: (1) acute psychologic/physical stress before the onset of chest pain, 2) disproportionately low release of cardiac enzymes with respect to the degree of the left ventricular dysfunction, 3) ischemic changes on the electrocardiogram (ECG), 4) no significant epicardial coronary artery disease, 5) transient akinesis or dyskinesia of the left ventricle, 6) apical ballooning with basal hyperkinesis on the left ventriculogram, 7) rapid resolution of the cardiac dysfunc tion, and 8) absence of recent head trauma, intracranial bleeding, pheochromocytoma, myocarditis, and hypertrophic cardiomyopathy.[7]

We introduce the case of a takotsubo syndrome in a patient who underwent general anesthesia for the reduction of a nasal fracture.

CLINICAL REPORT

On April 27, 2010, a 57-year-old white woman (American Society of Anesthesiologists status II, 56 kg) was involved a traffic car crash and, subsequently, was admitted to the Maxillo-Facial Unit of the Novara Major Hospital with a diagnosis of nasal fracture (Fig. 1). She had a history of hypertension, hepatitis, and bronchial asthma. She had no known drug allergies, and results of the preoperative laboratory tests were within the reference ranges; preoperative blood pressure and heart rate were 140/80 mm Hg and 74 beats/min, respectively.

General anesthesia was induced with propofol, and after the administration of rocuronium, the trachea was intubated with a 7-mm endotracheal tube; anesthesia was maintained with sevoflurane and fentanyl. Intraoperative monitoring consisted of ECG, noninvasive blood pressure measurement, and pulse oximetry. After a successful surgical reduction of the nasal fracture, the patient was extubated, but 2 hours after the end of the surgical procedure, she presented acute chest pain and hypertension; both conditions were rapidly controlled by esmolol and 2 consecutive sublingual administrations of nitroglycerine. Serial 12-lead ECG recordings demonstrated evolving anterolateral T wave inversion; postoperative cardiac enzymes



A B

FIGURE 2. A and B, A 12-lead ECG showing evolution of widespread ST/T wave changes throughout the anterior cheat leads without Q waves including dynamic ST elevation (A) and T wave inversion (B).

showed elevation of troponin I and creatine kinase MB (1.76 and 8 ng/mL, respectively; reference, <0.01 and <5.00 ng/mL, respectively; Fig. 2). A transthoracic echocardiogram demonstrated severely impaired left ventricular systolic function with akinesia of all mid and apical segments, hyperkinetic basal contraction, and left ventricular ejection fraction of 35%.

Transfer to the cardiothoracic center took place with emergency diagnostic coronary angiography and cardiac magnetic resonance image demonstrating normal epicardial coronary arteries, apical akinesia with typical ballooning, and basal hyperkinesias without signs of ischemic lesions; diagnosis of takotsubo cardiomyopathy was made (Figs. 3–5). On postoperative day 3, a test transesophageal echocardiography showed normal ventricular function with no segmental wall motion abnormality; on the same day, the troponin I level decreased to 0.04 ng/mL. The patient's postoperative course was uneventful, and on the postoperative day 5, she was discharged to home with aspirin, β-blocker, and angiotensin-converting enzyme inhibitor. The patient has been reviewed in the outpatient clinic and she is well; a subsequent echocardiograph showed a normal volume status with normal wall motion and no mitral regurgitation.

DISCUSSION

Takotsubo is a rare disorder; in a recent study, the annual incidence of this syndrome in a Western population is calculated to be 0.00006%.[7] Interestingly, in the Japanese population, the prevalence of this syndrome has been reported to be 1%.[8] There might be a genetic component that has not been yet discovered; the mean age at onset of this syndrome is 62 to 75 years, and it frequently occurs in postmenopausal women.

The pathophysiology of this condition is debatable; numerous hypotheses have been introduced: (1) catecholamine-mediated cardiotoxicity, (2) coronary vasospasm, (3) microvascular dysfunction, (4) left ventricular outflow tract obstruction, and (5) cardiac autonomic imbalance.[9] Catecholamine mediated cardiotoxicity is the most accepted mechanism; patients characteristically present with a preceding history of psychologic/physical stress leading to increased sympathetic activity with a direct catecholamine toxic effect on the cardiac myocytes.[10]

Results are, however, conflicting; some studies report elevated catecholamine levels in takotsubo cardiomyopathy, whereas in others, catecholamine levels are normal; it is more likely that the pathogenesis of this syndrome is multifactorial.[11] The overall prognosis of this condition is favorable; however, fatal complications such as cardiogenic shock and tachyarrhythmias can occur. Although the presence of a psychologic or physical stress is accepted as one of the main features of the takotsubo syndrome, some cases have been reported to occur without any triggering effect.[3]

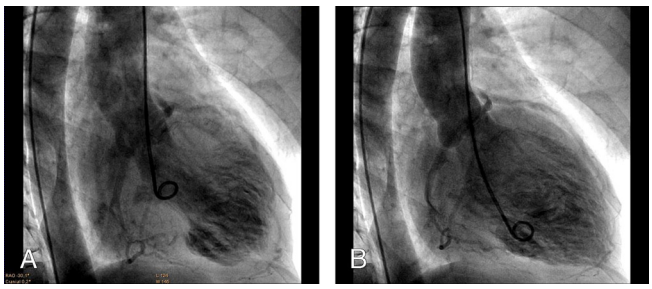


FIGURE 4. A and B, Contrast left ventriculogram showing akinesis of the apical region and hyperkinesis of the basal segments at end-systole (A) and end-diastole (B).

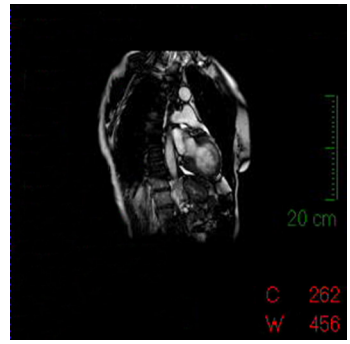


FIGURE 5. Cardiac magnetic resonance image showing left ventricular apical akinesis and basal hyperkinesis at end-systole without signs of ischemic lesions.

We theorize that stress caused by superficial anesthesia, insufficient analgesia, and surgical pain stimulus created a stressful event that caused a potentially life-threatening catecholamine release as basis of takotsubo syndrome in our patient. Because this condition leads to ventricular dysfunction with apical ballooning, the patients are potentially at a risk for ventricular thrombus formation and decisions regarding anticoagulation need to be discussed. Patients can also develop functional mitral regurgitation with hemodynamic failure; nowadays, no established guidelines exist.[5]

According to the literature, this syndrome has been frequently associated to emotional stress, sexual intercourse, and traumatic injury; it has been also described in patients with epileptic attacks, bronchial asthma exacerbations, dialysis, and during electrophysiological studies.

Wittstein et al[12] described 19 patients with ventricular dysfunction after sudden emotional stress who presented ECG, echocardiographic, and angiographic patterns similar to takotsubo cardiomyopathy; they identified a remarkable increase in catecholamine levels in such patients.

Tsuchihashi et al[2] described patients with transient apical ballooning associated with delivery, intubation, tracheotomy, lung biopsy, orthopedic surgery, colonectomy, and cholecystectomy.

It is evident that this syndrome is undistinguishable from the myocardial ischemia, and proper diagnosis is possible with angiographic evidence. The course is related to the entity of the left ventricular dysfunction, and signs of cardiac failure can be very severe, although the lesions are reversible.

The characteristic ECG features of takotsubo cardiomyopathy are nonspecific and include dynamic ST elevation (usually less than in acute anterior myocardial infarction) and/or T wave inversion typically throughout the anterior leads[13,14]; our patient also had elevated levels of cardiac biomarkers.

Transthoracic echocardiography can identify regional wall abnormalities such as akinesia of the apex and/or the midportion of the left ventricle. Typically, the area of dysfunction usually involves a larger territory than that supplied by 1 epicardial coronary artery.

Diagnostic coronary angiography needs to be performed in all patients to exclude obstructive epicardial CAD; ventriculography may demonstrate the apical ballooning that occurs in addition to hypercontraction of the basal segments.[15]

Cardiac magnetic resonance imaging provides information regarding functional involvement, chamber dimensions, and presence of intramyocardial edema; it also allows the physicians to exclude infarction and inflammatory processes.[16]

CONCLUSIONS

This syndrome, which presents the same clinical features of a ventricular failure, is probably underdiagnosed, but after the introduction of sophisticated cardiac imaging and coronary intervention, more cases are identified, and an unnecessary thrombolytic therapy can be spared.

This reversible condition, which is, to our knowledge, never mentioned after a craniomaxillofacial surgical procedure, should be considered in the diagnostic algorithm for all patients presenting with acute onset of chest pain, elevated cardiac biomarkers, and ischemic changes on the ECG after a general anesthesia.

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Maxillary Cementoblastoma in a Child

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Abstract: Cementoblastoma is a rare benign tumor that almost always occurs in the premolar or molar region and more commonly in the mandible than in the maxilla. We present a unique incisor maxillary cementoblastoma in an 11-year-old child not previously described. To our knowledge, only 2 maxillary cases, both related to canine teeth, were described in the international literature. Thus, the aim of this article was to discuss the clinical presentation, diagnosis, and subsequent treatment of a patient with a cementoblastoma in the anterior maxillary region.

Key Words: Cementoblastoma, odontogenic tumors, maxilla

Cementoblastoma is a rare benign tumor and comprises less than 1% to 6.2% of all odontogenic tumors.[1] This lesion is more frequent in young patients, with about 50% of the cases arising in younger than 20 years.[2] Approximately all benign cementoblastomas are intimately associated and partially enclosed to 1 or more roots of a single posterior mandibular erupted permanent tooth.[2,3] However, a few articles concerning maxillary lesions have been published, most of them forthcoming in the posterior site.[4]

We present a unique incisor maxillary cementoblastoma in a child not previously described. To date, only 2 maxillary cases, both related to canine teeth, were reported in the international literature.[4,5] Therefore, the aim of this work was to discuss the clinical

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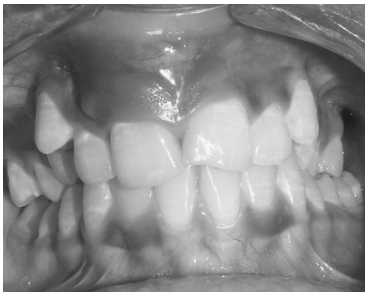

FIGURE 1. Intraoral appearance showing marked buccal expansion of the anterior portion of the maxilla.

presentation, diagnosis, and subsequent treatment of a patient with a cementoblastoma in the anterior maxillary region.

CLINICAL REPORT

An 11-year-old girl presented to the Stomatology Service, School of Dentistry, Catholic Faculty Rainha do Sertão, Quixadá, Brazil, with a slow-growing painless swelling of the anterior maxillary region that was noticed by the child's parents 5 years ago. Clinical examination showed a 3-cm, immobile, and bony hard swelling located in the anterior maxillary region (Fig. 1). The swelling was slightly painful, associated to incisor region. Upper anterior teeth were noncarious, periodontally healthy, nonmobile, and showing a normal vitality testing. Recovering mucosa was normal. Patient's medical history showed no evidence of systemic diseases, previous hospitalizations, or surgical procedures.

Radiographic examinations (Fig. 2) revealed a radiopaque mass attached to the root of the maxillary central incisor that was surrounded by a radiolucent halo. In addition, root resorption of the erupted central incisor was also detected. According to the clinical examination and radiologic findings, diagnosis of benign cementoblastoma was supposed. Because of the child's cooperative behavior, a decision of excising the lesion under local anesthesia was made.

In accordance with the patient's and her parents' desires, a conservative treatment was instituted, including enucleation followed by simultaneous endodontic management of the affected tooth. During surgical inspection, we observed an extensive root surface involvement and absence of cortical vestibular bone plate (Fig. 3). After enucleation, a high mobility of the associated tooth was noticed.

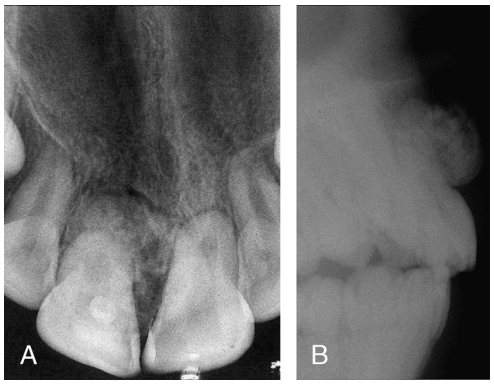

FIGURE 2. Radiologic features: radiopaque mass involving the vestibular root surface and apical region of the right central maxillary incisor (A, B).

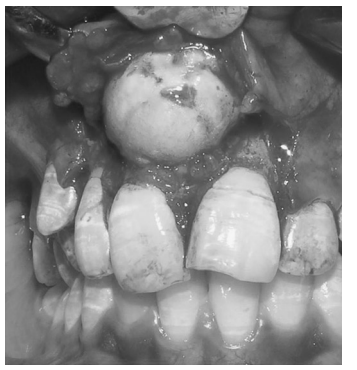

FIGURE 3. Intraoperative view showing a well-circumscribed mass attached to radicular tooth surface.

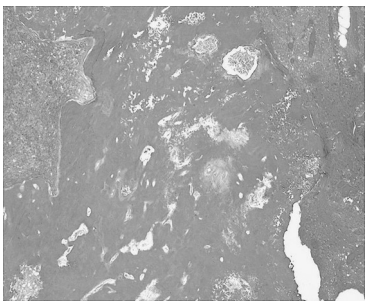

FIGURE 4. Photomicrography showing a cementum-like tumor tissue with entrapped cells in a fibrous stroma (hematoxylin-eosin stain, original magnification ×100).

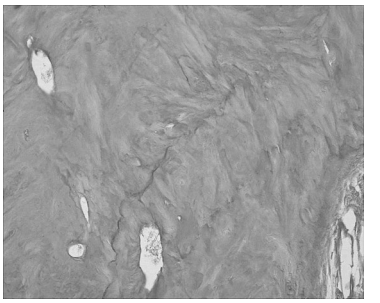

FIGURE 5. Photomicrography showing a cementum-like tissue with characteristic basophilic reversal lines (hematoxylin-eosin stain, original magnification ×400).

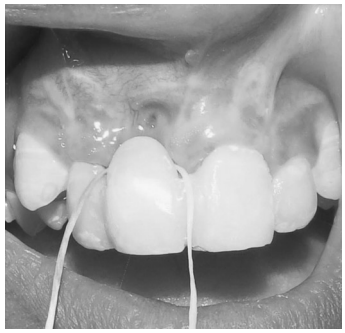

FIGURE 6. Clinical appearance 2 years after surgical procedure.

Because of this, a temporary teeth splintage was performed. Specimen was stored in 10% buffered formalin and sent for anatomopathologic examination. Microscopically (Figs. 4 and 5), tissue sections showed fragments of teeth closely associated to periapical cementum with thick layers of mineralized material, presenting trabecular basophilic organization and reverse lines interspersed by fibrovascular marrow tissue. Focal areas showed blast cells in margins of mineralized material. Other fragments exhibited connective tissue of variable density, with areas of moderate to intense inflammatory infiltrate, predominantly mononuclear. Thus, a benign cementoblastoma diagnosis was confirmed.

Following a 15-day period, patient returned with a local infectious process accomplished by worsening of tooth mobility, and analgesic and antimicrobial drugs were administered during 10 days. Subsequently, dental removal and curettage were performed under local anesthesia. For aesthetic and economic reasons, the crown of the extracted tooth was used to prepare an adhesive fixed dental prosthesis that is glass fiber reinforced. Follow-up examinations continued satisfactorily during a 2-year period (Fig. 6), and rehabilitation with dental implant will be performed after conclusion of the skeletal growth.

DISCUSSION

Cementoblastoma was first reported by Dewey[6] in 1927, but it was initially considered a cementum-producing disorder by the World Health Organization's classification of periapical odontogenic tumors together with periapical cemental dysplasia, cementifying fibroma, and gigantiform familial cementoma. However, the latest World Health Organization odontogenic tumors update recognized it as a unique true neoplasm of cementum origin, although its pathogenesis is still unknown.[7] Cundiff[8] reported an interesting case in which an odontogenic origin was supported. It was a radiographically well-described case concerning origin, development, and growth potential of a cementoblastoma that showed a 3-cm increase after a 4-year period.

We have hereby presented the first published case of cementoblastoma in which a permanent central incisor maxillary tooth was affected, with typical radiographic and histopathologic features. Most previously described cases involved mandibular molar and premolar teeth, with occasional maxillary reports.[3] The last known maxillary review series, performed by Hirai et al,[4] involved only 16 cases, of which 15 were in the posterior region. Thus, we are reporting the third case in the maxilla, which was preceded by the studies of Brocherious et al[5] and Hirai et al[4] reports.

Scarce epidemiologic data summarized in Table 1 showed a female predominance between the second and third decades of age. Clinically, swelling was a common feature together with slightly painful complaint. It has been well reported that cementoblastoma is a slowly growing neoplasia.[1–7] According to some authors,[9,10] considering that it develops by continuous deposition at a rate of approximately 0.5 mm/y, the estimated time of progression in the present case was 6 years, which was close to the report of the patient's parents. We believe that the delay in search for early diagnosis by our patient and parents was due to diverse factors, including asymptomatic development, despite late oral discomfort, socioeconomic status, and mainly geographical difficulties to access dental services.

Radiographically, radiopaque image was observed in all previous anterior maxillary cases. Regardless of anatomic site, radiographic appearance has been commonly described as a radiopacity surrounded by a radiolucent halo,[1–10] although Eversole et al[11] had reported an entirely radiolucent lesion. Brannon et al[3] showed 3 distinct image types: radiopaque (66.7%), mixed density (27.8%), and radiolucent (5.5%). Probably, interesting exophytic growth with concomitant vestibular bone resorption helped explain why a poor radiolucent line was encountered in this case. Furthermore, root relationship is another important finding that should be taken into account mainly for 2 aspects: diagnosis and treatment planning.

To diagnose the lesion, the radiographic appearance should be carefully studied. Differential diagnoses of radiopaque jaw masses range from reactive and/or benign process (idiopathic osteosclerosis, hypercementosis, focal sclerosing osteomyelitis, focal osseous dysplasia, fibrous dysplasia, odontoma, osteoid osteoma, and osteoblastoma) to uncommon malignant neoplasms (atypical osteosarcoma), depending on maturity lesion phase.[12,13] Clinical and

TABLE 1. Epidemiologic Data of Anterior Maxillary Cementoblastoma

						Radiographic Features				
Case	Author	Sex	Age, y	Clinical Symptoms	Course	Tooth	Root Involvement	Root Resorption	Size, cm	Management
1	Brocherious et al[5]	Female	26	Slightly painful swelling	1 y	Right canine	Apex, mesial portion	Yes	0.8	Enucleation with tooth removal
2	Hirai et al[4]	Female	15	Painless swelling	1 mo	Right canine	Apex	Yes	2.5	Enucleation with apicoectomy
3	Present case	Female	11	Slightly painful swelling	5 y	Central incisor	Apex, complete vestibular portion	Yes	3.0	Enucleation with tooth removal and prosthetic rehabilitation

radiographic aspects in our case were pathognomonic for cementoblastoma, especially because of its association with the root, although extremely rare location. This diagnosis was confirmed after histopathologic examination that revealed a real connection of neoplasm tissue with dental root. If these features have not been observed, a possible diagnosis of osteoblastoma could have been thought. It is an uncommon osteoid and bone-producing neoplasm, which is characterized by numerous active osteoblasts and bone trabeculae embedded by a rich vascularized delicate fibrous stroma without root relationship.[14,15]

There is a challenge in the optimal treatment in teeth affected by cementoblastomas. The most important question among dentists is whether to remove the compromised teeth. Previous authors have considered it as an innocuous, slow-growing, nonrecurrent neoplasm.[1,5,6] On the other hand, a large case series with literature review showed that recurrence was more common than formerly reported.[3] Thus, there are some who have reported satisfactory results with conservative management of these lesions, performing enucleation simultaneously with apicoectomy, followed by endodontic treatment,[4,16] whereas radical approach has been carried out by others.[2,5,14,15]

Treatment proposed for this case was dependent on some factors such as the age of the patient, anatomic site, root involvement, number of affected teeth, and the presence or absence of any pathology. In addition, we think that the psychological aspects, including wishes and expectations of the patient and those of their parents, should be considered when choosing the most appropriate treatment. Therefore, surgical removal was decided because of known recurrence after a simple curettage of this tumor.[3] Initially, we have planned a combined surgical and endodontic treatment for tooth maintenance because of patient age; aesthetic, functional, and psychological aspects; absence of other pathology; and principally the parents' desire. However, soon after surgical flap, complete cortical destruction and entire vestibular root surface connection were seen, setting as an exophytic and relatively aggressive lesion. This classification has been advocated when certain aspects are present, such as bony expansion, cortical plates erosion, teeth displacement, invasion into the pulp chamber and root canals, and extension to the adjacent teeth.[3] In addition, severe tooth mobility and lateral root resorption were observed after tumor separation of the root. Thus, a provisional adhesive prosthesis using the separated crown was made as an immediate and low-cost alternative. It will be maintained until the age of 17 or 18 years, a time when rehabilitation with endosseous implants can be considered.

Regardless of the extremely rare occurrence of anterior maxillary cementoblastomas, clinical dentists should take into consideration diverse aspects before treatment establishment. This case illustrates the importance of root involvement degree during management planning. Moreover, we propose a low-cost provisional alternative for functional and aesthetic rehabilitation until a definitive treatment can be made.

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Periodontal and Prosthetic Biologic Considerations to Restore Biological Width in Posterior Teeth

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Abstract: The biologic width is an essential dental space that always needs to be maintained to ensure periodontal health in any dental prosthetic restorations. An iatrogenic partial fixed prosthesis constructed in lower posterior teeth predisposed the development of

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subgingival caries, which induced violation of the biologic width in involved teeth, resulting in an uncontrolled inflammatory process and periodontal tissue destruction. This clinical report describes a periodontal surgical technique to recover a violated biologic width in lower posterior teeth, by crown lengthening procedure associated with free gingival graft procedure, to ensure the possibility to place a modified partial fixed prosthesis in treated area. The procedure applied to recover the biologic width was crown lengthening with some modifications, associated with modified partial fixed prosthesis to achieve health in treated area. The modified techniques in both surgical and prosthetic procedures were applied to compensate the contraindications to recover biologic width by osteotomy in lower posterior teeth. The result, after 4 years under periodic control, seems to achieve the projected goal. Treating a dental diseased area is necessary to diagnose, eliminate, or control all etiologic factors involved in the process. When the traditional methods are not effective to recover destructed tissues, an alternative, compensatory, and adaptive procedure may be applied to restore the sequelae of the disease, applying a restorative method that respects the biology of involved tissues.

Key Words: Diagnose, gingiva, dental caries, periodontitis

An adequate understanding of the relationship between periodontal tissues and prosthetic restorations is paramount, to ensure adequate function, form, aesthetics, comfort, and health of the entire dentition.[1] There may be various alternative ways to restore a diseased tooth, but just including correct diagnosis of the etiologic factors, the success of the treatment would be achieved.[2,3] Before the correction of the sequelae promoted by any disease, it is essential to eliminate or control all known etiologic factors modifying the pathogenic influential aspects of the contributing risk factors and improve the host resistance against etiologic factors.[1,4] The correction of the sequelae created by the disease may be much more complicated by applying restorative therapy than preventive control of the factors that conduct the disease process.[5] In various destructive diseases, the unique procedure to repair the sequelae that may be developed in diseased areas is to try establishing a compensatory and adaptive biologic procedure to recover the stability of diseased tissue.[6]

In this clinical report, some biologic aspects of dental prosthesis retreatment due to root caries and biologic width violation in the mandibular posterior region are discussed.

CLINICAL REPORT

A 45-year-old female individual was referred to the Department of Periodontology, School of Dentistry of Araçatuba, UNESP, Brazil, for retreatment partial fixed prosthesis in the left lower first premolar, left lower second premolar, and left lower first molar. Clinical and radiographic examination demonstrated inadequate partial fixed prosthesis fabricated by a general dentist and an insufficient keratinized attached gingiva width in the buccal side[7] (Fig. 1). These 3 unitary iatrogenic partial fixed prostheses that were fabricated with improperly subgingival margins adaptation due to overcontouring constructions were promoting bacterial plaque accumulation, gingival inflammation, and subgingival extension of dental caries into prosthetic restorations, leading to destruction of dental root tissues, which permitted the violation of biologic width (Fig. 2).

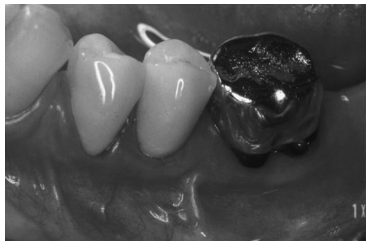

FIGURE 1. Clinical aspect showing inadequate partial fixed prosthesis.

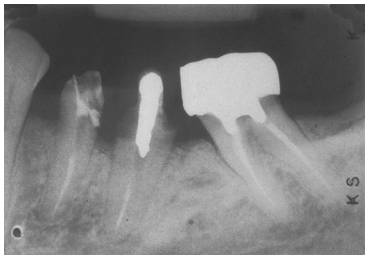

FIGURE 2. Caries violating biological width.

The objective of the treatment plan was to provide a basic biologic scheme for periodontal tissues to support the partial fixed prosthesis, which should be able to compensate dental root caries that promoted violation of biologic width. The first step was to remove the iatrogenic prosthesis to investigate the extension of dental root caries and examine the possibilities of eliminating it and, at the same time, to measure the real possibilities to recover the biologic width. The second step was teeth primary marginal preparation to receive temporary partial fixed prosthesis (Fig. 3). In the third step, a modified periodontal surgery was performed to recover biologic width via osteotomy and osteoplasty, by using at the same time 2 free gingival grafts (Figs. 4 and 5), to substitute the apically repositioned buccal flap[8–10] (Fig. 6). Three weeks after the surgical procedure, the fourth step was endodontic preparation, leaving at least 4 mm of the remaining gutta percha (Dentsply-Maillefer, Ballaigues, Switzerland), apically in the roots, to ensure that the seal of the root filling was not compromised and was sufficient to receive the prosthetic post and core.[11] Duplication of prepared endodontic channels with impression was obtained, to indirectly fabricate posts to retain the cores. The posts were cemented using zinc phosphate cement (SS White, Rio de Janeiro, Brazil), and another temporary fixed prosthesis set was fabricated (Fig. 7). All objectives established were achieved in the provisional phase before continuing with the final restoration. In the fifth step, after 60 days of periodontal surgery, the definitive partial fixed prostheses were fabricated as porcelain fused to metal crown. The maintenance phase period is keeping along 4 years, and

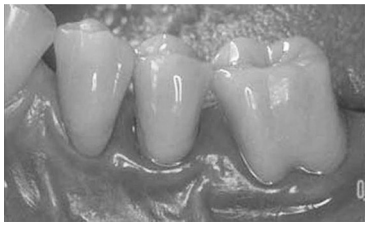

FIGURE 3. Radiographic aspect showing caries invading the biological width and inadequate prosthetic posts inside root channels.

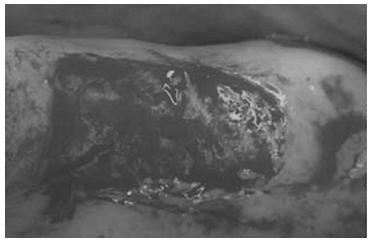

FIGURE 4. Provisional prosthesis.

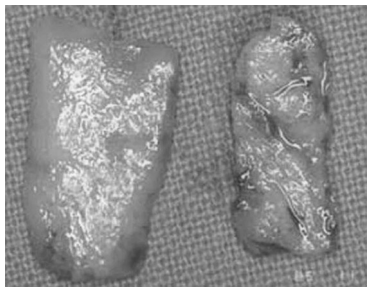

FIGURE 5. Clinical aspect of the periodontal surgical area after osteotomy, to recover biological width.

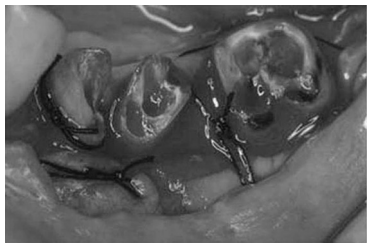

FIGURE 6. Donor site.

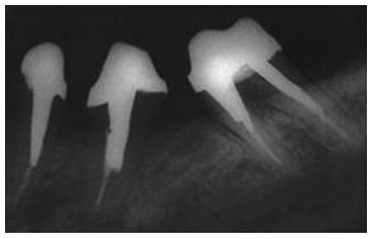

FIGURE 7. Free gingival graft and free connective tissue graft.

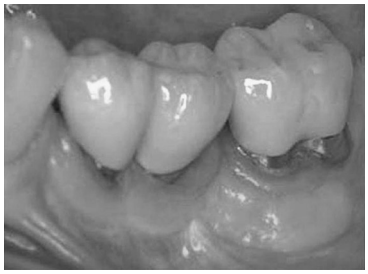

FIGURE 8. Both free gingival grafts sutured in position.

the interval of recall visits is around 6 months, determined by the nature of the case (Figs. 8 and 9).

DISCUSSION

When an iatrogenic decayed dental prosthesis is detected, the usual action taken is retreatment to provide a reconstruction of prosthetic crown with similar dimension and form as the natural tooth, to preserve the biology of periodontal tissues, functional articulation-occlusal relationship, and aesthetics.[12] Iatrogenic overcountoring prosthesis may promote plaque accumulation, periodontal disease, and subgingival caries, which may compromise dental tissue in varying degree.[2,3] The circumstance and the level of promoted damage in the tooth define the restorations around the remaining tooth structure.[6,13] In this case, when carious tissue was removed, a significant amount of dental structure was irreversibly lost, and it was realized that the repreparation of prosthetic margin could be located, invading the biologic width. This space is a natural defense of the periodontium, which will strive to maintain a certain distance between any restoration and alveolar crestal bone.[14] The biologic width is a healthy tooth space around 3 mm, located above underlying alveolar bone until the cementoenamel junction, which always needs to be respected in any prosthetic tooth margin preparation, to receive a prosthesis that preserves healthy tissues. In prosthetic restorations, the prepared tooth margin is considered as cementoenamel junction, and consequently, the biologic width becomes the space between the crest of alveolar bone and prosthetic prepared tooth margin. In this space, the components of biologic width are located, as depth of the histologic sulcus was a mean of 0.69 mm; junctional epithelium mean, 0.97 mm (0.71–1.35 mm); and supracrestal connective tissue attachment mean, 1.07 mm (1.06–1.08 mm).[15]

Whether the biologic width is violated because of the depth of prosthetic margin preparation, it is impossible to maintain periodontal tissues health, and some periodontal alterations could occur as follows: unpredictable bone loss under prepared prosthetic margin, or gingival recession and localized bone loss, or localized

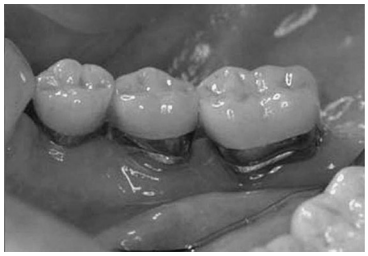

FIGURE 9. Radiographic aspect of endodontic root channel preparations to receive the core.

gingival hyperplasia and bone loss.[14] To recover biologic width, the main predictable procedures are tooth orthodontic extrusion and crown lengthening, which is a surgical procedure performed to expose a normal height of tooth structure by osseous resection, to properly restore the biologic width space.[8,9,16,17] With any surgical procedure, there are contraindications, and crown lengthening periodontal surgery is not an exception. In posterior lower molar teeth, one of the unique limitations to perform crown lengthening periodontal surgery is the furcation region that may not be exposed by bone resection because of the anatomic complexity of these areas, which permits easy bacterial plaque accumulation and consequently may induce the development of periodontal disease.[18] Then, a probable exposure of the furcation area would stimulate search for other alternative procedures and sometimes may be achieved by an adaptive and compensatory method, to permit accommodation of biologic width. In this case, the strategy to perform the crown lengthening periodontal surgery was based on recovering the maximum space of biologic width around compromised teeth, respecting furcation areas, or at least recovering around 1 mm of root space above the underlying alveolar bone, to permit repair of the supracrestal gingival connective tissue attachment. Although it was impossible to recover the normal space of biologic width, the attempt was to prepare a finishing prosthetic margin in remaining teeth structure to receive a modified dental core, after endodontic root channel preparation.[19] The gold alloy dental cores were shaped in such manner to produce a collar that embraces dental margin 2 to 3 mm around the teeth. The collar of the core permits a firm polish against the teeth, and instead of a gap localized between the core and the teeth, the improved adaptation may promote the thinnest interfacial cement line that would permit the adaptation of junctional epithelium and positioning of gingival sulcus without relevant modification in biologic width environment.[12] The free gingival graft also helps to compensate a fit between the collar core and prepared finishing prosthetic margin in remaining tooth structure by a significant augmentation of keratinized gingiva, attached in the bone and in the tooth roots.[20] The increased amount and quality of connective tissue attachment induced by free gingival graft could promote a pressure to press gingival sulcus and junctional epithelium against the teeth, leading to a corresponding decrease in probing depth.[21] The crown lengthening and free gingival graft periodontal surgery were performed at the same time. These were necessary because the apically repositioned flap, which is normally applied in crown lengthening periodontal surgery, is generally difficult to be sutured in external oblique ridge on the buccal side of mandibular posterior region.[9] Despite the difficulty, it is easier to suture the free gingival graft in the mandible posterior region than the apically repositioned flap, because the gingival graft is free, without any type of tension. These surgical procedures may achieve the necessity of establishing the health in that compromised area, by improving the ability of local area to resist against the pathogenicity of etiologic factors in restarting the disease.

After 60 days of crown lengthening and free gingival graft periodontal surgery, a duplication of prepared teeth with impression was performed to obtain a workable model for the definitive partial fixed prosthesis construction. The partial fixed prostheses were constructed as porcelain fused to metal crowns with margin finish and fit positioned supragingivally, which did not promote aesthetic limitations because the prostheses were located in the posterior area. The individual is in periodic maintenance phase mainly to prevent any failure in those adaptive procedures, which can induce periodontal disease, a complex disease that, dependent on multifactorial etiologic agents, is shown to be destructive.[22] The etiology of periodontal disease is an interaction between bacteria and risk factors as systemic factors and/or local environment factors, which could be caries, improper subgingival prosthesis margin adaptation, and overcontouring prosthesis construction as in this specific case.[1,4]

Then, to keep periodontal disease under control, it is very important to establish plaque control and to diagnose, eliminate, or control other etiologic risk predisposing factors of periodontal disease and/or to perform stable, biologic, and confident compensatory adaptive procedures to maintain the periodontal and tooth tissue in healthy conditions.

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Simpson-Golabi-Behmel Syndrome Associated With Cleft Palate

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Abstract: We report a very rare case of anomaly in the maxillofacial region. This case is a patient with a cleft palate who had Simpson-Golabi-Behmel syndrome. This X-linked symptom was first described by Simpson et al in 1975 and is characterized by prenatal and postnatal overgrowth, as well as visceral and skeletal anomalies. The syndrome consists of a distinctive facial appearance with wide nasal bridge, anteverted nostrils, wide-open mouth, enlarged tongue, and large protruding maxilla and jaw. The cleft palate was repaired surgically using the push-back method.

Key Words: Simpson-Golabi-Behmel syndrome, cleft palate, overgrowth, visceral anomaly, skeletal anomaly

Simpson-Golabi-Behmel syndrome belongs to the group of overgrowth syndromes. Simpson et al[1] were the first to report this syndrome, which has the following features: broad stocky appearance, characteristic facies (large protruding jaw, broad nasal bridge, upturned nasal tip), macroglossia, and broad, short hands and fingers. These patients have normal intelligence. Golabi and Rosen[2] and Behmel[3] reported more patients, and Garganta and Bodurtha[4] published a review of the syndrome. Patients with Simpson-Golabi-Behmel syndrome often attain a height of more than 195 cm. Xuan et al[5] mapped the gene (Xq25-q27) related to Simpson-Golabi-Behmel syndrome. Pilia et al[6] identified breakpoints in 2 female patients with X/autosomal translocations.

CLINICAL REPORT

A 14-day-old male was referred to the Oral and Maxillofacial Surgery Department on January 4, 2005, for examination of his palate. The neonate was delivered vaginally after a normal, full-term pregnancy; at birth, he weighed 4148 (2.4) g and was 53 (1.9) cm tall. He required respiratory care after the delivery. The family history was unremarkable except that his mother was hospitalized for treatment of a threatened premature birth. The patient had a micropenis and a cleft palate. Simpson-Golabi-Behmel syndrome was diagnosed owing to the presence of somatic overgrowth and the characteristic facial appearance, including hypertelorism, macroglossia, central tongue groove, and cleft palate (Fig. 1), as well as his *GPC3* gene examination. We delayed the surgery of his palate because of his poor feeding for several months after birth. At the age of 1.5 years, he weighed 12 (1.8) kg and was 53 (1.9) cm tall, and plastic surgery for the cleft palate was performed using the push-back method; surgery and the postoperative course were uneventful. At 2 years, he showed moderate overgrowth and a mild developmental delay (Fig. 2).

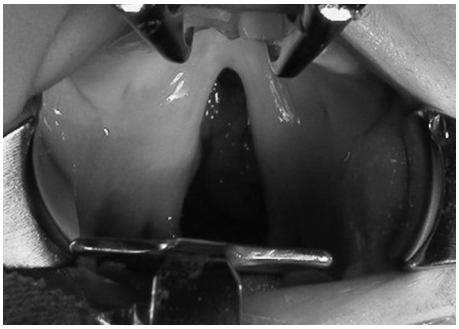

FIGURE 1. Cleft palate.

DISCUSSION

Simpson-Golabi-Behmel syndrome belongs to the group of syndromes that are characterized by overgrowth, characteristic facies, and multiple anomalies. The first case was described in 1975 by Simpson et al.[1] In 1996, Pilia et al[6] proved that Glypican 3 in Xq26/autosome was the gene responsible for the Simpson-Golabi-Behmel syndrome. The characteristic facial appearance included a large protruding jaw, a broad nasal bridge, an upturned nasal tip, hypertelorism, macrocephaly, macrostomia, macroglossia, a high-arch palate, and a short neck. Furthermore, patients with Simpson-Golabi-Behmel syndrome can have congenital heart disease (ventricular septal defects, pulmonary artery stenosis), heart block (atrioventricular block), hepatomegaly, inguinal hernia, umbilical hernia, and broad, short fingers. Simpson-Golabi-Behmel syndrome should be differentiated from other diseases wherein overgrowth occurs, such as Beckwith-Wiedemann, Pallister-Killian, Sotos, and Perlman syndromes (Table 1).[7]

Simpson-Golabi-Behmel syndrome is similar to Beckwith-Wiedemann syndrome, which is characterized by umbilical hernia,

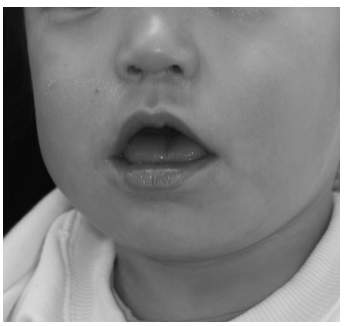

FIGURE 2. The characteristic facial appearance at the age of 2 years (a large protruding jaw, a broad nasal bridge, an upturned nasal tip, macrostomia, macroglossia, and a short neck).

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TABLE 1. Differential Diagnosis of Genetic Overgrowth Syndromes

Syndrome	Gene Location	Inheritance	Possible Sonographic Findings	Other Aids to Diagnosis
Beckwith-Wiedemann	11p15	Variable; sporadic in 85% of cases but can be autosomal Dominant; also can be caused by uniparental disomy and imprinting defects	Macrosomia, polyhydramnios, omphalocele, enlarged tongue, placentomegaly, long umbilical cord, enlarged echogenic kidneys, pancreatic cystic dysplasia	Karyotype but <1% of cases have cytogenetically detectable abnormalities; examination of parents by clinical geneticist
Pallister-Killian	Tetrasomy 12p	Sporadic	Polyhydramnios, rhizomelic micromelia, congenital heart disease, diaphragmatic hernia, increased nuchal translucency, facial abnormalities	Karyotype; confirm diagnosis with FISH probes for 12p
Sotos	*NSD1* on 5q35	Sporadic in 95% of cases	Brain abnormalities, macrocephaly	Fetal MRI; Karyotype, FISH for 5q35, NSD1 sequencing
Perlman	Unknown	Autosomal recessive	Polyhydramnios, macrosomia, visceromegaly, enlarged echogenic kidneys, cystic hygroma	
Simpson-Golabi-Behmel	*GPC3* on Xp26	X-linked recessive	Polyhydramnios, omphalocele, cystic hygroma, diaphragmatic hernia, enlarged or cystic kidneys, agenesis of the corpus callosum, Dandy-Walker malformation, and macrosomia, placentomegaly	Elevated MSAFP level, examination of mother by clinical geneticist, determine fetal sex, *GPC3* sequencing

FISH indicates Fluorescence in situ hybridization; MRI, magnetic resonance imaging.

macroglossia, and a large body. Although Simpson-Golabi-Behmel and Beckwit-Wiedemann syndromes have different causal genes, they have a similar mechanism, which is related to 1 GF2 protein, and have many similar clinical symptoms.[8]

Some patients who were previously thought to have Beckwith-Wiedemann syndrome may have Simpson-Golabi-Behmel syndrome. In patients with Beckwith-Wiedemann syndrome, the rate of growth decreases slowly from childhood, and their height is within reference limits. However, in patients with Simpson-Golabi-Behmel syndrome, overgrowth continues after childhood.[9] An adult patient with Simpson-Golabi-Behmel syndrome was found to have macroglossia. In dealing with patients with Simpson-Golabi-Behmel syndrome, one of the most important issues is a risk of upper airway obstruction by macroglossia. In particular, craniofacial surgery including dental treatment, patients with Simpson-Golabi-Behmel syndrome require careful preoperative airway management as well as management of heart dysfunction and abnormal glucose metabolism.

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Retrobulbar Lipofilling to Correct the Enophthalmos

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Abstract: In this article, we describe an alternative procedure to restore the retrobulbar volume in enophthalmic patients. We report

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the case of a patient with a late enophthalmos we submitted to retrobulbar lipofilling to correct the defect. The preoperative assessment and the surgical technique are described in detail. The volume of fat injected was 3.2 mL, with a satisfying increase in exophthalmometry measurements. The procedure was well tolerated without complications. Retrobulbar lipofilling for enophthalmos appears to be a safe alternative technique for orbital volume enhancement. It avoids the use of alloplastic materials and allows to obtain good cosmetic results with an easy technique and minimal donor-site morbidity.

Key Words: Lipofilling, enophthalmos, retrobulbar fat grafting, orbital volume defect

Among maxillofacial fractures, an important role is represented by the orbital fracture. Clinical experience shows that, despite an accurate surgical reconstruction, the ocular postoperative position is still largely unpredictable. The causes of the incorrect eyeball position after an orbital fracture repair are not well defined at the moment. However, enophthalmos may often appear as a late adverse effect. An increase in the orbital cavity volume and an atrophy of the soft retrobulbar tissues are deemed the most significant causes of this condition.

During the last years, autologous fat transplantation is growing in popularity. It is nowadays currently used by many plastic and maxillofacial surgeons. It is a safe, cheap, and simple procedure that avoids the adverse effects and the potential risks of the use of alloplastic materials.

In this article, we describe the case of a patient who had a blow-out fracture, with a late enophthalmos, following the reconstruction of the orbital floor. We used structural fat graft to correct the post-surgical deformity, carefully evaluating, before surgery, the volume to inject. Fat tissue, harvested and centrifuged, was injected in the retrobulbar space, obtaining a satisfactory improvement of eyeball projection.

CLINICAL REPORT

A 46-year-old man, after a severe facial trauma, reported multiple fractures of the right orbitomaxillary complex. He was first operated on to reduce the fracture of his right orbital floor and zygomatic arc, and then he underwent a functional septoplasty and an iliac crest bone graft of the right maxilla for a posttraumatic bone atrophy. After these surgical procedures, he developed a late enophthalmos of the right eyeball (Fig. 1), so we planned a retrobulbar lipofilling to correct the defect.

FIGURE 1. A 46-year-old man with late enophthalmos after an orbital floor reconstruction of a blow-out fracture.

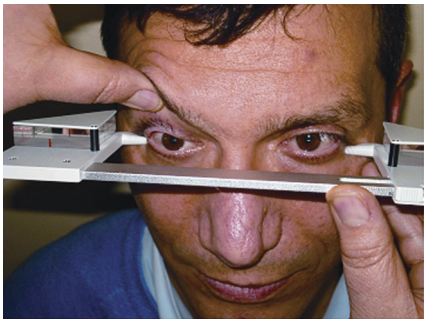

FIGURE 2. Quantification of the right-eyeball enophthalmos through the Hertel exophthalmometer.

Preoperative Assessment

First, a general clinical evaluation of soft periorbital and bony tissues was performed. Clinical evaluation was followed by a precise revision of patient's anamnesis focused on the trauma process and surgical history. As usual, preoperative photographs were taken. It was useful not only as baseline records but also as valuable assets in treatment planning.

An ophthalmologic preoperative assessment was requested to quantify the enophthalmos through the Hertel exophthalmometer (Fig. 2). This instrument quantifies the difference in the position of the eyeball between the affected side and the healthy side. A decrease of 2.4 mm of the right-eyeball projection was recorded.

To quantify the volume of adipose tissue to inject, an injection of physiological solution was performed through the conjunctiva into the muscle cone region (Fig. 3). After obtaining the same eyeball projection of the contralateral side (verified with the exophthalmometer), it was established to inject a fat volume of 2.6 mL.

The preoperative assessment was completed with a three-dimensional computed tomography (CT) scan, which gave us a detailed visualization of the soft and bony tissues. The CT images allowed us to precisely evaluate the in vivo changes of orbital volume dimension. Moreover, using a lateral orbital view, we could carefully define the deepness into which it was possible to inject the fat without damaging the optical nerve and the associated structures.

Intraoperative Procedures

Sixteen milliliters of fat was lipoaspirated from the abdominal wall through an umbilical access. The harvested tissue was centrifuged for

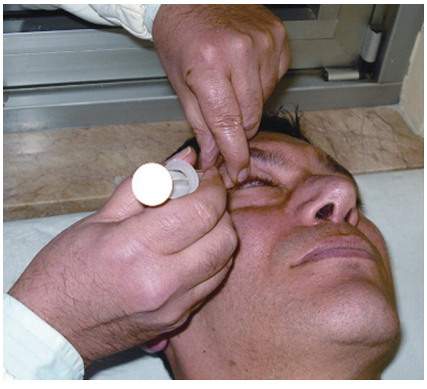

FIGURE 3. A physiological solution injection was performed transconjunctivally to quantify the volume of tissue to transplant.

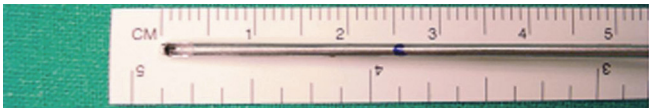

FIGURE 4. A blunt-tipped cannula 1 mm in diameter with a marking at 2.5 cm from the tip was used to inject the purified fat.

3 minutes at 3000 revolutions per minute. A blunt-tipped cannula 1 mm in diameter was used to inject the purified fat tissue through a 2-mm linear access of the inferior orbital rim (Fig. 4). The cannula was inserted, sliding over the orbital floor, until the established deepness of 2.5 cm (as showed on the cannula, Fig. 5). We injected the fat as micrografts in a radial arrangement. Assuming a resorption of about 20% to 30%, an extra volume of 0.6 mL was injected to determine a slight overcorrection (total volume injected, 3.2 mL).

With the use of the exophthalmometer, we could check intraoperatively the improvement of eyeball projection just after the transplant (Fig. 6). We verified a complete clinical resolution of the enophthalmos.

Postoperative Controls and Results

We experienced no early complications after the surgical procedure. During the follow-up, aesthetic and functional outcomes were evaluated. Photographs were taken at each visit. The first control was done in the first postoperative day (Fig. 7); the second one was after 3 weeks, and the third after 3 months.

An accurate physical examination was performed to search for any inflammatory or hemorrhagic process. To promptly show the possible resorption of the grafted fat we also checked the stability of the ocular position with the use of the exophthalmometer. An accurate ocular examination was performed to analyze visual acuity, visual field, pupillary function, and extraocular muscle function. An examination of the fundus oculi was also made to find possible damages to the optic nerve.

Stable results at the third postoperative month are described by many authors who used this technique.[1–4] The difference between eyeball projection of the 2 sides was 0.3 mm after 3 weeks (Fig. 8) and 0.7 mm after 3 months (Fig. 9). We recorded no abnormalities during ocular examination. All eyeball movements were maintained. Between the first and the last control, a low grade of fat resorption was noted. We expected it, so thanks to the slight overcorrection, fat resorption did not result in clinical effects. The resolution of the enophthalmos and a noticeable improvement of the aesthetic appearance were finally obtained.

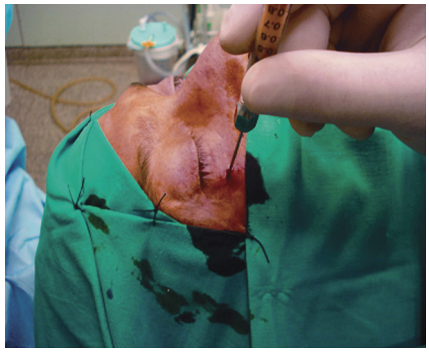

FIGURE 5. The cannula was inserted sliding over the orbital floor until the established deepness of 2.5 cm.

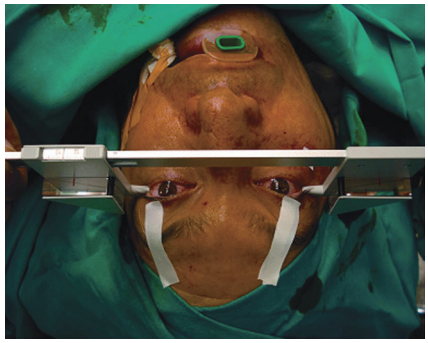

FIGURE 6. Intraoperative assessment of the correction obtained with the use of the exophthalmometer.

DISCUSSION

The most common maxillofacial fractures are the orbital fractures, which represent more than 40% of cases.[5] These fractures can involve the orbital floor as blow-out fractures or the medial orbital wall in some complex comminuted fractures. It is demonstrated that an accurate preoperative evaluation and a precise surgical technique allow restoring the preoperative anatomic condition.

Clinical experience demonstrates that functional and aesthetic sequelae can appear even after a satisfactory surgical reconstruction. Although knowledge of the postoperative sequelae of orbital floor fractures is still incomplete, it is well known that the most common consequence is the enophthalmos, defined as an eyeball moving backward or downward in the orbital cavity.

Even if there is a lack of dedicated reports, the analysis of the posttraumatic enophthalmos mechanisms is mandatory to choose a successful treatment.

Posttraumatic enophthalmos can be classified as early or late. Early enophthalmos is directly associated with the trauma, appearing immediately after. On the contrary, late enophthalmos appears later, generally after the orbital repair.

In the enophthalmic orbital cavity, the volume increases. In the early type, the anatomic changes are located behind the eyeball and consist of an inferior dislocation of the floor or a transversal expansion of the orbit.[6] A linear relationship between enophthalmos stage and volume increase has been observed.[7–10]

The causes of late enophthalmos after orbital reconstruction might not follow the same mechanism. Some authors suggested that

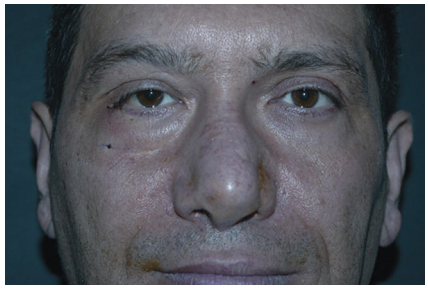

FIGURE 7. First postoperative control after 1 day.

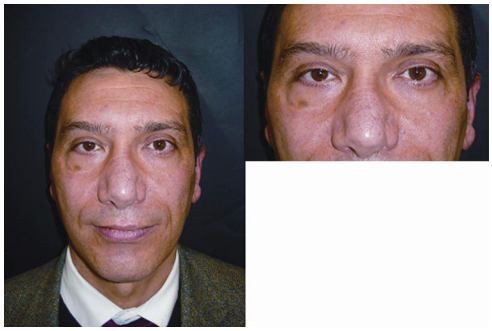

FIGURE 8. The second postoperative control after 3 weeks. A noticeable improvement of the aesthetic appearance was obtained.

alterations of the soft tissues have a decisive role in the enophthalmos after orbital reconstruction. The most important factor seems to be the retrobulbar fat tissue atrophy stage.[11] Studies have documented a loss of intraconal fat in the enophthalmos genesis. Manson et al,[12] calculating the fat volume in CT sections, underlined a 5% retrobulbar fat reduction. This hypothesis was confirmed by Ramieri et al,[6] who stated that the posterior fat is often reduced, fragmented, and back dislocated by scarring tissue. Ilankovan and Soames[13] stated that, as retrobulbar fat represents about 70% of the orbital volume, fat tissue atrophy and necrosis have a significant role in patients with enophthalmos. Driven by these pieces of evidence, we decided to apply lipofilling technique to restore the atrophied tissue.

Autologous fat grafts either as free fat grafts or as dermal fat transplants have been used since the end of the 19th century to correct various soft-tissue defects. Lipofilling, also known as autologous fat transplant, has been investigated for a long time especially as a natural filler for aesthetic and reconstructive purposes. Neuber[14] first used a fat autograft to correct some facial defects in 1893. He reported a 20% to 90% graft resorption, so these defects require multiple fillings to get a specific target. In 1950, Peer[15] formulated the theory on adipose cell survival after fat filling. His assumptions were supported by the presence of adipose stem cells in the lipoaspired fat. In 1995 and 1997, Coleman[16,17] described his method for fat filling. This technique, defined as atraumatic liposuction with injection of purified fat, was codified into 3 steps: harvesting, purification by centrifugation, and injection. Subsequently, Ersek et al[18] published several works confirming Coleman's results.[18–20]

Lipofilling has been primarily used to reconstruct minor soft-tissue defects. Then the indication has been enhanced for larger soft-tissue defects or to improve facial contour. Thanks to the recent

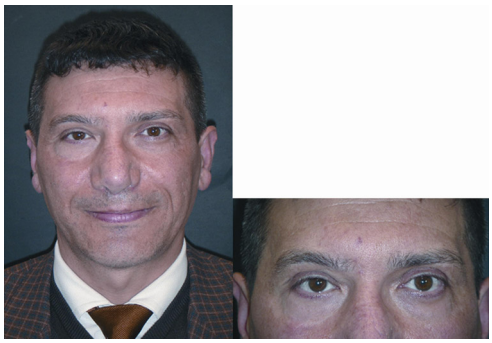

FIGURE 9. The third postoperative control after 3 months. The patient is satisfied with the result obtained.

published works on its effectiveness, reliability, and therapeutic potential, autologous fat graft is nowadays growing in popularity.

Concerning specifically the use of fat autograft for the treatment of enophthalmos, this application has not been largely studied. In 1994, Hunter and Baker[21] described the injection of a nonpurified fat into the retrobulbar muscle cone either in patients with healthy eyeball or in those with anophthalmic sockets. They obtained overall good results, but in 64% of cases, multiple injections were necessary.

More recently, Hardy et al[22] published a retrospective study on 12 patients with anophthalmic and enophthalmic orbital cavity. They used Coleman's technique to get an orbital volume improvement. Good results were obtained; a further injection was necessary in only 1 case. A strict Coleman technique has been the key point of the successful results obtained.

The low number of reports about this application can be explained by the common use of synthetic allograft materials for soft-tissue improvement and by the unpredictable long-term results of lipofilling.

In the treatment of late posttraumatic enophthalmos, autologous, homologous, heterologous, and alloplastic materials can be used. Among autologous materials, iliac crest and costal cartilage fragments have been experimented[23]; among homologous materials, the most used are non–cross-linked and cross-linked collagen; among heterologous materials, the lyophilized dura is the most commonly used, and among alloplastic materials, silastic and hydroxyapatite blocks are used.

Sometimes, the use of these materials can be harmful for the patient. Because of its high resorption grade, non–cross-linked homologous collagen[24] gave unsatisfactory results. The use of cross-linked collagen (Zyplast) in the orbital zone, even with a low capacity in inducing tissue inflammation and reabsorption, has a documented risk of blood vessels occlusion, which can lead to a severe visual damage.[25] Hydroxyapatite ceramic blocks have a demonstrated major stability, inertia, and availability.[26]

Even if cases with satisfactory results have been reported, there are some disadvantages with the use of these materials. The surgical technique, not always easy to perform, the risk of infection, and the possible immunologic reactions are the main drawbacks. Therefore, fat filling seems to be an advantageous technique for the treatment of enophthalmos.

At present, the greatest inconvenience is the unpredictable long-term outcome of the graft, related to the extremely variable rate of fat resorption.[27–29] An inadequate vascularization of the transplanted cells seems to be the reason of this problem. Many studies suggested that the surviving portion of the transplanted fat corresponds to that part in contact with the recipient site. In fact, the recipient-site vascularization is able to guarantee trophic support to that part of the graft just after the injection.

Considering these pieces of evidence, we think that a strict Coleman technique is mandatory to improve long-term graft vitality and to avoid the need for multiple injections in different surgical procedures.

This case confirmed us that lipofilling, even with this unusual indication, has features close to those of an ideal filler. It is readily available, abundant, and inexpensive; it is autologous and therefore lacks host immune response; it is safe and noncarcinogenic, and it is easily acquired with a minimally invasive procedure and has no scarring effects, allowing good functional and aesthetic results.

CONCLUSIONS

Retrobulbar lipofilling in a patient with enophthalmos, previously operated on for the surgical reduction of a blow-out fracture,

revealed itself as a safe and effective technique to improve the orbital volume. We believe that this technique is a simple and reproducible alternative to the classic techniques of orbital volume improvement with alloplastic materials. We consider retrobulbar autologous fat transplant the treatment of choice for the management of late enophthalmos.

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Orthodontic-Aided Extraction of Impacted Third Molar to Improve the Periodontal Status of the Neighboring Tooth

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Objectives: In the present clinical report, we describe the management and the long-term (3-year) outcome of a periodontally compromised lower second molar healed by orthodontic-aided extraction of the neighboring impacted third molar.

Methods: A healthy 21-year-old woman referred signs and symptoms of pericoronitis of impacted tooth 48 and periodontal injury on the distal aspect of tooth 47. The wisdom tooth was surgically exposed, and an orthodontic appliance was anchored to the neighboring teeth to stimulate eruption. After 5 months, third molar could be easily extracted.

Results: Three years after extraction, clinical and radiographic controls revealed a complete healing of the periodontal defect.

Conclusions: Orthodontic-aided extraction of impacted third molars may improve the periodontal status of the neighboring tooth. This protocol is not free from drawbacks and limitations and should be

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applied only when third-molar extraction is associated with a concrete risk of postoperative complications.

Key Words: Orthodontic therapy, impacted third molar, periodontal healing

Lower third molars represent the most frequent retained teeth among the adult population, and their failed eruption may result in pericoronitis, cysts, caries, root resorption, and periodontal problems to the neighboring tooth. Surgical removal of such impacted third molars is not lacking of complications, including alveolar osteitis, infections, hemorrhage, damage to adjacent teeth, bone fractures, and lingual or inferior alveolar nerve injury.[1] Furthermore, the extraction may cause bone loss and development (or aggravation) of periodontal pockets on the distal root surface of the adjacent second molar.[2,3] Such periodontal problems may jeopardize the prognosis of the second molar and may require further surgical interventions to be corrected.[4] An interesting therapeutic approach aimed to reduce complications of this type of surgeries is the orthodontic-aided extraction of impacted third molars. By this treatment, the retained tooth is surgically exposed and connected to an orthodontic appliance to be displaced and, successively, more easily and safely extracted in a second surgical stage.[5–7] In the present clinical report, we describe the management and the long-term (3-year) outcome of a periodontally compromised lower second molar healed by orthodontic-aided extraction of the neighboring impacted third molar.

CLINICAL REPORT

A healthy 21-year-old woman was referred to the Department of Dentistry of the Policlinic Hospital Second University of Naples (Italy) with signs and symptoms of pericoronitis of the impacted right mandibular third molar associated with the presence of periodontal injury on the distal aspect of tooth 47. In particular, periodontal pocket depths (PPDs) of 4 and 8 mm on the distal-buccal and distal-lingual aspect, respectively, of tooth 47 were measured. The radiographic examination showed a well-defined radiolucency surrounding the crown of the third molar, which appeared mesioangulated and almost completely retained into the bone, with its mesial cusps lying in close contact with the distal root of the second molar and, apically, with the mandibular canal (Fig. 1). Because of the position of the third molar and the periodontal involvement of the neighboring tooth, we proposed to the patient an orthodontic-aided extraction of the wisdom tooth by a 2-stage surgical approach.

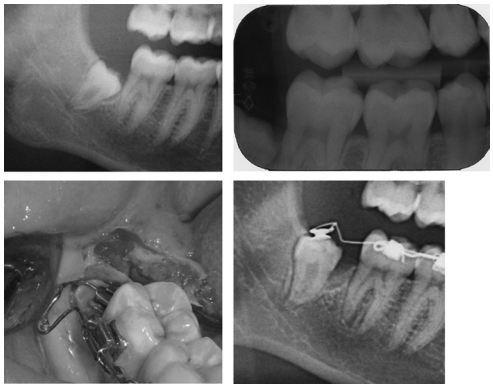

FIGURE 1. Preoperative panoramic x-ray (A) and periapical x-ray (B) showing the impacted tooth 48 lying in close contact with the distal root of the neighboring second molar. Intraoperative image of the orthodontic appliance bonded to the exposed crown of the impacted third molar (C). Panoramic x-ray 5 months after the orthodontic treatment (D).

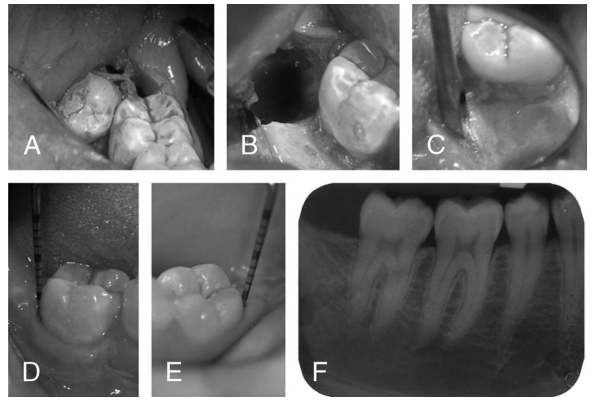

FIGURE 2. The impacted tooth was surgically exposed (A) and carefully extracted (B). A well-defined bone cortical margin at the distal aspect of tooth 47 is visible (C). Three years after third-molar extraction, physiological probing depth values are measured on both the buccal (D) and lingual (E) aspects of the tooth 47. The periapical radiographic image confirms the complete reformation of bone and the elimination of any periodontal defect at the distal aspect of tooth 47 (F).

Briefly, an accurate periodontal treatment was carried out preoperatively, and a stable anchorage was obtained by orthodontic brackets and a sectional stainless steel wire applied to the right lower premolars and molars. Subsequently, the crown of the impacted tooth 48 was surgically exposed, the bone covering the crown was gently remodeled, and a slight odontoplasty was carried out by rotating instruments. A root planing of the distal root of tooth 47 was carried out, and an orthodontic bracket was bonded on the third molar crown and connected to a cantilever anchored to the neighboring teeth, which was activated to stimulate tooth eruption. Finally, flaps were repositioned and sutured. After 5 months, once the movement of tooth 48 was radiographically verified, the cantilever was made passive, and the third molar was extracted. No neurologic complications occurred during the surgery, and the healing was uneventful. After 3 years, the PPDs were 1 and 3 mm on the distobuccal and distolingual sides, respectively, of tooth 47, and the radiographic examination revealed a prominent bone apposition with a restitutio ad integrum of the crestal bone distal to the second molar (Fig. 2).

DISCUSSION AND CONCLUSIONS

The main part of the studies agreed that third-molar extraction may be associated to an intraosseous defect at the distal aspect of the second molar, although several factors have been shown to affect occurrence, severity, and prognosis of this complication.[2,3] Such factors include the presence itself of a preoperative periodontal defect, the amount of residual periodontal structures before surgery, the size of contact-area third/second molar, the presence of a root resorption, the presence of a pathologic pericoronal follicle, and other variables such as patient age, plaque scores, and PPD values. In our case, a deep periodontal pocket was present before surgery, as clinically and radiographically evidenced; the patient was young and with a good oral hygiene; an accurate periodontal treatment was carried out before and during surgery, and these factors may

have had a role in the clinical outcome achieved. The possibility to minimize the risks of complications of impacted third-molar extraction by preventively repositioning the impacted tooth is particularly fascinating. Other approaches proposed include coronectomy,[8] by which the crown of impacted third molars is removed and the roots leaved submerged, or the surgical removal of the mesial portion of third-molar crown to create adequate space for its mesial migration.[9] However, such techniques are not free from complications and, differently from third-molar orthodontic-aided extraction, are not able to actively affect the periodontal structures of the neighboring tooth. In fact, orthodontic forces may promote bone apposition and reduce periodontal defects by putting in tension periodontal fibers.[10] In 1996, Checchi et al[5] proposed the application of a preoperative orthodontic extrusion in high-risk cases to move impacted third molars away from the mandibular canal, thus limiting the occurrence of neurologic complications. Successively, the favorable periodontal implications of this approach have been emphasized.[6,7] In particular, in the study of Hirsch et al,[6] 18 patients were treated, resulting in a significant ($P < 0.001$) PPD reduction of 6.1 ± 1.6 and 5.5 ± 1.4 mm on the buccal and lingual sides, respectively, in line with the values of PPD reduction obtained in our case. The orthodontic extrusion of impacted third molar allows improving the periodontal status of the second molar, avoiding further corrective surgeries. Furthermore, this procedure, moving the tooth away from the mandibular canal, reduces the risk of lower alveolar nerve injury and facilitates tooth extraction in the second stage surgery. On the other hand, it is a time-consuming (at least 3 months) and more expensive treatment than simple extraction, and it involves 2 surgical phases. The first-stage surgery may be very complex, especially in deeply impacted molars, and, occasionally, also the second stage may be difficult, especially when the mandibular ramus interferes with the third-molar extraction. Furthermore, a good compliance of patient to periodic controls and to a more difficult daily oral hygiene practice is required, together with patient tolerance to the uncomfortable orthodontic device, which may cause soft-tissue lesions. In conclusion, within the limits of a clinical report, it appears that orthodontic-aided extraction of impacted third molars may be used to stabilize and improve the periodontal status of the neighboring tooth, apart from moving the third molar away from the mandibular canal to reduce the risks of neurologic complications. This protocol, which is not free from drawbacks and limitations, should be applied only when there are concrete risks of intraoperative/postoperative complications, as evidenced by an accurate clinical and radiographic examination, and patient offers good compliance to the treatment plan and to the maintenance therapy.

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Metastatic Esthesioneuroblastoma Secreting Adrenocorticotropic Hormone in Pediatric Patients

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Abstract: The purpose of this article was to report a pediatric case of secondary cervical esthesioneuroblastoma involving the parapharyngeal lymph nodes. A 3-year-old boy came to our clinical observation because of a right lymphonodal mass evidenced by nuclear magnetic resonance and a diagnosis of Cushing syndrome associated with ectopic adrenocorticotropic hormone secretion, moon face, central obesity, asthenia, and hirsutism. At the age of 10 months, the patient underwent endoscopic surgery for asportation of the World Health Organization stage IV esthesioneuroblastoma. At 38 months of age, the patient underwent right parapharyngeal lymphadenectomy with surgical access by a double mandibulectomy. After surgery, serum ACTH, cortisolemia, and urinary excretion of cortisol were within the reference range. Blood pressure was recorded at 110/70 mm Hg. Moon face disappeared, as well as central obesity and hirsutism. Clinical report is presented together with brief review of literature.

Key Words: Pediatric esthesioneuroblastoma, parapharyngeal lymph nodes metastasis, ectopic ACTH, Cushing syndrome

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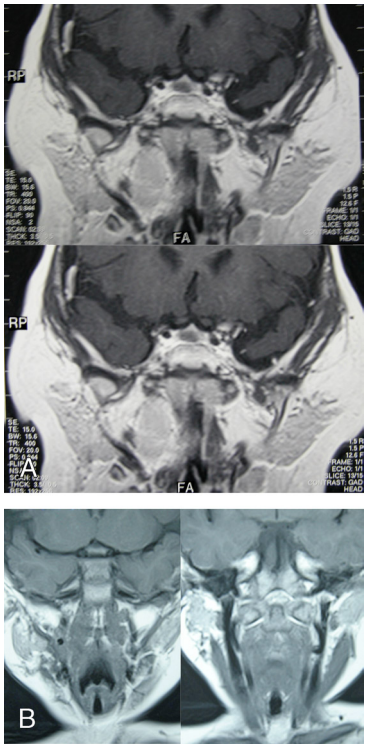

FIGURE 1. A and B, Preoperative nuclear magnetic resonance showing parapharyngeal mass.

Also known as neuroblastoma of the olfactory nerve, esthesio neuroblastoma (ENB) is a rare malignant tumor of the nasal cavity. Esthesioneuroblastomas originate from the olfactory epithelium that is situated at the roof of the nasal cavity in the cribriform plate, upper part of the superior turbinate and upper third of the nasal septum.[1–5] This is why ENBs typically involve the cribriform plate and the upper part of the nasal cavity.[1,3]

The basal cells of the olfactory neuroepithelium are presumably the progenitor of the ENB. No known causal factor exists for this tumor,[6] although diethylnitrosamine injections can induce tumors in hamsters at the site of the olfactory epithelium.[7] No hereditary patterns have been described for this neoplasm, and there is no apparent racial predilection.

Recently, it has been postulated that ENB is a member of the Ewing sarcoma peripheral neuroectodermal tumor family,[8] but immunohistochemical studies have not supported this hypothesis.[9–11] The presence of trisomy 8 has been found in ENB.[12]

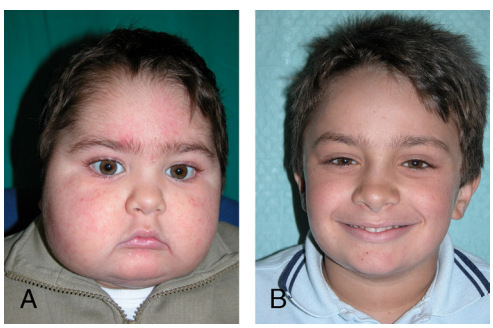

FIGURE 2. A and B, Preoperative and postoperative photographs of the patient.

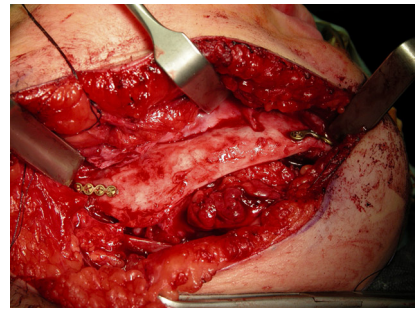

FIGURE 3. Intraoperative photograph after closure of access mandibulectomy.

The tumor typically involves the nasal cavity with extension to ethmoid sinus, anterior skull base, and orbit. The development of an ENB outside to the region in which olfactory epithelium exists is extremely rare.[13]

In addition to frequent local recurrence, olfactory neuroblastoma has also been shown to metastasize aggressively. Although the site of metastasis is widely variable and often atypical, the most common site of metastasis is the cervical lymph node.[14,15] However, the patterns of lymph node metastasis in olfactory neuroblastoma have not been well described.

Retropharyngeal lymph nodes represent the first echelon of the lymphatic drainage for the nasopharynx and the posterior pharyngeal wall.[16,17]

Ectopic adrenocorticotropic hormone (ACTH) secretion is the cause of 10% to 18% of cases of Cushing syndrome.[18–21] The most common malignancy to produce ectopic ACTH syndrome (EAS) as a paraneoplastic syndrome is small-cell lung cancer that accounts for as much as 50% of the causal tumors of EAS.[18,22,23] Carcinoids and pancreatic neuroendocrine tumors are also known to be common causes of EAS; less frequently, breast cancer,[24] colon cancer,[25,26] and prostate cancer[27] are reported to be causative.

Ectopic ACTH syndrome caused by an olfactory neuroblastoma is extremely rare; there have been only 6 cases reported in literature.[28–32] No cases in pediatric age have ever been previously reported.

CLINICAL REPORT

A 3-year-old boy came to our clinical observation because of a right lymphonodal mass evidenced by nuclear magnetic resonance (Fig. 1A). The patient has been sent to our operative unit in Istituto Gaslini (Genova), where the child was followed because of Cushing syndrome, started at 28 months of age. Appearance at presentation includes moon face (Fig. 2A), central obesity, asthenia, and hirsutism.

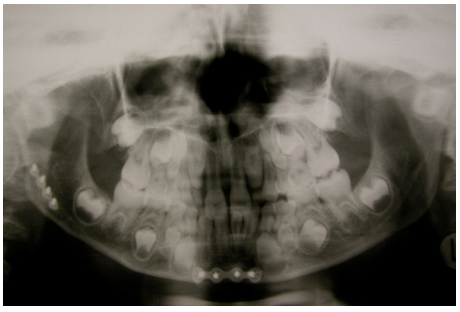

FIGURE 4. Postoperative orthopantomography.

TABLE 1. Kadish ENB Staging System

Stage	Description
A	tumor limited to the nasal cavity
B	tumor involves the nasal and paranasal cavities
C	tumor extends beyond the nasal and paranasal cavities

His blood pressure was 160/120 mm Hg. Laboratory findings includes high levels of serum ACTH (1930 pg/mL), cortisol (36.5 μg/dL), and 24-h urine cortisol (720 μg/dL), which was measured by immunoradiometric assay. A consultancy between neuroradiologists and neurosurgeons executed at Istituto Gaslini excluded that the level of serum ACTH could be related by a pituitary adenoma.

Fundamental issue, the patient underwent at the age of 10 months endoscopic surgery for asportation of a World Health Organization stage IV ENB at Graz Hospital (personal communication, Prof Stammberger, May 2003). After surgery, the patient did not execute radiotherapy but did adjuvant chemotherapy by means of RMS 96 protocol (ifosfamide, vincristine, adriamycin, and actinomycin D).

Notably, at the time of the first surgical act, a cortisolemia of 720 ng/mL was recorded, and moon face could be evidenced, even if there was no diagnosis of Cushing syndrome.

On these bases, we executed at 38 months of age right parapharyngeal lymphadenectomy with surgical access by a double mandibulectomy (Figs. 1B, 3, 4). Histologic analysis showed a lymphonodal ENB metastasis, with dimensions of 10 × 9 × 6 mm. The mass was positive for staining with neuron-specific enolase, chromogranin A, and protein gene product 9.5. Twelve months after surgery, serum ACTH, cortisolemia, and urinary excretion of cortisol were within the reference range. Blood pressure was recorded at 110/70 mm Hg. Moon face disappeared (Fig. 2B), as well as central obesity and hirsutism.

DISCUSSION

Representing about 3% of all malignant nasal tumors, ENB is an uncommon malignancy of the nasal vault, thought to be originating from the olfactory epithelium. During the 70 years after the first description of the tumor by Berger et al,[2] in 1924, 945 cases have been reported in the literature.[33]

Esthesioneuroblastoma affects male and female patients with similar frequency and can be found in all age groups,[33,34] with a bimodal peak in the second and sixth decades of life.[35]

Since its first descriptions,[2,36] the histogenesis of this neoplasm has been of debate. This controversy has given rise to a variety of names including ENB, olfactory neuroepithelioma, olfactory neuroblastoma, and others. Several anatomic origins of ENB have been suggested, including the sympathetic fibers of the anterior nasal cavity, the neuroectodermal cells of the sphenopalatine ganglion, the olfactory placode, and the vomeronasal organ.[36–42] It was not until Obert et al[43] demonstrated that the tumor originated from the superior nasal vault that agreement was needed, and it was concluded that the tumor was of neuroectodermal origin and more from the olfactory epithelium. Early attempts to categorize these neoplasms based on gene expression at a molecular level have been not correlated with clinical outcomes.

TABLE 2. Biller ENB Staging System

Stage	Description
T1	tumor in the nasal cavity and sinuses (except sphenoid) with or without erosion of the anterior cranial fossa bone
T2	periorbital or anterior cranial fossa extension
T3	brain involvement with resectable margins
T4	unresectable tumor

TABLE 3. Modified TNM Staging System

Stage	Definition
Tumor	
T1	tumor involving the nasal cavity and/or paranasal sinuses (excluding sphenoid), sparing the most superior ethmoidal cells
T2	tumor involving the nasal cavity and/or paranasal sinuses (including the sphenoid), with extension to or erosion of cribriform plate
T3	tumor extending into orbit protruding into anterior cranial fossa, without dural invasion
T4	tumor involving the brain
Node	
N0	no cervical lymph node metastasis
N1	any form of cervical lymph node metastasis
Metastasis	
M0	no metastasis
M1	distant metastasis

There is no specific symptom for ENB, as for most nasal and paranasal malignant diseases. The average delay between the appearance of the first symptom and the diagnosis is 6 months.[34] The most common symptoms are a unilateral nasal obstruction, followed by epistaxis, and anosmia.[34,44]

Traditionally, ENB is staged, as suggested by Kadish et al.[45] These authors divided the lesions into 3 stages: stage A in which the tumor is limited to the nasal fossa, stage B in which the tumor extends to the paranasal sinuses, and stage C in which the tumor extends beyond the paranasal sinuses (Table 1). Currently, there are no epidemiological, molecular, or pathologic prognostic indicators that are more valuable in predicting the recurrence of the disease or biologic behavior than the Kadish staging system.[46] The main shortcoming of the classification of Kadish et al[45] is that the language of defining stage C is too broad. Recognizing these inadequacies, a proposed classification is based on the TNM system, which is predicated on computed tomographic and magnetic resonance imaging findings. The study of ENB has clearly suffered from a uniform, effective staging system. Two other staging systems, the method of Biller et al[47] and the method of Dulguerov and Calcaterra,[34] have also been described. In 1992, Dulguerov and Calcaterra[34] criticized the Biller staging system (Table 2) because cribriform plate involvement was assumed in all cases and because craniotomy was required for accurate staging. As a result, they proposed a modified TNM system based on computed tomographic and/or magnetic resonance imaging findings (Table 3).

Morita et al[35] published a series of 49 patients treated at the Mayo Clinic over a 39-year period; they performed numerous treatment techniques that evolved over this long evaluation period. In seeking to determine prognostic factors, they found that the pathologic grading

TABLE 4. Hyams Histopathologic Grading System

Grade	LA Preservation	Mitotic Index	Nuclear Polymorphism	Fibrillary Matrix	Rosettes	Necrosis
I	+	zero	none	prominent	HW	none
II	+	low	low	present	HW	none
III	+/−	mod	mod	low	FW	rare
IV	+/−	high	high	absent	none	frequent

FW indicates Flexner-Wintersteiner; HW, Homer-Wright; LA, lobular architecture; mod, moderate; +, present; +/−, present/absent.

system proposed by Hyams et al[48] (Table 4) provided a statistically significant method of predicting outcome compared with the staging system of Kadish et al.[45]

Surgical treatment of ENB necessitates en bloc resection of the tumor with negative surgical margins. Some authors perform endoscopic surgery in selected cases,[3] and most authors propose adjuvant

TABLE 5. Incidence of Cervical Lymph Node Metastasis in Patients With ENB Collected From Various Centers Around the World

Reference	Year	Institution	Period	No. Patients	Lymph Node Metastasis, n (%)	Remarks
Schmidt et al[51]	1990	Henry Ford Hospital	1979–1989	4	2 (50)	The diagnosis was always supported by immunohistochemical studies
Beitler et al[52]	1991	Memorial Sloan-Kettering Cancer Center	1975–1985	14	4 (28.5)	The diagnosis was supported by histochemical and ultrastructural studies
Davis and Weissler[53]	1992	University of North Carolina	1972–1991	4	4 (100)	
Dulguerov and Calcaterra[34]	1992	UCLA Medical Centre	1970–1990	24	4 (16.6)	
Harrison and Lund[54]	1993	Institute of Laryngology and Otology	1976–1990	20	1 (5)	The diagnosis was supported by immunohistochemical studies in 10 patients[55]
Morita et al[35]	1993	Mayo Clinic	1951–1990	49	10 (20.4)	Electron microscopy or immunohistochemical studies were performed in selected cases
Zappia et al[56]	1993	University of Michigan	1978–1989	21	4 (19)	
Slevin et al[57]	1996	Christie Hospital	1984–1993	9	3 (33.3)	The diagnosis was always supported by immunohistochemical studies and in 2 cases by ultrastructural investigations
Koka et al[44]	1998	Gustave-Roussy Institute	1980–1995	40	11 (27.5)	
Levine et al[46]	1999	University of Virginia	1976–1998	35	9 (25.5)	
Pickuth et al[58]	1999	Martin Luther University	1986–1998	22	5 (22.7)	
Eriksen et al[59]	2000	Odense University Hospital	1977–1997	13	2 (15.3)	The diagnosis was supported by immunohistochemical studies
Resto et al[60]	2000	Johns Hopkins Hospital	1981–1998	27	9 (33.3)	The diagnosis was supported by immunohistochemical and ultrastructural studies
Chao et al[61]	2001	Washington University	1976–1996	25	5 (20)	
Simon et al[62]	2001	University of Iowa	1978–1998	13	2 (15.3)	The diagnosis was supported by immunohistochemical studies in 5 cases, one of which required electron microscope examination
Total				320	75 (23.4)	

Here are included only the largest and the most recent series reported in literature partly because the diagnosis was often supported by histochemical, immunocytochemical, and/or ultrastructural investigations.[63]

UCLA indicates University of California, Los Angeles.

radiotherapy after craniofacial resection. However, others recommend radiotherapy even after resection of T1 lesions with negative surgical margins.[49]

Esthesioneuroblastoma is a malignant neoplasm with a likelihood for cervical node metastasis.[44] Historically, the suspicion of neck metastases was not associated with ENB. The incidence of metastatic disease has been reported to vary, probably depending on the accuracy of the diagnosis and on the follow-up. To carefully evaluate the incidence of neck metastases from ENB, we collected data from only the largest and most recent series reported in the literature, in which the diagnosis was often supported by histochemical, immunocytochemical, and/or ultrastructural investigations.

Overall, ENB is a tumor with a known propensity for local recurrence and lymphatic and hematogenous dissemination.[50] Table 5 shows data compiled from 15 institutions worldwide regarding the incidence of synchronous and metachronous lymph node metastasis. Given the high percentage of patients who develop cervical nodal disease (23.4%) without considering the presence of lymph node micrometastasis,[64] treatment of the clinically negative neck may be warranted.[60] Although the overall incidence of neck node metastases is more than 20% in ENB, most surgeons do not consider elective neck dissection to be part of the initial treatment at this time. This is because these cervical node metastases occur over a long period and not during the first 1 or 2 years. It is quite likely that one may be able to treat them at a later date when they are clinically apparent. If cervical nodal disease is present initially, or develops subsequently, then a selective neck dissection should be performed. The relatively high frequency of occurrence of lymph node metastases is sufficient to refute claims that ENB is a "low-grade" malignant tumor that seldom metastasizes, as previously pointed out by Bailey and Barton.[65] Indeed, Beitler et al[52] found cervical lymph node metastases to be as common as local recurrence.

Although the treatment of advanced ENB and the role of standard chemotherapy as a treatment have not been clearly delineated, case studies have shown that for Kadish stage C and high Hyams grade, cisplatin-based chemotherapy is active, and that high-grade advanced ENB is sensitive to platinum-containing chemotherapy regimens.[66] In addition, neoadjuvant chemotherapy has been shown to be effective in tumor burden reduction and in improving survival and outcome when combined with surgery and radiation.[67–70]

Cushing syndrome is caused by exogenous glucocorticoids, hypothalamic-pituitary disorders, primary adrenal disorders, and ectopic secretion of ACTH. This is usually caused by small-cell carcinomas of the lung, carcinoid tumors, pancreatic islet cell tumors, medullary thyroid carcinomas, and rarely, paragangliomas. The diagnosis of EAS secondary to ENB was established based on the presence of ACTH seen on immunohistochemical staining of the tumor, the disappearance of symptoms, and a decrease and normalization of plasma ACTH and cortisol levels after resection of the tumor.[71]

In international scientific literature, 6 cases of EAS related to ENB have previously been reported.[29–32] The present case is the first ever reported in a patient of pediatric age.

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Clinical Management of Peripheral Ameloblastoma

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Abstract: Peripheral ameloblastoma is a rare epithelial odontogenic tumor, limited to the soft tissues of the gingiva or oral mucosa. Peripheral ameloblastoma represents approximately 2% to 10% of all

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ameloblastomas. It is always considered to be benign, but occasionally it may be locally aggressive or with malignant potential. In this article, we report 3 new cases of benign peripheral ameloblastoma and further discuss the clinical management of this disease.

Key Words: Peripheral ameloblastoma, clinical management, prognosis

Peripheral ameloblastoma (PA) is a rare epithelial odontogenic tumor, limited to the soft tissues of the gingiva or oral mucosa, representing approximately 2% to 10% of all ameloblastomas.[1] Peripheral ameloblastoma is always considered to be benign, but occasionally it may be locally aggressive or with malignant potential. This tumor was first described by Kuru[2] in 1911 and systematically reviewed with 160 cases by Philipsen et al[1] in 2001. Up to now, about 41 new cases have been reported after the 160 cases,[3–26] and 7 cases have been reported as malignant PA. We report 3 new cases of benign PA and discuss the clinical management of this disease.

CLINICAL REPORTS

From March 2006 to June 2009, there were 10 patients with PA treated at our Department of Oral and Maxillofacial Surgery, Ninth People's Hospital, School of Medicine, Shanghai Jiao Tong University. Among the 10 patients, 7 patients were treated by outpatient treatment, and 3 patients were treated by hospitalization. For the patients with outpatient treatment, the follow-up record could not be found because they took their clinical history with them and were always lost after treatment with good results. The incomplete information of these patients was obtained at the Department of Oral Pathology. Complete patients' information could be reviewed only in the 3 patients treated by hospitalization. Among the 10 patients, there were 9 men and 1 woman; their ages ranged from 34 to 81 years, with a mean of 46.9 years. Three lesions occurred in the maxillary gingiva, and 7 lesions occurred in the mandibular gingiva. We report the 3 cases with complete information and discuss the management of PA.

Patient 1

A 38-year-old man complained a small and painless mass enlarging slowly in his palatal gingiva of the left first maxillary premolar for 6 years. Six years before his presentation, he found a small mass in his palatal gingiva of the left first maxillary premolar without any symptoms. Four years before his presentation, he received tooth extraction because of the loosening of the left first maxillary premolar in his local hospital, followed by fixed-denture restoration. The small mass was not treated. Recently, he felt slight pain of the mass. Medical history was noncontributory. Physical examination revealed a mass with papilla on its surface, 1.5 × 1.0 cm, soft, mobile, with clear boundary without tenderness in his palatal gingiva of the left first maxillary premolar (Fig. 1). The neighboring teeth were stable with fixed denture. The routine laboratory tests were all within reference ranges. Computed tomographic scan showed the increased thickness of the soft tissue according to the palatal gingiva of the left first maxillary premolar. Computed tomography enhancement scanning showed that the density of the thickening tissue was slightly increased. However, the bony structure was intact without any absorption (Fig. 2). Pathological biopsy was performed confirming the diagnosis of ameloblastoma. Surgical resection was performed under general anesthesia. During the operation, the left lateral incisor, cuspid, and second premolar were extracted, and the mass could be separated from the bony surface completely. Then, the alveolar bone was removed (Fig. 3). Finally, the wound was packed with iodoform gauze. The wound healed well without any complications. The postoperative pathologic examination confirmed the diagnosis of ameloblastoma (Fig. 4). During the follow-up period of 26 months, the wound had healed well, and no tumor recurrence occurred (Fig. 5).

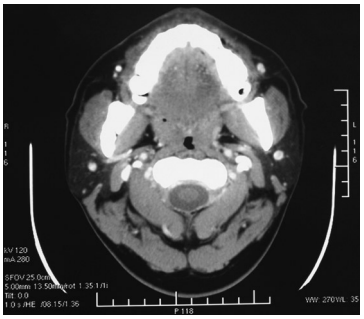

FIGURE 2. Patient 1. Computed tomographic scan shows the intact bony structure of the maxilla without any absorption.

Patient 2

A 47-year-old man complained a small and painless mass enlarging slowly in his right mandibular premolar region for 2 years. Two years before his presentation, he found a small mass with a diameter of 0.3 cm in his right mandibular premolar region; both the right mandibular first and second premolars had been lost for a long time. The mass grew larger gradually without any symptoms. Medical history was noncontributory. Physical examination revealed a mass

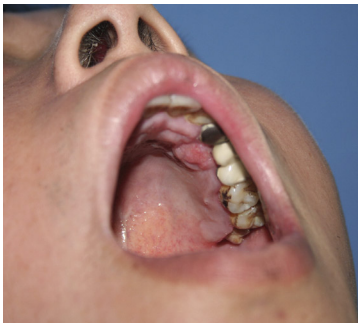

FIGURE 1. Patient 1. Front view of the mass in the palatal gingiva of the left first maxillary premolar.

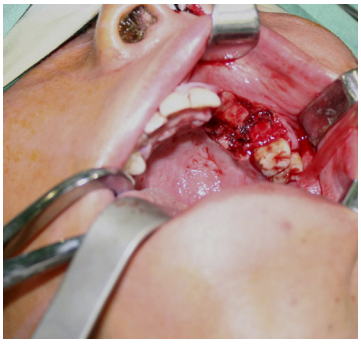

FIGURE 3. Patient 1. Surgical resection of the mass with removal of alveolar bone.

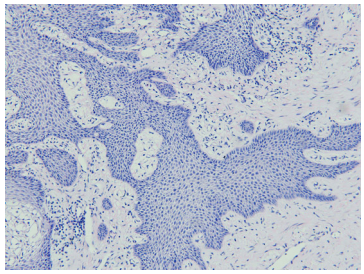

FIGURE 4. Patient 1. The postoperative pathologic examination confirmed the diagnosis of PA (hematoxylin-eosin staining, original magnification ×200).

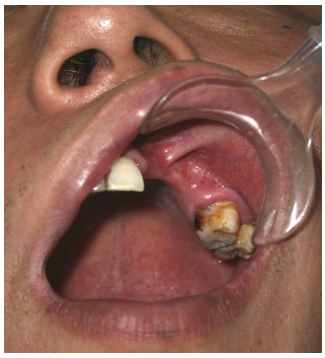

FIGURE 5. Patient 1. Twenty-six months after surgery, the wound healed well without tumor recurrence.

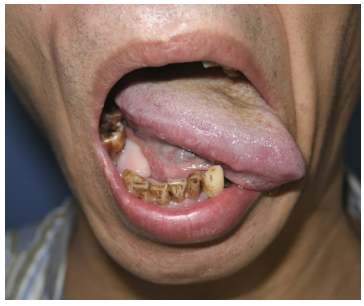

FIGURE 6. Patient 2. Front view of the mass in the right mandibular premolar region.

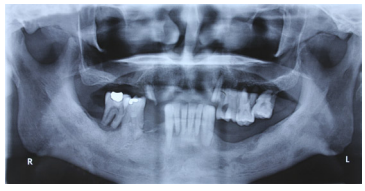

FIGURE 7. Patient 2. Panoramic radiograph shows the increased thickness of the soft tissue in the right mandibular premolar region, without obvious bone absorption by the mass.

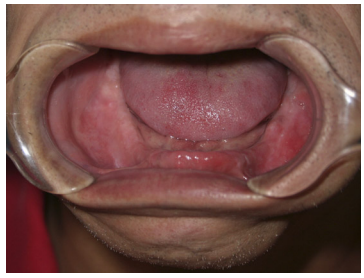

FIGURE 8. Patient 2. Twenty-seven months after surgery, the wound healed well without tumor recurrence.

2.0 × 1.0 cm, hard, immobile, with clear boundary without tenderness in his right mandibular premolar region (Fig. 6). The routine laboratory tests were all within reference ranges. Panoramic radiograph showed the bony absorption of the mandible and maxilla; in the right mandible, there was root absorption of the right mandibular second molar, and root fracture of the right mandibular first molar could be observed; the increased thickness of the soft tissue in the right mandibular premolar region could also be observed with a small piece of bony-like structure in this region (Fig. 7). The clinical diagnosis of this mass was epulis. Surgical resection of the mass was performed with extraction of the residual roots and loosening teeth. The frozen section of the mass showed the diagnosis of ameloblastoma, and further extended resection of the soft tissue and the alveolar bone was performed. The wound was closed directly with the buccal slipping flap. The wound healed well without any complications. The postoperative pathologic examination confirmed the diagnosis of ameloblastoma. During the follow-up period of 27 months, the wound had healed well, and no tumor recurrence occurred (Fig. 8).

Patient 3

A 57-year-old man complained a small and painless mass enlarging slowly in his right mandibular premolar region for 1 year. One year before his presentation, he found a small mass with a diameter of 0.2 cm in his right mandibular premolar region without any symptoms. Then, the mass grew larger gradually. Medical history was noncontributory. Physical examination revealed a mass with granules on its surface, 1.5 × 1.2 cm, moderate in hardness, immobile, with clear boundary without tenderness in the lingual side of the right mandibular second premolar region. The routine laboratory tests were all within reference ranges. Panoramic radiograph showed no bony absorption of the mandible or root absorption of the right mandibular second premolar and first molar. The clinical diagnosis of this mass was papilloma of the gingiva. Surgical resection of the mass was performed. The frozen section of the mass showed the possible diagnosis of ameloblastoma. So, further extended resection of the soft tissue and the alveolar bone with extraction of right mandibular second premolar and first molar was performed. The wound was closed directly with the buccal slipping flap. The wound healed well without any complications. The postoperative pathologic examination confirmed the diagnosis of ameloblastoma. During the follow-up period of 38 months, the wound healed well, and no tumor recurrence occurred.

DISCUSSION

Up to now, there are 204 cases of PA reported in the literatures, including our 3 cases with complete information. The most cases (96.6%) of PA are benign, and only 7 cases (3.4%) of PA have been reported to be malignant.

Among the 196 benign PAs, 65% of PAs occurred in males, and 35% of PAs occurred in females; the male-female ratio is 1.8:1. The ages of the patients ranged from 9 and 92 years, with a mean age of 51.9 years. The most common site of PA is mandibular gingiva (66.0%), followed by maxillary gingiva (28.2%), buccal mucosa (5.3%), and floor of mouth (0.5%).

Among the 7 malignant PAs, there are 4 males and 3 females. The ages of the patients range from 40 to 83 years, with a mean of 65.4 years, which is larger than that of the patients with benign PAs. Three lesions (42.9%) occurred in the mandibular gingiva, including one involving the floor of mouth. Four lesions (57.1%) occurred in the maxillary gingiva.

The clinical manifestation of benign PA is always known as painless, sessile, firm, and of exophytic growth, with relatively smooth surface; in some cases, it is granular or pebbly or papillary. Its color is always normal just like the adjacent mucosa, but is sometimes red. The x-ray examinations always show negative findings on bone involvement, at most pressure resorption at the bone margin. Without pathologic biopsy, it is very difficult to make the correct diagnosis. It is always diagnosed as epulis, benign tumor, papilloma, granuloma, and so on.

Conservative supraperiosteal surgical excision with adequate disease-free margins is the treatment of choice for the benign PA. The recurrent rate is always low (<20%), because of incomplete removal of the lesion. In our report, all of the 3 cases underwent surgical resection of the mass with extraction of the teeth with compact relationship to the mass; no tumor recurrence was found during the follow-up period. For the malignant PA, radical surgery is recommended, because of tumor invasion and metastasis, and long-term follow-up is necessary.

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Ganglioneuroma of the Base of the Skull

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Abstract: Ganglioneuromas are rare benign tumors originating from the ganglion cells of the sympathetic and parasympathetic nervous system. Ganglioneuromas in the base of skull are extremely rare. In this article, we describe a case of primary ganglioneuroma below the foramen ovale observed in a 38-year-old man. The lesion

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was asymptomatic. The patient underwent surgical intervention for diagnostic and therapeutic purposes. Craniomaxillofacial surgery was conducted by the oral and maxillofacial surgical team and the neurosurgical team. Combined frontotemporal–preauricular infratemporal approach was used to expose the lesion. Cerebrospinal fluid leakage and facial paralysis did not occur postoperatively. The results of histopathologic examination indicated that the excised lesion was a ganglioneuroma. Clinical follow-up was done, and no recurrence has been observed up to now.

Key Words: Ganglioneuromas, skull base, craniomaxillofacial surgery

Ganglioneuromas are well-differentiated benign tumors originating from the ganglion cells of the sympathetic and parasympathetic nervous system.[1] They consist of a variable mixture of ganglion cells, Schwann cells, and nerve fibers; the ganglion cells may resemble primitive neuroblasts or mature nerve cells.[2] They most commonly occur in the posterior mediastinum, retroperitoneum, and occasionally in the adrenal gland.[3–6] Unusual sites include the neck,[7,8] bone,[9] and intestine.[10] Presentation of a ganglioneuroma in the base of the skull is extremely rare, and only 2 cases have been reported in English literature.[11,12] In this article, the diagnosis and treatment of a 38-year-old man with a primary ganglioneuroma below the foramen ovale are described.

CLINICAL REPORT

A 38-year-old man was referred to our department for uncomfortable feeling in the throat. The results of ear, nose, and throat examination were within normal limits. No systemic symptoms were found after the physical examination. All laboratory values were within reference ranges. However, computed tomography (CT) and magnetic resonance imaging (MRI) showed an uneven nonenhanced mass measuring 2 × 2.5 cm below the right foramen ovale (Fig. 1). Computed tomography and MRI coronal images demonstrated that the foramen ovale was dilated by the tumor (Fig. 1B, D). From the physical examination and imaging results, a clinical diagnosis of benign tumor (schwannoma) or low potential malignancy (fibrosarcoma) was made.

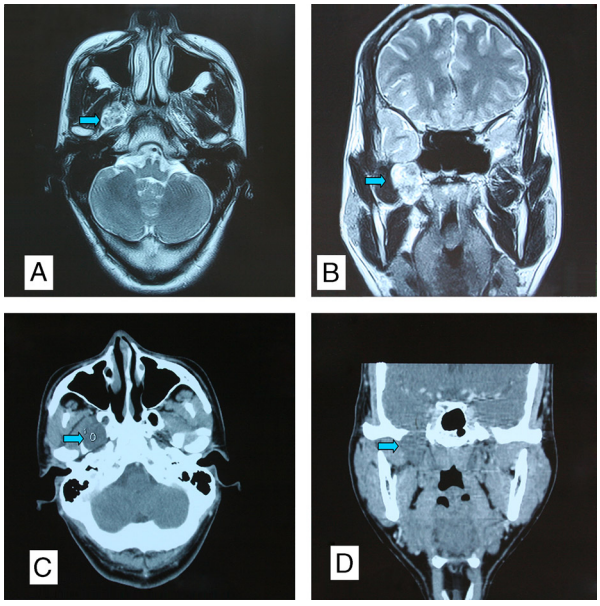

FIGURE 1. A, Axial T2-weighted MRI shows an uneven tumor in the skull base. B, Coronal T2-weighted MRI shows a tumor below the right foramen ovale. C, Axial CT scan with contrast enhancement shows a nonenhanced mass measuring 2 × 2.5 cm in the skull base. D, Coronal CT scan with contrast enhancement shows that the foramen ovale was dilated by the tumor (arrow shows the mass).

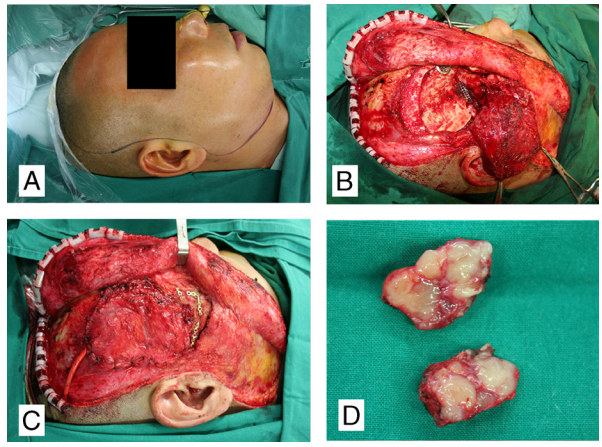

FIGURE 2. A, Combined frontotemporal–preauricular infratemporal approach was used to expose the lesion. B, Defect of the skull base was repaired with titanium mesh, and craniofacial skeleton removed for exposure was stabilized with titanium plates and screws. C, Zygomatic arch was stabilized with titanium plates and screws. D, Cut surface of the tumor.

Because the site of the tumor is below the foramen ovale, fine needle aspiration was painful, and the cytology was inconclusive. The patient was encouraged to have a surgical intervention for diagnostic and therapeutic purposes. Craniomaxillofacial surgery was conducted by the oral and maxillofacial surgical team and the neurosurgical team. Combined frontotemporal–preauricular infratemporal approach was used to expose the lesion (Fig. 2A). The incision begins in the frontal scalp; curves behind the hairline; and turns downward, with a preauricular extension; continues anterior to the tragus; surrounds the mandibular angle; then extends inferiorly to the neck along a skin crease. To expose the temporal fossa and

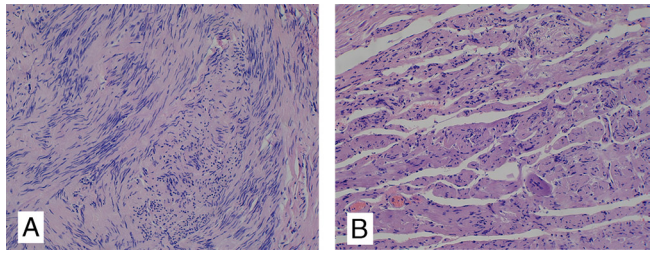

FIGURE 3. A, Microscopic examination reveals a predominance of neurofibroma-like areas composed of Schwann cells exhibiting scant spindled cytoplasm, wavy dark nuclei, and inconspicuous nucleoli (hematoxylin-eosin, original magnification ×200). B, Microscopic examination reveals ganglion cells exhibit abundant eosinophilic cytoplasm, large vesicular nuclei, and prominent nucleoli (hematoxylin-eosin, original magnification ×200).

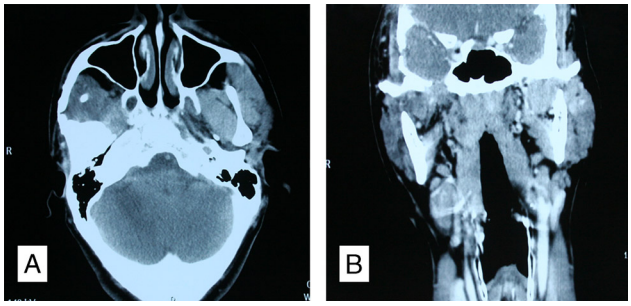

FIGURE 4. No sign of recurrence of the lesion 8 months after operation, based on CT scan with contrast enhancement: axial image (A) and coronal image (B).

protect the facial nerve, zygomatic arch transection and temporal musculofascial flap are created. Parts of temporal bone and sphenoid bone were removed with high-speed drill to expose the mass. The encapsulated tumor was entirely excised, and the facial nerve integrity was preserved. The frozen section of the tumor indicated that the mass was a benign neurogenic tumor. As a result, craniofacial skeleton and zygomatic arch removed for exposure were stabilized with titanium plates and screws. The defect of the skull base was repaired with titanium mesh (Fig. 2B, C). Temporal musculofascial flap was used to fill up the operative cavity. Cerebrospinal fluid leakage and facial paralysis did not occur postoperatively.

Histologically, the tumor was well circumscribed with a fibrous capsule. The cut surface was gray-white without necrosis and hemorrhage (Fig. 2D). Microscopic examination reveals a predominance of neurofibroma-like areas composed of Schwann cells exhibiting scant spindled cytoplasm, wavy dark nuclei, and inconspicuous nucleoli (Fig. 3A). Ganglion cells exhibit abundant eosinophilic cytoplasm, large vesicular nuclei, and prominent nucleoli, and they are seen throughout the tumor, often in small aggregates surrounded by ill-defined fascicles of Schwann cells (Fig. 3B).

The man has been kept under oncological follow-up. Based on CT scan with contrast enhancement, there was no sign of recurrence of the lesion 8 months after operation (Fig. 4A, B).

DISCUSSION

Ganglioneuromas are benign neurogenic tumors originating in the sympathetic and parasympathetic nervous system. These lesions frequently develop in the posterior mediastinal or retroperitoneal sites and occasionally in the adrenal gland.[3–6] According to reported rates of distribution, ganglioneuromas affect both sexes equally and have a slightly higher incidence in women.[13,14]

Hypotheses for the pathogenesis of benign ganglioneuromas include the spontaneously or artificially induced maturation of neuroblasts from a neuroblastoma into distinct ganglion cells, separation of the remaining cells from the embryonic neural crest, and necrosis of neuroblasts at an early stage of tumor development.[13,15,16]

The development of ganglioneuromas is characteristically silent because they do not release excessive catecholamine or steroid hormones.[17] These slow-growing, noninvasive tumors are usually identified during imaging studies performed for other reasons or when they affect the cranial and sympathetic nerves and cause dysfunction.[17,18] They are usually identified at the late stage of the disease. The appearance of clinical symptoms depends on the tumor size and site.[17,18]

On imaging studies, ganglioneuromas appear as large, round-lobulated, solid heterogeneous masses. Maweja et al[19] reviewed 8 cases of ganglioneuromas and suggested that ganglioneuromas have the following characteristics: some presence of calcifications (2.4%–60%), absence of vessel involvement, and a nonenhanced attenuation of less than 40 Hounsfield units on CT; and low nonenhanced T1-weighted signal, a slightly high and heterogeneous T2-weighted signal, and a late and gradual enhancement on dynamic MRI.

Because this skull base tumor was asymptomatic, it may lead to a significant number of misdiagnoses. The most common clinical differential diagnosis includes schwannoma, meningioma, and fibrosarcoma. Schwannoma arises almost exclusively from sensory nerves such as vestibular nerve, trigeminal nerve, and hypoglossal nerve. Enhanced CT scan demonstrates uneven enhancement of the tumor with smooth boundary.[20] Meningioma is encapsulated and attach to the dura, from which they derive their blood supply. On nonenhanced CT scans, meningioma is isodense or slightly hyperdense compared with the brain.[21] Contrast enhancement is strong and homogeneous; margins are distinct. Hyperostosis of the underlying bone is common and can be appreciated radiographically. On MRI, meningioma is isointense or hypointense, when compared with normal brain on both T1-weighted and T2-weighted imagings.[21,22] Fibrosarcoma is a soft-tissue tumor that arises from bone, periosteum, scalp, or dura. It is often accompanied by bony destruction, showing a regular but discrete lytic radiographic image.[23]

It is generally suggested that the best treatment of ganglioneuromas is complete excision via surgery or microsurgery depending on the site of the lesion.[2,5,7,10] Radiotherapy should be avoided because of the risk of radiation-induced neoplastic transformation of the lesion.[24] The prognosis of the ganglioneuromas is good.[2,5,7,10] The transformation into malignant peripheral nerve sheath tumor is extremely rare. Well-documented cases of ganglioneuroma metastasis have not been reported in the literature. This clinical report suggests that ganglioneuromas are rare nonmetastatic tumors that should be considered in the differential diagnosis of lesions of the base of skull.

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Treatment of Temporomandibular Joint Ganglion Cyst

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Abstract: Ganglion cysts of the temporomandibular joint are very rare and always misdiagnosed as synovial cyst, parotid gland tumor, or other cystic lesions. They present with pain, swelling, or dysfunction. Image studies could facilitate to identify the tumor mass from the adjacent soft tissue, but a definitive diagnosis could be made from the pathologic report.

A 59-year-old woman presented to the clinics with a chief complaint of a painless swelling mass in the right preauricular region of 3-month duration. Computed tomography was performed, which showed a small radiolucent lesion adjacent to the right condyle. Local excision was performed, and the specimen was sent for histologic examination.

Microscopic examination showed a cystic space walled by dense fibrous connective tissue without epithelial or endothelial lining. Immunohistochemical staining of these lining cells showed positivity for vimentin and negativity for cytokeratin. These findings were consistent with the diagnosis of ganglion cyst.

Ganglion cysts present as unilobulate or multilobulate cysts that arise from the collagenous tissue and is filled with highly viscous fluid. It does not communicate with the joint cavity. In contrast, synovial cyst is a true cyst lined by cuboidal or flattened cells from the synoviocytes and is filled with gelatinous fluid. It may or may not communicate with the joint cavity. Excision is the treatment of choice of symptomatic cystic lesions. Incomplete excision of these lesions may cause further recurrence or infection. Thus, injection of hydrocortisone or aspiration may be considered as an alternative management.

Key Words: Ganglion cyst, temporomandibular joint, synovial cyst

Ganglion cysts are the most common benign cystic tumors of the hand. They can present as unilobulate or multilobulate cysts that arise from the collagenous tissue and are filled with highly viscous fluid that contains hyaluronic acid, albumin, globulin, and glucosamine.[1]

Ganglion cysts of the temporomandibular joint (TMJ) are very rare.[2–11] They are difficult to be diagnosed and always misdiagnosed as synovial cyst, parotid gland tumor, or other cystic lesions. They present as a painful lesion or swelling and may cause dysfunction. Image studies could facilitate to identify the tumor mass from the adjacent soft tissue, but a definitive diagnosis could be made until the intraoperative observation or pathologic report confirms the diagnosis.

In this report, we present a case of ganglion cyst of the TMJ and review the literature of this scarce entity.

CLINICAL REPORT

A 59-year-old woman presented to the plastic surgery outpatient department of China Medical University Hospital with a chief complaint of a painless swelling mass in the right preauricular region of 3-month duration. Her medical history was significant for thyroid

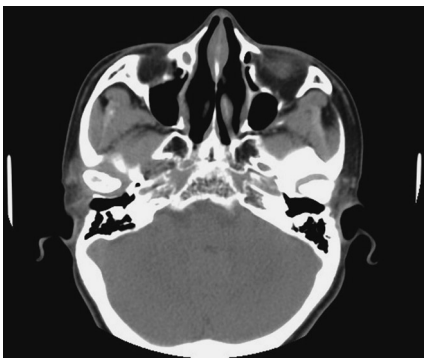

FIGURE 1. The transverse view of computed tomography showing a small radiolucent lesion adjacent to the right condyle.

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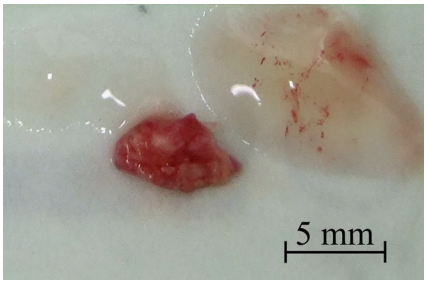

FIGURE 2. A cystic lesion with gelatinous fluid content, which favors a ganglion cyst.

cancer with regular follow-up in the general surgery outpatient department of China Medical University Hospital, and no history of traumatic injury was reported.

Physical examination showed a preauricular hard and firm mass situated 1.5 cm anterior to the tragus. The mass was 6 × 4 mm and had grown slowly within half a year. She had normal mouth opening without any deviation, joint noise (crepitus), or numbness sensation. The mass did not change in position or size while opening the mouth. An osteoma was ruled out by computed tomography, which showed a small radiolucent lesion adjacent to the right condyle (Fig. 1). Therefore, under the impression of a soft-tissue nodule, surgical intervention was carried out.

Under general anesthesia, a preauricular approach to the TMJ was used and carried down to the tumor. A cystic lesion of approximately 1 cm was found, and it was located adjacent to the lateral portion of the condyle. After dissection of the cystic lesion carefully, there was no communication with the inner articular space. Unfortunately, the surface of the mass ruptured, and there was some gelatinous fluid that leaked (Fig. 2). The mass was then excised completely for biopsy, and the wound was closed in layers.

The specimen was fixed in formalin and sent for microscopic examination. Hematoxylin-eosin–stained sections showed a cystic space walled by dense fibrous connective tissue without epithelial or endothelial lining (Fig. 3). Immunohistochemical staining of these lining cells showed negativity for cytokeratin (Fig. 4) and positivity for vimentin (Fig. 5). These findings were consistent with the diagnosis of ganglion cyst.

Six-month postoperative follow-up showed normal mandibular joint function and no recurrence of preauricular swelling, pain, or facial nerve injury.

DISCUSSION

Synovial cyst is first described by Baker in 1885.[12] It is a true cyst lined by cuboidal or flattened cells from the synoviocytes and is

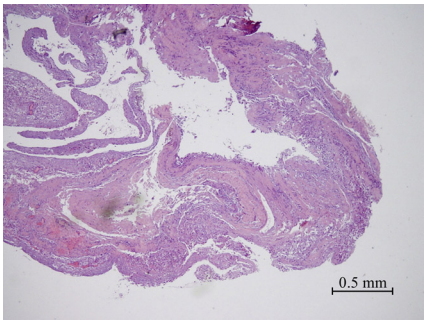

FIGURE 3. Photomicrograph of the cystic-like lesion demonstrating fibrous connective tissue (hematoxylin-eosin stain).

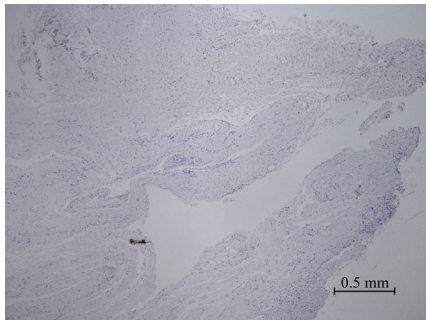

FIGURE 4. Photomicrograph of tissue stained immunohistochemically with cytokeratin showed negative stain.

filled with gelatinous fluid.[13] It may or may not communicate with the joint cavity. The pathogenesis of the synovial cyst may arise from the embryonic displacement of synovium and herniation of the synovial lining into the surrounding tissues.[14] It is associated with traumatic and inflammatory process.[5] The terms *synovial cyst* and *ganglion cyst* have been used interchangeably and are considered to be synonymous in the past.

Ganglion cyst is a pseudocyst with fibrous tissue lining that arises from myxoid degeneration and cystic softening of the collagenous tissue of a joint capsule.[15] It contains clear, high-viscosity gelatinous fluid rich in hyaluronic acid and other mucopolysaccharides.[16] Different from synovial cyst, it does not communicate with the adjacent joint cavity.[17]

Gaisford et al[2] presented a synovial cyst of the TMJ in 1969. Cysts of the TMJ are scarce. They are more common in females, with a female-male ratio of 3:1, and often present with preauricular swelling and TMJ pain. Moreover, most patients had this pathologic lesion on the right side.[8] As there are many anatomic structures in the preauricular area, the differential diagnosis of TMJ tumor includes parotid mass, ganglion cyst, synovial cyst, retention cyst, sebaceous cyst, benign cervical lymphoepithelial cyst, and benign vascular or neural tumor.

Clinical presentation such as swelling, tenderness over the preauricular area, limitation of mouth opening, or noise cannot help us identify which disease it belongs to. Therefore, other radiographic picture could help in diagnostic workup.

Ultrasound was first recommended by Lopes et al.[18] They found that the cystic lesion showed an anechoic picture, and this could differentiate it from the parotid gland tumor. Then, Bonacci et al[5] clarify an anechoic lesion for ganglion cyst and hypoechoic lesion for synovial cyst. However, Nablieli et al[8] stated a case of ganglion cyst that has been shown to be hypoechoic later. These conflicting

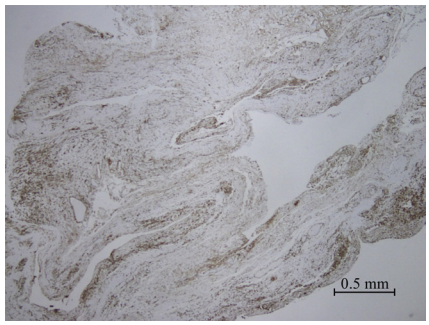

FIGURE 5. Photomicrograph of tissue stained immunohistochemically with vimentin showing positive stain.

reports concluded that ultrasound is not reliable in the diagnosis; however, it can be performed safely and easily.

Computed tomography and magnetic resonance imaging are the most useful diagnostic tools, which are suggested literatures.[19–21] The relationship between the lesion and the condyle could be shown clearly. These could help the in diagnosis and serve as a useful guide in the succeeding surgical treatment. Although there are so many diagnostic tools, an accurate diagnosis is not usually made preoperatively, and a definitive diagnosis could be made only until intraoperative observation or postoperative pathologic examination is performed.

Immunohistochemical results of the lining of these lesions in the study of Nablieli et al[8] were helpful in final diagnosis. Ganglion cyst shows a positive reaction to vimentin, a mesenchymal marker, but negative to cytokeratin, an ectodermal marker and vice versa for synovial cyst. Therefore, immunohistochemical staining could be one of the useful tools to differentiate the cysts.

CONCLUSIONS

Excision is the treatment of choice of symptomatic cystic lesions. Incomplete excision of these lesions may cause further recurrence or infection. Then, injection of hydrocortisone or aspiration may be considered as an alternative management.

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Adenoid Cystic Carcinoma Associated With Mucous Retention Cyst of the Parotid Gland

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Abstract: Mucous retention cysts of the parotid gland are rare, and a coexistent adenoid cystic carcinoma is even an unusual occurrence. Such coexistent adenoid cystic carcinomas with mucous retention cyst of the parotid gland are difficult to diagnose clinically and, at times, stage difficulty in their management. We report a rare case of adenoid cystic carcinoma associated with mucous retention cyst of the parotid gland with its diagnostic and management dilemma in a 14-year-old adolescent girl.

Key Words: Mucous retention cyst, parotid gland cyst, adenoid cystic carcinoma, parotid tumor

Intraparotid cystic lesions comprise approximately 5% of all salivary gland lesions, and many of them represent cystic components of neoplasms.[1,2] Most of them are unilateral, simulating parotid tumor and require surgical removal for diagnostic purpose. Cystic masses within the parotid gland present problems in diagnosis and in treatment. Accurate diagnosis of such lesions at times is difficult, and there may be a necessity for surgical removal of the entire lesion for the histopathologic examination. The mucous retention cysts of

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FIGURE 1. Preoperative photographs.

the parotid gland are rare, and association of adenoid cystic carcinoma is probably an extremely rare occurrence. This article reports an unusual presentation of adenoid cystic carcinoma associated with mucous retention cyst of the right parotid gland with its diagnostic and management dilemma in a 14-year-old adolescent girl.

CLINICAL REPORT

A 14-year-old girl presented with a recurrent unilateral parotid swelling that was aspirated several times by other surgeons for histologic examination with no results. An incision and drainage of the swelling was attempted elsewhere that left the patient with a scar over the preauricular region without regression of the lesion. The patient reported to us with a 3 × 2-cm firm swelling that is nontender with shiny skin over the lesion and with a scar in the right preauricular region (Fig. 1). No other nodules or masses were palpated. Facial nerve dysfunction was not evident.

Ultrasonographic examination reported with evidence of multiple nodular discrete lesions in the right parotid region suggestive of parotid group lymphadenopathy. The right parotid gland showed an enlarged intraparotid lymph node (Fig. 2).

With an informed consent, under general anesthesia, superficial parotidectomy sparing the branches of facial nerve by careful dissection was executed. The tissue margin clearance was confirmed with frozen sections before the wound was closed in layers. The tissue was sent for histopathologic examination that reported on hematoxylin-eosin staining as mucous retention cyst of the right parotid gland with features of an adenoid cystic carcinoma in the connective tissue capsule of the cystic lesion (Fig. 3). The patient is kept under regular follow-up and observation with no signs and symptoms of recurrence or facial nerve dysfunction even after 2 years of postoperative period (Fig. 4).

DISCUSSION

Mucous retention cysts of the parotid gland have been rarely reported.[2] The association of adenoid cystic carcinoma with the

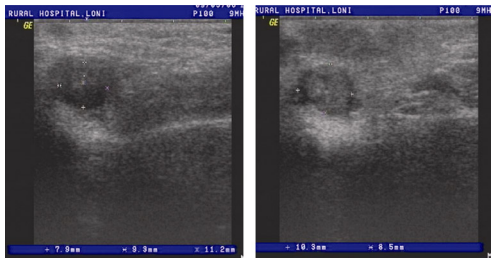

FIGURE 2. Ultrasonography images.

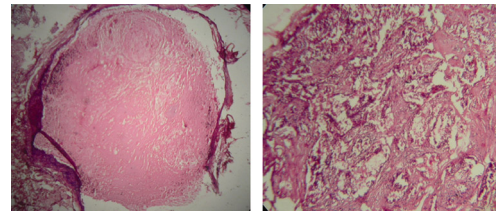

FIGURE 3. Histopathologic photomicrographs (hematoxylin-eosin, original magnifications ×10 and ×40).

mucous retention cyst in the parotid gland is probably the extremely rare case to be reported.

In instances of cysts originating from the parotid gland, it is very difficult to identify the site of origin.[3] An understanding of the clinical and histologic differential diagnosis of cystic parotid lesions will lead to better management of these lesions.

The association of mucoepidermoid carcinoma with mucous retention cyst of the parotid gland is well documented in the literature.[3–5] Similarly, adenoid cystic carcinoma associated with salivary duct cyst in the sublingual gland has also been reported.[6–9] Surgical excision of the parotid masses with a sufficient margin of normal tissue has been shown to be the most acceptable method of managing lesions of the salivary gland.[2] In the present case, histopathologic examination revealed cystic lesion lined by epithelium comprised of collagen fibers interspersed with fibroblasts, endothelial lined vascular spaces surrounding the epithelium, as well as darkly stained cells resembling the cells of adenoid cystic carcinoma, cystic lumen filled with mucoid material. All these features are suggestive of mucous retention cyst with features of adenoid cystic carcinoma in the connective tissue capsule of the cystic lesion.

Histopathologically, the primary lesion was clearly the mucous retention cyst, and the accompanying tumor, adenoid cystic carcinoma, should be considered secondary. No literature reports on the incidence of mucous retention cyst of the parotid gland transforming into adenoid cystic carcinoma. Management of such lesions sways the challenge to the surgeon because the malignant component is associated with the cystic lesion. The entire mass along with some part of the adjacent normal tissue has to be removed and sent for frozen section for margin clearance.[2] Postoperative radiotherapy can be reserved depending on the type and grade of malignancy that is associated and the surgical margins. The present case had clear surgical margins and had no signs of recurrence even after a follow-up

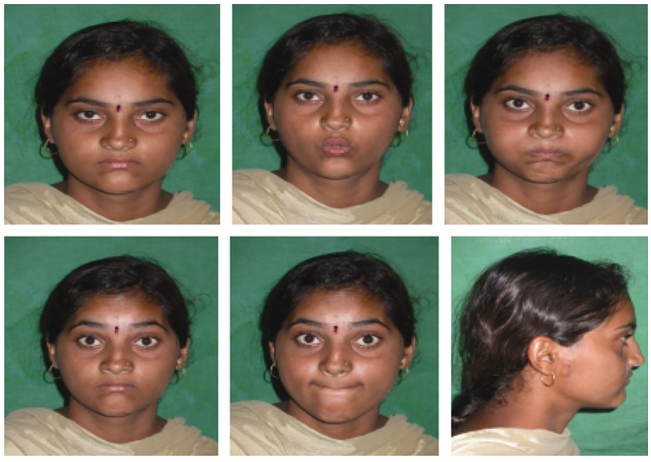

FIGURE 4. Postoperative photographs.

of 2 years. Careful monitoring and regular follow-up are mandatory in such presentations.

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Conservative Treatment of Odontogenic Myxoma

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Abstract: Odontogenic myxomas (OMs) are nonencapsulated rare benign tumors that can occur in gnathic bones. They are locally invasive and have a high recurrence rate. Radiologically, OMs show a multilocular (in the majority of cases) or unilocular radiolucency, with either distinct or poorly defined margins. Histopathologically, OMs are characterized by spindle-, wedge-, or stellate-shaped cells loosely arranged in an abundant mucoid background. Myxomas are mainly asymptomatic. Radical surgery, excision, and enucleation followed by curettage of the surrounding bony tissue have all been advocated as treatment options. This study presents a successful case of conservative treatment of OMs with a 5-year follow-up.

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Key Words: Odontogenic tumors, myxoma, conservative treatment

Odontogenic myxomas (OMs) are nonencapsulated rare benign tumors that can occur in gnathic bones.[1] They appear to be odontogenic in origin, related to malformations or missing teeth.[2] They are characterized by mucoid or gelatinous gray-whitish tissue that replaces the cancellous bone and expands the cortex.[3] Odontogenic myxomas represent from 3% to 9% of all odontogenic tumors.[4,5] They are locally invasive and have a high recurrence rate, ranging from 10% to 43%.[3] There is a preference for females, in a proportion ranging from 1:1.5 to 1:4.[1] Most patients are between the second and the fourth decades of life.[3] The mandible is the most frequently affected anatomic region (2.5:13), with a preference for the posterior mandibular body, angle, and ramus areas.[3] In the maxilla, the alveolar and zygomatic processes are the most common sites, with frequent invasion of the tumor into the maxillary sinus.[2]

Radiologically, OMs show a multilocular (in the majority of cases[6]) or unilocular radiolucency, with either distinct or poorly defined margins.[3] The tumors may also occur with "soap-bubble," "tennis racket string," or "honeycomb" patterns.[7] Histopathologically, these lesions are characterized by spindle-, wedge-, or stellate-shaped cells, many of which have long fibrillar processes that tend to intermesh,[1] loosely arranged in a abundant mucoid background.[3] Myxomas are mainly asymptomatic. The lesion grows slowly and may be present several years before consultation is sought, occasionally attaining sufficient size to produce considerable facial deformity.[2] The involved teeth may be malpositioned or mobile and are usually viable.[7,8]

Radical surgery, excision, and enucleation followed by curettage of the surrounding bony tissue have all been advocated as treatment options.[1] This study presents a successful case of conservative treatment of OM with a 5-year follow-up.

CLINICAL PRESENTATION

A 23-year-old woman came to the Bucomaxillofacial Ambulatory of the Federal University of Espirito Santo (Brazil), on February 8, 2001, complaining of a mass located in the premolar and molar region of the left maxilla. Clinical examination revealed a soft-tissue mass measuring about 2.5 cm in length, causing facial asymmetry. The overlying mucosa was normal. Radiological examination showed a poorly circumscribed unilocular radiolucent

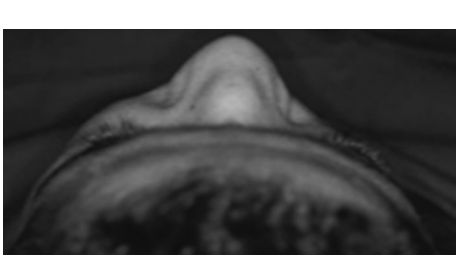

FIGURE 1. Initial clinical assessment.

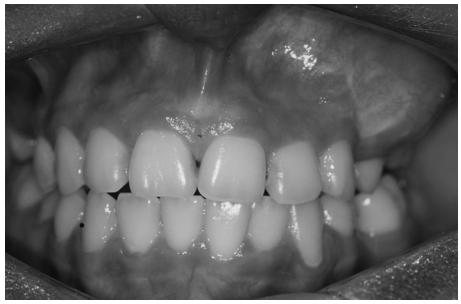

FIGURE 2. Intrabuccal initial clinical assessment.

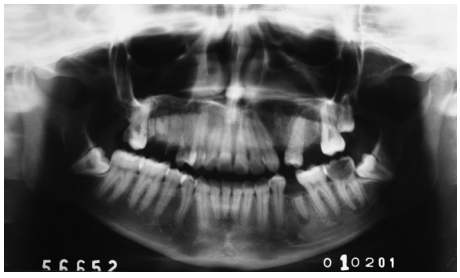

FIGURE 3. Initial panoramic radiograph.

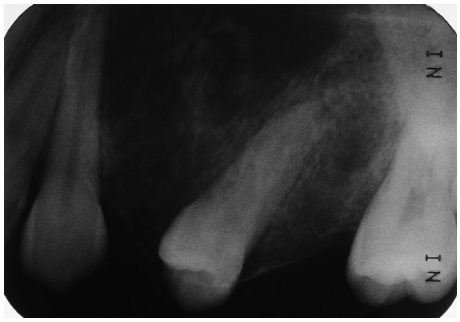

FIGURE 4. Initial periapical radiograph showing a poorly circumscribed unilocular radiolucent lesion, from the left superior lateral incisor to the left superior second molar, involving the left maxillary sinus, with a soap-bubble pattern.

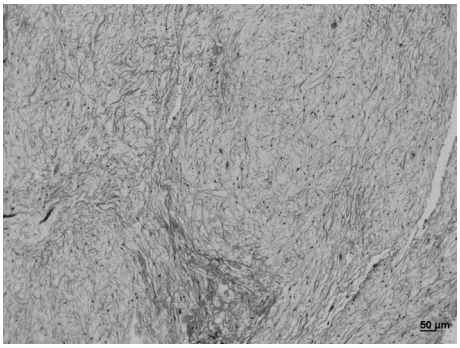

FIGURE 5. At low magnification, myxoid stroma with randomly oriented stellate, spindle-shaped, and round cells with a few fine collagen fibers.

lesion, from the left superior lateral incisor to the left superior second molar. The lesion involved the left maxillary sinus and showed a soap-bubble pattern (Figs. 1–4). An incisional biopsy was performed, after administration of local anesthesia, and the tissue sample (1.7 × 1.5 × 0.3 cm) was sent for histological evaluation. This evaluation showed loosely arranged spindle- and stellate-shaped cells arranged in a myxoid fibrous stroma (Figs. 5 and 6). On the basis of these histological findings, the final diagnosis was odontogenic myxoma.

The removal of the lesion was planned with a conservatory resection and a wide curettage of normal surrounding tissue, with general anesthesia and nasopharyngeal intubation. A mucoperiosteal flap, extending from the left superior canine to the left superior second molar, was reflected. The tumor was totally removed with a cleaver and a mallet, followed by curettage and probing. The left superior second premolar was also removed. After cleaning the surgical site with physiologic NaCl solution, the closure was realized with 3-0 Vicryl sutures.

The patient's progress was followed monthly for the first 6 months and for the next 5 years with yearly panoramic radiograph requests, with no sign of recurrence (Fig. 7).

DISCUSSION

Odontogenic myxoma is a rare odontogenic benign tumor from mesenchymal tissue,[1] with aggressive and invasive behavior leading

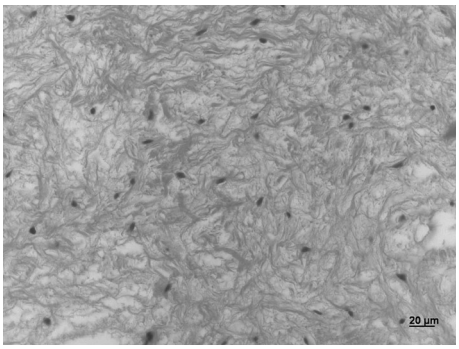

FIGURE 6. The cells dispersed with long, fine, and slightly eosinophilic cytoplasmic processes. The upper part shows collagen fibers.

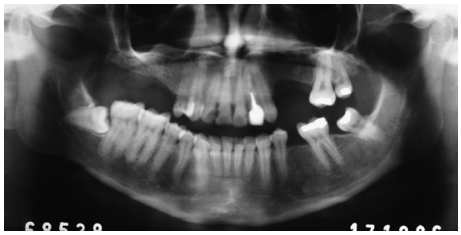

FIGURE 7. Patient's last yearly panoramic radiograph request in 2006, with no sign of recurrence.

to its high recurrence tendency.[4] Odontogenic myxoma presents common characteristics to many other osseous pathologic findings, such that clinical and radiographic examinations are not sufficient for diagnosis, necessitating histologic analysis.

Complete surgical removal of OMs by conservative treatment can be difficult, because, unlike most benign neoplasms, they are not encapsulated, and the myxomatous tissue infiltrates the surrounding bony tissue without its immediate destruction.[9]

Although radical surgery appears to eradicate the tumor completely, it also causes significant functional and aesthetic disturbance.[10] In addition , radical surgery should be carried out in the hope of preventing recurrence, but this is not always successful.[2] An alternative treatment involves enucleation of the lesion with a wide curettage of normal tissue or a generous amount of apparently uninvolved surrounding tissue or even peripheral ostectomy, with the advantage of preserving vital structures and maintaining oral function. This method can also be used to cure recurrent myxomas following conservative treatment.

Long-term follow-up is necessary. The patient should be followed up closely for the first 2 years, because this is the period during which the neoplasm is most likely to recur, although sometimes recurrence may appear much later.[3]

With this successful clinical case, it is possible to encourage more research to confirm what surgical treatment is more effective in each case. Because of the rarity and indefinite etiology of OMs, research of this nature will be limited, but encouraged by others authors.

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Long-Standing Postsurgical Lower Cranial Nerve Palsy Mimicking a Pharyngolaryngeal Submucosal Mass in an Elderly Patient

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Abstract: Changes that occur as a natural part of senescence in the complex action of deglutition predispose to dysphagia and aspiration. This dysfunction is worsened in patients with preexisting anatomic or functional alteration such as in case of a postsurgical lower cranial nerve palsy. We present the case of a 72-year-old woman who underwent surgical resection of a right jugulotympanic tumor 33 years ago, resulting in lower cranial nerve palsy, and came to our attention referring a 4 months' history of progressive dysphagia in which a pharyngolaryngeal submucosal mass was suspected.

Key Words: Dysphagia, lower cranial nerve palsy, elderly

Dysphagia features different causes; it can be due to anatomic or functional alteration of the deglutitory phases that can follow systemic pathologic findings, local diseases, or surgery. It becomes an increasing problem in the elderly, because of changes that occur as a natural part of senescence.[1] In particular, jugular foramen tumor surgery often leads to lower cranial nerve (IX–XII) iatrogenic damage. Clinically, it results in pharyngeal hypesthesia and reduction in palatal activity (cranial nerve IX), vocal cord palsy, hoarseness, and eventually severe dysphagia and aspiration (cranial nerve X), shoulder disability (cranial nerve XI), and paresis of 1 side of the

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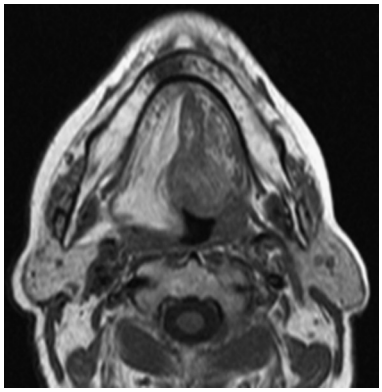

FIGURE 1. Magnetic resonance imaging spin echo T1 sequences in the axial plane.

tongue (cranial nerve XII).[2] We present the case of a 72-year-old woman who in 1976 underwent surgical resection of a right jugulotympanic tumor that resulted in lower cranial nerve palsy and came to our attention referring a 4-month history of progressive dysphagia in which a pharyngolaryngeal submucosal mass was suspected.

CLINICAL REPORT

A 72-year-old woman with a 4 months' history of progressive dysphagia presented to our center with signs suggestive of a laryngeal mass. About 33 years ago, the patient underwent surgical resection of a right jugulotympanic tumor that resulted in lower cranial nerve palsy. No smoking, alcohol abuse, familial history of pharyngolaryngeal cancer was found during history taking. Clinical examination showed right tongue paresis with ipsilateral palatal palsy. At fiberoptic pharyngolaryngoscopy, right vocal cord palsy with ipsilateral base of the tongue hypertrophy was observed; respiratory airway space was sufficient, and asymmetry of the pharyngeal walls was present, as for the presence of a right swelling with no evidence of mucosal alterations. The head and neck magnetic resonance imaging (MRI) scan showed an area with signal intensity on T1 and T2 sequences similar to fat in the right tongue, in relation to fat involvement of the right tongue (Figs. 1–3). On the basis of these findings, a pharyngolaryngeal mass was excluded, and diagnosis of dysphagia due to senescence in a patient with long-standing lower cranial nerve palsy was made.

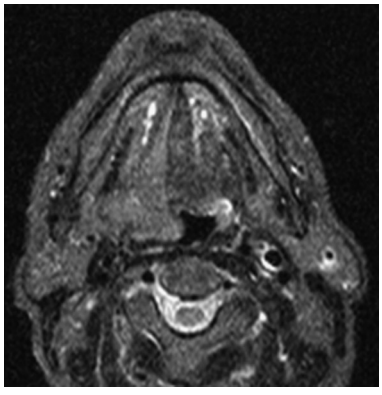

FIGURE 2. Magnetic resonance imaging spin echo T2 fat-suppressed sequences in the axial plane.

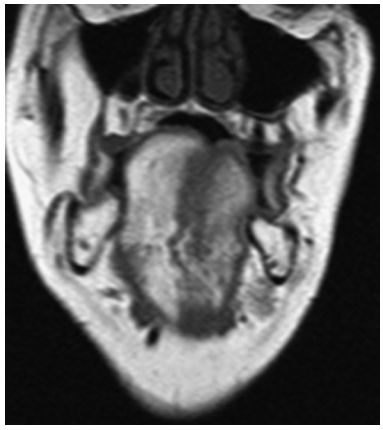

FIGURE 3. Magnetic resonance imaging spin echo T1 sequences in the coronal plane.

DISCUSSION

Some submucosal slow-growing masses can cause pharyngolaryngeal obstruction and consequent dysphagia or dyspnea, in most of elderly patients. Among them, we can consider some differential diagnoses, as illustrated in Table 1. Lipoma is a benign lesion rarely involving the larynx and hypopharynx; because of its slow growth, it usually presents in the elderly with progressive dysphagia, hoarseness, and sometimes dyspnea.[3] About 19% of the amyloidoses involve the head and neck, and the larynx is the most commonly affected area.[4] They are firm lesions that usually occur in the supraglottic region. Paragangliomas usually present in the same way as other laryngeal tumors, with voice changes, airway compromise, and hemoptysis, and can affect patients in all decades of life, with preference for women (3:1).[5] Inflammatory myofibroblastic tumor or inflammatory pseudotumor is extremely rare, and it may easily be misinterpreted as a malignant tumor because of its clinical findings, severe symptoms, and rapid proliferation.[9] Schwannoma (neurilemmoma) of the larynx is a rare benign tumor thought to arise from the internal branch of the superior laryngeal nerve after it penetrates the thyrohyoid membrane, usually involving the false vocal cord and the aryepiglottic fold.[6] Lymphomas primary to the larynx are mainly non-Hodgkin lymphoma and are predominantly located in the supraglottic region; age at occurrence is around the seventh decade.[7] The average patient age at presentation of a laryngeal solitary fibrous tumor is 52 years (29–73 years), with a male preponderance (male-female ratio of 5:1). All the documented cases of laryngeal solitary fibrous tumors were located in the supraglottic region.[8] In our case, all these lesions were excluded by imaging. Other lesions such as cartilaginous tumors or a rhabdomyoma can present with progressive dysphagia, also if rare.[10,11] Surgical management of jugular foramen tumors is complex and difficult; most dangerous complications are the postoperative lower cranial nerves deficits.[2] An isolated cranial nerve IX palsy is minimal, but it usually occurs with cranial nerve X damage. In this case, severe dysphagia with aspiration can be observed.[2,12] An associated cranial nerve XII injury increases functional loss and affects rehabilitation.[12] In effect, imaging shows fat involvement of the right tongue, without evidence suggestive of lesions. On the other side, changes

TABLE 1. Submucosal Pharyngolaryngeal Masses Causing Dyspnea

Differential Diagnosis	Endoscopic Appearance	Imaging
Lipoma[3]	Yellowish submucosal or polypoid mass	Computed tomography (CT) scan: low attenuation and densitometric value less than water MRI: high-signal lesion in T1; fat saturation sequences to confirm diagnosis
Localized amyloidosis[4]	Submucosal mass	CT: no specific findings MRI: prolonged T2 relaxation time and signal-intensity characteristics similar to those of skeletal muscle on T1- and T2-weighted images; peripheral enhancement in the region of the amyloid deposits
Paraganglioma[5]	Vascular-appearing mass	CT: intensive contrast enhancement MRI: "pepper" image on pre-contrast T1
Schwannoma[6]	Submucosal bulge; "pseudofixation" of the ipsilateral cricoarytenoid joint may occur	CT: absence of an infiltrative pattern, a spheric lesion lying medial to the thyroid ala, no cartilage erosion, and slight heterogeneous enhancement with contrast
Lymphoma[7]	Polypoid or smooth submucosal supraglottic mass, nonulcerated, without these characteristics to be specific	CT: solid hypodense mass
Solitary fibrous tumor[8]	Submucosal mass	CT: well-defined isodense mass in the submucosal region MRI: mixed-intensity lesion on the T1- and T2-weighted MR images. A T2-weighted MR image showed a central, round, and low-signal intensity area within the mass; heterogeneous enhancement
Inflammatory myofibroblastic tumor/pseudotumor[9]	Contour bulging, firm, glistening submucosal mass	CT: well-enhancing mass MRI: isointense lesion. T1-weighted images and contrast-enhanced study demonstrating a well-enhanced mass
Chondroma, low-grade chondrosarcoma[10]	Submucosal mass with or without larynx hypomobility	CT: calcified tumor involving 1 or more cartilages

that occur as a natural part of senescence in the complex action of deglutition predispose to dysphagia and aspiration.[1] In fact, elderly patients have reduced lingual pressure reserve necessary to drive pharyngeal swallowing, and the increased connective tissue within the body of the tongue leads to the necessity of multiple tongue movements to hold the bolus posteriorly.[13,14] In addition, a reduction of the pharyngeal reserve may lead to silent aspiration also in healthy patients.[15] The most common causes of dysphagia in the elderly patients are cerebrovascular and neuromuscular diseases, such as Parkinson disease, amyotrophic lateral sclerosis; medication (most frequently causing xerostomia, mental status changes, dyskinesia, gastroesophageal reflux, or esophagitis); and cricopharyngeal dysfunction.[1]

In our patient, neuromuscular and cerebrovascular diseases and the hypothesis of medication-induced dysphagia were excluded. Simple functional and anatomic changes naturally induced by senescence in deglutition process have led to worsening of symptoms due to lower cranial nerve palsy.

CONCLUSIONS

Otolaryngologists should be aware that long-standing lower cranial nerve palsy can lead to anatomic alteration mimicking laryngeal mass; in elderly patients without specific risk factors for pharyngolaryngeal cancer, functional causes of dysphagia primarily have to be considered. Performing skull base surgery, surgeons have to consider that dysfunction such as dysphagia may present later.

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Alternative Transoral Approach for Intranasal Tooth Extraction

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Abstract: Intranasal ectopic eruption is an uncommon condition. Even if an intranasal tooth can be completely asymptomatic, sometimes a variety of nasal signs and symptoms may be associated, ranging from mild nasal congestion to recurrent epistaxis and purulent rhinorrhea. As a consequence, surgical removal is often required. Endoscopic extraction of the intranasal tooth has been reported to present several advantages with respect to traditional surgical approaches and thus recommended as routine treatment in such cases. However, when a tooth is impacted next to the nasal floor, an alternative approach could be needed. We suggest an alternative transoral approach to perform extraction of intranasal teeth, aimed at avoiding excessive bone removal to reach the nasal floor area and preventing the complications related to traditional intraoral buccal or palatal approach. It could represent a reliable alternative to traditional removal in the Oral Surgery Department.

Key Words: Intranasal tooth eruption, supernumerary tooth

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The presence of supernumerary teeth may cause a variety of pathologic effects and usually requires surgical removal. The prevalence of this abnormality ranges from 0.15% to 1% in permanent dentition, with predilection of 2:1 for male sex; in deciduous dentition supernumerary prevalence ranges from 0.3% to 0.66%.[1] The etiology is thought to be related to the dichotomy of the tooth bud or to a local, independent, conditioned hyperactivity of the dental lamina. Furthermore, other factors, such as traumas and hereditary factors such as Gardner syndrome, palatine clefts, and cleidocranial dysplasia can be involved.[1,2]

Supernumerary teeth are often located in the upper arch and in permanent dentition,[1,3] leading to several complications such as prolonged retention of deciduous or permanent teeth, ectopic eruption, malocclusion, cyst development, root resorption of adjacent permanent teeth, aesthetic alterations.[2–4]

In more cases, however, intranasal ectopic eruption can occur, leading to atypical signs and symptoms, including rhinosinusitis, nasal obstruction, and general nasal discomfort.[3]

Recently, endoscopic surgical removal of such intranasal teeth have been reported to be very effective and to cause less morbidity than traditional surgical approaches.[3] The aim of the present work was to report an alternative transoral surgical approach to remove an intranasal ectopic tooth.

CLINICAL REPORT

A 15-year-old boy was referred to our Oral Surgery Department with a history of recurrent rhinitis and intermittent pain in the anterior region of the upper arch. Furthermore, the patient reported 3 episodes of monolateral suppurative rhinorrhea. Previous otolaryngology consultation led to show via rhinoscopic examination a mild inflammation of the nasal floor, without major abnormalities. However, the otolaryngologist asked for a dentistry consultation, because of the persistence of pain in the premaxilla area. Intranasal examination failed to show dental or mucosal pathologic lesions, and orthopantomographic examination showed the presence of a supplementary tooth in 13–23 region, which, on computed tomography, appeared to be a supplementary inverted tooth partially erupted in the nasal floor (Fig. 1).

The patient was then hospitalized, and surgical removal of intranasal tooth under general anesthesia was planned. A novel transoral approach to the nasal floor was performed: A paramarginal incision

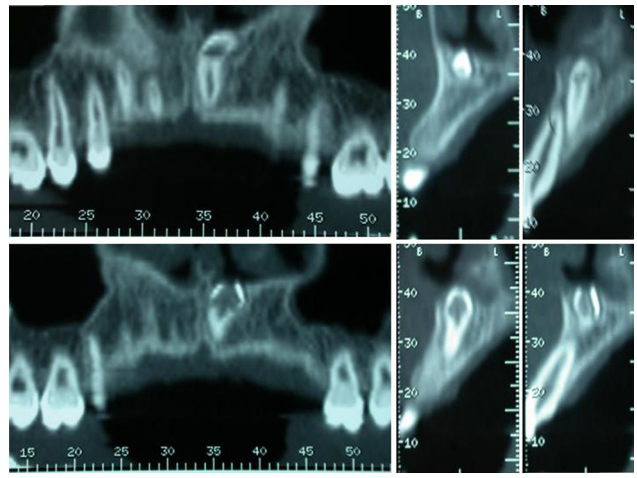

FIGURE 1. Computed tomography of the premaxilla showing a partially erupted tooth in the floor of the left nose.

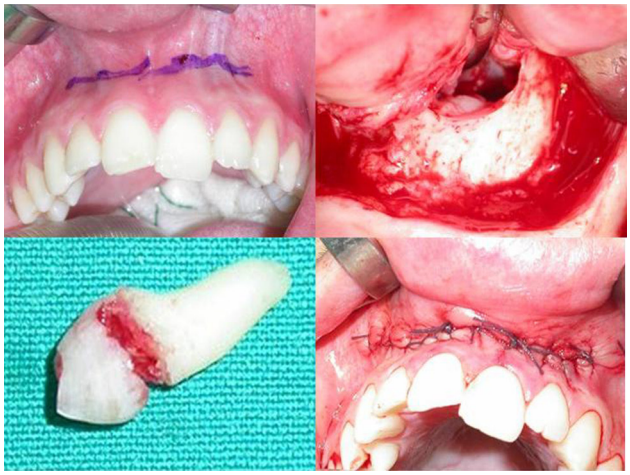

FIGURE 2. Surgical steps of the transoral approach to nasal floor.

in 13–23 region was drawn to visualize the anterior nasal spina and the nasal pear hole. The impacted tooth was visualized by a conservative bone approach and then divided, allowing the dislocation and extraction of the crown (Fig. 2). After the root's removal, curettage and disinfection of surgical pocket with povidone-iodine (Betadine) were performed.

The patient was followed up a week later for suture removal and at 1 to 2 and 3 months after surgery. After 12 months, the patient was free of symptoms and signs of mucosal involvement, with x-ray and computed tomography scan showing nasal bone healing of the involved region.

DISCUSSION

Supernumerary teeth are well known to cause several complications such as failure of eruption, displacement of the adjacent permanent teeth (especially permanent incisors),[2] crowding caused by supplementary lateral incisors,[2] dentigerous cyst formation,[2] permanent teeth's root resorption.[2] Treatment depends on the type and position of supernumerary teeth, on the potential complications,[2] on the patient age, and on the stage of permanent eruption.[1,4]

TABLE 1. Indications for Monitoring With and Without Supernumerary Tooth Removal

Indications for supernumerary teeth removal
• Central incisor eruption has been delayed or inhibited
• Altered eruption or displacement of central incisors is evident
• There is dentigerous cyst development
• A mechanical complication is present (diastema, rotation, ulceration, root resorption of permanent teeth)
• Neuralgic pain
• Infective complications are present (pericoronitis)
• Active orthodontic alignment of an incisor in close proximity to the supernumerary is envisaged
• Supernumerary presence would compromise secondary alveolar bone grafting in cleft lip and palate patients
• The tooth is present in bone designed for implant placement
• Spontaneous eruption of the supernumerary has occurred
Indications for monitoring without supernumerary removal
• Satisfactory eruption of related teeth has occurred
• No active orthodontic treatment is envisaged
• There is no associated pathologic finding
• Removal would prejudice the vitality of the related teeth

Best time for supernumerary teeth removal has been reported to be at 8 to 9 years of age, because patient management is easier, and the type of anesthesia is less invasive.[3] It is advisable, however, to postpone surgical removal until the root formation of the permanent adjacent teeth.[4]

Indications for surgical removal of supernumerary teeth were differently discussed[2] and are resumed in Table 1. Currently, there is agreement on the need of extraction in case of complications.[2,4] However, when intranasal ectopic eruption occurs, major nasal signs and symptoms can develop, including unilateral obstruction, localized ulceration, nasal oral fistula, pain and epistaxis, external deviation of the nose, cacosmia, septal perforation, and rhinolithiasis,[5,6] which almost always require surgical removal.

Smith et al[7] found in literature 41 cases of intranasal supernumerary eruption in subjects with age ranging from 3 to 62 years, and Lee[5] reported that intranasal teeth are frequently supernumerary, but they may also be permanent element.

Intranasal teeth eruption is mainly related to palatine cleft and displacement of dental bud because of traumas, cysts, or odontogenic or rhinogenic infections.[8] These elements are often located on the nasal floor, approximately between the anterior and posterior portion of naris.[5] Also in our case, ectopic supernumerary element was partially erupted in the nasal floor. This condition had caused during the past 3 years several episodes of monolateral suppurative rhinorrhea and persistent pain in the anterior region of the upper arch.

With regard to surgical approach, intranasal tooth extraction is often carried out by a transpalatal or transnasal approach, which are burdened by relative high morbidity and postsurgical complications.[3] Since 1980, endoscopic nasal surgery offered many advantages, such as good illumination, clear visualization, avoidance of injury to surrounding mucosa, and precise dissection with preservation of neighboring structures.[3,5] However, when the impacted tooth is next to the nasal floor and partially erupted, an endoscopic approach is not possible. In such cases, a minimally invasive intraoral surgery should be performed, avoiding excessive palatal or buccal bone removal to reach the nasal floor area and preventing the complications related to traditional buccal or palatal approach (potential nasopalatine bundle damage, intraoperative bleeding, and hypesthesia of the anterior one third of the palate).

Our transoral surgical approach to the nasal floor, on the contrary, leads to a less invasive procedure, with clear visualization of supernumerary tooth, less bone removal from the buccal or palatal plate, and no risk of nasopalatine bundle damage.

It can represent a reliable alternative surgical approach to reach the nasal floor in these types of tooth impaction in the Oral Surgery Department. However, in our experience, extraoral approaches are suitable also in other pathologic findings regarding inclusive teeth, such as cleidocranial dysostosis.[9]

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Diagnosis and Surgical Treatment of a Nasolabial Cyst

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Abstract: Nasolabial cysts are uncommonly diagnosed nonodontogenic soft tissue lesions located close to the nasal alar region of the face, presenting as extraosseous swelling in the region of the nasolabial fold. Nasolabial cysts are likely to remain undetected unless and until they become infected or are associated with facial deformity. Histologically, it is lined with nonkeratinized squamous epithelium or, more frequently, with respiratory-type cylindrical epithelium with goblet cells. The aim of this article was to present and discuss the surgical management of a case of nasolabial cyst and to briefly review the literature.

Key Words: Nasolabial cyst, fissural cysts, respiratory-type columnar epithelium, goblet cell

Nasolabial cysts are uncommonly diagnosed nonodontogenic soft tissue lesions located close to the nasal alar region of the face, presenting as extraosseous swelling in the region of the nasolabial fold.[1–5] They are usually unilateral, and they are often confused with other fissural or odontogenic cysts.[6] Nasolabial cysts are likely to remain undetected unless and until they become infected or are associated with facial deformity. Therefore, they may be more frequent than previously thought.[7]

They characteristically present as a swelling of the upper lip adjacent to the nasal alae of the nose, producing a very typical facial asymmetry.[6,8] Patients may complain of pain if the cyst becomes infected.[6,8]

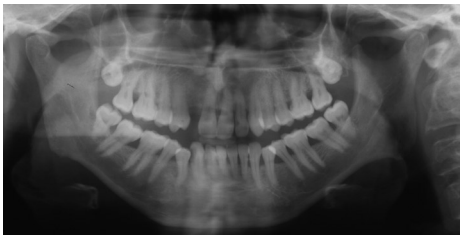

FIGURE 1. Panoramic radiograph showing bony rarefaction and deformity of the lateral and anterior edges of the nasal floor in the periapical region of the first and second left upper maxillary incisors and upper left canine.

They seem to be developmental, rather than inflammatory, in origin.[9] However, several theories on the pathogenesis of nasolabial cyst have been advanced, and their etiology is still debated.[10] The 2 more accredited hypotheses are that they either are fissural cysts arising from the epithelial rests in fusion lines of the globular, lateral, nasal, and maxillary processes or originate from remnants of the embryonic nasolacrimal ducts.[1,2] Histologically, it is lined with nonkeratinized squamous epithelium or, more frequently, with respiratory-type cylindrical epithelium with goblet cells.[5,8]

The aim of this article was to present and discuss the surgical management of a case of nasolabial cyst and to briefly review the literature.

CLINICAL REPORT

A 47-year-old man was referred to the Division of Maxillofacial Surgery of the University of Turin, Turin, Italy, for evaluation and treatment of a unilateral swelling of unknown etiology in the left nasolabial region. He had noticed a slight swelling in that area several years before, and since then he referred a couple episodes of painful,

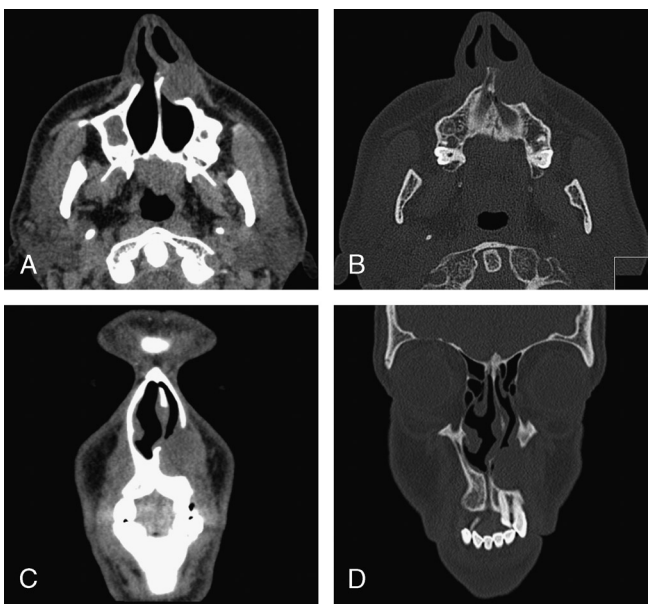

FIGURE 2. Axial (A, B) and coronal (C, D) CT scans revealing a well-demarcated, low-density cystic lesion lateral-anterior to the piriform aperture, with remodeling of the underlying anterior maxilla.

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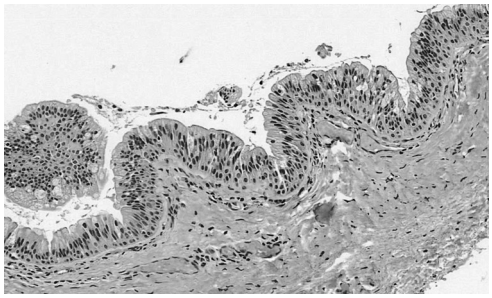

FIGURE 3. Epithelial lining of the cyst composed of pseudostratified columnar cells with scattered goblet cells (hematoxylin-eosin stain, original magnification ×10).

gradual increase in swelling, resolved by antibiotic therapy. His medical history was unremarkable.

Physical examination revealed a painless, soft, round, and nontender swelling beneath the left alar base. Panoramic radiograph showed bony rarefaction and deformity of the lateral and anterior edges of the nasal floor in the periapical region of the first and second left upper maxillary incisors and upper left canine (Fig. 1). The computed tomography (CT) scan revealed a nonodontogenic cyst in the nasolabial area (Fig. 2).

The lesion was removed under general anesthesia. An intraoral incision on the upper alveolar ridge was performed, and after the elevation of a mucoperiosteal flap, the lesion was easily removed from the bone. It presented tight adhesions with the overlying nasal mucosa, but it was peeled intact from it: During this operation, the mucosa of the nasal floor was perforated and consequently repaired by primarily absorbable suture.

The lesion, containing yellowish serous fluid, was sent for histopathologic examination, which revealed that the cyst walls were lined by pseudostratified columnar epithelium with scattered goblet cells (Fig. 3). Therefore, the lesion was diagnosed as nasolabial cyst.

Postoperative course was uneventful. Postoperative follow-up at 6 months showed good healing without evidence of recurrence.

DISCUSSION

Nasolabial cysts are uncommon developmental entities that account for 0.7% of all maxillofacial cysts.[8,11] They are usually diagnosed in the fourth or fifth decade of life with a noticeable predilection for women, with female-to-male ratio of 3:1.[1,4,6,7,9,11] Most nasolabial cysts are unilateral; only 10% are bilateral.[8,9]

The etiopathogenesis of this lesion remains controversial. Two main theories, which may not be mutually exclusive, have been proposed. The first hypothesis suggests that nasolabial cysts develop from embryonic nasal respiratory epithelium retained in the mesenchyme after fusion of the maxillary, medial, and lateral nasal processes.[2,3,6,7,9,12] Therefore, nasolabial cyst would be considered as a fissural cyst.[6] However, the lack of evidence for embryonic epithelial entrapment in this region would lower the reliability of this hypothesis.[4,10]

Instead, according to the second (more accepted) theory, persisting epithelial remnants of the embryonic nasolacrimal duct located between the maxillary and nasal processes would originate nasolabial cysts.[2,3,7,9,10,12] The nasolacrimal duct first develops from the nasolacrimal groove. Because of development of the maxillary prominence in nasal direction and expansion in frontal direction, this groove deepens and finally is transformed into a canal following fusion of the frontal and maxillary prominences.[13] Epithelial remnants of the nasolacrimal groove might therefore persist just deep to the junction of the nasal ala with the lip.[12] This hypothesis is supported by the fact that cystic walls of the nasolabial cyst are lined by the same epithelium of the nasolacrimal duct: the pseudostratified columnar epithelium.[3,4] Furthermore, it also could be confirmed by the location of the nasolabial cyst, which is exclusively at the floor of the nose anteriorly to the opening of the nasolacrimal duct.[8,10]

The clinical features of nasolabial cyst are typical. In fact, this lesion presents as a slow-growing, fluctuant, painless, soft tissue swelling between the upper lip and nasal aperture, producing facial asymmetry with elevation of the nasal ala and inferior turbinate and obliteration of the nasolabial fold.[1,4,6–12] Many nasolabial cysts probably remain undetected for several years because of its slow growth until they become infected and painful or are associated with facial asymmetry.[4] If infected, the cyst may rupture spontaneously and may spontaneously drain into the oral cavity or nose, which could explain the variation in size encountered by some patients, as ours.[8] When it is infected, a cyst enlarges rapidly, becomes tender, and may be mistaken for an abscess of the nasal floor.[7] Furthermore, the cysts may determine pressure erosion of the underlying bone and may erode the maxillary alveolus, as in our patient.[7,12]

Finally, a nasolabial cyst could be erroneously considered as a dental or periodontal abscess, but odontogenic or nonodontogenic cysts (such as nasopalatine duct and globulomaxillary cysts) and soft tissue masses including benign tumors should also be kept in mind for the differential diagnosis,[3,12] which can be perfected, thanks to imaging examinations.

As for imaging, conventional radiographs may show bony rarefaction and deformity of the lateral and anterior edges of the nasal floor as in our case, although these findings may not be present in all patients, nor are they specific for the disease.[7] Computed tomography scans reveal a well-demarcated, low-density cystic lesion lateral-anterior to the piriform aperture, without signs of bony invasion but with the possibility of remodeling of the underlying anterior maxilla.[1,2,7,11] Nasolabial cysts appear on magnetic resonance imaging as homogeneous intermediate-intensity T1 signals and homogeneous hyperintensity T2 signals.[1,2,7] Because of its lower cost, CT can be preferred to magnetic resonance imaging in the assessment of a suspected nasolabial cyst.

Anyway, the conclusive diagnosis of the lesion is obtained by the correlation of clinical, radiologic, and histopathologic examinations. Microscopically, this lesion is lined by nonciliated columnar epithelium associated with basal cells and mucous-producing cells (goblet cells),[6] although occasionally a stratified squamous epithelium can be encountered.[7,12] Intraoperatively, a nasolabial cyst appears to be made up of a thick fibrous capsule containing mucoid or yellow serous fluid.

The classic and preferred treatment of nasolabial cysts is surgical excision via a sublabial incision. Our patient preferred to undergo intervention under general anesthesia, although a local anesthesia was proposed. The cyst wall should be carefully dissected from the surrounding tissues and removed, paying attention to the tight attachment of the cyst with the nasal mucosa: this can lead to perforation of the floor of the nose that anyway just requires nasal repair of the defect with primarily absorbable suture, as in our case.[3]

Transnasal endoscopic marsupialization seems to be a simple and effective alternative,[8] whereas other proposed options (simple aspiration, injections with a sclerosing agent, destruction by cautery) are associated with high recurrence rates.[7]

CONCLUSIONS

Nasolabial cyst is an uncommon benign cystic lesion that may be encountered incidentally or in conjunction with a soft tissue mass near the nasal alae. Computed tomography scans are the diagnostic tool of choice. Histopathology reveals nonciliated columnar epithelium and

mucus-producing cells. The treatment of choice is surgical enucleation, which presents low recurrence rates. Finally, nasolabial cyst should be considered in the differential diagnosis of soft tissue vestibular swelling in nasal vestibule, nasal base, and sublabial areas.

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Witch Nose: An Embarassing Metaphor for Nasal Tip Dermoid Cysts

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Abstract: Nasal dermoid cysts are one of the most frequent congenital pathologic lesions of the nasal area. These lesions may have intracranial extensions without any clinical evidence, which can be explained by the pathophysiologic development of the lesion, thought to be related to a delay or pause during the separation period of the dura and skin during embryogenesis. This factor is independent from the location and size of the lesion and may lead surgeons to misdiagnoses or inadequate treatments.

In this article, 2 cases of nasal dermoid cysts localized at the tip of the nose are presented. Although these lesions can be seen anywhere from the root to the columella, nasal tip location is rare in previously published series. In the cases presented in this study, both patients are school-aged and are faced with social and psychologic problems, as their friends call them "witch nose," in reference to the lesion at the tip of the nose. Vertical tip incision for complete excision of these lesions was performed after accurate radiologic imaging of the brain and possible intracranial extension. No complications or recurrence was seen in either patient.

Although the treatment of dermoid cysts is surgical, operative planning should be made after complete physical and radiologic examinations. As the lesion may lead to psychologic and social problems because of its appearance, especially in children, therapy should not be delayed.

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Key Words: Nasal tip, dermoid cyst, congenital anomaly

Various congenital pathologic lesions may be localized in the nasal region and lead to serious complications when left untreated or inadequately excised. Congenital nasal masses are present in 1 of every 20,000 to 40,000 live births.[1,2] The differential diagnoses of nasal masses in children include many diseases ranging from infectious lesions (subcutaneous abscess, Pott tumor) and facial trauma sequels to benign (angioma, neurofibroma, odontogenic tumor) and malignant (neuroblastoma, teratoma) neoplasms. Of these, gliomas, encephaloceles, and nasal dermoid sinus cysts (NDSCs) are the most frequent.[3–5] These lesions may present anywhere in the nose from the root to the columella.

We present 2 nasal dermoid cases, both of which are located at the tip of the nose. Because of this rare location, both children were called "witch" by their friends. The lively imagination of childhood unfortunately resulted in the avoidance of social activities and development of introverted behavior due to the teasing.

CLINICAL REPORT

Patient 1

A 4-year-old girl presented with a firm, noncompressible, and nonpulsating midline mass at the tip of the nose. There was no

FIGURE 1. Lateral view preoperatively and 8 months postoperatively. The 4-year-old girl does not resemble a ''witch'' anymore.

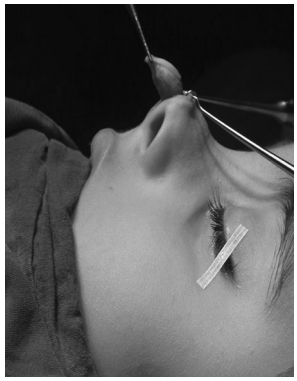

FIGURE 2. The cyst was excised through a vertical incision at the tip of the nose.

evidence of trauma or infection in the history. A 4-year-old girl presented with a firm, noncompressible, and nonpulsating midline mass at the tip of the nose. There was no evidence of trauma or infection in the history. This 1 × 1.5-cm mass was present at birth and increased in size with the age of the child. Although the family was distressed by the nature of the lesion, the child complained of being isolated because of the appearance of her nose and being called "witch" by her friends (Fig. 1). Because the direct radiographs and computed tomography imaging showed no sign of bony defect or intracranial extension, we planned to reach the lesion with a vertical incision at the tip and completely excise it. In the course of the operation, the mass was identified well surrounded under the skin and followed upward. The cyst extended cranially and posteriorly between the left upper lateral cartilage and septum. The lesion was removed completely with a blind-ending tract (Fig. 2). The skin was closed in 2 layers.

Patient 2

A 6-year-old boy was referred to our clinic with a nontransilluminating mass at the tip of the nose. The covering skin was thin and telangiectasic. No dimple or pits were observed on the nose. The lesion had been present since birth and was gradually growing. The child had started school that same year and had been nicknamed "witch nose" by the class (Fig. 3). This embarrassing name resulted in a regression of his social skills and school performance, and he was reluctant to go to school. The imaging methods revealed no sign of deeper extension. The cyst was restricted at the tip, and the lesion was excised easily through a 1-cm median vertical incision at the tip.

The pathologic examinations of both specimens confirmed the preoperative dermoid cyst diagnoses (Fig. 4). No complication or recurrence was seen in either patient in the follow-up period of 2 years in the first case and 8 months in the second case.

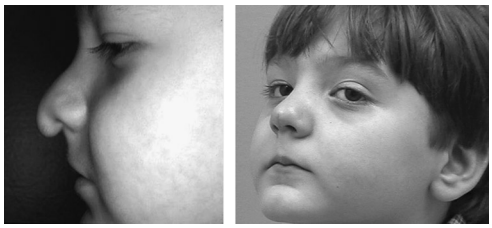

FIGURE 3. Lateral view preoperatively and 2 years postoperatively. The mass at the tip of the nose was removed without a conspicuous scar.

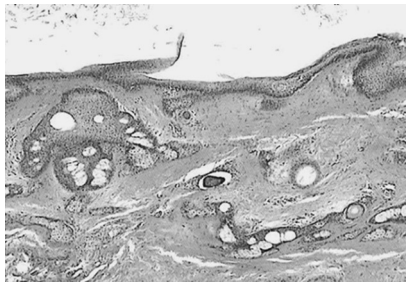

FIGURE 4. Typical microscopic view of a dermoid cyst under ×40 magnification and hematoxylin-eosin staining. The cyst wall is lined with multilayered squamous epithelia and contains numerous pilosebaceous units.

DISCUSSION

Congenital NDSCs are well-known causes of dorsal nasal midline masses and pits. They account for 11% to 12% of head and neck dermoids and 1% to 2% of overall dermoids of the body.[3,6,7] Of these, lesions at the tip of the nose are rare cases. Rahbar et al[8] reported only 3 cases of nasal tip dermoid cysts in their series of 42 cases. In the series of Pensler et al,[9] only 3 of 32 lesions were located at the tip.

These congenital lesions may be noticeable at birth or stay unrecognized until late childhood or adulthood. They can occur anywhere in the nose from the root to the columella and present as small draining pits, infected sinuses, or distinct subcutaneous masses.[3,8,10] Each of these forms may have intracranial extensions without any clinical evidence, which can lead to misdiagnoses and inadequate treatments.[11,12]

Although most of the lesions are sporadic and isolated, some familial cases and associated malformations have been reported. Concomitant anomalies may include an absent third ventricle, spinal anomalies, tracheoesophageal fistula, preauricular sinuses, and any facial clefts. Wardinsky et al[13] reported a 41% incidence of craniofacial malformations associated with NDSCs. In the series of Ghestem et al,[14] the incidence of malformative anomalies was 26%. Posnick et al[15] noted a 14% associated craniofacial abnormality. Our cases did not have any additional anomalies.

Nasal dermoid sinus cysts are believed to occur as a result of an abnormality during embryogenesis, and much has been written about their pathogenesis. The most widely known theories were proposed by Bland-Sutton in 1893 (superficial sequestration theory), Grunwald in 1910, and Littlewood[16] in 1961 (trilaminar theory).[7] Grunwald's theory was later named "prenasal theory" by Pratt[17] and "cranial theory" by Bradley.[18] Of these, Grunwald's study is currently the most popular and generally accepted one.

According to Grunwald's theory, there is a small fontanelle (fonticulus nasofrontalis) between the developing frontal and nasal bones. Between the nasal bones and cartilage capsule, prenasal space extends from the cranial base to the nasal tip. Early in embryogenesis, dura projects from the foramen cecum and comes into direct contact with the skin through the prenasal space. Normally, this dural diverticulum regresses and closes during development. But if the dura fails to separate from the skin during recession because of an ossification delay or pause, dermal attachments follow this course, and a pathway that is lined by ectoderm is formed. This results in an incomplete closure of the cranial sutures and

herniation of the cranial contents. Depending on the patency of the diverticulum and its contents, the resultant lesion can be a dermal sinus, dermoid cyst, glioma, or encephalocele.[1,2,5,9,11,17]

Clinically, encephaloceles are herniations of cranial contents through bony defects in the skull containing meninges with or without brain or any other central nervous system (CNS) tissue.[19] They are classified as anterior or posterior according to their location. Anterior sincipital encephaloceles may occur around the root of the nose and produce hypertelorism. These lesions pulsate and expand with compression on the jugular veins (positive Furstenberg test).[2,20] Some lesions may protrude through the nostrils, and cerebrospinal fluid leakage may occur.

Gliomas develop in a similar manner as encephaloceles, but they do not have meningeal continuity. Fifteen percent to 20% have a fibrous band that connects them to the intracranial space. Seventy percent of nasal gliomas appear as external masses, and the covering skin tends to be telangiectatic.[5] Furstenberg test is always negative. Because of the closure of the intracranial extension, these lesions are rarely associated with cerebrospinal fluid leakage.

Dermoid cysts are derived from ectoderm and mesoderm; they are lined with squamous epithelia and contain adnexial structures such as hair follicles, sweat glands, sebaceous glands, lymphoid tissue, and cartilage.[15] Forty-five percent have intracranial connections.[11,14] Furstenberg test is negative. Lesions may present as a soft, nontender swelling that is usually mobile but attached to surrounding structures. A dimple on the nose with sebaceous fluid discharge with or without protruding hair may be pathognomonic for nasal dermoid or dermal sinus. This sinus may reach to the CNS through the embryologic remnant and bony defect. Usual sites of intracranial entrance are the cribriform plate, foramen cecum, and anterior crista galli.[14]

Although a bifid crista galli and widened foramen cecum are the signs of intracranial extensions in computed tomography, Pensler et al[9] reported false-positive postoperative results after radiologic examinations of 32 patients. Rahbar et al[8] showed 3 false-positive and 1 false-negative radiologic findings in a series of 35 patients.

The treatment of nasal dermoids is proposed to be surgical. Complete excision of the lesion is required, because any epithelial remnant may lead to recurrence. Other modalities such as curettage, drainage, aspiration, or part excision result in 30% to 100% recurrence.[2,3,5,7–12,15,18,21,22]

Kelly et al[21] proposed 4 principles of surgical approach to nasal dermoids: (1) lesion must be completely excised; (2) contrast dye must be weighed against potential liabilities; (3) surgery must be performed as early as possible; and (4) careful planning is necessary.

Pollock[22] proposed that any surgical approach for nasal dermoids should fulfill these criteria: (1) provide an excellent access to a midline cyst and allow osteotomies in a complex case; (2) allow adequate exposure to the deep nasal structures and skull base; (3) provide adequate exposure for the reconstruction of the dorsum and any deformed portion; and (4) result in an acceptable scar.

Many authors have recommended various surgical incisions, such as midline vertical, transverse, lateral rhinotomy, inverted U, external rhinoplasty, and endoscopic processes.[7,8,22–25] Most of them advocate combining the procedure with an intracranial approach in patients with radiologically proven intracranial extension or when such a situation is recognized intraoperatively.

Bilkay et al[23] used an open rhinoplasty approach in 4 of 8 patients and reported excellent exposure of the dorsum and complete excision of the lesions without any visible scar. They saw no growth disturbance with age. They also used midline vertical incisions in 3 patients. Rahbar et al[8] preferred a paracanthal incision for a lesion in the glabellar area that might not be completely excised via an external rhinoplasty incision. Weiss et al[24] proposed transnasal endoscopic excision of the lesion when minimal or no cutaneous involvement was found.

Rohrich et al[7] preferred the open rhinoplasty approach because of (1) ease of exposure, (2) better aesthetic result, (3) adequate exposure of the dorsum, (4) controlled osteotomies, (5) easy reconstruction, and (6) wide exposure of the upper cartilages and septum.

We used nasal tip vertical incisions in our patients, as they presented with relatively small lesions located at the tip. Because no intracranial extensions could be seen in the preoperative imaging, we did not require additional approaches. In patient 1, we easily reached the nasal septum and completely excised the tract of the lesion through this small excision. During the follow-up period, no widening of the scar or bad aesthetic outcome was observed. Therefore, we propose the use of small vertical nasal tip incisions in cases that present with nasal columellar or tip lesions without intracranial or deep nasal extensions.

In conclusion, although the nasal region may be the site of various congenital lesions, nasal tip dermoids are rare. In many cases, dermoids cysts are restricted to where the cyst primarily appeared, but the lesion may have an extension to the CNS through cranial bony defects. This point must always be considered, and the lesion must never be underestimated. Operative planning should be made after accurate, complete physical and radiologic examinations. Therapy should not be delayed, as the lesion may become infected or cause psychologic and social problems, especially in children.

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Platelet-Rich Fibrin and Piezoelectric Surgery: A Safe Technique for the Prevention of Periodontal Complications in Third Molar Surgery

Emanuele Ruga, DDS, Cesare Gallesio, MD, DDS, Paolo Boffano, MD

Purpose: The surgical removal of impacted mesioangular mandibular third molars (3Ms) may produce trauma to the soft tissues and bony structures. In particular, healing of postextractive socket may cause periodontal defects at the distal root of the second molar. The aim of the present prospective study was to assess the outcomes of a surgical protocol to remove 3Ms including the use of ultrasound bone surgery devices and platelet-rich fibrin (PRF) as a grafting material.

Patients and Methods: Twenty-eight impacted mandibular 3Ms were removed. Fourteen 3Ms were removed by piezoelectric osteotomy and PRF application. Instead, the 14 3Ms of the control group were removed by piezoelectric osteotomy but without PRF application. Probing depth in the distal position for all the mandibular second molars was registered before and 6 months after surgery.

Results: No complications were encountered. In the study group, alveolar socket fulfillment was rated as sufficient in 4 cases and adequate in the remaining 10 cases, whereas in the control group, it was rated as insufficient in 3 cases, sufficient in 4 cases, and adequate in 7 cases. Mean preoperative periodontal probing in the control group was 3.78 mm, whereas in the study group, it was 3.93 mm.

Six months after surgery, mean periodontal probing was 3.28 mm in the control group and 3.07 mm in the study group.

Conclusions: Combined action of PRF and piezoelectric surgery can be considered a safe and fine technique for third molar surgery and alveolar socket healing.

Key Words: Platelet-rich fibrin, piezoelectric surgery, periodontal defects, third molars, surgical extraction, postextraction sockets

The surgical removal of impacted mandibular third molars may produce a significant degree of trauma to the soft tissues and bony structures of the oral cavity, potentially resulting in a significant inflammatory reaction, patient pain, and discomfort.[1]

Moreover, healing of postextractive socket requires a large amount of time and may cause periodontal defects at the distal root of the second molar. This may determine difficulties in hygiene maintenance by the patient and potentially compromises periodontal health of second molar.[2,3]

Several authors have proposed using piezoelectric surgical devices to perform osteotomies and ostectomies as an alternative to rotary instruments.[1,4–16] The use of ultrasonic instruments would allow to obtain a more precise and conservative cut action, minimizing trauma on hard tissues and facilitating healing process.

The use of graft materials and platelet concentrate has been proposed to improve socket hard and soft tissue regeneration stimulating and accelerating soft tissue and bone healing and to prevent periodontal defects.[3,17,18] The use of graft materials or platelet concentrates is reported to reduce the distal probing depth and attachment level of the lower second molar.[3,17] There is actually a very large number of proposed platelet concentrates.[19–21]

Platelet-rich fibrin (PRF) is a new platelet concentrate concept (without addition of thrombin during preparation) that may be used to enhance bone generation in postextraction sockets and residual cyst cavities and to promote wound epithelialization.[22–26] Platelet-rich fibrin derives from a natural and slow, progressive polymerization occurring during centrifugation.[27] It is a second-generation

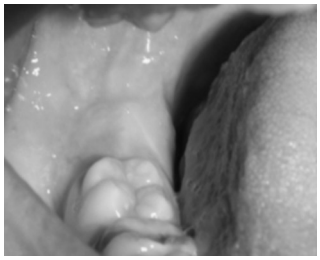

FIGURE 1. Preoperative image of the right retromolar trigon region.

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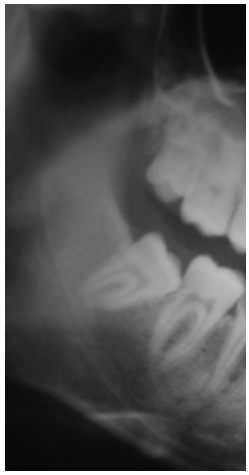

FIGURE 2. Detail of preoperative panoramic radiograph showing the mucosal mesioangular impaction of the right mandibular third molar.

immune and platelet concentrate collecting on a single fibrin membrane all the constituents of a blood sample favorable to healing and immunity: in particular, it is a fibrin matrix in which growth factors (PDGF-ββ, TGF-β1, VEGF, and insulin-like growth factor 1), leukocytic cells, and their cytokines (interleukin 1β [IL-1β], IL-6, IL-4, and tumor necrosis factor α) are enmeshed.[22–26]

The aim of the present prospective study was to investigate and assess the outcomes of a surgical protocol to remove impacted mandibular third molars including the use of ultrasound bone surgery devices and PRF as a grafting material.

MATERIALS AND METHODS

Between January 2009 and December 2009, 14 healthy patients (8 females and 6 males) underwent surgical extraction of 28 bony or mucosal impacted mandibular mesioangular third molars due to prophylactic reasons. Patients' age ranged from 17 to 42 years (mean age, 29.5 y).

The study group included 14 left or right mandibular third molars: they were removed by piezoelectric ostectomy, and postextraction sockets were filled with PRF. Instead, the remaining 14 contralateral third molars were extracted without PRF application.

The study was conducted in accordance with the standards of the Declaration of Helsinki. The patients were informed about the aim and design of the study, and written consent was obtained.

Patients with immunologic diseases, unstable diabetes mellitus, ongoing chemotherapy or radiotherapy, uncontrolled periodontal

FIGURE 4. Platelet-rich fibrin membranes are created in the PRF box.

disease, a smoking habit of 20 cigarettes or more per day, or a history of drug abuse had been excluded. The inclusion criteria had been a blood concentration of thrombocytes within the reference range and the presence of mandibular bilateral mesioangular bony or mucosal impacted third molars.

Time necessary for the tooth extraction and complications was registered. Preoperative panoramic radiographs were obtained. Preoperative measurement of the probing depth was made using a Marquis periodontal probe in the distal position for all the mandibular second molars, and the values of probing depth were registered.

A standardized surgical procedure was performed in all cases. The mandibular third molars of which the extraction sockets would receive PRF treatment were selected randomly and were operated on first. The contralateral third molars of each patient were extracted 3 months later the same surgical procedure, but without PRF application in the postextraction sockets (Figs. 1 and 2).

In all cases, local nerve block anesthesia of inferior dental, lingual, and buccal nerves was induced with two 1.8-mL capsules of 2% mepivacaine containing 1:100,000 adrenaline (Carboplyina; Dentsply Italia, Rome, Italy). A buccal triangular mucoperiosteal flap that was limited distally to the second molar was raised (Szmyd flap).[17] The osteotomy necessary to remove a sufficient amount of bone to easily reach impacted third molar and to facilitate its luxation was performed using an ST 70 sword tip of an OSADA ENAC 10W piezoelectric unit (Osada Electric Co, Ltd, Tokyo, Japan). Then, the third molar was luxated and removed. Alveolar socket was irrigated with an isotonic NaCl 0.9% solution, and it was finally washed with a clindamycin solution (Fig. 3).

In the study group, during surgery, as described by Toffler et al,[28] 18 to 54 mL (2–6 tubes) of whole blood was drawn into 9-mL glass-coated plastic tubes without anticoagulant and immediately centrifuged (PRF Process, Nice, France) at 2700 rpm for 12 minutes. The result was a fibrin clot located in the middle of a mass of acellular plasma, with a maximum number of platelets and more than half

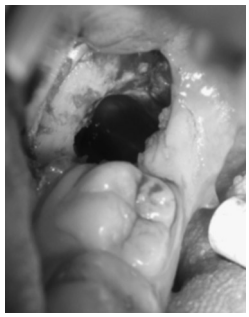

FIGURE 3. Intraoperative image of the postextraction alveolar socket.

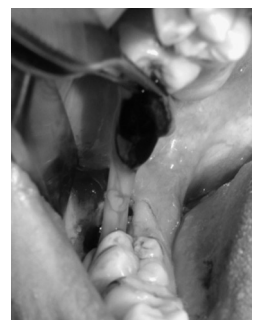

FIGURE 5. Platelet-rich fibrin membranes are positioned in the postextraction alveolar socket.

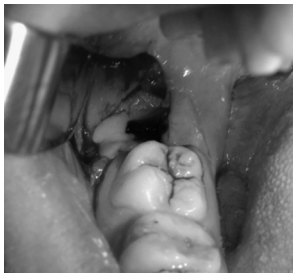

FIGURE 6. Appearance of the alveolar socket that was filled with PRF membranes.

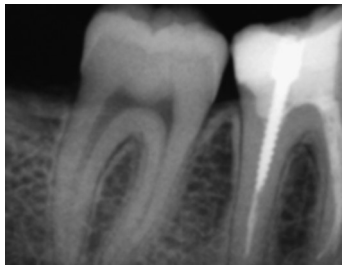

FIGURE 8. Periapical radiograph at 6 months confirms an adequate bone regeneration.

of the leukocytes caught in the mesh of fibrin. The clot was removed from the tube, and the attached red blood cells were shaved off and discarded. The clots were then placed on a grid in the PRF box (PRF Process) and compressed by a cover (masher) to create a fibrin membrane (Fig. 4). Alveolar postextraction sockets of study group cases were finally filled with PRF membranes (Figs. 5 and 6).

Primary closure of the flap was performed using a 3-0 silk suture. All patients were prescribed amoxicillin-clavulanate 1 g and ibuprofen 400 mg twice daily for 5 days and 0.12% chlorhexidine gluconate mouth rinse twice daily for 2 weeks. Sutures were removed 7 days after intervention. In this occasion, pain was assessed by a visual analog scale (VAS) score of 10 and reported to clinician (Fig. 7). On the VAS, the leftmost end represented absence of pain (score, 0) and the rightmost end indicated the most severe pain (score, 10).

Patients were monitored for more than 6 months after surgery. Six months after surgery, intraoral radiographs were obtained with a paralleling technique using the Rinn system (Fig. 8). Furthermore, periodontal probing of distal area of second molars and alveolar socket fulfillment were registered. Excessive pain, edema, cheek swelling, or any local or general complications were registered.

RESULTS

No significant difference in the surgical trauma associated with surgical extraction were found between the study and control groups. Surgical time was slightly increased in the study group because of PRF preparation and application. No general or local complications were encountered both in the study and control groups.

In the study group, grade of alveolar socket fulfillment that was assessed 6 months postoperatively was rated as sufficient in 4 cases and adequate in the remaining 10 cases of 14 interventions. Instead, in the control group, the grade of fulfillment was rated as insufficient in 3 cases, sufficient in 4 cases, and adequate in the remaining 7 cases.

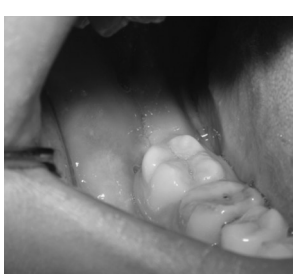

FIGURE 7. Postoperative image (10 d after surgery).

Mean preoperative periodontal probing of distal area of second molars in the control group was 3.78 mm, whereas in the study group treated with PRF, a mean value of 3.93 mm was found.

Six months after surgery, average periodontal probing was 3.28 mm in the control group and 3.07 mm in the study group. By VAS scale, patients revealed a mean value of 4.92 in the study group, whereas a mean value of 5.42 was found in the control group.

DISCUSSION

Platelet-rich fibrin of Choukroun et al is an autologous fibrin matrix, in which a large quantity of platelet and leukocyte cytokines are embedded.[19–26,29,30] As Dohan et al[19,21,24–26] and Choukroun et al noted,[22,23] thanks to a completely natural polymerization (without any modifiers), PRF is a totally autologous material prepared extemporaneously.

Platelet-rich fibrin of Choukroun et al[31] could be considered an easily and quickly prepared bioactive surgical additives for regulation of inflammation and enhanced healing.

Platelet-rich fibrin would promote wound healing and serve as an immune organizing node that regulates inflammation and provide wound protection thanks to the presence of growth factors (PDGF-ββ, TGF-β1, VEGF, and insulin-like growth factor 1) and inflammatory cytokines (IL-1β, IL-6, IL-4, and tumor necrosis factor α).[24,31,32] Therefore, PRF has a supportive effect on the immune system, being able to stimulate defense mechanisms, which might reveal useful in the case of wound infection.[24,31,32]

Furthermore, PRF may act as a supportive matrix for bone morphogenetic protein too,[32] and it has been shown to release the growth factors TGF-β1, PDGF-ββ, and VEGF and thrombospondin 1 for 7 days after its preparation.[31,33] Its molecular structure with low thrombin concentration is an optimal matrix for migration of endothelial cells and fibroblasts. It permits a rapid angiogenesis and an easier remodeling of fibrin in a more resistant connective tissue.[23] All of these factors suggest that PRF might be useful to promote wound healing.[32]

Opposite to other platelet concentrates, during preparation of PRF, the venous blood is collected in a dry glass or glass-coated plastic tubes without anticoagulant. On contact with the glass surface and during the centrifugation, the venous blood coagulates immediately and natural fibrin slow polymerization occurs without addition of thrombin and $CaCl_2$. Because of this slow natural and progressive polymerization, the homogenous three-dimensional organization of PRF is thought to lead to formation of equilateral junctions (particularly favorable to cell migration and soluble molecule retention), instead of bilateral junctions associated with thick fibrin polymers constituting a rigid fibrin network, between fibrin fibrillae. This should enable entrapment of growth factors from platelets and cytokines from leukocytes into fibrin matrix.[25,31]

Although Gürbüzer et al[31] found in their study that PRF does not seem to increase scintigraphically detectable enhanced bone healing within the extraction sockets of soft tissue impacted mandibular third molars 4 weeks after surgery, Chang et al[34] recently demonstrated that PRF can stimulate osteoblasts' proliferation. In fact, the activation of p-ERK and OPG expression by PRF, thus respectively promoting an up-regulation of cell proliferation in human osteoblastic cells and inhibiting osteoclastogenesis, suggests a potential role for new bone formation and regeneration.

In the current study, both in the control group and in the study group, we observed a reduction of periodontal probing depth, but this reduction was more noticeable in the study group: from 3.92 to 3.07 mm. This result may be compared with other studies, in which platelet concentrates reduce the distal probing depth and attachment level of the distal region of the lower second molar.[3,17]

Even if working time was slightly increased in the study group because of PRF preparation and application, several advantages were observed. In fact, in the study group, less pain was referred, and hygienic maintenance was facilitated by a more rapid alveolar socket fulfillment; consequently, distal periodontal condition was improved also. The healing processes seemed to be accelerated, and the patient's postoperative comfort seemed to be positively influenced.

Therefore, although we are aware of the limits of our clinical study and the small number of cases in our study and control groups, we agree with Simonpieri et al[29,30] about the several advantages offered by PRF. In fact, they claimed that PRF plays an important mechanical role (enabling a biomaterial to be maintained and protected against moderate parasitic forces); that together with allogenic bone, it facilitates cellular migration, vascularization, and survival of the graft; that it may create a perpetual process of healing (thanks to cytokines, transforming growth factor β1, and insulin-like growth factors); and that it has an important role in immunity too (because of the presence of leukocytes and cytokines into the network of fibrin).[29,30]

In conclusion, the combined action of PRF and piezoelectric surgery can be considered a safe and fine technique for third molar surgery and alveolar socket healing. In agreement with Choukroun et al[23] and Simonpieri et al,[29,30] PRF can be considered as a healing biomaterial. In fact, it seems to accelerate physiologic healing, enhancing bone healing in a controllable and relatively long-term effective way, although further randomized clinical studies would be needed to deepen the knowledge of this biomaterial.

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Effect of Rectal Diclofenac and Acetaminophen Alone and in Combination on Postoperative Pain After Cleft Palate Repair in Children

Seyed Mohammad Mireskandari, MD, Jalil Makarem, MD

Abstract: Acetaminophen and diclofenac are prescribed as postoperative analgesic agents in children. However, the efficacy of their combination is not studied sufficiently. We compare the analgesic effects of acetaminophen, diclofenac, and their combination after cleft palate surgery. In this randomized clinical trial, 120 children (1.5–5 y) who were scheduled for cleft palate repair were studied. Children were randomized to receive placebo, acetaminophen (40 mg/kg), diclofenac (1 mg/kg), or acetaminophen (40 mg/kg) plus diclofenac (1 mg/kg) rectally just after surgery. Acetaminophen (30 mg/kg) and diclofenac (1 mg/kg) were administered every 8 hours until 48 hours. Postoperative pain was assessed regularly with the Children Hospital of Eastern Ontario Pain Scale, and rescue analgesia was provided if scores were 7 or greater. Time to the first prescription of meperidine, total postoperative meperidine consumption, and adverse effects were the main outcomes.

After surgery, pain scores were higher in placebo than in other groups in all time intervals. In the first 12 hours, pain scores in the combined group were less than those in the acetaminophen ($P < 0.05$) and diclofenac ($P < 0.05$) groups. Postoperative meperidine consumption was the highest in placebo and was the least in combined group ($P < 0.05$). It was significantly higher in the acetaminophen group than in the diclofenac group ($P < 0.05$). Time to the first prescription of meperidine was significantly different among all groups. Adverse effects were comparable among groups.

Rectal acetaminophen plus diclofenac was found to be the most effective in pain control. However, both rectal acetaminophen and diclofenac were more effective than placebo, whereas diclofenac was more effective than acetaminophen.

Key Words: Analgesics, nonnarcotic, acetaminophen, anti-inflammatory agents, nonsteroidal, diclofenac, congenital, hereditary, and neonatal diseases and abnormalities, musculoskeletal abnormalities, cleft palate, minors, preschool child

Repair of the cleft palate commonly is performed in 1- to 2-year-old children.[1] Manipulation the soft and hard palate causes a lot of pain after this surgery.[2] Inadequate analgesia can cause restlessness, agitation, hemorrhage, and delay in wound healing in these preverbal patients.[3] Therefore, it is necessary to study different pain management regimens after surgical correction of the cleft palate. Rectal nonsteroidal anti-inflammatory drugs (NSAIDs)[4–6] or rectal acetaminophen[5–8] is prescribed in children for postoperative analgesia, but there are limited evidence in the efficacy of their combination in children. However, in adults, it has reported that this combination provided better postoperative analgesia than either drug given alone.[8–10] The analgesic effects of combined rectal diclofenac and acetaminophen after surgery of the cleft palate were not studied previously.

In this randomized, double-blinded, placebo-based study, we compared the analgesic effects of rectal acetaminophen, rectal diclofenac, and their combination on the postoperative period of cleft palate surgery.

MATERIALS AND METHODS

The study protocol was approved by the ethics committee of Tehran University of Medical Science. All parents provided informed consent before enrollment in the study. This randomized double-blinded trial was performed from February 2008 to August 2010 in a referral children hospital in Tehran.

Selection criteria included children aged between 1.5 and 5 years and American Society of Anesthesiologists (ASA) class I and II who were scheduled for cleft palate surgical repair. Children with a history of bleeding or thrombocytopenia, allergy to nonsteroid pain remedies (especially diclofenac and acetaminophen), asthma, digestive system disorders (specifically gastric ulcer), renal diseases, taking opioid or noninflammatory drugs before surgery, hepatic

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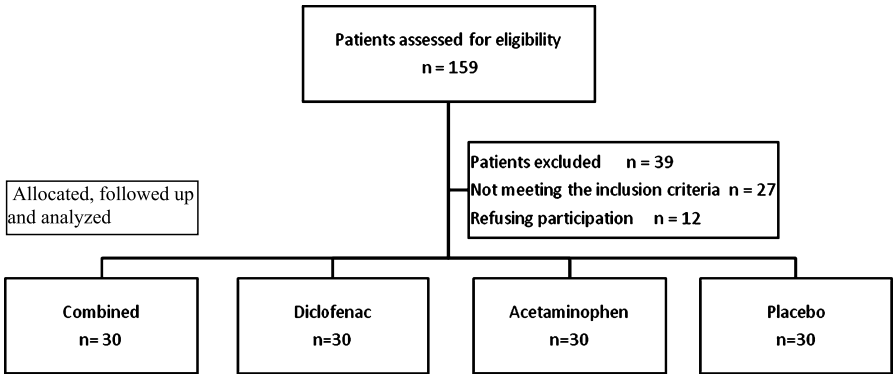

FIGURE 1. Flowchart of this prospective randomized study.

disorders, central nervous system diseases, hypovolemia, hypotension, and active bleeding were excluded from the study.

Included children according to these criteria were allocated in 4 equal groups by block randomization method: group 1, rectal placebo (Pharma Chemie, Tehran, Iran); group 2, rectal acetaminophen 40 mg/kg (Aburaihan Pharmaceutical Co, Tehran, Iran); group 3, rectal diclofenac 1 mg/kg (Pharma Chemie); and group 4, combination of rectal diclofenac 1 mg/kg and rectal acetaminophen 40 mg/kg. Rectal placebo, diclofenac, and acetaminophen were administered in the operating room at the end of surgery and every 8 hours until 48 hours. The repeated doses of rectal diclofenac and acetaminophen were 1 and 30 mg/kg, respectively.

Rectal diclofenac and acetaminophen were administered by a nurse who was not involved in the care of the patients. Parents, the physicians, and the nurses who were involved in assessing the patients and collection of data were blinded to the study groups.

All children were managed in the same manner in the preoperative, operative, and postoperative periods, except for the kind of intervention. Children were fasted overnight with an opportunity to drink clear fluids until 2 hours before surgery. Before transferring children to the operating room, intravenous access was established using a 22-gauge catheter inserted in the surgical ward 45 minutes after the application of EMLA cream (AstraZeneca, Sodertalje, Sweden). Children were premedicated with midazolam (0.4 mg/kg) that was administered orally 30 minutes before induction and intravenous fentanyl (2 μg/kg) 5 minutes before induction of anesthesia. Lactated Ringer's serum (7 mL/kg) was infused before inducing anesthesia.

After preoxygenation with 100% O_2, anesthesia was induced with sodium thiopental (5 mg/kg). Tracheal intubation was facilitated by an intravenous injection of atracurium (0.5 mg/kg). Anesthesia was maintained with a mixture of 50% N_2O and 1.5% to 2% isoflurane. Thereafter, patients received additional atracurium and fentanyl as required. Intraoperative monitoring consisted of capnography, pulse oximetry, noninvasive blood pressure monitoring, Fi_{O_2}, and electrocardiography. One of the staff surgeons who use similar surgical techniques performed the surgery. At the end of the surgery, the wound was infiltrated by a mixture of bupivacaine (0.25%) and epinephrine (1:200,000) 1 mL/kg. Neuromuscular blockade was reversed by neostigmine 40 μg/kg and atropine 20 μg/kg. After extubation and administration of the first dose of rectal acetaminophen and/or diclofenac, children were transferred to the postanesthesia care unit for continuous monitoring of vital signs and assessment of pain. After 120 minutes, they were transferred to the children surgery ward.

A trained nurse, who was blinded to the analgesic treatment prescribed, recorded the postoperative pain score using the Children Hospital of Eastern Ontario Pain Scale (CHEOPS) in the 1, 2, 6, 12, 24, 36, and 48 hours after surgery. The CHEOPS is an observational pain scale consisting of 6 behavioral components that provide a global score ranging from 4 to 13.[11] Patients with a score of 7 or higher received intramuscular meperidine (1 mg/kg) as rescue analgesia. Adverse events such as nausea, vomiting, itching, and any unusual bleeding were recorded.

The main outcome of the study was 25% decrease in meperidine consumption in the combination group in comparison with the groups of diclofenac or acetaminophen alone. A minimum of 26 children was required to achieve a significance level of 0.05 with a power of 80%. To accommodate for dropouts, we aimed to randomize at least 30 children in each of the groups.

TABLE 1. Basic Characteristics

	Placebo	Acetaminophen	Diclofenac	Combined	*P*
Age, mean (SD), y	2.3 (1)	2.1 (0.8)	2.2 (1.1)	2.0 (5)	0.64
Weight, mean (SD), kg	14.5 (3.2)	14.4 (4.4)	14.0 (4.5)	14.1 (3.8)	0.94
Sex (female), n (%)	14 (46.7)	16 (53.3)	15 (50)	13 (43.3)	0.88
ASA class II, n (%)	5 (16.7)	3 (10)	3 (10)	4 (13.3)	0.84
Duration of surgery, mean (SD), min	81.7 (30.1)	89.2 (31.8)	88.2 (29.4)	87.3 (29.9)	0.78
Duration of anesthesia, mean (SD), min	119.2 (32.5)	128.2 (21.5)	124.7 (29.3)	126.0 (30.6)	0.66
Intraoperative fentanyl, mean (SD), μg/kg	3.1 (0.8)	3.2 (0.8)	3.1 (0.7)	2.9 (0.9)	0.38
Hard palate plasty (yes), n (%)	19 (63.3)	22 (73.3)	20 (66.7)	21 (70)	0.86

There were 30 patients per group.

TABLE 2. Time to the First Prescription of Opioid, Postoperative Meperidine During the First 24 Hours, and 48 Hours

	Placebo	Acetaminophen	Diclofenac	Combined	*P*
Time to the first prescription of meperidine, mean (SD), min	**50 (11.7)**	**62 (19.5)**	**92 (12.5)**	**140 (21.7)**	**<0.001**
Total meperidine dose during 24 h, mean (SD), mg	**79.8 (8.8)**	**72.3 (9.5)**	**60.0 (9.2)**	**44.0 (8.8)**	**<0.001**
No. doses during 24 h, mean (SD)	**5.6 (0.6)**	**5.1 (0.7)**	**4.2 (0.6)**	**3.1 (0.6)**	**<0.001**
Total meperidine dose during 48 h, mean (SD), mg	**97.1 (13.9)**	**88.3 (8.3)**	**70.7 (10)**	**51.0 (9.5)**	**<0.001**
No. doses during 48 h, mean (SD)	**6.8 (0.9)**	**6.2 (0.6)**	**4.9 (0.7)**	**3.6 (0.7)**	**<0.001**

There were 30 patients per group. Time to the first prescription of opioid: all pairwise comparisons among groups were significant ($P < 0.05$). Total meperidine dose and number of doses during 24 and 48 hours after surgery: all pairwise comparisons among groups were significant ($P < 0.05$).

Only the available CHEOPS pain scores were used for assessment. The average of the CHEOPS pain scores in each group was calculated at 1, 2, 6, 12, 24, 36, and 48 hours after surgery. The continuous variables are presented as mean (SD). The Kolmogorov-Smirnov test for goodness of fit to normal distribution was performed, and normality was obtained for all measurements. One-way analysis of variance test was used for comparison of the means of continuous variables. Bonferroni test was used for multiple comparisons. Categorical variables are given as counts, and group comparisons were made with χ^2 test. All calculations were performed with SPSS version 16 (SPSS, Inc, Chicago, IL). $P < 0.05$ was considered to indicate statistical significance.

RESULTS

Of 159 patients assessed for eligibility, 39 patients were excluded (not meeting the inclusion criteria [n = 27] or refusing participation [n = 12]; Fig. 1).

One hundred twenty patients were randomized, and all the randomized patients completed the study; therefore, data from 120 patients, 30 in each group, were analyzed. The groups were comparable regarding to the age, weight, sex, ASA class, duration of anesthesia, duration of surgery, and the frequency of hard palatoplasty (Table 1).

As presented (Table 2), the total dose of postoperative meperidine administration and the number of doses during the first 24 and 48 hours after surgery were significantly different in pairwise comparison among the 4 groups. In both periods, the highest dose was in the placebo group, and the lowest dose was in the combined group. In the diclofenac group, meperidine was administered lower than in the acetaminophen group.

Time to the first prescription of opioid in the placebo group was significantly lower than in the acetaminophen, diclofenac, and combined groups. In comparison, between each 2 groups, the differences in time to the first prescription of opioid were significant.

As depicted (Table 3), the average of the CHEOPS pain scores during the first 48 hours after surgery was compared between groups in 8 time intervals. In all postoperative intervals (except at recovery), pain scores were significantly different among 4 groups (all $P < 0.05$), and in the combined group, the pain score was less than that of the placebo ($P < 0.05$). At 1, 2, and 6 hours after surgery, pain scores in the combined group were less than those in the acetaminophen group ($P < 0.05$), and at 6 and 12 hours after surgery, pain scores in the diclofenac group were significantly less than those in the placebo group ($P < 0.05$).

Adverse effects consisted of nausea, vomiting, and itching, but there was no significant difference in the incidence of adverse effects between the groups (Table 4). No unusual bleeding after surgery was reported. Treatment intervention for adverse effects was needed only in 5 of the patients for nausea or vomiting.

DISCUSSION

In this study, we found that the analgesic effect of combined rectal diclofenac and acetaminophen was significantly higher than that of placebo and either alone after cleft palate surgery. We showed that the mean pain scores were significantly lower in the combined group than in the other groups at 1, 2, 6, and 12 hours after surgery.

TABLE 3. Postoperative CHEOPS Pain Scores

	Placebo	Acetaminophen	Diclofenac	Combined	*P*
Recovery	**7.9 (2.4)**	**7.5 (3.1)**	**6.9 (3.6)**	**6.8 (2.8)**	**0.57**
1 h	**7.5 (2.0)**	**7.2 (2.3)**	**6.7 (0.8)**	**5.5 (1.5)**	**<0.001**
2 h	**6.7 (1.1)**	**6.6 (1.7)**	**6.2 (0.7)**	**5.3 (0.8)**	**<0.001**
6 h	**6.6 (1.3)**	**6.1 (1.1)**	**5.8 (0.9)**	**5.1 (0.9)**	**<0.001**
12 h	**6.5 (1.0)**	**5.9 (1.2)**	**5.7 (0.9)**	**5.0 (0.7)**	**<0.001**
24 h	**5.9 (1.4)**	**5.6 (1.3)**	**5.2 (1.0)**	**4.9 (1.3)**	**0.02**
36 h	**5.6 (0.9)**	**5.3 (0.7)**	**5.1 (0.7)**	**4.7 (0.9)**	**<0.001**
48 h	**5.2 (0.9)**	**5.1 (0.6)**	**4.9 (0.6)**	**4.6 (0.9)**	**0.21**

There were 30 patients per group. In all hours, placebo versus combined ($P < 0.05$). At 1, 2, 6, and 12 hours: acetaminophen versus combined ($P < 0.05$), diclofenac versus combined ($P < 0.05$). At 6 and 12 hours: placebo versus diclofenac ($P < 0.05$). All the other pairwise comparisons among the groups were not significant ($P > 0.05$).

TABLE 4. Adverse Effects in Groups

	Placebo	Acetaminophen	Diclofenac	Combined	*P*
Nausea/vomiting, n (%)	8 (26.7)	**9 (30)**	8 (26.7)	**10 (33.3)**	**0.93**
Itching, n (%)	**4 (13.3)**	**2 (6.7)**	3 (10)	**1 (3.3)**	**0.54**

No significant differences between groups.

Meperidine consumption was significantly lower in the combined group than in the placebo, acetaminophen, and diclofenac groups in the first 24 and 48 hours after surgery. Time to the first opioid prescription was significantly longer in the combined group than each of the placebo, acetaminophen, and diclofenac groups.

It is showed that twice-daily rectal diclofenac (1 mg/kg) effectively controlled pain after cleft palate repair.[12] However, the analgesic effect of a single dose of rectal acetaminophen (40 mg/kg) in children undergoing cleft palate repair was weak.[13] According to our knowledge, the analgesic effect of combined acetaminophen and NSAIDs on cleft palate repair has not been studied yet.

There are several studies with conflicting results about the analgesic efficacy of acetaminophen and NSAIDs during the postoperative period in children. The type of surgery may be an important factor. Rectal acetaminophen (40 mg/kg) provided a good analgesia for 6 hours after adenotonsillectomy in children.[7] With rectal acetaminophen, postoperative pain was sufficiently controlled after herniorrhaphies, whereas it could not provide an effective analgesia after orchidopexies.[14] After major surgeries in young infants, acetaminophen does not have an additional analgesic effect when added to morphine infusion.[15] In a previous report, the effect of diclofenac (2–3 mg/kg per 24 hours) was the same as a high-dose acetaminophen (90 mg/kg per 24 hours) for providing analgesia after pediatric tonsillectomy.[16]

Reports about the efficacy of their combination in children are limited and controversial, too. Children who received the rectal diclofenac (1 mg/kg)–acetaminophen (40 mg/kg) combination experienced a lower level of pain and a decreased need for morphine compared with children receiving each drug alone after inguinal hernia repair.[17]

Combining rectal acetaminophen with rectal ibuprofen did not improve pain control in the immediate postoperative period after adenoidectomy in children.[4] After appendectomy in children, concurrent administration of diclofenac (1 mg/kg) reduced cumulative morphine consumption significantly, but no additive effect of acetaminophen (15–20 mg/kg) was detected.[18]

Another possible factor for these controversies is the route, dose, and time of administration. Both acetaminophen and diclofenac have been administered from the rectal route. But their pharmacokinetic is really different. Absorption of acetaminophen from the rectum is slow, irregular, and unpredictable.[19–21] Therefore, its bioavailability is not consistent. Peak plasma concentration occurs in an average of 2 to 3 hours after insertion of rectal suppository of acetaminophen.[22,23] Administration of rectal acetaminophen with a dose of 45 mg/kg did not reach to the analgesic serum level.[23] Although diclofenac is rapidly and well absorbed via rectal suppositories,[24] its absorption half-life is 0.613 hours, with a lag time of 0.188 hours.[25]

In previous studies, administration of low-dose rectal acetaminophen was introduced as the cause of an inefficient analgesic effect.[13,18] Studies suggest that a dose of 80 to 100 mg/kg acetaminophen per day can be given safely postoperatively for 2 to 3 days.[26–28] Therefore, high-dose acetaminophen should be administered for postoperative pain control, as we did in the current study.

The mechanisms of acetaminophen and NSAIDs in pain control are different, so theoretically, their combination should be more efficient than each of them alone, as it has been showed in adults. However, in children, the reports are not sufficient for a consistent conclusion.

Acetaminophen potently inhibits brain cyclooxygenase[26,29]; also, it has peripheral effects by blocking impulse generation within the bradykinin-sensitive chemoreceptors responsible for the generation of afferent nociceptive impulses. Several reports have shown that acetaminophen could inhibit substance P–mediated hyperalgesia and reduce nitric oxide generation involved in spinal hyperalgesia induced by substance P or *N*-methyl-d-aspartate.[30] Another way for acetaminophen antinociceptive effects is through the opioidergic system, modulating dynorphin release in the central nervous system.[31]

Diclofenac is an NSAID with an approximate relative cyclooxygenase 1/2 specificity ratio of 1.[32] Nonsteroidal anti-inflammatory drugs such as diclofenac inhibit prostaglandin synthesis, mainly peripherally but also centrally.[33,34]

It has been suggested that NSAIDs are more effective for relieving postoperative pain after orthopedic surgery because, in this condition, prostanoid activity is increased prominently. In orthopedic surgeries in children, the combination of ketoprofen and acetaminophen was significantly more effective than each of them given alone. However, the result of this study was not achieved in other soft tissue surgeries.[35]

The interaction of NSAIDs and acetaminophen's effects with opioids is different. In an animal model, it is depicted that the combination of acetaminophen and morphine is additive; however, the combination of diclofenac and morphine is synergistic.[36]

In our study, no significant differences in adverse effects were detected between groups, and the most frequent adverse effects were nausea and vomiting similar to some previous reports.[18,35] Although diclofenac, in comparison with acetaminophen, resulted in a lower incidence of nausea and vomiting in pediatric patients after tonsillectomy,[16] and it was associated with lower opioid consumption.

In the current study, we concluded that the combination of rectal acetaminophen and diclofenac is more effective in pain control than each of this drug given alone after cleft palate repair. Pain score and meperidine consumption were significantly decreased in the combined group. However, adverse effects were comparable in the 4 groups.

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Iatrogenic Arteriovenous Fistula of the Superficial Temporal Artery After Manual Reduction of Temporomandibular Joint Dislocation

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Abstract: A 33-year-old man fell from a height and was referred to our hospital. Physical examination showed a swelling in the left preauricular region without laceration. No thrill or bruit was detected

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at this time. A face x-ray and a computed tomographic scan showed a left temporomandibular joint (TMJ) dislocation, Le Fort I fractures, and a mandibular body fracture. Left TMJ dislocation was treated by manual reduction. Two days after admission, a swelling in the left preauricular region progressed, with thrill and bruit. Left external carotid artery angiograms showed an arteriovenous fistula with a dilated pouch near the left TMJ. The fistula was fed by the left superficial temporal artery and drained into the left superficial temporal vein. The fistula was successfully embolized using Tornado coils. This is the first case of an arteriovenous fistula of the superficial temporal artery after manual reduction of TMJ dislocation.

Key Words: Arteriovenous fistula, superior temporal artery, temporomandibular joint dislocation, manual reduction

Arteriovenous fistulas (AVFs) of the superficial temporal artery (STA) are extremely rare, and there have been only occasional published reports.[1–10] Most such AVFs are caused by trauma and rarely after an iatrogenic manipulation.[4–9] We report the first case of an AVF of the STA after manual reduction of temporomandibular joint (TMJ) dislocation.

CLINICAL REPORT

A 33-year-old man fell from a height and was referred to our hospital. The patient had a history of schizophrenia treated with antipsychotics. Vital signs were as follows: blood pressure, 86/44 mm Hg; pulse rate, 136 beats/min; and respiratory rate, 24 breaths/min. Physical examination showed a swelling in the left preauricular region without laceration. No thrill or bruit was detected at this time. The Glasgow Coma Scale score was 15. A face x-ray and a computed tomographic (CT) scan showed a left TMJ dislocation, Le Fort I fractures, and a mandibular body fracture (Figs. 1A, B). Other x-ray examinations showed a pelvic fracture, a right femoral fracture, and a right tibial fracture. A brain CT scan was normal. An abdominal CT scan revealed retroperitoneal hemorrhage. Left TMJ dislocation was treated by manual reduction. Rapid transfusion was administered, and emergent transcatheter arterial embolization for retroperitoneal hemorrhage was performed. Two days after admission, a swelling in the left preauricular region progressed, with thrill and bruit. Left external carotid artery angiograms showed an AVF with a dilated pouch near the left TMJ (Figs. 2A, B). The fistula was fed by the left STA and drained into the left superficial temporal vein.

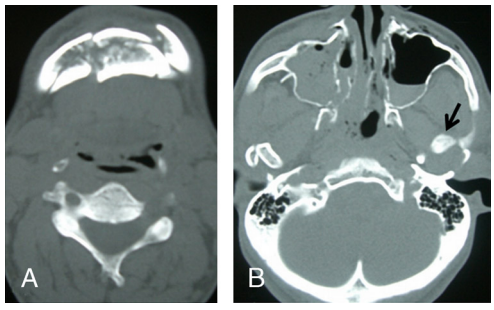

FIGURE 1. A and B, Face CT scan showing a left TMJ dislocation (arrow), Le Fort I fractures, and a mandibular body fracture.

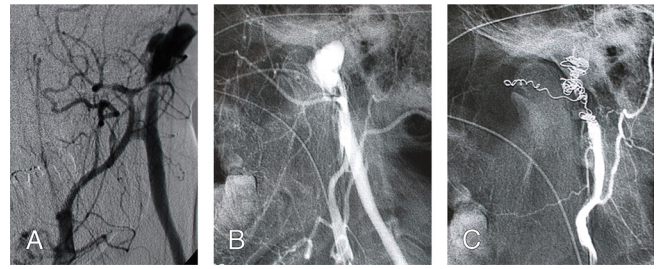

FIGURE 2. Left external carotid artery angiogram, anteroposterior (A) and lateral (B) views, showing a superficial temporal AVF with a dilated pouch near the left TMJ. Left lateral external carotid artery angiogram (C) after embolization using Tornado coils, revealing no residual arteriovenous shunting.

The fistula was embolized using Tornado coil (Cook, Bloomington, IN) (Fig. 2C). After this procedure, thrill and bruit disappeared. Facial fractures were treated conservatively. On day 20, the patient underwent surgeries for right femoral and tibial fractures. The patient was transferred to another hospital on day 27 with mild disability.

DISCUSSION

Arteriovenous fistula of the STA is a rare pathology.[1] Most such AVFs are caused by trauma and rarely after an iatrogenic manipulation.[1–10] Iatrogenic etiologies, which have been reported in the literature, include craniotomy, hair transplantation, and temporomandibular arthroscopy.[4–9]

Moreover, the STA follows a long and tortuous course over the frequently injured temporal bone and is thus susceptible to injury.[4] The STA is particularly vulnerable where it crosses the sharp edge of the superior temporal line, which has no underlying cushion of muscle.[4] However, AVF of the STA near the TMJ is extremely rare.[5,10]

There has been no reported case of AVF of the STA after manual reduction of TMJ dislocation. Our patient did not show signs of AVF on admission. However, 2 days after manual reduction of the dislocated TMJ, the patient presented with AVF of the STA near the TMJ. We concluded that the AVF was caused by manual reduction of TMJ dislocation because of its delayed onset.

The indications for treatment include improvement of the disfigurement, prevention of hemorrhage, and relief of symptoms such as headache and tinnitus.[1] Treatment may consist of surgical excision or endovascular embolization.[1] In our patient, investigation by transarterial angiography was combined with embolization.

In conclusion, we suggest that careful attention should be paid to the development of delayed AVF of the STA after manual reduction of TMJ dislocation.

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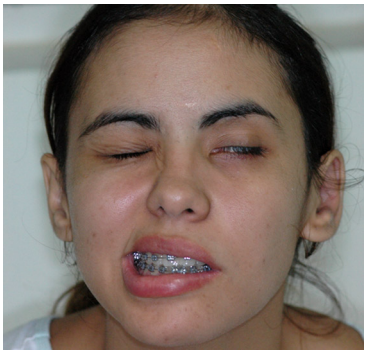

FIGURE 1. Initial picture: loss of control of facial muscle motor function on the left with deviation of the buccal rima to the right, left facial edema.

Unilateral Facial Paralysis Caused by Ramsay Hunt Syndrome

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Abstract: The Ramsay Hunt syndrome is a rare disease caused by an infection of the geniculate ganglion by the varicella-zoster virus. The main clinical features of the syndrome are as follows: Bell palsy unilateral or bilateral, vesicular eruptions on the ears, ear pain, dizziness, preauricular swelling, tingling, tearing, loss of taste sensation, and nystagmus. We describe a 23-year-old white woman, who presented with facial paralysis on the left side of the face, pain, fever, ear pain, and swelling in the neck and auricular region on the left side. She received appropriate treatment with acyclovir, vitamin B complex, and CMP nucleus. After 30 days after presentation, the patient did not show any signs or symptoms of the syndrome. At follow-up at 1 year, she showed no relapse of the syndrome.

Key Words: Ramsay Hunt syndrome, varicella-zoster virus, syndrome, geniculate ganglion

The Ramsay Hunt syndrome (RHS) was described for the first time by the neurologist James Ramsay Hunt in 1907, who defined it as an inflammation of the geniculate ganglion, with compromise of cranial nerve VII pair (facial nerve) and eventually of the cranial nerve VIII pair (cochlear nerve), caused by an infection by the varicella-zoster virus (VZV).[1] This virus is a member of the herpesvirus family, responsible for 12% of the cases of facial nerve paralysis capable of remaining latent in other ganglions of the head and neck.[2–4] It appears with greater frequency and dissemination in patients with neurological complications and HIV-positive and immunosuppressed patients,[5,6] presenting increasing incidence with age.

The main clinical manifestations are as follows: unilateral or bilateral peripheral facial paralysis, vesicular eruptions on the auricular pavilion, diminished audition, buzzing, nausea, otalgia, vomiting, nystagmus, lacrimation, and diminishment of taste in the anterior third of the tongue.[1,7,8] Thus, facial paralysis includes 3% to 12% of all the facial paralyses,[7,9,10] being the second most common cause of atraumatic peripheral facial paralysis.[11]

The diagnosis of RHS is basically made by the clinical signs and symptoms. There was great advance in the treatment with the development and application of antiviral agents, particularly acyclovir, which is effective for the prevention of replication of a variety of viral particles, including VZV.[10,12] Some studies have suggested the association of acyclovir with corticosteroids for a more effective treatment.[13] The possibility has been related to a complete improvement in the clinical condition when treatment is instituted in up to 3 days from the onset of the first symptoms. With early treatment, complete recovery was observed in 70% to 86% of cases, whereas only around 20% of nontreated patients improved completely.[14]

Although facial paralysis is a rare complication, the purpose of this report was to aid the dentist in distinguishing RHS from other

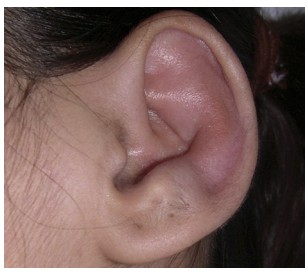

FIGURE 2. Initial picture: hyperemia of the left external auditive pavilion.

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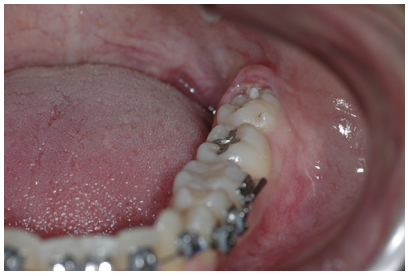

FIGURE 3. Initial picture: discrete gingival inflammation in the distal region of the third mandibular molar.

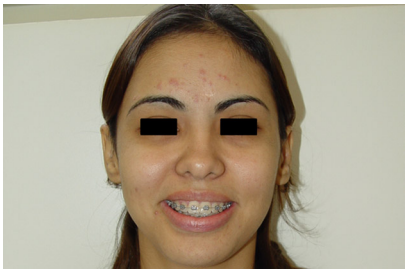

FIGURE 5. One-year follow-up.

diseases or complications, to institute effective and early antiviral therapy.

CLINICAL REPORT

The patient, a 23-year-old leukoderm woman, sought attendance at the First Aid Station of Santa Casa de Araçatuba with the complaint of difficulty in moving the left side of her face that began on waking. She also informed that she was very worried, because in the previous evening she had been submitted to surgery for the extraction of tooth no. 38 under local anesthetic, and believed this to be the cause of her problem. On clinical examination, loss of control of facial muscle motor function on the left was observed, with deviation of the buccal rima to the right, otalgia, fever, facial edema on the left, and hyperemia of the left external auditive pavilion (Figs. 1 and 2).

On intraoral examination, a discrete gingival inflammation was observed in the distal region of the third mandibular molar (Fig. 3). Ultrasound examinations of the parotid gland were requested, to discard the possibility of paralysis due to anesthesia in the region of the parotid gland. The examinations presented no alterations. In conjunction with the evaluations by the hospital's specialties of otorhinolaryngology, neurology and dermatology, the diagnosis of RHS was made. The following medication treatment was instituted: acyclovir 500 mg oral (every 6 hours in the first 7 days and afterward 500 mg every 24 hours for a further 7 days), intramuscular vitamin B complex (1 ampoule every 2 days), and CMP nucleus (every 8 hours for 30 days). The patient was also informed that there was no direct relationship between the extraction and all the signs and symptoms she presented.

On the fifth day of development, the left auditive pavilion presented multiple vesicular coalescent lesions and several ulcerated lesions, characteristic signs of the syndrome (Fig. 4). On the 10th day, there was discrete diminishment of the facial paralysis, absence of otalgia, diminishment of the edema in the auricular region and neck, and exacerbation of the vesicular lesions, and on the 15th day, the facial paralysis was discrete, and the vesicular lesions were in a state of healing.

After 1 month, the patient presented complete recovery of facial motor function, without pain complaints and absence of lesions on the auditive pavilion, and after 1 year, there was no sign or symptom of recurrence of the syndrome (Fig. 5).

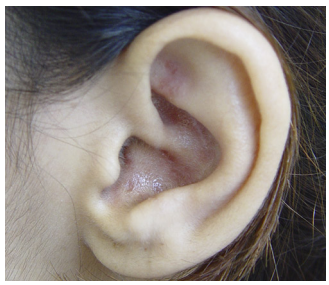

FIGURE 4. Left auditive pavilion with multiple vesicular coalescent lesions and several ulcerated lesions.

DISCUSSION

Ramsay Hunt syndrome occurs in individuals who previously had chickenpox and is caused by the reactivation of VZV. In the case presented, the patient related that she had chickenpox at the age of 6 years. Other systemic diseases may be associated with the reactivation of VZV, such as diabetes, leukemia, AIDS, lipomas, and malignant diseases.[15,16]

The traditional medication treatment of RHS is basically with corticosteroids and antiviral agents. Acyclovir is an effective antiviral agent against the replication activity of the virus. It is activated by the action of the viral quinase thymidine enzyme that phosphorylates the monophosphate of acyclovir into acyclovir triphosphate. This inhibits the polymerase of viral DNA and interrupts the synthesis of the virus DNA and its replication.[10,17,18]

Kinishi et al,[19] in their study, showed that the results of therapy with acyclovir in the resolution of the facial paralysis were effective, showing improvement in the function of the nerves, observed by means of excitability tests. However, an increase in the resistance of the virus to therapy with acyclovir has been related.[20,21]

The parenteral dose of acyclovir is 15 mg/kg per day, and the oral dose is 800 mg 4 times a day, because absorption of acyclovir in the gastrointestinal tract is only from 15% to 30% of the dose ingested. In a clinical report,[3] the option was to use the oral dose of the drug, which in the proportion of milligrams per weight was 500 mg 4 times a day.

Murakami et al[22] related the treatment of 48 patients with oral acyclovir and 32 patients with intravenously administered acyclovir and when analyzing the results observed that there was no significant difference between the 2 treatments proposed, enabling 81% of complete recovery of the patients, with the advantage that the oral treatment reduces the need for hospitalizing the patient.

Coadjuvant therapy with steroids may be useful in the treatment of facial paralysis in RHS.[23] However, some authors contraindicate this therapy, especially in patients with periocular lesions, fearing the dissemination of the infection by VZV.[23,24] The use of corticoid in this clinical cause did not cause any other complication, other than those initially mentioned.

CMP nucleus was also used for functional recovery of the neuromuscular system. It is composed of cytidine-5-monophosphate disodium, uridine-5-triphosphate trisodium, and hydroxocobalamin

acetate. According to the manufacturer (Lab Gross SA, Rio de Janeiro, Brazil), cytidine is a nucleotide that actively intervenes in the metabolism of the nervous system, becoming a coenzymatic factor that is absolutely necessary for the maintenance and regeneration of nervous structures. The action of cytidine is complemented by uridine. Hydroxocobalamin acetate is the more active form and has a more prolonged effect of vitamin B_{12}, having neurotrophic and analgesia action in all painful neuritic manifestations.

The authors conclude that the clinical characteristics of RHS described in the literature allow the dentist to make a differential diagnosis with other pathologies or complications, enabling an early and effective antiviral therapy to be instituted; the medication treatment instituted was effective for the case related; the interaction with professionals from other specialties allowed early diagnosis and consequently a more effective treatment with fewer possibilities of sequelae.

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Proptosis, Congestion, and Secondary Glaucoma Due to Carotid-Cavernous Fistula After Embolization

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Abstract: Carotid-cavernous fistulas (CCFs) are traumatic or spontaneously occurring communications between the carotid artery and the cavernous sinus. Carotid-cavernous fistulas can be due to a direct connection or indirect connections between the carotid artery system and the cavernous sinus. According to the etiologic classification, they may be of traumatic or spontaneous origin, and according to the angiography classification, they may be of direct or dural. Most CCFs are of spontaneous origin, and these are reported as frequently self-healing lesions. Spontaneous CCFs are mostly secondary to arteriosclerotic changes, which explains the increased ratio of elderly patients. Traumatic CCFs are usually of high-flow type and need intervention. The symptoms are various usually correlated to the size and type of venous

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drainage. The most frequent symptoms on presentation are proptosis, conjunctivitis, and chemosis; however, this picture may be complicated by optic nerve edema, cranial nerve palsies, and intracranial hemorrhage. We report a case with right low-flow dural CCF, which has worsened after angiography and recovered totally soon after endovascular embolization process.

Key Words: Carotid-cavernous fistula, proptosis, cerebral angiography

Carotid-cavernous fistulas (CCFs) are abnormal communications between the carotid artery system and the venous cavernous sinus. CCFs may be high flow (usually direct fistulas), and these types of fistulas may arise on trauma or spontaneously. Some CCFs are low-flow dural fistulas, and they usually occur spontaneously.[1] High-flow fistulas usually require endovascular intervention,[2,3] but some low-flow fistulas may resolve spontaneously.[4,5] In some cases, low-flow CCFs may need embolization because of consistent complaints. These patients usually present with proptosis, edema of the eyelids, chemosis, prominent conjunctival vasculature, reduced eye movement, pain, and some other findings.[2,4]

We report a case with spontaneously occurring low-flow CCF with prominent ophthalmic findings such as proptosis, swelling of the upper eyelid, chemosis, engorgement of conjunctival vessels, decreased visual acuity, and increased intraocular pressure, which has been resolved with embolization of the fistula.

CLINICAL REPORT

A 62-year-old woman, with no prior trauma or medical history other than well-going hypertension, has presented with mild right orbital edema with prominent swelling of the upper eyelid, diplopia, decreased visual acuity, and mild chemosis. A careful and detailed taken history revealed that she has been admitted to another center with the complaint of eyelid edema and red eye and has been treated for allergic conjunctivitis for 3 months. On entry to the outpatient clinic, we have done full ophthalmoscopic examination. Her right upper eyelid was edematous, whereas there was little restriction in abduction of the right eye (Fig. 1). There was diplopia in full right gaze. Corrected visual acuity was 20/30 in the right eye and 20/20 in the left eye. There was no afferent pupillary defect. Intraocular pressures were 38 mm Hg in the right eye and 13 mm Hg in the left eye measured with an applanation tonometer. Biomicroscopic examination revealed chemosis and prominent conjunctival corkscrew vessels (Fig. 1). There was no additional anterior or posterior segment pathology. Examination result of the left eye was normal. Antiglaucomatous medication including a prostaglandin analog and a combined preparation of dorzolamide and timolol maleate has been administered for the next 2 weeks, but the lowest measured intraocular pressure has been recorded as 32 mm Hg. Orbital and cranial magnetic resonance imagings (MRIs) were taken, demonstrating that there were right proptosis with the enlargement of extraocular muscles and dilated superior ophthalmic vein (Figs. 2 and 3). Selective cerebral angiography was planned and performed, revealing right low-flow type D CCF (Fig. 4). Immediately after the angiography, her findings have worsened, such as eyelid edema and chemosis has increased (Fig. 5) and bruit was begun to be heard on auscultation, which was explained as increased flow in the fistula after intervention. Embolization has been performed, and after that, the patient's symptoms have been resolved. One month later, she has been examined for follow-up when eyelid edema and chemosis were absent, whereas intraocular pressure has declined to 14 mm Hg without medication. Proptosis and diplopia due to limited abduction of the right eye have been recovered after the procedure (Fig. 6).

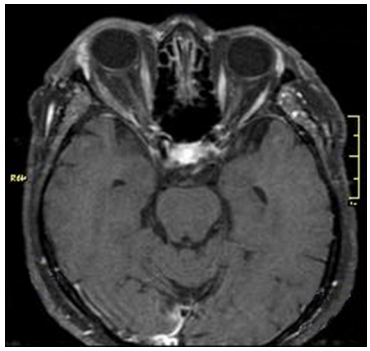

FIGURE 2. Orbital MRI of the patient on the first presentation.

DISCUSSION

Indirect CCFs are reported to be frequently, spontaneously resolving disorders.[5] Unlike direct CCFs, these cases have a more subtle

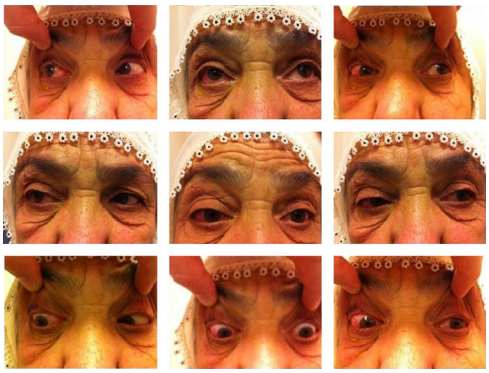

FIGURE 1. Patient with proptosis and conjunctival hyperemia on the first presentation.

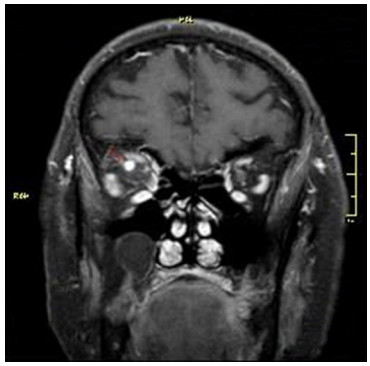

FIGURE 3. Prominent superior ophthalmic vein observed in orbital MRI.

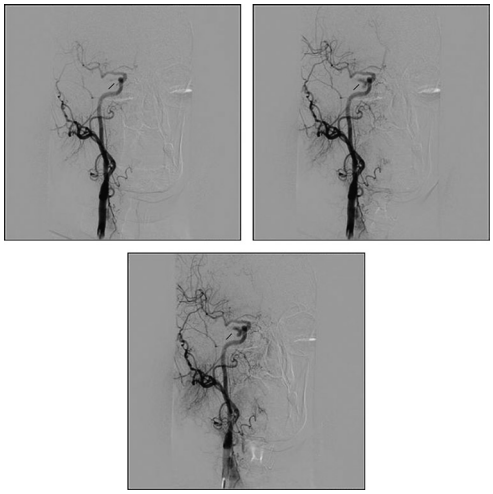

FIGURE 4. Selective angiogram showing the type D CCF mainly on the right internal carotid artery.

presentation and are usually clinically silent with minor symptoms such as conjunctival hyperemia and mild chemosis. However, there may be a need for endovascular intervention in case that signs of orbiculo-ocular venous hypertension, such as reduced eye movements, diplopia, increased intraocular pressure, retinal venous hemorrhage, optic neuropathy, and so on, are present.[2,4] Our case has applied with the signs such as recurrent eyelid edema, mild chemosis, conjunctival engorged vessels, diplopia on extreme lateral gaze, and minimal proptosis in the right eye. On further examination, intraocular pressure was measured to be as high as 32 mm Hg despite maximal antiglaucomatous medication including a prostaglandin analog and a combination of dorzolamide and timolol maleate. In such circumstances, a series of orbital and cranial MRIs were taken, and cerebral angiography was carried out.

Low-flow indirect CCFs are more common in women older than 50 years, with a ratio of 7:1 female predominance.[6] Our patient was a 62-year-old woman with no kind of trauma history. Bilateral selective angiography is the criterion standard in these patients to demonstrate the anatomy of the fistula and the extent of collateral flow from the contralateral hemisphere, which is crucial to decide the occlusion of the involved internal carotid artery (ICA). Our patient has undergone angiography that has revealed low-flow type D CCF mainly filling from the right ICA. However, immediately after angiography, signs of orbital congestion have enhanced, and on examination, severe chemosis and limitation of ocular movements have been observed, visual acuity has dropped to 20/40, and intraocular pressure was measured as 35 mm Hg with antiglaucomatous medication. There was bruit on auscultation that has not been there before angiography. Some authors have explained the worsening

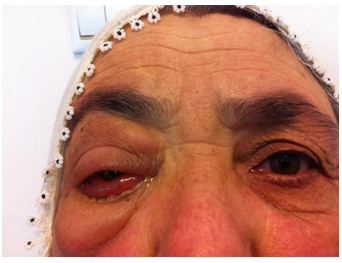

FIGURE 5. Worsening of the patient after angiography.

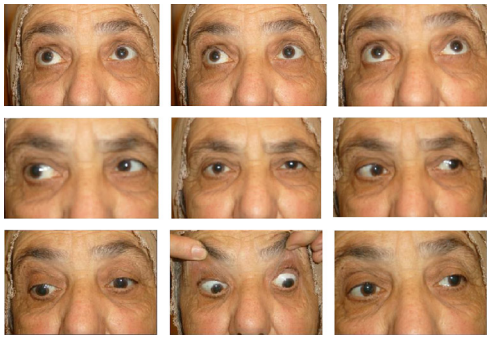

FIGURE 6. Recovery of the patient after endovascular embolization of CCF.

condition after a successful angiography as transient and due to induced stasis or local thrombosis in the region of the fistula or blame the contrast material to induce the closure of the fistula.[5–8] We have explained this situation in our case as an increase of flow in the fistula due the change of endovascular dynamics after angiography, because of that, the bruit was not present before angiography. In a week, embolization of the fistula was performed in another center, and the patient has returned to our clinic 2 weeks after this procedure.

The patients who have undergone embolization process were reported to be recovered soon after the procedure.[2] Our case was readmitted to our clinic 2 weeks after a successful embolization procedure. This procedure has enabled her to recover from all signs of orbital congestion and cured the increased intraocular pressure disorder.

In conclusion, indirect CCFs are frequently spontaneously resolving disorders, but in case of indicating ophthalmic signs, endovascular intervention should be performed. Worsening right after the angiography may be a sign of thrombosis or stasis and sometimes may be a sign of increase of flow in the fistula just like our case. This probability must be kept in mind because it may be necessary to perform the endovascular embolization earlier than planned.

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One-Stage Treatment of Acquired Facial Deformity Caused by Severe Unilateral Condylar Hyperplasia

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Abstract: Condylar hyperplasia (CH) is a rare and self-limiting process manifesting between the first and third decades of life. It causes facial asymmetry and derangement of occlusion. The management involves resection of the condylar head and orthognathic surgery followed by orthodontic treatment. We present a 33-year-old man with spontaneous onset of CH during a span of 10 years. This was managed with resection of the condyle alone, which dramatically improved facial asymmetry in addition to restoration of the occlusion within a few months of follow-up. Therefore, orthognathic surgery or orthodontic treatment was not needed.

Key Words: Condylar hyperplasia, condylar resection

Condylar hyperplasia (CH) was first described in 1836 as an overgrowth of the mandibular condyle, and no comparable pathologic abnormality has been described in any other joint.[1] It is a rare and self-limiting process; the most common age group of occurrence is between the ages of 11 and 30 years and it may manifest as an acceleration of growth in young patients, which arises at the time of physiologic development, or as a growth spurt in adults.[2–4] There are 2 types of classifications: one by Obwegeser and Makek and the other by Wolford et al. Obwegeser and Makek proposed 2 types of CH based on its radiographic and clinical characteristics: (1) hemimandibular elongation, which occurs in the horizontal plane, and (2) hemimandibular hyperplasia, which occurs in the vertical plane. Hemimandibular elongation is characterized by a lengthened condylar neck resulting in a shift of the body of the mandible. The mandible is usually skewed laterally, causing deviation of the chin and the lower dental centerline, including a posterior crossbite. Hemimandibular hyperplasia, on the other hand, is a downward bowing of the lower border of the mandible with an increased height of the ramus, a lateral open bite, but no shift in the centerline.[5] The pathogenesis of CH affecting the temporomandibular joint remains obscure, with factors ranging from a reactive growth response, trauma, Y-linked, or autosomal dominant trait. The condyle and ramus can also be affected, showing enlargement caused by hemimandibular hyperplasia and condylar tumors.[6–10] Wolford et al[11] based their classification on the frequency of occurrence, the types of jaw deformity, and the treatment best suited in the disease. In their classification, they noted that CH type 1 is the most frequently occurring form and involves an accelerated growth rate of the "normal" growth mechanism of the mandibular condyle, with relatively normal architecture of the condyle but elongation of the condylar head, neck, and mandibular body. This type, with a predominant horizontal growth vector, causes the mandible to grow forward ahead of the maxilla, creating a class III occlusal and skeletal relationship; although occasionally, a vertical growth vector may occur for CH type 1, causing mandibular prognathism. The onset of accelerated mandibular growth usually occurs during puberty, and the mandibular growth can continue into the mid 20s but is self-limiting. This mandibular overgrowth can cause major jaw and facial deformities. Type 2 occurs unilaterally and involves enlargement of the condylar head; usually, the condylar neck increases in thickness and the vertical height of the mandibular ramus and body increases on the ipsilateral side, often accompanied by a compensatory downward growth of the ipsilateral maxilla.[4] Type 2 can occur at any age and is not self-limiting and can be caused by benign tumors such as osteochondroma, osteoma, or other rare forms of condylar enlargement.

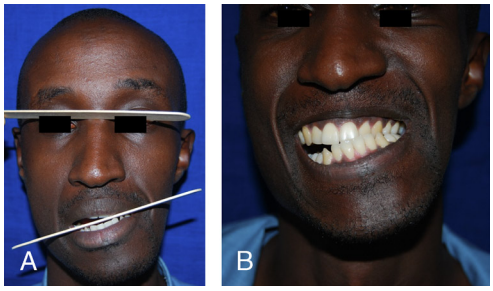

FIGURE 1. A, Severe derangement of occlusion—cant of the occlusion. B, Crossbite and RT posterior open bite.

CLINICAL REPORT

A 33-year-old man presented to the Oral and Maxillofacial Clinic at the University of Nairobi with complaints of progressive facial deformity and changes in his facial appearance for approximately 10 years. At the age of 23 years, he started to notice that his lower jaw was deviating to the left (LT). It was asymptomatic, and there was no prior history of trauma or recurrent ear infection. His medical

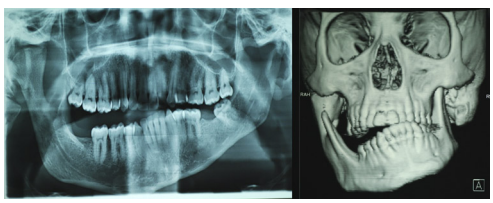

FIGURE 2. Plain radiograph–orthopantomogram and three-dimensionally reformatted image showing condylar hyperplasia and RT posterior open bite.

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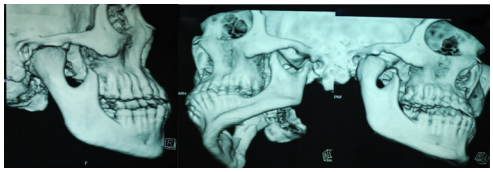

FIGURE 3. Three-dimensionally reformatted image of the RT CH and LT condylar hypoplasia.

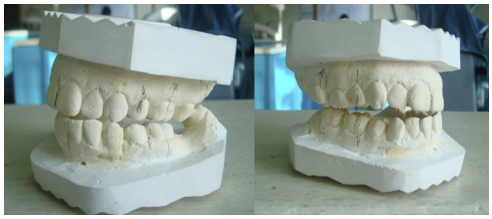

FIGURE 4. Pretreatment study models.

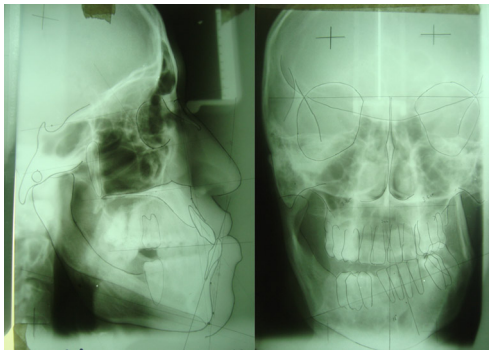

FIGURE 5. Tracing of the cephalogram and posteroanterior view.

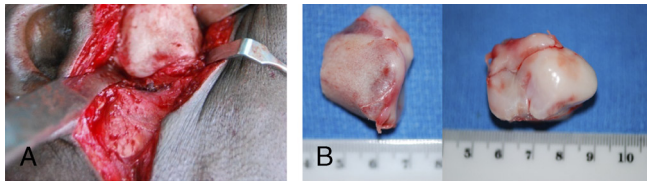

FIGURE 6. A, Right preauricular approach with temporal extension exposing the enlarged condyle. B, Resected specimens (lateral and superior view of the expanded condylar and hyperplastic cartilage).

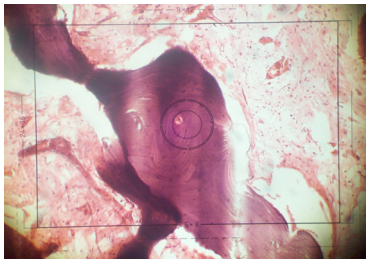

FIGURE 7. Histopathologic examination revealing an increased cellularity of undifferentiated mesenchyme layer and the cartilage shows the presence of columns and, at the border with the marrow, endochondral ossification features of juvenile growth pattern.

history was unremarkable, and there was no family history of skeletal disease or any other chronic illness. He also denied use of tobacco and alcohol. He had undergone orthodontic treatment with fixed appliances due to crowding of teeth during his teenage years. On examination, he was in a fairly good general condition. On extraoral examination, the patient had a concave facial profile with severe facial asymmetry; the chin was shifted to the LT. He had lip competence; however, the LT commissure was shifted superiorly and the right (RT) inferior (Fig. 1A). On mouth opening (35 mm), the lower jaw deviated to the LT, and the abnormal prominence of the RT condyle could be seen and palpated during its restricted movement. The swelling was hard and nontender. The RT condylar movement was restricted, and the mass was visible when the patient opened his mouth. Intraorally, there was severe derangement and canting of the occlusion. The mandibular teeth were shifted to the LT side, with a

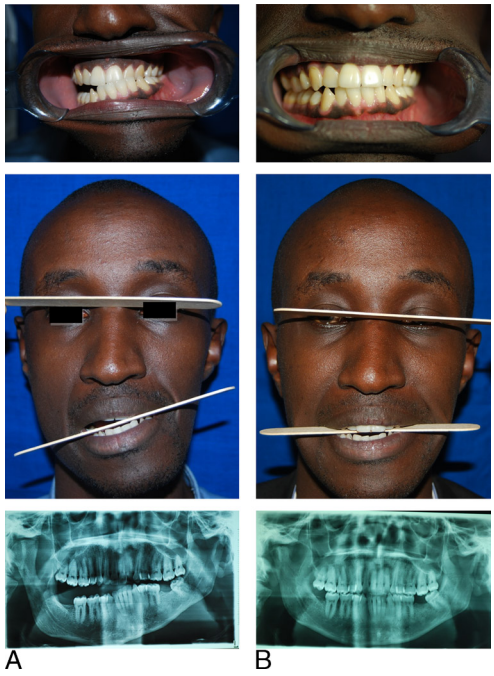

FIGURE 8. Preoperative (A) and postoperative (B) photographs and orthopantomograms illustrating the remarkable change in occlusion and the occlusal cant after condylectomy.

crossbite/reverse bite on the same side; whereas on the RT side, there was a posterior open bite. The net result of growth was a translational rotation of the mandible to the LT. The patient also had congenital LT abducent (cranial nerve VI) palsy and LT eyelid ptosis (Fig. 1B). Investigations included a panoramic view, cephalogram, computed tomographic scan (axial, coronal), and three-dimensionally reformatted images (Fig. 2). The radiographic images showed an enlarged condyle, ramus, angle, and body of the RT side of the mandible (Fig. 3). Orthognathic workup included clinical photograph, study models, and cephalometric analysis (Figs. 4 and 5). Analysis of the study models revealed prognathism anteroposteriorly at the incisor level and vertical overbite of 2 mm. On the RT side, there was a scissor bite and a crossbite on the LT posteriorly with a RT shift of the dentalmidline.

Orthognathic workup developed a problem list as follows:
The comprehensive management plan thus was staged.

Stage 1 was to undertake removal of the growth center and restore temporomandibular joint function via mechanics of the functional matrix.

Stage 2 was to reevaluate the patient and consider preorthognathic surgery, orthodontic treatment, and orthognathic surgery (LT advancement and RT side setback).

Under nasotracheal intubation, a preauricular incision was made with a temporal extension. The temporal fascia was exposed, and T-incision was made over the zygomatic arch and the head and neck of the condyle (Fig. 6). Precaution was taken to preserve the temporal branch of the facial nerve. A condylectomy was performed to remove the hyperplastic condylar head; the meniscus was spared, and the bony edges of the residual neck were smoothened. The wound was closed in layers with the Portovac drain in situ to minimize the drainage of the hematoma. Histopathologic examination confirmed that the enlargement was a hyperplastic process (Fig. 7). The patient was followed up, and in 3 months, the mandible attained acceptable symmetry and the occlusal cant reduced considerably. The patient was satisfied and desired no further treatment (Fig. 8).

This is a case of severe jaw asymmetry due to severe unilateral CH, which was managed through surgery alone. Cosmetic and functional needs were achieved to the satisfaction of the patient. The fact that no additional orthodontic and orthognathic surgeries were done saved the patient both the expenses and time. This case demonstrates the need for careful consideration of all the options that can be offered to patients with these types of problems.

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Usefulness of Vascularized Galeal Frontalis Myofascial Flap as Treatment for Postoperative Infection in Frontal Sinus Fracture

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Abstract: Inappropriate treatments of frontal sinus fractures may lead to serious complications, such as mucopyocele, meningitis, and brain abscess. Assessment of nasofrontal duct injury is crucial, and nasofrontal duct injury necessitates sinus obliteration. For sinus obliteration, autogenous fat, muscle, bone, pericranium grafts, and alloplastic materials, such as Surgicel, hydroxyapatite (Bone Source), and methyl methacrylate, are commonly used. However, autogenous tissue grafts and alloplastic materials cannot prevent infection, which is the leading cause of complications of frontal sinus surgery. A good vascular supply is the mainstay of resistance against infection.

A 21-year-old man had nasofrontal duct injury and frontal sinus fracture by motorcycle accident. He underwent cranialization and frontal sinus obliteration with autologous bone graft and hydroxyapatite. On 16 days postoperatively, the patient had a high temperature with wound infection in the glabella. Thus, he was treated with sinus obliteration with vascularized galeal frontalis myofascial flap. He was followed up postoperatively for 1 year without infection.

The authors experienced a postoperative infection in frontal sinus fracture, which was treated with vascularized galeal frontalis

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myofascial flap and achieved satisfactory result; thus, we presented our case with reviews of the literature.

Key Words: Frontal sinus, infection, sinus obliteration, galeal frontalis myofascial flap

The complications that can occur after frontal sinus fractures are chronic frontal headaches, sinus infections, and contour deformities of the forehead, and inappropriate treatments of frontal sinus fractures may lead to serious complications, such as mucopyocele, meningitis, and brain abscess.

In case of frontal sinus fracture with nasofrontal duct injury, the technique in current use is the removal of the frontal sinus mucosa with sinus obliteration using autogenous tissues such as fat, bone, or alloplastic materials, such as Surgicel or hydroxyapatite. However, there is a higher complication rate because of no vascularity.[1] To obliterate the frontal sinus, the vascularized flaps are known to be the best method, followed by autologous bone, other autologous tissues, and alloplastic materials.[2]

If there is contaminated frontal sinus fracture, concern about ascending infection from the nasopharynx, or postoperative frontal sinus infection, frontal sinus obliteration with well-vascularized autologous graft is needed to prevent a recurrence of the infection.

The authors experienced a postoperative infection in frontal sinus fracture, which was treated with vascularized galeal frontalis myofascial flap and achieved satisfactory result; thus, we presented our case with reviews of the literature.

CLINICAL REPORT

A 21-year-old man presented with frontal sinus fracture with nasofrontal duct injury and multiple facial fractures by motorcycle accident (Fig. 1). He had cerebrospinal fluid leakage and got conservative care. On 10 days after the injury, he underwent cranialization and frontal sinus obliteration with autologous bone graft and hydroxyapatite.

After exposure of the site with multiple facial bone fractures by bilateral temporal, subciliary, and gingivobuccal incisions, open reduction and internal fixation was done with absorbable and nonabsorbable fixators. To correct the frontal sinus fracture site, cranialization was done by craniectomy of the anterior wall of the frontal sinus, removal of the fractured posterior table carefully not to damage the dura mater, and removal of the frontal sinus mucosa totally. The nasofrontal duct was obstructed with the bone fragment removed from the posterior wall of the frontal sinus; the remnant dead space was filled with the bone fragment removed from the posterior wall of the frontal sinus and with fibrin glue. The anterior wall of the frontal sinus was covered and fixed with absorbable plates, and to correct the depression of the frontal sinus, cranioplasty using hydroxyapatite was done. On 16 days postoperatively, the patient had a high temperature with pus discharge from the operative wound on the frontal area, and the *Klebsiella pneumoniae* was cultured from the wound. We did multiple irrigations and treated him with antibiotics, but postoperative infection was not controlled. The authors thought that the cause of infection was foreign materials. He underwent reoperation to remove hydroxyapatite and absorbable plates. After debridement of the cause of infection (Fig. 2), the nasofrontal duct was occluded with a pericranium graft. Then, reconstruction of the frontal sinus was done by sinus obliteration using vascularized galeal frontalis myofascial flap (Fig. 3). There was no evidence of recurrence or any serious complications.

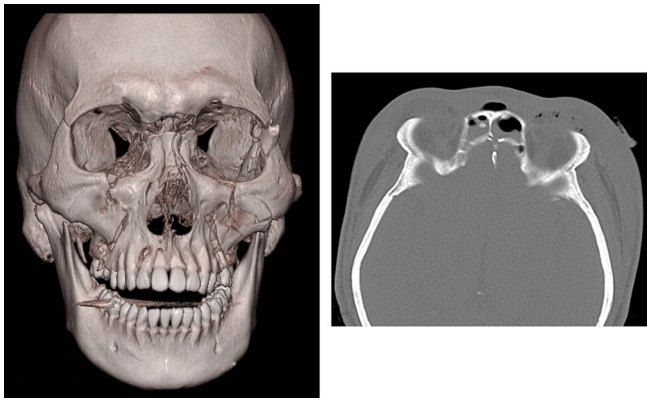

FIGURE 1. Facial bone three-dimensional computed tomographic scan (left) and an axial view showing the nasofrontal duct injury and the frontal sinus fracture (right).

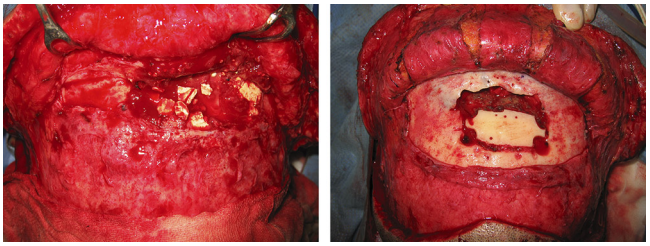

FIGURE 2. Infected bone graft and Bone Source after cranialization (left) and after debridement of the infected bone graft, Bone Source, and unhealthy bone (right).

DISCUSSION

Frontal sinus fractures constitute 5% to 15% of maxillofacial fractures, with motor vehicle accidents being the most common. Approximately 75% of the cases have associated orbital, nasal, and/or other midfacial fractures. In addition, approximately 20% of patients are unconscious at the time of their initial evaluation, and 20% have cerebrospinal fluid leaks.

The complications that can occur after frontal sinus fractures are chronic frontal headaches, sinus infections, and contour deformities of the forehead, and inappropriate treatment of frontal sinus fractures may lead to serious complications, such as mucopyocele, meningitis, and brain abscess. Most of the complications are caused by inadequate restoration of the barrier between upper airway and intracranial contents.[1]

The surgical plan should be based on the presence of fracture of the anterior and/or posterior wall, damage of the dura mater, and

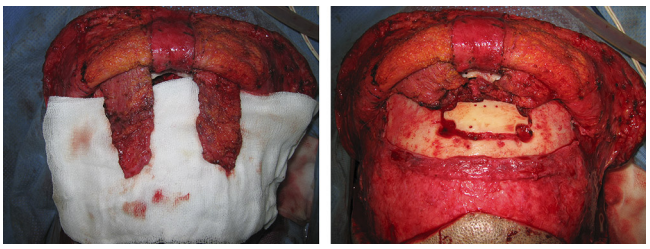

FIGURE 3. Intraoperative view. Elevation of the bilateral galeal frontalis myofascial flap (left) and insetting of the bilateral galeal frontalis myofascial flap (right).

injury of the nasofrontal duct. Assessment of nasofrontal duct injury is crucial, and intraoperative exploration is necessary.

The diagnosis of nasofrontal duct injury necessitates sinus obliteration. Sinus mucosa is removed, and the nasofrontal duct is plugged with a pericranium graft or allogenous bone graft. After that, the frontal sinus is obliterated with autogenous tissues or foreign plastic materials. For sinus obliteration, there are several graft options. Some of the most commonly used ones are autogenous fat, dermal fat, muscle, cartilage, bone, pericranium grafts, and alloplastic materials such as Surgicel, silicone, hydroxyapatite, methyl methacrylate, and bone wax. Another method of sinus obliteration is osteoneogenesis.[1] Among the various graft options, which one is used to obliterate the frontal sinus is important in treatment and associated with postoperative complications.

Mickel et al[3,4] did frontal sinus obliteration using autologous fat, muscle, and bone and spontaneous osteogenesis in an uninfected feline frontal sinus model, and all methods studied were effective without complications. They said that if frontal sinus obliteration was done using a strictly standardized operative technique, including complete removal of sinus mucosa using loupe magnification, removal of inner bony cortex, and total occlusion of nasofrontal duct, all methods studied would be effective in sinus obliteration. However, there were donor-site morbidities when autologous tissue transplantation was used. Thus, they said that spontaneous osteogenesis may be the best choice for frontal sinus obliteration. However, there is limitation for this study because cat frontal sinus is quite small than human frontal sinus and the cat frontal sinus was not fractured, so it is different from clinical situations. Donald and Ettin[5] did frontal sinus obliteration using autologous fat in cats in which all of either the anterior or the posterior frontal sinus wall was removed. They reported that the rate of postoperative complications such as mucocele, mucopyocele, and infections is 44%.

Wolfe and Johnson[6] reported frontal sinus obliteration using cancellous iliac bone in the management of the late complications of mucocele and pyocele, and there were no complications such as infections. They said that there is faster osteogenesis of cancellous iliac bone, so there were no complications. However, none of the autogenous obliteration methods previously mentioned provide vascularity to the sinus. In addition, these autogenous obliterations and the methods using alloplastic materials take time to engraftment, and tissue absorption happened. Moreover, the alloplastic materials cannot prevent postoperative infection. Because of these reasons, a vascularized flap is needed to obliterate the frontal sinus.

Xie at al[2] said that reconstruction with obstruction of the nasofrontal duct by vascularized tissue was found to be the best treatment, followed by osteogenesis, other autologous tissue graft, and alloplastic alternative.

A pericranial flap and galeal frontalis myofascial flap are the vascularized tissue grafts in frontal sinus obliterations. The pericranial flap is a composite flap consisting of the pericranium, and the overlying loose areolar tissue receives rich blood supply from the supraorbital, supratrochlear, occipital, posterior auricular, and the superficial temporal arteries.

Parhiscar and Har-el[7] reported good result in frontal sinus obliteration with the pericranial flap in 10 patients with frontal sinus mucocele, mucopyocele, frontal sinus osteomyelitis, and frontal sinus fracture. Thaller and Donald[8] did frontal sinus obliterations with the pericranial flap without complications in 14 patients with frontal sinus fracture.

The pericranial flap has advantages such as reducing the infection risk, no morbidities of donor site, and no additional incisions. However, there is excessive bleeding when dissection and thickness of the flap is thin. A galeal frontalis myofascial flap we used is also a vascularized flap like the pericranial flap and can be generally used for reconstruction of frontal sinus fractures or anterior cranial base fractures, treatment of infectious complications, and treatment of chronic illness of frontal sinus.

The galeal frontalis myofascial flap is elevated from the uninjured site; the galeal aponeurosis is outlined on the previously dissected scalp flap based on the supratrochlear artery, the supraorbital artery, or both. Dissection proceeds in a cranial-to-caudal direction, and the frontalis muscle is included in the flap. The base of the flap requires careful dissection to avoid injury to the supraorbital and supratrochlear vessels where they exit the frontal bone. The flap is then rotated into the sinus and secured using absorbable sutures or fibrin glue. If 1 flap is not enough to obliterate the sinus or when both sinuses are injured, 2 flaps are used. Recently, fibrin glue has also been used in fontal sinus obliteration. First, fibrin glue helps to fix the pericranial patch in the nasofrontal duct without any difficulty. Second, fibrin glue can also be used for fixation of the galeal frontalis flap. Third, fibrin glue fills any residual space between the galeal frontalis and the sinus. Fourth, fibrin glue also provides hemostasis for the remaining walls of the frontal sinus.

In our case, the patient presented with frontal sinus posterior wall fracture with nasofrontal duct injury, and he underwent cranialization and frontal sinus obliteration with autologous bone graft and hydroxyapatite, but postoperative infection was developed. Thus, he was treated with sinus obliteration with vascularized galeal frontalis myofascial flap. One flap was not enough to obliterate the sinus, so 2 flaps were used. There was no evidence of recurrence or other complications. Severely contaminated wounds, comminuted posterior wall fractures, or postoperative infections are better treated with the removal of remnant sinus contents with infected bone and sinus obliteration with the galeal frontalis flap or cranialization.[9]

There are several advantages of galeal frontalis myofascial flap. First, a good vascular supply is the mainstay of resistance against infection. Second, the volume of the flap is enough to fill the dead space of the frontal sinus. Third, the diameter of the flap is narrow, so rotation of the flap is easy. Fourth, additional operation time is short and can avoid additional complications to harvest donor site. Fifth, there is cost reduction using autologous graft. Sixth, when a contralateral galeal frontalis flap is used for sinus obliteration, the bone graft that is placed to reconstruct the anterior wall is supported by the overlying vascularized periosteum and the underlying vascular galeal flap. This is important for successful bone graft survival.[1]

There are also disadvantages of galeal frontalis myofascial flap. First, transient sensory loss covering the frontal region can occur. Second, contour deformity of the donor area can occur. Third, functional impairment of the frontalis muscle can occur. Fourth, if severely contaminated wound involves bilateral orbital and/or supratrochlear arteries, the galeal frontalis flap cannot be elevated. However, transient sensory loss over the frontal region lasting from 4 to 12 months, with an average duration of 6 months, can occur, and little or no significant contour deformity has been seen.

In our case, the patient had mild sensory loss of the frontal region and mild functional impairment of frontalis muscle, but he did not complain about these problems. In addition, there was mild depressed deformity on his forehead; thus, he underwent autologous fat graft, and the deformity was corrected. The authors experienced a postoperative infection in frontal sinus fracture, which was treated with vascularized galeal frontalis myofascial flap and achieved satisfactory result; thus, we presented our case with reviews of the literature.

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What Is the Importance of Radiology in Obstructive Sleep Apnea?

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Abstract: Lipomas are rare, and they rarely arise in deep soft tissue. Only few cases associated with obstructive sleep apnea (OSA) have been reported. Because of radiologic examinations (computed tomography and magnetic resonance imaging) in a case with OSA, a rarely observed large lipoma was encountered in the retropharyngeal region. In the case that was followed, the apnea-hypopnea index was measured. The radiologic examination of the upper airway of the case with OSA symptoms was presented.

Key Words: Retropharyngeal, lipoma, obstructive sleep apnea

Lipomas of the retropharyngeal region are very rare benign tumors often causing unspecific clinical symptoms.[1] Because of their slow growth rate, most lipomas reach large sizes without being noticed. The initial symptoms are most often related to the upper aerodigestive tract. The locations, where lesions develop, cause dysphagia and respiratory problems. A case of lipoma causing the OSA syndrome is discussed.

CLINICAL REPORT

A 44-year-old female patient applied to the otorhinolaryngology department, with complaints of snoring, dysphagia, and daytime sleepiness. The Epworth Sleepiness Scale was used to measure the general daytime sleepiness of the patient, and the patient's score was determined as 20 (sitting and reading, 3; watching television, 3; sitting inactively in a public place, 3; as a passenger in a car for an hour without a break, 3; lying down to rest in the afternoon when circumstances permit, 3; sitting and talking to someone, 1; sitting quietly after a lunch without alcohol, 3; and in a car, while stopped for a few minutes in traffic, 1).

The patient had had these complaints for the last 1 year, and the symptoms had become far more evident for the last several months. The nonsmoker patient described apnea during sleep.

Apart from pulsion on the median line on the right tonsil, no additional pathologic finding was observed during the otorhinolaryngologic examination. In the flexible nasopharyngolaryngoscopic examination, it was observed that the mass, which settled in the right nasopharynx, led to distortion in the supraglottis and constricted the airway. A lobule-contoured and low-density mass with dimensions of around 5 × 4 cm was observed in the CT examination. The well-defined, homogeneous, and low-density right retropharyngeal lesion on CT examination is observed in Figures 1A and B. The mass extended downward and showed close proximity to cervical vertebrae. The density characteristic was compatible with the lipomatous tissue.

In the magnetic resonance imaging (MRI), a lesion was detected in the high-intensity fat-suppressed sequences in the same localization in T1 and T2 images. A complete signal loss in the short T1 inversion recovery sequence was the evidence of lipomatous origin. Adipose tissue is observed with high signals in T1 and T2 images. The high signal from fat is suppressed in the short T1 inversion recovery sequence, thereby enabling to distinguish from fluid-filled structures (Figs. 2A, B and 3). When all these findings are evaluated, it is proved that the retropharyngeal lesion is of lipomatous origin.

First, the fine-needle aspiration biopsy and then total excision of the mass were recommended to the patient; however, the patient did not accept any interventional procedures because of the high morbidity of the operation. In the polysomnographic findings of the

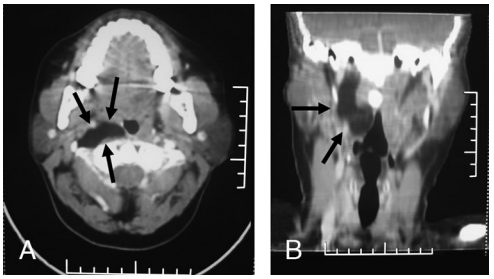

FIGURE 1. The lipoma on axial (A) and coronal (B) reformatted CT scans in 2009 (arrows).

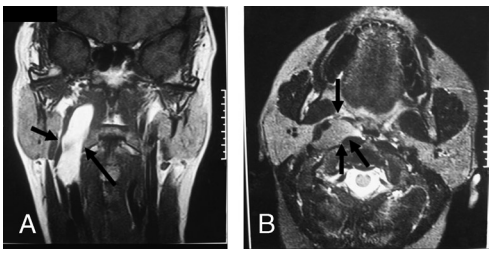

FIGURE 2. The retropharyngeal lipoma on coronal T1 MRI image (A) and axial fat-suppressed image (B) in 2009 (arrows).

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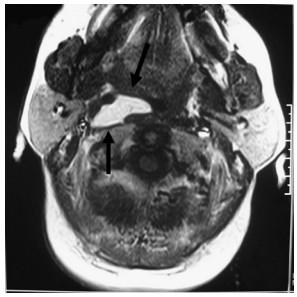

FIGURE 3. The lipoma on axial T1 MRI image in 2010 (arrows).

patient that were recorded for 1 night, the apnea-hypopnea index (maximum apnea time, 38 seconds; hypopnea time, 55 seconds; microarousal index, 167/h), the mean oxygen saturation (Sa_{O_2}), the lowest Sa_{O_2}, and the time of sleep with Sa_{O_2} were measured as 20, 97%, 90%, and less than 90%, respectively.

The patient was clinically observed at regular intervals, and no variation in the size or characteristic of the mass was observed in the radiologic examination that was performed some 24 months later. The case was presented with literature findings.

DISCUSSION

Retropharyngeal lipomas are quite rare, and some 13% of all lipomas are observed in the head and neck region.[2] Before retropharyngeal lipomas reach large dimensions and are diagnosed, they manifest themselves with symptoms, such as dyspnea, snoring, and obstructive sleep apnea (OSA).[1–3] Obstructive sleep apnea is a disease that occurs with the functional stenosis of the pharynx and leads to sleep disorder. Some anatomic, pathologic lesions, such as palate hypertrophy and uvular or tongue hypertrophy, are found in 80% of the cases with OSA. Multiple anatomic abnormalities and neuromuscular dysfunction lead to airway obstruction. Tumors of the retropharyngeal space are quite rare, and they seldom cause OSA.[4] In this case, the severe obstruction of the high retropalatal airway is the real cause of OSA.

Lipomas are tumors of mesenchymal origin. These lesions, often derived from subcutaneous localization, are thinly encapsulated and do not contain cellular atypia.[5] Their subtypes can be listed as follows: well differentiated, myxoid, round cell, dedifferentiated, and pleomorphic.

In its diagnosis, fine-needle aspiration biopsy might be applied to the selected cases. Nevertheless, it should be known that it is rather difficult to distinguish from well-differentiated liposarcomas.[5] Therefore, the importance of imaging methods for diagnosis further increases.

In lipoma cases, clinical findings might be attributed to other reasons because the mass grows slowly. The sleep apnea of our case was a 1-year complaint, and its diagnosis was delayed, for no imaging method had been applied. The aim of this presentation was to emphasize the importance of diagnostic radiology in the neck potential space. The radiology of soft tissue and fascial planes is rather complex. Especially in those regions where biopsy cannot be applied, it is of great importance to distinguish the normal tissue from the pathologic lesion. Our case stresses the importance of radiology to determine the tissue character and the place of radiology in the final diagnosis.

If the surgical risk is below comorbidity in cases with retropharyngeal lipoma, surgical treatment might be applied. Because surgical comorbidity was high in our case, treatment was not applied. However, the use of continuous positive airway pressure was considered appropriate at a pressure of 6.5 mbar. In the 24-month follow-up, it was found that the mass size had not increased. Malign transformation is low in lipomas, and a rapid change in the symptoms and hypertrophy during follow-up should raise suspicion for malign transformation.

We presented the case with obvious OSA that was caused by retropharyngeal lipoma. Together with our case, some 8 cases causing OSA were presented in the literature (Table 1).[4–9]

It has been reported that, if the radiologic diagnosis is compatible with lipoma in the lipomatous lesions of the retropharyngeal region, clinical and radiologic follow-up might be adequate in patients who reject an operation or are not suitable for an operation because the surgical excision of the mass in this region is difficult, the lipoma does not have any metastatic potential, and the malign transformation is debatable.[8]

Computed tomography is an examination that is easy to apply for head and neck masses. Contrast enhancement patterns enable to distinguish malign masses. However, it is difficult to distinguish inflammation. In the soft tissue, MRI provides more information than CT.

In conclusion, in cases that are detected to have OSA, it is useful to radiologically examine the upper airway in detail. Radiologic close follow-up is necessary in cases from which it is impossible to obtain histopathologic specimens.

TABLE 1. The Cases of Lipomas in the Literature With OSA Symptoms

Case No.	Sex	Age, y	Diagnosis	Follow-Up, y	Operation	Size, cm	References
1	M	36	CT	—	Yes	8	Aland[6]
2	M	56	—	—	Yes	?	Di Girolamo et al[7]
3	M	64	FNAB-CT	5	No	?	Hockstein et al[5]
4	F	73	CT	—	Yes	5	Piccin and Sorrenti[4]
5	M	40	CT	—	Yes	11.7	Namyslowski et al[1]
6	M	75	FNAB-CT, MRI	2	No	6	McNeill et al[10]
7	M	75	CT	2	No	12.5	Lakadamyali et al[8]
8	F	11					Gong et al[9]
9	F	50	CT-MRI	2	No	5	Present case

FNAB indicates fine-needle aspiration biopsy.

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Intradiploic Epidermoid Cyst of the Temporal Bone: Is It the Same as or Different From Cholesteatoma?

Dong-Hee Lee, MD, PhD

Abstract: Intradiploic epidermoid cyst is a benign lesion that is derived from the ectodermal cells of the cranium. It is a rare tumor, representing less than 1% of all primary intracranial tumors. It can be located in any part of the skull, but frontal and parietal bones are the most common sites for the cysts. It occurs from the first to the seventh decade of life. Because it is a slowly growing tumor, it is usually discovered incidentally and may remain asymptomatic for many years. Rarely, it grows intracranially to produce brain compression or undergo malignant change. For treatment, it is important to completely remove the capsule of the cysts to avoid recurrence. I describe a 25-year-old man with an intradiploic epidermoid cyst of the temporal bone, who underwent tumor removal and simple mastoidectomy. I also describe the unique radiologic findings of my case, which differed from those of cholesteatoma.

Key Words: Epidermoid cyst, mastoid, temporal bone, mastoidectomy

Intradiploic epidermoid cyst accounts for less than 1% of primary intracranial tumors. It can be located in any part of the skull, but the most common sites are the frontal, parietal, and occipital regions. It occurs from the first to the seventh decade of life. It is usually discovered incidentally and may remain asymptomatic for many years.[1–3]

Among the intradiploic epidermoid cysts of the skull, that of the temporal bone is rare, with about 20 cases having been reported worldwide in the English-language literature.[4–8] Most of the reports state that it is different from cholesteatoma of the middle ear, which is a kind of epidermoid (inclusion) cyst. Clark et al[9] suggest, however, that intradiploic epidermoid cyst and congenital cholesteatoma are the same disease entity because they are indistinguishable histopathologically as well as radiologically. They also suggest that clarifying the nomenclature would help to avoid confusion when the subject is researched in the literature.

Recently, I had a case involving an epidermoid cyst in the temporal bone in a young male adult, which was accompanied with central perforation of tympanic membrane (TM). Contrary to the report of Clark et al,[9] the radiologic and macroscopic findings of this case were different from those of cholesteatoma. In this case report, I describe the clinical, radiologic, and histopathologic aspects of my case and state the differences from cholesteatoma. This case is unique because the epidermoid cyst here showed 2 radiologic differentiations from cholesteatoma, and these aspects can be helpful for clinicians.

CLINICAL REPORT

A 25-year-old man was transferred to our hospital from a primary clinic; his chief complaint was right progressive hearing loss for

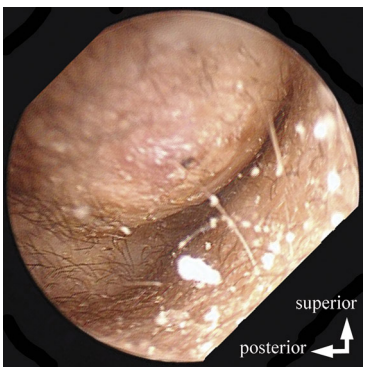

FIGURE 1. Otoscopic images of right EAC. Bulging mass obstructed the EAC and made it impossible to examine the TM preoperatively.

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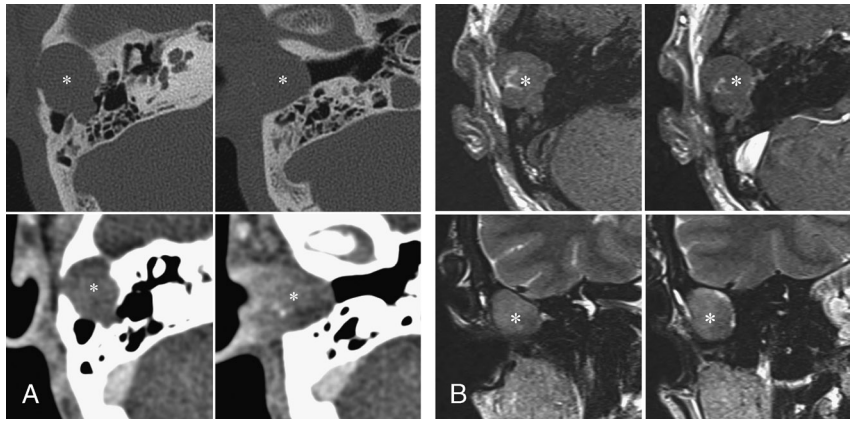

FIGURE 2. A, Computed tomographic images of right temporal bone before (upper 2 panels) and after contrast enhancement (lower 2 panels). A huge mass of soft tissue density (asterisk) is located at the entrance of EAC and destructed surrounding squama as well as mastoid parts of right temporal bone. B, This mass is isosignal on T1- (left upper panel) and T2-weighted (lower 2 panels) images and not enhanced by gadolinium–diethylenetriamine pentaacetic acid (right upper panel).

several years. Seven or 8 years ago, he underwent middle ear surgery on both ears at another hospital, but he did not remember the exact methods and type of surgery. He went through closed reduction for nasal bone fracture 8 years ago. He did not have any other medical or surgical history. After a retrospective review of the physical examination and computed tomography (CT), I found that he had received myringoplasty or tympanoplasty (Farrior type I) through the postauricular approach using tragal perichondrium on both ears.

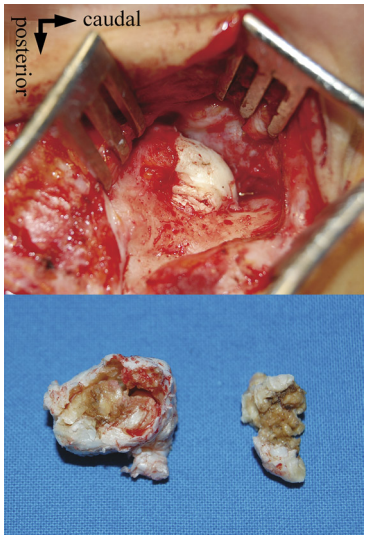

FIGURE 3. Photographs of the surgical field and the cystic mass. After the periosteum of the mastoid was elevated, a lightly colored mass was located where Macewen suprameatal triangle was (upper panel). The mass was well encapsulated and filled with yellow greasy and pearl-like whitish materials, which looked differently from cholesteatoma matrix (lower panel).

On the physical examination, his left auricle, external auditory canal (EAC), and TM were normal. However, a huge, firm, soft tissue mass covered by normal skin was found at the entrance of the EAC. It looked like it arose from the posterosuperior portion of the cartilaginous EAC. His right TM was not visible because the mass obstructed the EAC completely (Fig. 1).

Temporal bone CT showed that a 1.7-cm, ovoid, nonenhancing mass of soft tissue density occupied all cartilaginous EAC and the lateral half of bony EAC. It showed that the squama and mastoid parts of the temporal bone were destroyed by pressure erosion on adjacent bone (Fig. 2A). On coronal images of CT, the mass looked like it was not connected to the brain because thin bony walls were found between a mass and intracranial cavity.

On temporal magnetic resonance images (MRI), a 1.7-cm, ovoid mass showed isosignal intensity on T1- and T2-weighted images and was not enhanced by gadolinium–diethylenetriamine pentaacetic acid. Magnetic resonance image showed that its margins were delineated very well and that there was no connection or involvement of the brain parenchyma (Fig. 2B).

Resection of the tumor and simple mastoidectomy was planned and undertaken under general anesthesia. I used the postauricular approach. After elevation of the mastoid periosteal flap, a huge, lightly colored, firm mass was found at the location of Macewen suprameatal triangle (Fig. 3). With care taken so as not to tear the capsule of the mass, the mastoid cortex around the mass was drilled out carefully using diamond burrs. After the mass was removed completely without its rupture, I found that the mass destroyed some mastoid air cells and a portion of bony EAC, which were in direct contact with the mass. However, the air cells that were far from the mass looked normal. I also found that the tegmen mastoideum and the mastoid antrum were not involved. After complete removal of the mass and vigorous irrigation of the surgical wound, the defect of mastoid cortical bone and bony EAC was reconstructed using bone cement. At the last stage of the operation, a central perforation was found on his left TM, which was not found preoperatively because of the mass obstructing the EAC. Myringoplasty was performed using the superficial temporal fascia in underlay grafting fashion.

The firm, cystic mass was well encapsulated by a thin wall. Its content was mixed with yellow greasy material and pearl-like keratin

material. Histopathologic findings showed that the cystic structure was lined by squamous epithelium containing laminated keratin material. The pathologic findings were consistent with the diagnosis of epidermoid cyst.

DISCUSSION

Epidermoid cyst is an uncommon, benign, and slow-growing lesion, and it develops from ectopic ectodermal tissue. It can be congenital or acquired. The former relates to the implantation of the ectoderm at the time of closure of the epithelial fusion lines. The latter is usually caused by posttraumatic inclusion of the epithelium.[1,10] Arana et al[3] reported that 24% of epidermoid cysts are associated with trauma history. My patient had a history of middle ear surgery on the same side. A typical myringoplasty or tympanoplasty via the postauricular approach includes meatal incision, which in my case was undertaken where the epidermoid cyst was located. Therefore, I supposed that in my case there might be a correlation between mechanical trauma on the EAC skin during myringoplasty or tympanoplasty and the origin of the lesion.

The differential diagnosis of epidermoid cyst in this area includes cholesteatoma, dermoid cyst, hemangioma, hydatid cyst, eosinophilic granuloma, aneurysmal bone cyst, giant cell reparative granuloma, meningioma, hemangioma, facial neurilemoma, and acoustic neuroma. Among these, cholesteatoma is the most difficult disease to differentiate clinically, radiologically, and histopathologically from epidermoid cyst, because the 2 diseases have similar CT findings (mass of soft tissue density, pressure erosion on adjacent bone, and no contrast enhancement), similar MRI findings (low signal intensity in T1-weighted image, high signal intensity in T2-weighted image, and no enhancement), and similar histopathologic findings (a cyst lined by squamous epithelium and containing laminated keratin material).[4–7,11,12] Considering that the 2 diseases share the same pathogenesis, this is not surprising. For that reason, some authors suggest that epidermoid cyst and cholesteatoma are one and the same disease entity.[9]

However, I arrive at 2 different findings through a retrospective review of CT and MRI. The difference in CT findings is that epidermoid cyst has a rounder, well-demarcated contour, and its boundary between adjacent bones is clearer and smooth. Cholesteatoma also mostly shows a sharply marginated, expansile mass, but its margin is usually irregular and scalloped and accompanied by varying degrees of bony erosion. This arises because cholesteatoma tends to be more invasive into the surrounding bones and air cells. In addition, cholesteatoma destroys the bone through osteoclastic bone resorption and chronic inflammation, rather than through pressure necrosis.[13] In contrast, epidermoid cyst has a well-marginated, round mass with smooth margin. Because it erodes adjacent bones through pressure necrosis, the borderline between cyst and bone tends to be smooth and regular, and the degree of bony erosion seems to be uniform.

The difference in MRI findings is the higher frequency of high signal intensity on T2-weighted images in cholesteatoma. Warren et al[13] reported that all of 9 cases diagnosed with congenital mastoid cholesteatoma showed isosignal or slightly high signal intensity on T1-weighted images and high signal intensity on T2-weighted images. However, my case showed isosignal intensity on T1- and T2-weighted images. This coincides with the report of Hong et al[14] that subcutaneous epidermal cyst shows variable low-signal intensity on T2-weighted images. More frequent inflammation and the resultant increase of water content within a cyst may be why the high signal intensity of T2-weighted images is more consistent in cholesteatoma than in epidermoid cyst.

Complete surgical excision of epidermoid cyst is the treatment of choice. It is very important to remove it with whole cystic walls because the remnant cystic wall always results in a recurrence. Its recurrence rate has been reported at 8.3% to 25%.[3,7] Because large intradiploic epidermoid cyst of the temporal bone can often involve the intracranial cavity, careful dissection of the cyst from the surrounding skull bone and dura mater is important to prevent postoperative intracranial complication.

Abscess formation, hemorrhage, and malignant transformation have been reported as complications of epidermoid cyst.[3,5,8,15] Repeated incomplete resection and frequent episodes of infection within a tumor have been especially known to increase the risk of malignant transformation.[3] Newly identified contrast enhancement or abrupt increase of the tumor size strongly indicates malignant change in epidermoid cysts.[15]

As stated as above, I described my case of epidermoid cyst in the temporal bone and stated 2 radiologic differences of epidermoid cyst from cholesteatoma. Readers should keep in mind that these differences are not absolute. However, it is to be hoped that these radiologic differences can be helpful for surgeons.

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Intraoral Sublabial Approach for Anterior Skull Base Juvenile Ossifying Fibroma

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Abstract: Juvenile ossifying fibroma is a rare fibro-osseous neoplasm seen in the pediatric age group of 5 to 15 years. It has been recognized as a separate histopathologic entity among the fibro-osseous group of lesions. Occasionally, it may grow aggressively and extend to involve the orbits and skull base, resulting in serious cosmetic and functional problems. We present a case of a juvenile ossifying fibroma arising below the skull base of a 9-year-old boy. This case demonstrates that cemento-ossifying fibroma in case of maxilla grow to considerable size before causing symptoms because of the remodeling of the surrounding bony compartment and intraoral sublabial approach can be used to excise lesion completely even at the skull base.

Key Words: Juvenile ossifying fibroma, sublabial incision, skull base, orbit

Ossifying fibroma is an uncommon benign fibro osseous tumor of the craniofacial bones. One of its forms, described in children and adolescents, is aggressive (juvenile) ossifying fibroma. Most commonly, it occurs in children younger than 15 years and involves paranasal sinuses and the orbit. Conventional ossifying fibroma occurs in the tooth-bearing areas of the jaws and may be odontogenic in origin in that it arises from tissues of the periodontal membrane. The juvenile variants arise outside the tooth-bearing areas, either in the jaws or in the craniofacial bones in younger people, and have a tendency toward aggressive behavior. It should be noted that this aggressive behavior is probably more related to anatomic site than to any intrinsic biologic properties. Lesions can grow and permeate relatively unrestricted through the paranasal bones and sinuses, resulting in large lesions that are very difficult to remove surgically.

We report a case of Juvenile ossifying fibroma of the left anterior skull base, which was excised through an intraoral sublabial approach.

CLINICAL REPORT

A 9-year-old boy was referred to our head and neck unit with gradual left eye proptosis of 2 months in duration and fullness of cheek of 1 month in duration. It was also associated with diplopia. There was no history of epistaxis or nasal obstruction. On examination, there was proptosis of the left eye with fullness of the left cheek. Extraocular muscle movements were normal. There was no other significant history. The computed tomographic scan of the paranasal sinuses revealed a nonenhancing expansile, lobulated heterogeneous mass with multiple internal calcifications arising from the left maxilla. It measured 3.79 × 2.76 cm. It was extending to the orbit and retro-orbital region posterioly causing proptosis of the left eye (Fig. 1). Hence, radiologic diagnosis of ossifying fibroma was made, which was confirmed on histopathology. The lesion was approached intraorally by a sublabial incision starting from the frenulum to the last molar (Fig. 2). Partial resection of the anterolateral wall of left maxilla was done to allow access to the anterior skull base. The tumor was resected completely by this approach (Fig. 3). Three years of his postoperative follow-up did not reveal any recurrent lesion.

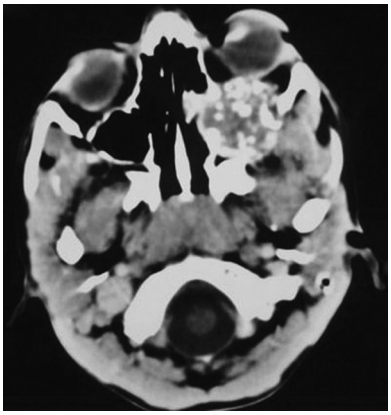

FIGURE 1. Computed tomographic scan of the paranasal sinuses (axial view) showing a nonenhancing expansile, lobulated heterogeneous mass with multiple internal calcifications. The mass has extended to the orbit, causing proptosis.

DISCUSSION

Cemento-ossifying fibroma belongs to the fibro-osseous lesions according to the 1992 World Health Organization classification.[1] Also, according to the World Health Organization's classification, a lesion with aggressive growth in young patients younger

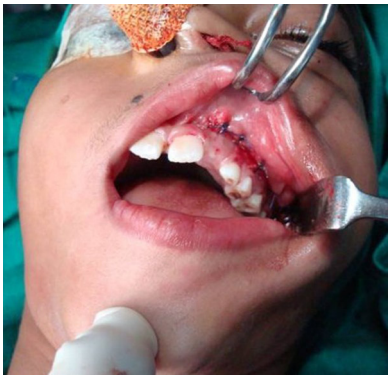

FIGURE 2. Clinical photograph showing closure of the sublabial incision used for the excision of the tumor.

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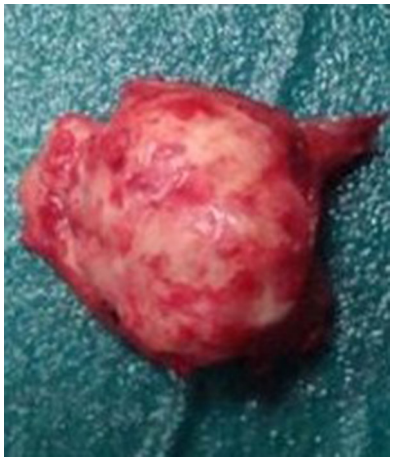

FIGURE 3. Clinical photograph of specimen showing excision in toto of the juvenile ossifying fibroma.

than 15 years is known as juvenile ossifying fibroma. It is distinguished from the conventional ossifying fibroma by early age of onset (5–15 y) and aggressive behavior. Bony pattern and high chance of recurrence also form features of the juvenile ossifying fibroma. Nasal obstruction, exophthalmos, and, rarely, intracranial extension can be associated with those lesions arising within the paranasal sinuses and orbit.[3] The aggressive behavior of the juvenile ossifying fibroma seems to be related to the site of occurrence rather to the biologic behavior of the lesion. The ossifying fibromas in the maxillary and frontoethmoidal areas appear to be large and aggressive because of the unimpeded growth in these sites.[3] In fact, no definitive predictor variable could be found on histopathology, which can determine their aggressive behavior or their tendency to recur.[4] These lesions usually arise in the paranasal sinuses and extend to involve the surrounding structures.[5]

When the orbital bones and paranasal sinuses are involved, an exophthalmos or bulbar displacement may be observed.[3,6] In our case, the lesion had extended to the orbit and retro-orbital lesion causing exophthalmos (Fig. 2). The radiologic features are variable. Depending on the site and the extent of ossification, the unilocular or multilocular lesion may be radiolucent or mixed radiolucent or radiodense, but there is always a distinct radiodense border; it may show invasion and erosion of the surrounding bone.[3,6] Such cases may require disfiguring surgery, and an additional reconstructive surgery to restore its shape and function.

The definitive treatment of juvenile ossifying fibroma is surgery. Both conservative and radical resections have been reported in the literature. Slootweg and Muller[6] recommend a conservative surgical approach when managing the juvenile ossifying fibroma. Literature search does not show ossifying fibromas to be fatal, although they may be locally aggressive. Although no cases of malignant transformation have been reported in literature, juvenile ossifying fibroma is very aggressive locally and has a high recurrence rate (30%–58%) when not adequately treated.[5,7] Although many authors suggest radical en bloc resection rather than conservative surgery[8,9] in the present case, a conservative procedure was performed because we favor organ-sparing surgery in case of the pediatric age group. We were successful in removing the mass in toto using the intraoral sublabial approach, although it was extending to the anterior skull base. A radical external approach would have resulted in a scar on the face and any form of maxillary resection would have given child bad functional results. Also, the radical nature of the bone resection would have impeded with the normal growth of the facial skeleton. Hence, the conservative approach should be adopted unless it is extremely necessary to do many bony resections dictated by the extent of the lesion. The success of this conservative approach is demonstrated by the absence of recurrence after 3 years of follow-up in the present case.

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White-Eyed Medial Wall Blowout Fracture Mimicking Head Injury Due to Persistent Oculocardiac Reflex

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Abstract: White-eyed medial wall blowout fracture associated with muscle entrapment is rare. It may present with symptoms consistent with an intracranial injury, delaying the diagnosis and putting the patient at risk for permanent damage. A case of an isolated white-eyed medial wall fracture associated with persistent bradycardia on abduction secondary to oculocardiac reflex as well as limited abduction mimicking sixth-nerve weakness is presented. Patients with white-eyed medial wall blowout fracture with muscle entrapment

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can present with oculocardiac reflex symptoms, pain, diplopia, and strabismus in the absence of any signs on ocular examination except for abnormal motility. Computed tomography imaging of the orbit should be performed to confirm the diagnosis, followed by immediate surgical intervention to avoid ischemia and permanent injury.

Key Words: White-eyed, medial wall blowout fracture, oculocardiac reflex

White-eyed medial wall blowout fracture (WMB) associated with muscle entrapment is exceedingly rare and tends to occur in young patients after accidental trauma.[1–5] Muscle entrapment occurs when the soft tissue is incarcerated as the "trapdoor" fragment of the fracture returns to its original position.[4] There is usually restriction in ocular movement, severe pain, and diplopia with the absence of edema or subconjunctival hemorrhage (hence the "white eye").

Orbital fractures may present with symptoms consistent with an intracranial injury (eg, nausea and vomiting), delaying diagnosis and compromising success of repair because of prolonged soft tissue entrapment and ischemia. The radiographic features of these fractures are also minimal.[1–4,6,7] It is recommended that WMB be managed with immediate surgical intervention.[8,9]

To our knowledge, only 1 case of WMB exists in the literature that presented with oculocardiac reflex symptoms (nausea and vomiting—no bradycardia or tachycardia).[8] We report a case of a 12-year-old boy who presented with an isolated left white-eyed medial wall fracture associated with persistent bradycardia on abduction secondary to the oculocardiac reflex.

CLINICAL REPORT

A 12-year-old boy presented to the emergency department of a neighboring hospital with a sudden onset of nausea, vomiting, sleepiness, and diplopia after kneeing himself in the left eye while jumping on a trampoline. Computed tomography (CT) imaging of the head was performed because his symptoms were suggestive of an intracranial process. The preliminary CT reading was reported as normal, and the patient was admitted for observation. After being reviewed the next day, the official CT reading reported an isolated left orbital medial wall fracture (Fig. 1). The patient was referred to our unit on the third day after trauma and had persistent oculocardiac reflex symptoms. On examination, his vision was normal (6/4.8 OD, 6/6 OS). There was no evidence of periorbital ecchymosis or subconjunctival hemorrhage, and examination was negative for infraorbital paresthesia, dystopia, or enophthalmos. His pulse was 56 beats/min, but decreased on left gaze (abduction). He also developed nausea on attempted abduction. He had marked restriction of adduction (−4) and abduction (−3) in his left eye with no restriction of upward and downward gaze (Fig. 2A). Optic nerve function as well as dilated fundus examination was normal.

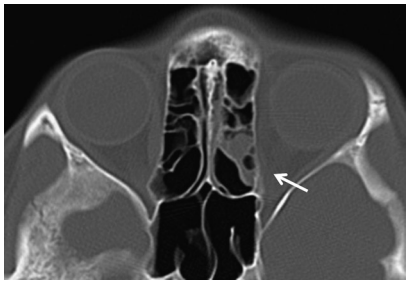

FIGURE 1. An axial CT image showing an isolated left orbit medial wall fracture (arrow).

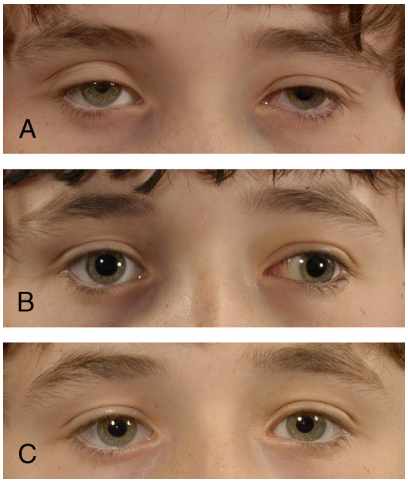

FIGURE 2. A, Preoperative. Patient unable to open his eyes fully as a result of nausea and vasovagal symptoms due to the oculocardiac reflex. He has a mild left divergent squint in primary position with restriction of adduction and abduction. B, One week postoperative. Patient no longer has vasovagal symptoms and no sign of oculocardiac reflex, feels well, and is able to open eyes fully. Left divergent squint worse secondary to swelling. Adduction and abduction of left eye have improved. C, One month postoperative. Resolution of swelling and squint with full eye movements.

He underwent transcaruncular and inferior fornix approach medial wall fracture repair on the same day. Entrapped tissue was freed, and the medial wall defect was repaired with a Medpor sheet (Porex Corporation, Newnan, GA). Forced duction test was normal postoperatively. His oculocardiac reflex symptoms resolved on the first postoperative day with a heart rate of 68 beats/min. The patient continued to improve at 1 week and 1 month postoperatively, with decreased swelling, no vasovagal symptoms, and no signs of oculocardiac reflex (Figs. 2B, C). At 6-week follow-up, abduction and adduction of the left eye were full.

DISCUSSION

There is a high prevalence of ocular injury in children, mostly due to accidents or assault. Despite this, orbital fractures in this population are underreported, resulting in limited information available.[10] In a series reviewing 96 cases of pediatric orbital wall fracture, it was noted that the location pattern of fractures was similar to that observed in adults (orbital floor and medial wall).[10] However, the pediatric age group (<18 years) is at a higher risk of presenting with a white eye associated with muscle entrapment.[11] These patients usually complain of headache and oculocardiac reflex symptoms (nausea, vomiting, and irritability), which may increase the likelihood of a delayed diagnosis.

In 1998, Jordan et al[4] coined the term *white-eyed blowout* and described it as an orbital floor fracture with upward gaze restriction secondary to an entrapped inferior rectus muscle in a trapdoor fracture. It was described as *white eye* because of the absence of any

obvious injury to the eye, no enophthalmos, and no (or minimal) radiographic evidence of bone displacement. Patients may complain of sudden diplopia, limited adduction secondary to an entrapped medial rectus muscle, and possible limited abduction with retraction on attempted abduction known as "pseudo-Duane's retraction syndrome."[11] There is also associated risk of ischemia of the entrapped tissue (muscle or tissue) resulting in necrosis and stimulation of the oculocardiac reflex.[11] The presentation is remarkable in that the eye is quiet with no evidence of soft tissue injury. That, in association with oculocardiac reflex symptoms, can very easily misguide the emergency department physician toward the suspicion of brain injury, thus resulting in a delayed diagnosis. Therefore, knowledge of this entity is necessary, and an ocular motility examination is essential. A delayed diagnosis can result in ischemic muscle and necrosis secondary to entrapment (compartment syndrome), diplopia secondary to restriction, and oculocardiac reflex complications, namely, decreased heart rate and heart block.[5] A recent study and review found that the main reason for surgical delay was in the time it took before a qualified ophthalmologist capable of performing the surgery was consulted.[12]

Isolated medial orbital wall fractures associated with medial rectus muscle entrapment are uncommon, with a total of 52 cases reported in the literature.[5,8,11] Only one of these was associated with oculocardiac reflex symptoms, with no mention of bradycardia with horizontal gaze.[8] Our case is unique in terms of the predominance of oculocardiac reflex symptoms, which were first mistaken for symptoms of intracranial injury, and in the decrease in heart rate on abduction. Oculocardiac reflex symptoms of nausea, vomiting, and irritation combined with limitation of horizontal gaze raised concern of intracranial injury at presentation. The diagnosis can be made with a detailed history, physical examination including pulse rate during gaze, and adequate coronal CT imaging reviewed by experienced clinicians.[6]

The dynamics of medial wall fractures have not been studied as extensively as orbital floor fractures, but it is postulated that similar mechanisms apply. The hydraulic theory proposes that soft tissue displacement secondary to trauma fractures the walls, whereas the buckling theory proposes that posteriorly transmitted forces secondary to force at the bony rim result in fracture of the orbital walls.[13,14] Because the medial wall has no bony rim, it is possible that hydraulic mechanisms may contribute to medial wall fractures. Bones in children are more elastic and thicker than in adults; therefore, hydraulic forces would result in a crack forming a trapdoor rather than a displaced bone fracture as in adults.[11] Children, therefore, may be more susceptible to medial wall trapdoor fractures in comparison to adults.

This case report demonstrates that WMB can present primarily with symptoms mimicking those of head injury leading to a delayed or incorrect diagnosis. In addition to oculocardiac reflex symptoms of nausea and vomiting, those of bradycardia on attempted gaze should be looked for.

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