Left Frontal Meningioangiomatosis Associated with Type IIIc Focal Cortical Dysplasia Causing Refractory Epilepsy and Literature Review

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Key words
- Focal cortical dysplasia
- Meningioangiomatosis
- Refractory epilepsy
- Surgery

Abbreviations and Acronyms
MRI: Magnetic resonance imaging

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■ BACKGROUND: **We report the surgical management of a lesional drug-resistant epilepsy caused by a meningioangiomatosis associated with a type IIIc focal cortical dysplasia located in the left supplementary motor area in a young male patient.**

■ CASE DESCRIPTION: **A first anatomically based partial surgical resection was performed on an 11-year-old under general anesthesia without intraoperative mapping, which allowed for postoperative seizure control (Engel IA) for 6 years. The patient then exhibited intractable right sensatory and aphasic focal onset seizures despite 2 appropriate antiepileptic drugs. A second functional-based surgical resection was performed using intraoperative corticosubcortical functional mapping with direct electrical stimulation under awake conditions. A complete surgical resection was performed, and a left partial supplementary motor area syndrome was observed. At 6 months postoperatively, the patient is seizure free (Engel IA) with an ongoing decrease in antiepileptic drug therapy.**

■ CONCLUSIONS: **Intraoperative functional brain mapping can be applied to preserve the brain function and networks around a meningioangiomatosis to facilitate the resection of potentially epileptogenic perilesional dysplastic cortex and to tailor the extent of resection to functional boundaries.**

INTRODUCTION

Meningioangiomatosis is a rare, benign developmental lesion frequently associated with refractory epilepsy.[1-4] It has recently been described as histopathologically associated with International League Against Epilepsy type IIIc focal cortical dysplasia.[5] The complete surgical resection of the epileptogenic brain tissue can be an efficient treatment for patients with meningioangiomatosis-associated refractory epilepsy.[2] However, surgical management of such epileptogenic brain lesions located within eloquent brain areas poses a particular surgical challenge because of the higher risk of postoperative neurologic and cognitive deficits. Similar to diffuse glioma cases, functional-based surgical resection using intraoperative functional brain mapping with cortico-subcortical electrostimulation under awake conditions can improve the benefit-to-risk balance for these patients. We report the case of a young male patient harboring a lesional drug-resistant epilepsy caused by a meningioangiomatosis associated with a type IIIc focal cortical dysplasia located in the left supplementary motor area; it was treated with functional-based surgical resection with intraoperative cortico-subcortical functional mapping under awake conditions.

CASE DESCRIPTION

A right-handed 11-year-old boy had a history of right sensitive focal onset epileptic seizures for 2 years and aphasic seizures for 1 month. The results of a physical examination were unremarkable, and magnetic resonance imaging (MRI) demonstrated an atypical intracerebral mass in the left supplementary motor area (Figure 1). Antiepileptic drug therapy with oxcarbazepine was introduced, allowing for seizure control. A multidisciplinary meeting in the paediatric neurosurgical centre proposed a surgical resection. A first anatomically based partial surgical resection using intraoperative MRI-based neuronavigation (Surgiscope; ISIS, Grenoble, France) was performed using general anesthesia without intraoperative functional brain mapping in a pediatric neurosurgical center. The initial neuropathologic analysis suggested the diagnosis of focal cortical dysplasia with insufficient histopathologic features to propose a precise diagnosis of type I or II focal cortical dysplasia, according to the focal cortical dysplasia classification available at the time[6] (Figure 2). There were no postoperative complications or neurologic deterioration, except mild disturbance of thin movements of the distality of the right upper limb. Seizure control (Engel IA) was obtained during the next 6 postoperative years. At 17 years old, the patient experienced

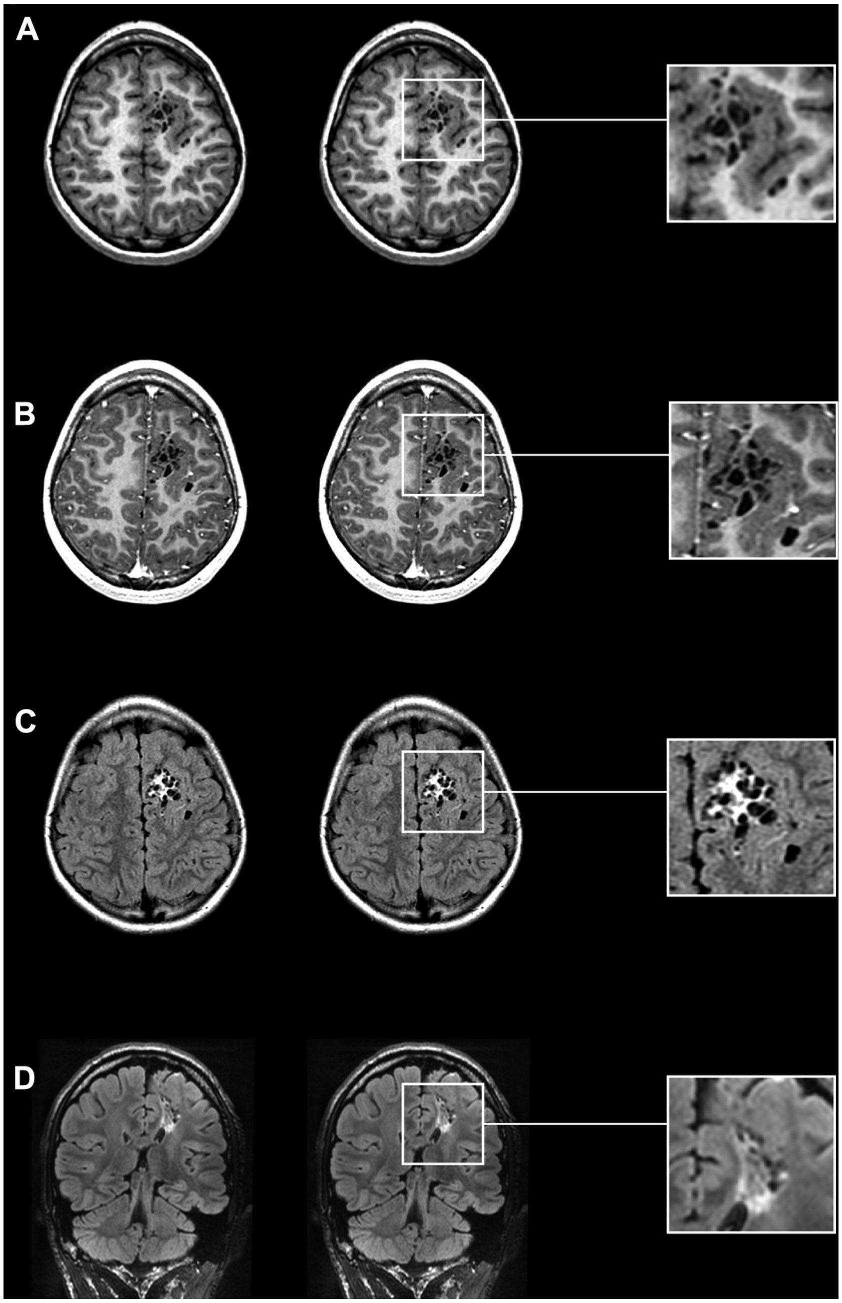

Figure 1. Preoperative magnetic resonance imaging findings of a left frontal meningioangiomatosis. The T1-weighted (**A**) non–contrast-enhanced and (**B**) contrast-enhanced sequences demonstrate a hypointense mass with a slight pial enhancement next to the abnormal cortex (see enlarged focus). (**C**, **D**) The fluid attenuated inversion recovery sequence demonstrates a cortico-subcortical signal increase, with an adjacent multicystic mass within the left supplementary motor area and precentral gyrus. The hypersignal tapers to the ventricle wall (transmantle sign) and is associated with a cortical thickening and a blurring of the grey-white matter interface. There is no mass effect or vasogenic edema.

generalized onset tonic-clonic seizures requiring a higher dose of oxcarbazepine. At 19 years old, the patient presented with intractable right sensatory and aphasic focal onset seizures (2 seizures per week in the preceding 3 months) despite 2 appropriate antiepileptic drugs (lacosamide, oxcarbazepine). There was no progression on imaging follow-up. A multidisciplinary meeting between epileptologist of the paediatric neurosurgical centre and neurosurgeon of the adult neurosurgical centre proposed an extended, functional-based surgical resection with intraoperative cortico-subcortical functional mapping under awake conditions.[7,8] A preoperative multimodal and functional MRI was performed (**Figure 3**). A highly specific anatomic sulcal criterion of focal cortical dysplasia in the central region—previously described as the "power button sign"—was identified on the left central sulcus.[9] Intraoperatively, the surgical resection was continued until cortical and subcortical eloquent sites were identified, defining the functional limits of the resection (**Figure 3**). There were no intraoperative complications, except one 10-second long motor focal onset seizure, which stopped after ice-cold serum application and which did not alter the surgical procedure. Postoperatively, a left partial supplementary motor area syndrome with motor impairment and mutism, but without other language disturbance, was observed and a dedicated rehabilitation was started at postoperative day 1. There were no postoperative complications and postoperative MRI showed a complete surgical resection of the meningioangiomatosis (**Figure 3**). The patient was discharged at postoperative day 6 to a rehabilitation center. The initial neuropathologic analysis proposed the diagnosis of a left frontal meningioangiomatosis (**Figure 2**). A blind neuropathologic review performed for the present study proposed the diagnosis of meningioangiomatosis associated with type IIIc focal cortical dysplasia, according to the International League Against Epilepsy classification of focal cortical dysplasia available at that time.[10] At 6 months postoperatively, the patient has controlled seizures under 2 appropriate antiepileptic drugs (lacosamide, oxcarbazepine) with an ongoing decrease in lacosamide. The 6-month postoperative cognitive and language evaluation confirmed functions similar to the preoperative evaluation. He is still participating in physical rehabilitation to achieve neurologic recovery of his

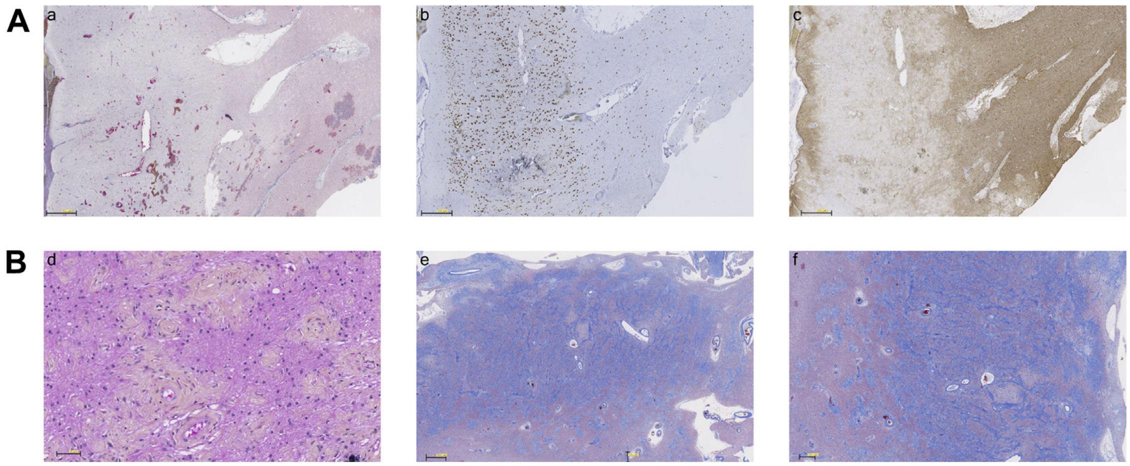

Figure 2. Neuropathologic findings concerning (**A**) first and (**B**) second surgical resections. (**A**) The histopathologic specimen showed an enlargement of Virchow-Robin spaces, without vascular proliferation or fibrosis (*a*, Masson trichrome). Spindle cells within the spaces were positive for vimentin but negative for GFAP and EMA (data not shown). Selective labelling of neuronal cell bodies using antibodies directed against NeuN (*b*) revealed a small cortical ribbon (hypoplasia). Adjacent neocortex revealed architectural disturbances without enlarged dysmorphic neurons or balloon cell. Neurons were observed in the molecular layer, and heterotopically in white matter. (*c*) GFAP labelling revealed astrogliosis. Scale bar, 500 μm. (**B**) Masson trichome staining showed a plaquelike intracortical desmoplastic lesion (*d*), with hyalinized blood vessels and more cellular foci with perivascular spindled cells and scattered psammoma bodies (*e* and *f*; hematoxylin, phoxine, and saffron).

left supplementary motor area syndrome (mild disturbances of thin movement of the distality of right upper and lower limbs and of bimanual coordination).

DISCUSSION

Meningioangiomatosis is a rare, benign, developmental epileptogenic lesion involving the leptomeninges and brain cortex; it is characterized by vascular proliferation and calcifications. Meningioangiomatosis occurs sporadically[2] or in association with a type II neurofibromatosis, in which case it is frequently multifocal and asymptomatic.[11] Meningioangiomatosis can be associated with other developmental lesions, such as focal cortical dysplasia[3,5] or meningiomas.[12] Meningioangiomatosis occurs in children and young adults with a long-standing history of epileptic seizures. Complete surgical resection can be an efficient treatment for patients with refractory epilepsy, as in focal cortical dysplasia.[13-16]

The etiopathogeny of meningioangiomatosis is poorly understood. Three main hypotheses have been proposed: 1) meningioangiomatosis represents a hamartomatous lesion that subsequently undergoes degenerative changes; 2) meningioangiomatosis represents a vascular malformation with secondary meningothelial cell proliferation; or 3) meningioangiomatosis represents an extension of an overlying meningioma that spreads along Virchow-Robin spaces.[3,17,18] We performed a comprehensive literature review with the description of 175 cases of meningioangiomatosis, including the present one[1-4,11,12,17,19-70] (Table 1). The association of a meningioangiomatosis together with a focal cortical dysplasia, which has been reported in 14 cases (8%), including the present one, provides into the understanding of the etiology of meningioangiomatosis. One can assume that these 2 distinct lesions share a common pathophysiology that favors a developmental etiology.[3,17,18] However, the low reported rate of focal cortical dysplasia is possibly due to the difficulties in ascertaining such a diagnosis in the context of meningioangiomatosis, as reported in the present observation. To our knowledge, we report for the first time the "power button sign," which is an anatomic sulcal variation described as a highly specific criterion of focal cortical dysplasia in the central region.[9] We suggest that the anatomic sulcal variation defined as the "power button sign" can be observed in developmental malformations associated with focal cortical dysplasia, including meningioangiomatosis.

We found 109 patients (62.3%) with a meningioangiomatosis-associated refractory epilepsy who may have benefitted from surgical management to achieve seizure control (Engel IA). In addition, we found 71 patients harboring a meningioangiomatosis located within eloquent brain areas (40.8%).[74] These observations favor the referral of such patients to highly specialized centers where they could benefit from functional-based surgical resection for epileptic purposes using intraoperative cortico-subcortical functional mapping under awake conditions.

Concerning the surgical management, a functional-based surgical resection using intraoperative cortico-subcortical functional mapping under awake conditions was proposed to the patient, after clear explanations about the benefit-to-risk ratio, especially the risk of partial recovery of the left supplementary motor area syndrome, the risk of language disturbances, and the need for prolonged cognitive and physical rehabilitation. The multidisciplinary meeting proposed a surgical resection with the following

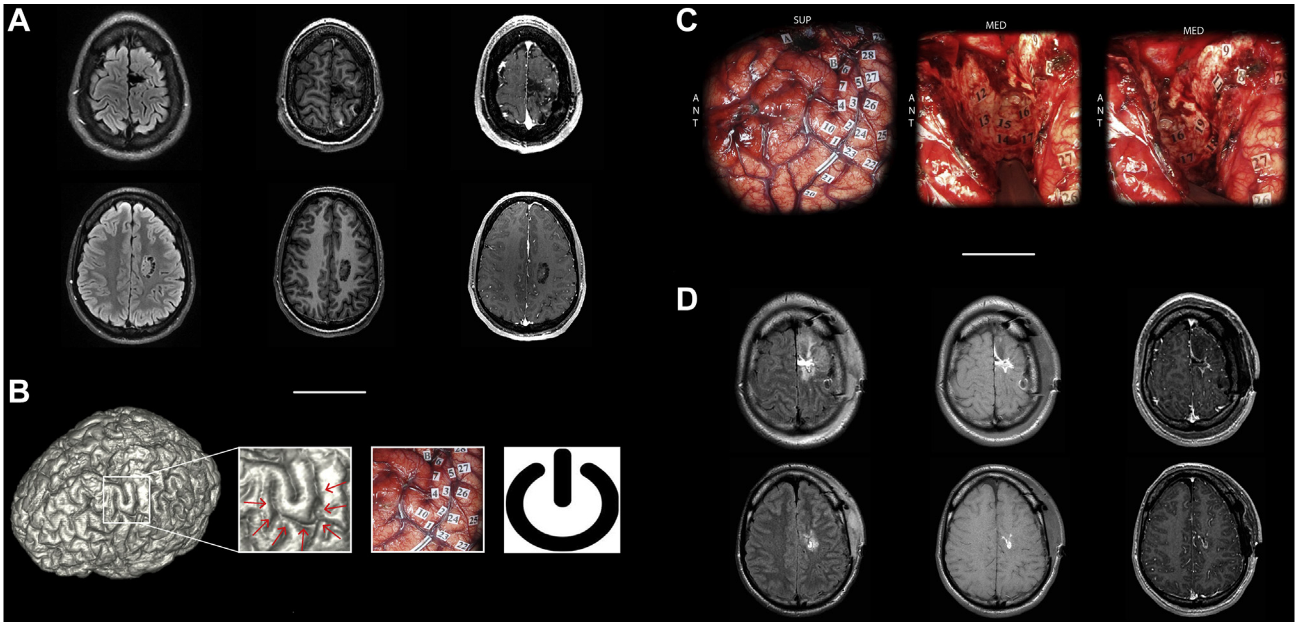

Figure 3. Perioperative magnetic resonance imaging (MRI) findings and intraoperative findings of the functional-based surgical resection with intraoperative cortico-subcortical functional mapping under awake conditions. (**A**) Preoperative MRI with fluid attenuated inversion recovery (*left*), T1-weighted non–contrast-enhanced (*middle*), and T1-weighted contrast-enhanced (*right*) sequences demonstrating the first resection cavity. (**B**) Preoperative functional MRI with 3D reconstruction (*left*) demonstrating an atypical sulcal pattern defined by the interposition of a pre-central sulcal segment between the central sulcus and one of its hook-shaped anterior ascending branches called the "power button sign" (*red arrows*) by analogy to the power symbol (*right*). (**C**) Intraoperative photograph (*middle*) confirms this atypical sulcal shape and shows the cortical functional organization of the primary motor areas within the left central sulcus. Intraoperative photographs before (*left*) and after (*middle*, *right*) surgical resection. Intraoperative functional boundaries were defined through the identification of eloquent sites by cortico-subcortical electrical bipolar stimulation mapping during awake craniotomy. The locations of eloquent sites were marked intraoperatively by numbered tags. Tags A, B, and C delimit the meningioangiomatosis, and the linear tag (1 cm in length) delimits the central sulcus. Cortical direct electrical stimulation of that area elicited reproducible involuntary movements (tags 1, 2, 3, 4, 5, 6, and 7), reproducible speech arrest during naming task (tag 10), and reproducible paresthesias (tags 20, 21, 22, 23, 24, 25, 26, 27, 28, and 29), according to the sensorimotor somatotopy. Subcortical direct electrical stimulation of that area elicited reproducible right leg (tags 8, 9, 11, 18, and 19) and right arm (tags 16, 17) involuntary movements, reproducible latency during naming task (tags 14 and 15), and reproducible saccadic eye movements (tags 12 and 13). The surgical resection was performed without opening of the left lateral ventricle. (**D**) Postoperative MRI with fluid attenuated inversion recovery (*left*), T1-weighted non–contrast-enhanced (*middle*), and T1-weighted contrast-enhanced (*right*) sequences demonstrate the complete surgical resection of the meningioangiomatosis.

aims: 1) symptomatic, to achieve epileptic seizure control, which is directly correlated to the extent of resection[2,13,75] and to increase the chance of antiepileptic drug withdrawal; 2) diagnostic, to ascertain the diagnosis when confronting such an atypical imaging presentation for an isolated focal cortical dysplasia, according to our neuroradiologic analysis; 3) curative, especially for a benign brain lesion that can be responsible for refractory seizures. The occurrence of an early postoperative left supplementary motor syndrome was expected and is associated with good outcomes following dedicated rehabilitation with the participation of the contralateral supplementary motor area.[76-78]

The present findings should be interpreted with caution given the limited follow-up, especially regarding postoperative seizure control and Engel classification that requires a 2-year-long postoperative follow-up. The long-term functional results may be affected by the follow-up time frame of 6 postoperative months, the potential increase of seizures in the early postoperative period, the potential continued withdrawal of antiepileptic drugs during longer postoperative follow-up, and the potential for delayed postoperative seizures.

CONCLUSION

The surgical resection of an epileptogenic brain lesion located within eloquent brain areas poses a particular surgical challenge because of the higher risk of postoperative neurologic and cognitive deficits. To minimize the risk of damage to eloquent areas and their connecting pathways, the use intraoperative cortico-subcortical functional electrostimulation mapping using an awake procedure is recommended during glioma surgery in these brain regions.[7,8,79-81] Applying the same rationale, intraoperative functional mapping can be applied to map and preserve the brain function and networks around a meningioangiomatosis to facilitate the resection of the potentially epileptogenic perilesional dysplastic cortex and to tailor the extent of resection to functional boundaries.[2,82]

Table 1. Review of Meningioangiomatosis-Associated Focal Cortical Dysplasia

References	Country	Number of Patients	Focal Cortical Dysplasia (n)	Refractory Epilepsy (n)	Eloquent Area* (n)
Roux et al., 2018 (present case)	France	1	1	1	1
Iorgulescu et al., 2018[71]	USA	4	0	2	2
Anand et al., 2017[72]	USA	3	0	3	1
Nascimento et al., 2016[73]	Canada	1	0	1	1
Donovan and Thavapalan, 2016[19]	USA	1	0	1	0
Yust-Katz et al., 2016[20]	Israel	1	n/a	0	1
Motevalli et al., 2016[21]	Iran	1	0	1	n/a
Bulut et al., 2015[22]	Turkey	1	0	0	0
Zhang et al., 2015[23]	China	14	0	12	5
Grabowski and Prayson, 2015[3]	USA	16	10	12	8
Sun et al., 2015[24]	China	3	0	1	2
Aw-Zoretic et al., 2015[25]	USA	1	0	1	0
Fu et al., 2015[26]	China	1	0	1	1
Mukae et al., 2014[4]	Japan	2	2	2	2
Li et al., 2014[27]	China	1	0	1	0
Jeon et al., 2013[18]	Korea	8	0	8	6
Feng et al., 2013[2]	China	10	0	10	4
Shah et al., 2013[28]	USA	1	0	0	1
Abdulazim et al., 2013[29]	Germany	1	0	0	1
Jamil et al., 2012[30]	USA	1	0	1	1
Jansen et al., 2012[31]	Belgium	1	0	1	0
Marzi et al., 2012[32]	Italy	1	0	0	0
Cui et al., 2012[12]	China	1	0	1	0
Barbosa-Silva et al., 2012[33]	Brazil	1	0	1	1
Shi et al., 2011[34]	China	1	0	0	1
Kashlan et al., 2011[35]	USA	1	0	1	1
Alexiou et al., 2011[1]	Greece	1	0	1	0
Chen et al., 2010[36]	China	1	0	0	1
Yao et al., 2009[16]	China	7	0	6	n/a
Rokes et al., 2009[38]	USA	1	0	0	0
Kim et al., 2009[39]	Korea	1	0	1	1
Kim et al., 2009[40]	Korea	9	1	8	n/a
Ishihara et al., 2009[41]	Japan	1	0	0	1
Fedi et al., 2009[42]	Australia	1	0	0	1
Saad et al., 2009[43]	USA	1	0	1	1
Wang et al., 2006[44]	China	5	0	2	4
Omeis et al., 2006[11]	USA	2	0	0	2
Giulioni et al., 2006[45]	Italy	1	0	1	0

*According to Chang EF, Smith JS, Chang SM, et al. Preoperative prognostic classification system for hemispheric low-grade gliomas in adults. *J Neurosurg.* 2008;109: 817-824.
†In this study, the topography of the meningioangiomatosis was not described for 9 patients.

Continues

Table 1. Continued

References	Country	Number of Patients	Focal Cortical Dysplasia (n)	Refractory Epilepsy (n)	Eloquent Area* (n)
Deb et al., 2006[46]	India	1	0	1	0
Kobayashi et al., 2006[47]	Japan	1	0	1	0
Jallo et al., 2005[48]	USA	6	0	6	3
Wixom et al., 2005[49]	USA	1	0	1	1
Perry et al., 2005[17]	USA	24	n/a	n/a	11†
Krolczyk and Prayson, 2003[50]	USA	1	0	0	1
Koutsopoulos et al., 2003[51]	Greece	1	0	1	1
Kuchelmeister et al., 2003[52]	Germany	1	0	0	0
Seo et al., 2003[53]	Korea	1	0	1	0
Savargaonkar et al., 2003[54]	USA	3	0	0	2
Kim et al., 2002[56]	Korea	2	0	1	2
Kim et al., 2002[55]	Korea	5	0	3	n/a
Savargaonkar et al., 2001[57]	USA	2	0	0	1
Scroop et al., 2000[58]	Australia	1	0	1	0
Mut et al., 2000[59]	Turkey	1	0	1	0
Jallo et al., 2000[60]	USA	1	0	0	0
Izycka-Swieszewska et al., 2000[61]	Poland	1	0	1	1
Park et al., 1999[62]	Korea	1	0	1	1
Tacconi et al., 1997[63]	United Kingdom	1	0	1	1
Kollias et al., 1994[64]	USA	1	0	0	1
Fujimoto et al., 1993[65]	Japan	1	0	n/a	1
Kim et al., 1993[66]	Korea	1	0	1	1
Blumenthal et al., 1993[67]	USA	1	0	1	0
Aizpuru et al., 1991[68]	USA	4	0	4	2
Louw et al., 1990[69]	Canada	2	0	0	0
Ogilvy et al., 1989[70]	USA	1	0	1	1
Data analysis	—	175	14—8%	109—62.3%	71—40.8%

*According to Chang EF, Smith JS, Chang SM, et al. Preoperative prognostic classification system for hemispheric low-grade gliomas in adults. *J Neurosurg.* 2008;109: 817-824.
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REFERENCES

1. Alexiou GA, Moschovi M, Stefanaki K, Siozos G, Hatzigiorgi C, Prodromou N. Meningioangiomatosis in a 5-year-old boy presenting with intractable seizures. *Pediatr Neurosurg*. 2011;47:143-146.

2. Feng R, Hu J, Che X, Pan L, Wang Z, Zhang M, et al. Diagnosis and surgical treatment of sporadic meningioangiomatosis. *Clin Neurol Neurosurg*. 2013;115:1407-1414.

3. Grabowski MM, Prayson RA. Focal cortical dysplasia in meningioangiomatosis. *Clin Neuropathol*. 2015;34:76-82.

4. Mukae N, Suzuki SO, Morioka T, Murakami N, Hashiguchi K, Shigeto H, et al. ILAE focal cortical dysplasia type IIIc in the ictal onset zone in epileptic patiens with solitary meningioangiomatosis. *Epileptic Disord*. 2014;16:533-539.

5. Batra A, Prayson RA. Meningioangiomatosis associated with focal cortical dysplasia and neurofibrillary tangles. *Clin Neuropathol*. 2013;32:37-41.

6. Palmini A, Najm I, Avanzini G, Babb T, Guerrini R, Foldvary-Schaefer N, et al. Terminology and classification of the cortical dysplasias. *Neurology*. 2004;62(6 suppl 3):S2-S8.

7. Pallud J, Mandonnet E, Corns R, Dezamis E, Parraga E, Zanello M, et al. Technical principles of direct bipolar electrostimulation for cortical and subcortical mapping in awake craniotomy. *Neurochirurgie*. 2017;63:158-163.

8. Pallud J, Rigaux-Viode O, Corns R, Muto J, Lopez Lopez C, Mellerio C, et al. Direct electrical bipolar electrostimulation for functional cortical and subcortical cerebral mapping in awake craniotomy. Practical considerations. *Neurochirurgie*. 2017;63:164-174.

9. Mellerio C, Roca P, Chassoux F, Danière F, Cachia A, Lion S, et al. The power button sign: a newly described central sulcal pattern on surface renderig MR images of type 2 focal cortical dysplasia. *Radiology*. 2015;274:500-507.

10. Blümcke I, Thom M, Aronica E, Armstrong DD, Vinters HV, Palmini A, et al. The clinicopathologic spectrum of focal cortical dysplasias: a consensus classificaion proposed by an ad hoc Task Force of the ILAE Diagnostic Methods Commission. *Epilepsia*. 2011;52:158-174.

11. Omeis I, Hillard VH, Braun A, Benzil DL, Murali R, Harter DH. Meningioangiomatosis associated with neurofibromatosis: report of 2 cases in a single family and review of the literature. *Surg Neurol*. 2006;65:595-603.

12. Cui H, Shi H, Chen X, Wang W, Lai R, Han A. Clinicopathological features of meningioangiomatosis associated with meningima: a case report with literature review. *Case Rep Oncol Med*. 2012;2012:296286.

13. Roessler K, Kasper BS, Heynold E, Coras R, Sommer B, Rampp S, et al. Intraoperative magnetic-resonance tomography and neuronavigation during resection of focal cortical dysplasia type II in adult epilepsy surgery offers better seizure outcomes. *World Neurosurg*. 2018;109:e43-e49.

14. Chassoux F, Landré E, Mellerio C, Turak B, Mann MW, Daumas-Duport C, et al. Type II focal cortical dysplasia: electroclinical phenotype and surgical outcome related to imaging. *Epilepsia*. 2012;53:349-358.

15. Fauser S, Essang C, Altenmüller D-M, Staack AM, Steinhoff BJ, Strobl K, et al. Long-term seizure outcome in 211 patients with focal cortical dysplasia. *Epilepsia*. 2015;56:66-76.

16. Yao K, Mei X, Liu X, Duan Z, Liu C, Bian Y, et al. Clinical characteristics, pathological features and surgical outcomes of focal cortical dyspasia (FCD) type II: correlation with pathological subtypes. *Neurol Sci*. 2014;35:1519-1526.

17. Perry A, Kurtkaya-Yapicier O, Scheithauer BW, Robinson S, Prayson RA, Kleinschmidt-DeMasters BK, et al. Insights into meningioangiomatosis with and without meningioma: a cinicopathologic and genetic series of 24 cases with review of the literature. *Brain Pathol*. 2005;15:55-65.

18. Jeon TY, Kim JH, Suh Y-L, Ahn S, Yoo S-Y, Eo H. Sporadic meningioangiomatosis: imaging findings with histopathologic correlations in seven patients. *Neuroradiology*. 2013;55:1439-1446.

19. Donovan DJ, Thavapalan V. Pediatric meningeal tumors of the sylvian fissure region without dural attachment: a series of three patients and review of the literature. *Surg J (N Y)*. 2016;2:e31-e36.

20. Yust-Katz S, Fuller G, Fichman-Horn S, Michaeli N, Inbar E, Lukman J, et al. Progressive diffuse meningioangiomatosis: response to bevacizumab treatment. *Neurology*. 2016;86:1643-1644.

21. Motevalli D, Kamalian N, Tavangar SM. Meningioangiomatosis in an otherwise healthy 13 year-old boy: a case report with emphasis on histopathological findings. *Iran J Pathol*. 2016;11:291-295.

22. Bulut E, Mut M, Soylemezoglu F, Oguz KK. Meningioangiomatosis of the cerebellum: radiopathologic characteristics of a case. *Acta Neurochir (Wien)*. 2015;157:1371-1372.

23. Zhang C, Wang Y, Wang X, Zhang JG, Li JJ, Hu WH, et al. Sporadic meningioangiomatosis with and without meningioma: analysis of clinical differences and risk factors for poor seizure outcomes. *Acta Neurochir (Wien)*. 2015;157:841-853 [discussion: 853].

24. Sun Z, Jin F, Zhang J, Fu Y, Li W, Guo H, et al. Three cases of sporadic meningioangiomatosis with different imaging appearances: case report and review of the literature. *World J Surg Oncol*. 2015;13:89.

25. Aw-Zoretic J, Burrowes D, Wadhwani N, Ryan M. Teaching NeuroImages: Meningioangiomatosis. *Neurology*. 2015;84:e9-e10.

26. Fu Y, Sun ZH, Zhang J, Liu HT. Cystic meningioangiomatosis with enhancing mural nodule on MRI and no calcification on CT. *Neurol Neurochir Pol*. 2015;49:58-60.

27. Li P, Cui G, Wang Y, Geng M, Wang Z. Multicystic meningioangiomatosis. *BMC Neurol*. 2014;14:32.

28. Shah A, Korya D, Larsen BT, Torres M, Drake K, La Wall J. Meningioangiomatosis: a rare presentation with progressive cortical blindness. *Neurology*. 2013;81:511-512.

29. Abdulazim A, Samis Zella MA, Rapp M, Gierga K, Langen KJ, Steiger HJ, et al. Meningioangiomatosis in a patient with progressive focal neurological deficit-case report and review of literature. *Br J Neurosurg*. 2013;27:253-255.

30. Jamil O, Ramkissoon S, Folkerth R, Smith E. Multifocal meningioangiomatosis in a 3-year-old patient. *J Neurosurg Pediatr*. 2012;10:486-489.

31. Jansen K, Sciot R, Lagae L. Apnea as the sole manifestation of a seizure attributable to meningioangiomatosis of the temporal lobe in an infant. *Pediatr Neurol*. 2012;47:144-146.

32. Marzi S, De Paulis D, Ricci A, Taddei G, Dehcordi SR, Coletti G, et al. Meningioangiomatosis without neurofibromatosis type 2. *World J Oncol*. 2012;3:127-133.

33. Barbosa-Silva E, Dellaretti M, de Carvalho GTC, Pereira JL, Botrel L Jr, Pittella JE, et al. Meningioangiomatosis without neurofibromatosis simulating encephalitis in neuroimaging. *Surg Neurol Int*. 2012;3:34.

34. Shi H, Zhao S, Tian X, Li Z, Huang Q, Li B. Meningioangiomatosis-associated meningioma misdiagnosed as glioma by radiologic and intraoperative histological examinations. *Brain Tumor Pathol*. 2011;28:347-352.

35. Kashlan ON, Laborde DV, Davison L, Saindane AM, Brat D, Hudgins PA, et al. Meningioangiomatosis: a case report and literature review emphasizing diverse appearance on different imaging modalities. *Case Rep Neurol Med*. 2011;2011:361203.

36. Chen Y-Y, Tiang X-Y, Li Z, Luo B-N, Huang Q. Sporadic meningioangiomatosis-associated atypical meningioma mimicking parenchymal invasion of brain: a case report and review of the literature. *Diagn Pathol*. 2010;5:39.

37. Yao Z, Wang Y, Zee C, Feng X, Sun H. Computed tomography and magnetic resonance appearance of sporadic meningioangiomatosis correlated with pathological findings. *J Comput Assist Tomogr*. 2009;33:799-804.

38. Rokes C, Ketonen LM, Fuller GN, Weinberg J, Slopis JM, Wolff JEA. Imaging and spectroscopic findings in meningioangiomatosis. *Pediatr Blood Cancer*. 2009;53:672-674.

39. Kim SH, Yoon SH, Kim JH. A case of infantile meningioangiomatosis with a separate cyst. *J Korean Neurosurg Soc*. 2009;46:252-256.

40. Kim NR, Cho SJ, Suh Y-L. Allelic loss on chromosomes 1p32, 9p21, 13q14, 16q22, 17p, and 22q12 in meningiomas associated with meningioangiomatosis and pure meningioangiomatosis. *J Neurooncol*. 2009;94:425-430.

41. Ishihara M, Miyagawa-Hayashino A, Nakashima Y, Haga H, Takahashi JA, Manabe T. Intracerebral schwannoma in a child with infiltration along perivascular spaces resembling meningioangiomatosis. *Pathol Int*. 2009;59:583-587.

42. Fedi M, Kalnins RM, Shuey N, Fitt GJ, Newton M, Mitchell LA. Cystic meningioangiomatosis in neurofibromatosis type 2: an MRI-pathological study. *Br J Radiol*. 2009;82:e129-e132.

43. Saad A, Folkerth R, Poussaint T, Smith E, Ligon K. Meningioangiomatosis associated with meningioma: a case report. *Acta Cytol*. 2009;53:93-97.

44. Wang Y, Gao X, Yao Z-W, Chen H, Zhu JJ, Wang SX, et al. Histopathological study of five cases with sporadic meningioangiomatosis. *Neuropathology*. 2006;26:249-256.

45. Giulioni M, Acciarri N, Zucchelli M, Marucci G, Badaloni F, Calbucci F. Meningioangiomatosis involving the wall of the middle cerebral artery. *J Neurooncol*. 2006;78:105-106.

46. Deb P, Gupta A, Sharma MC, Gaikwad S, Singh VP, Sarkar C. Meningioangiomatosis with meningioma: an uncommon association of a rare entity—report of a case and review of the literature. *Childs Nerv Syst*. 2006;22:78-83.

47. Kobayashi H, Ishii N, Murata J-I, Saito H, Kubota KC, Nagashima K, et al. Cystic meningioangiomatosis. *Pediatr Neurosurg*. 2006;42:320-324.

48. Jallo GI, Kothbauer K, Mehta V, Abbott R, Epstein F. Meningioangiomatosis without neurofibromatosis: a clinical analysis. *J Neurosurg*. 2005;103(suppl 4):319-324.

49. Wixom C, Chadwick AE, Krous HF. Sudden, unexpected death associated with meningioangiomatosis: case report. *Pediatr Dev Pathol*. 2005;8:240-244.

50. Krolczyk S, Prayson RA. Pathologic quiz case: an 11-year-old boy with intractable seizures. Meningioangiomatosis. *Arch Pathol Lab Med*. 2003;127: e349-e350.

51. Koutsopoulos AV, Yannopoulos A, Stathopoulos EN, Evangeliou A, Panayiotides JG, Kafousi M, et al. Meningioangiomatosis with predominantly cellular pattern. *Neuropathology*. 2003;23:141-145.

52. Kuchelmeister K, Richter H-P, Kepes JJ, Schachenmayr W. Case report: microcystic meningioma in a 58-year-old man with multicystic meningioangiomatosis. *Neuropathol Appl Neurobiol*. 2003;29:170-174.

53. Seo DW, Park MS, Hong SB, Hong SC, Suh Y-L. Combined temporal and frontal epileptogenic foci in meningioangiomatosis. *Eur Neurol*. 2003;49: 184-186.

54. Savargaonkar P, Chen S, Bhuiya T, Valderrama E, Bloom T, Farmer PM. Meningioangiomatosis: report of three cases and review of the literature. *Ann Clin Lab Sci*. 2003;33:115-118.

55. Kim NR, Choe G, Shin S-H, Wang KC, Cho BK, Choi KS, et al. Childhood meningiomas associated with meningioangiomatosis: report of five cases and literature review. *Neuropathol Appl Neurobiol*. 2002;28:48-56.

56. Kim W-Y, Kim I-O, Kim S, Cheon J-E, Yeon M. Meningioangiomatosis: MR imaging and pathological correlation in two cases. *Pediatr Radiol*. 2002;32:96-98.

57. Savargaonkar P, Bhuiya T, Valderrama E, Farmer P. Scrape cytology of meningioangiomatosis: a report of two cases with diagnostic cytologic features. *Acta Cytol*. 2001;45: 1069-1072.

58. Scroop R, Voyvodic F, Sage MR. Meningioangiomatosis. *Australas Radiol*. 2000;44:460-463.

59. Mut M, Söylemezoğlu F, Firat MM, Palaoğlu S. Intraparenchymal meningioma originating from underlying meningioangiomatosis. Case report and review of the literature. *J Neurosurg*. 2000;92: 706-710.

60. Jallo GI, Silvera VM, Abbott IR. Meningioangiomatosis. *Pediatr Neurosurg*. 2000;32:220-221.

61. Izycka-Swieszewska E, Rzepko R, Kopczynski S, Franc Z, Szurowska E, Borowska-Lehman J. Meningioangiomatosis with a predominant fibrocalcifying component. *Neuropathology*. 2000;20: 44-48.

62. Park MS, Suh DC, Choi WS, Lee SY, Kang GH. Multifocal meningioangiomatosis: a report of two cases. *AJNR Am J Neuroradiol*. 1999;20:677-680.

63. Tacconi L, Thom M, Symon L. Cerebral meningioangiomatosis: case report. *Surg Neurol*. 1997;48: 255-260.

64. Kollias SS, Crone KR, Ball WS, Prenger EC, Ballard ET. Meningioangiomatosis of the brain stem. Case report. *J Neurosurg*. 1994;80:732-735.

65. Fujimoto K, Nikaidoh Y, Yuasa T, Nagata K, Ida Y, Fujioka M, et al. Meningioangiomatosis not associated with von Recklinghausen's disease—case report. *Neurol Med Chir (Tokyo)*. 1993;33: 651-655.

66. Kim YW, Choi WS, Lee J, Yang MH. Meningioangiomatosis—a case report. *J Korean Med Sci*. 1993;8:308-311.

67. Blumenthal D, Berho M, Bloomfield S, Schochet SS, Kaufman HH. Childhood meningioma associated with meningio-angiomatosis. Case report. *J Neurosurg*. 1993;78:287-289.

68. Aizpuru RN, Quencer RM, Norenberg M, Altman N, Smirniotopoulos J. Meningioangiomatosis: clinical, radiologic, and histopathologic correlation. *Radiology*. 1991;179: 819-821.

69. Louw D, Sutherland G, Halliday W, Kaufmann J. Meningiomas mimicking cerebral schwannoma. *J Neurosurg*. 1990;73:715-719.

70. Ogilvy CS, Chapman PH, Gray M, de la Monte SM. Meningioangiomatosis in a patient without von Recklinghausen's disease. Case report. *J Neurosurg*. 1989;70:483-485.

71. Iorgulescu JB, Ferris S, Agarwal A, Casavilca Zambrano S, Hill DA, Schmidt R, et al. Non-meningothelial meningeal tumours with meningioangiomatosis-like pattern of spread. *Neuropathol Appl Neurobiol*. 2018. https://doi.org/10.1111/nan.12481. [Epub ahead of print].

72. Anand R, Garling RJ, Poulik J, Sabolich M, Goodrich DJ, Sood S, et al. Sporadic meningioangiomatosis: a series of three pediatric cases. *Cureus*. 2017;9:e1640.

73. Nascimento FA, Kiehl T-R, Tai PC, Valiante TA, Krings T. Meningioangiomatosis: a disease with many radiological faces. *Can J Neurol Sci*. 2016;43: 847-849.

74. Chang EF, Smith JS, Chang SM, Lamborn KR, Prados MD, Butowski N, et al. Preoperative prognostic classification system for hemispheric low-grade gliomas in adults. *J Neurosurg*. 2008;109: 817-824.

75. Pallud J, Audureau E, Blonski M, Sanai N, Bauchet L, Fontaine D, et al. Epileptic seizures in diffuse low-grade gliomas in adults. *Brain J Neurol*. 2014;137(Pt 2):449-462.

76. Vassal M, Charroud C, Deverdun J, Le Bars E, Molino F, Bonnetblanc F, et al. Recovery of functional connectivity of the sensorimotor network after surgery for diffuse low-grade gliomas involving the supplementary motor area. *J Neurosurg*. 2017;126:1181-1190.

77. Duffau H, Lopes M, Denvil D, Capelle L. Delayed onset of the supplementary motor area syndrome after surgical resection of the mesial frontal lobe: a time course study using intraoperative mapping in an awake patient. *Stereotact Funct Neurosurg*. 2001; 76:74-82.

78. Russell SM, Kelly PJ. Incidence and clinical evolution of postoperative deficits after volumetric stereotactic resection of glial neoplasms involving the supplementary motor area. *Neurosurgery*. 2007; 61(suppl 1):358-368.

79. Sanai N, Berger MS. Operative techniques for gliomas and the value of extent of resection. *Neurotherapeutics*. 2009;6:478-486.

80. De Witt Hamer PC, Robles SG, Zwinderman AH, Duffau H, Berger MS. Impact of intraoperative stimulation brain mapping on glioma surgery outcome: a meta-analysis. *J Clin Oncol*. 2012;30: 2559-2565.

81. Freyschlag CF, Duffau H. Awake brain mapping of cortex and subcortical pathways in brain tumor surgery. *J Neurosurg Sci*. 2014;58:199-213.

82. Pallud J, Dezamis E. Functional and oncological outcomes following awake surgical resection using intraoperative cortico-subcortical functional mapping for supratentorial gliomas located in eloquent areas. *Neurochirurgie*. 2017;63:208-218.

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