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ORIGINAL PAPER

An unusual presentation of extramedullary plasmacytoma in testis and review of the literature

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Abstract Extramedullary plasmacytoma is a rare plasma cell neoplasm, and it is extremely uncommon in the testicles. We report a 73-year-old man with multiple myeloma presented with testicular plasmacytoma. He complained of left leg pain and scrotal swelling. Ultrasonography revealed testicular masses. Pathologic examination of the orchiectomy specimen showed plasmocytoma with kappa expression. Multiple lytic bone lesions were seen in bone survey scans, serum immunoelectrophoresis and bone marrow aspiration aided to the diagnosis of multiple myeloma. He received chemotherapy, melphalan and prednisolone, and palliative radiotherapy. He succumbed to disease after 8 months.

Keywords Testis · Plasmacytoma · Multiple myeloma · Extramedullary plasmacytoma

Introduction

Extramedullary plasmacytoma (EMP) is a plasma cell neoplasm of soft tissue without bone marrow involvement [1]. It constitutes about 3% of all plasma cell tumors with a male to female ratio of 3:1 [2]. EMP occurs as a solitary plasmacytoma or involvement of multiple myeloma (MM). The disease may involve a wide variety of anatomic sites; approximately 85% is seen in head and neck region, especially in the respiratory or the gastrointestinal tract. Other regions such as pancreas, spleen, urinary bladder, thyroid, breast and testicles are rarely involved [3–5]. About 15% of plasmacytomas may progress to MM [3, 6]. Testicular involvement of EMP is extremely uncommon. We report here a case of MM presented with testicular plasmacytoma.

Case

A 73-year-old man presented with left leg pain and testicular swelling. He had a history of night sweats and weight loss (7 kg in 3 months). Movement of the left leg was painful. A mass was palpable in the left testicle. Laboratory tests were as follows: hemoglobin 12,3 g/dl, hematocrit 37.5%, white blood cell 6.8 $\times$ 10^9/L, platelets 278 $\times$ 10^9/L, AST 28 U/L, ALT 34 U/L, creatinine 0.9 mg/dl, calcium 9.4 mg/dl, ALP 130 U/L, LDH 236 U/L, blood smear showed rulo formation of erythrocytes. Erythrocyte sedimentation rate was 66 mm/h. Serum levels of α-FP and β-HCG were normal. Scrotal ultrasound showed a left testicular mass of 6 cm $\times$ 5 cm with heterogeneous-hypoechoic pattern. The patient underwent high inguinal orchiectomy. Pathologic examination revealed atypical plasma cells that have eosinophilic cytoplasm and eccentric nuclei and exhibit mild pleomorphism. These immature and atypical plasma cells were infiltrating the interstitial tissue and separating the atrophic tubules (Fig. 1). Immunohistochemical staining was positive for kappa expression and negative for myeloperoxidase and

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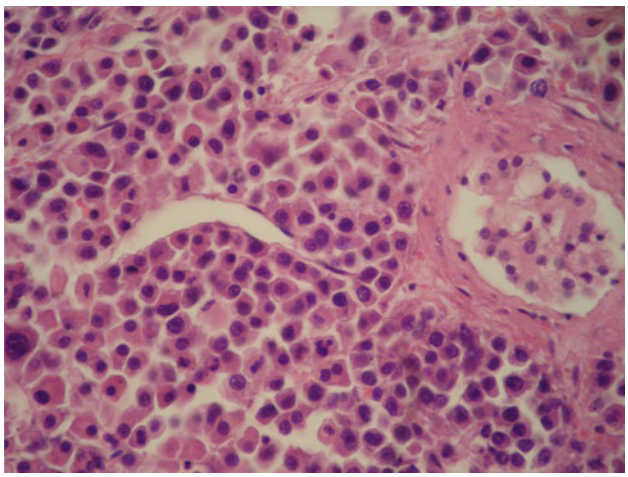

Fig. 1 Histopathology of testicular mass showed atypical plasma cells infiltrating the interstitial tissue and surrounding the atrophic tubules (HE ×200)

lambda expression. Skeletal survey showed lytic lesions in the right humeral head. Serum levels of IgA and β_2-microglobulin were 18.4 g/L and 6.6 mg/L, respectively. Serum protein electrophoresis revealed M protein spike in the gamma band. Bone marrow aspiration showed 40% plasma cells. He received chemotherapy consisting melphalan 5 mg/m^2/day and prednisolone 1 mg/kg/day for 7 days and palliative radiotherapy to the right shoulder. Eight months later, he died of progressive disease.

Discussion

MM is a disease presenting with neoplastic proliferation of monoclonal plasma cells that lead to lytic bone lesions, hypercalcemia, and renal impairment. Plasmacytomas rarely arise outside the bones because of the favorable microenvironment in the bone. The diagnosis of EMP depends upon the demonstration of extramedullary plasma cell tumor with no evidence of systemic signs and symptoms associated with MM [1]. EMP commonly involves the respiratory tract such as nasal cavity, paranasal sinuses, nosopharynx, and larynx as well as the gastrointestinal tract and lymph nodes. Testicular presentation is quite rare. Testicular involvement more often indicates a complication of leukemias and lymphomas. We reported here an IgA kappa MM with primary testicular plasmacytoma presentation.

Helwig et al. [7] reviewed 128 cases of plasmacytoma involving various organs; however, none of them had testicular localization. In another study, Hayes et al. [8] reported only one case with testicular plasmacytoma among 161 cases of EMP. Lewin and Mostofi [7] reviewed 6,000 tumors of the American Testicular Tumor Registry and found only seven cases with testicular plasmacytomas. Accordingly, only 3 of 2,700 testicular cancers in the English Testicular Tumor Registry were testicular plasmacytomas [9]. The rate of testicular and epididymal plasmacytomas among all primary and secondary testicular cancers is estimated to be approximately 0.03–0.1%. Testicular plasmacytomas consist 0.6–2.7% of all MM cases [7, 10].

Plasmacytoma of the testicle is seldom diagnosed as a primary event. In the study of Oppenheimer et al. [11], only 6 out of 37 cases were solitary plasmacytoma of the testicle, while others developed in patients with previously diagnosed MM or concurrently with MM. In patients with both MM and concurrent testicular involvement the primary, whether the skeletal or testicular lesion, is presently a matter of debate. However, the common opinion is that testicular plasmacytoma is a part of systemic disease process. In the study, with 34 MM cases having concurrent testicular involvement [12], the majority (20 patients) had a fatal outcome with progressive disease; some of them died of progression very soon after diagnosis (9–36 days). This patient died of progression 8 months after the diagnosis.

Immunoperoxidase staining on plasmacytomas with testicular involvement reveals an incidence of IgG, IgA, IgD, and light chains of 20, 46.7, 6.6, and 26.7%, respectively. This distribution is different in MM. Alexanian reported that IgA type constituted 25% of the total MM cases [13]. Immunoperoxidase techniques show that testicular plasmacytomas produce the same immunoglobulin idiotype as the disseminated disease clone [14]. In our case, the M protein was of IgA isotype and immunoperoxidase staining detected IgA kappa in the testicle.

No clear guidelines for treatment of EMP are available due to its rarity and variable presentation. However, treatment options include surgery, radiotherapy, chemotherapy, and combined modality treatment including surgery with radiotherapy or chemotherapy or both. EMP is highly radiosensitive and nearly all patients successfully achieve local control [15]. Considering high cure rate of EMP with radiotherapy, use of adjuvant chemotherapy is not recommended [2]. Less then 10% of patients have local recurrence of EMP. Any risk factors for development of MM are not described. After treatment, all patients with EMP should have close observation to detect local recurrences or progression to MM. There is limited experience in the treatment of isolated testicular plasmacytomas. In case of testicular plasmacytomas that arises in the course of disseminated disease, orchiectomy followed by chemotherapy or radiotherapy to the testicular mass are considerable options. Testicular plasmacytoma is extremely uncommon. Non-Hodgkin's lymphoma and plasmacytoma should be sought into consideration in the differential diagnosis of testicular tumors, and clinical findings are very valuable in decision-making.

References

1. Soutar R, Lucraft H, Jackson G, Reece A, Bird J, Low E. Samson D; Guidelines on the diagnosis and management of solitary plasmacytoma of bone and solitary extramedullary plasmacytoma. Br J Haematol. 2004;124(6):717–26.
2. Shih LY, Dunn P, Leung WM, Chen WJ, Wang PN. Localised plasmacytomas in Taiwan:comparison between extramedullary plasmacytoma and solitary plasmacytoma of bone. Br J Cancer. 1995;71(1):128–33.
3. Alexiou C, Kau RJ, Dietzfelbinger H, Kremer M, Spiess JC, Schratzenstaller B, et al. Extramedullary plasmacytoma: tumor occurrence and therapeutic concepts. Cancer. 1999;85(11):2305–14.
4. Strojan P, Soba E, Lamovec J, Munda A. Extramedullary plasmacytoma: clinical and histopathologic study. Int J Radiat Oncol Biol Phys. 2002;53(3):692–701.
5. Kyte RA, Greipp PA. Plasma cell dyscrasias: current status. Crit Rev Oncol Hematol. 1998;8:93–152.
6. Tsang RW, Gospodarowicz MK, Pintilie M, Bezjak A, Wells W, Hodgson DC, et al. Solitary plasmacytoma treated with radiotherapy impact of tumor size on outcome. Int J Radiat Oncol Biol Phys. 2001;50(1):113–20.
7. Levin SH, Mostofi FK. Symptomatic plasmacytoma of the testis. Cancer. 1970;25(5):1193–203.
8. Hayes DW, Bennett WA, Heck FJ. Extramedullary lesions in multiple myeloma; review of literature and pathologic studies. AMA Arch Pathol. 1952;53(3):262–72.
9. Gowing NF. Malignant lymphoma of the testis. Br J Urol. 1964;36(Suppl):85–94.
10. Chica G, Johnson DE, Ayala AG. Plasmacytoma of testis presenting as primary testicular tumor. Urology. 1978;11(1):90–2.
11. Oppenheim PI, Cohen S, Anders KH. Testicular plasmacytoma: a case report with immunohistochemical studies and literature review. Arch Pathol Lab Med. 1991;115(6):629–32.
12. Anghel G, Petti N, Remotti D, Ruscio C, Blandino F, Majolino I. Testicular plasmacytoma: report of a case and review of the literature. Am J Hematol. 2002;71(2):98–104.
13. Barlogie B, Alexanian R, Jagannath S. Plasma cell dyscrasias. JAMA. 1992;268(20):2946–51.
14. Avitable AM, Gansler TS, Tomaszevski JE, Hanno P, Goldwein MI. Testicular plasmacytoma. Urology. 1989;34(1):51–4.
15. Mayr NA, Wen BC, Hussey DH, Burns CP, Staples JJ, Doornbos JF, et al. The role of radiation therapy in the treatment of solitary plasmacytoma. Radiother Oncol. 1990;17(4):293–303.