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Case Report

Meningiomatosis with Meningioma: A Case Report

Nitishri Sinha, MD[1*], Olexsandr Yartym, MD[2], Emmanuel Uzoma Okoro, MD[3], Taras Havryliv, MD, FEBNS[4], Ingmar Blümcke, MD[5] and Volodymyr Smolanka, MD, PhD[6]

[1, 2, 3, 4, 6] Neurosurgery, University of Uzhhorod, Regional Center for Neurosurgery, Neurology and Psychiatry Kapushanska 24, Uzhhorod 88000, Uzhhorod- Ukraine.

[5] Neuropathology, Institute of Neuropathology, University Hospital, Erlange, Erlangen-91054, Erlangen-Germany.

***Corresponding Author:** Dr. Nitishri Sinha, Neurosurgery, University of Uzhhorod, Regional Center for Neurosurgery, Neurology and Psychiatry Kapushanska 24, Uzhhorod 88000, Uzhhorod- Ukraine.*

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Abstract

Meningioangiomatosis (MA) is a rare abnormality that may be occasionally associated with intracranial meningioma. The transitional variant of the presented meningioma is classified as a World Health Organization Grade I tumor. MA is a benign, epileptogenic and poorly studied brain lesion. In the present report, we describe a case of MA in conjunction with meningioma. We describe the case in a female child who initially presented with seizure onset and underwent complete resection of the lesion. Histopathological and immunohistochemistry have reported it as MA-M. Patients with MA associated with meningioma, after surgical resection often have a good post-operative prognosis.

Abbreviations: MA- Meningionangiomatosis, M- Meningioma, MA-M- Meningioangiomatosis-Meningioma

***Keywords:** Meningioangiomatosis, Meningioma, Epilepsy, Surgical Resection.*

Introduction

Meningioangiomatosis is a rare and benign meningio-vascular and hamartomatous lesion which may or may not occur with Neurofibromatosis type 2 [1,2]. It is distinguished by nodular, plaque-like growth within the leptomeninges and the cerebral cortex. It shows features of both meningioma as well as angioma. Sporadic MA can clinically present as uncontrollable seizures or vague neurological symptoms (headache and nausea). In most cases, seizures are the most common complaint. Therefore, MA is an important pathological differential condition in epilepsy surgery [3,4,5]. Sporadic MA can often combine with a neoplastic lesion, most commonly a meningioma (MA-M) [2]. Previous studies show MA to be a benign, non-neoplastic lesion, while genetic and molecular studies conclude that MA-M is neoplastic in nature [2,6,7]. However, the relative comparison between MA-M and pure MA has been poorly studied.

Herein, we report a case of MA-M found in a non-NF2 patient presenting with convulsions. We briefly elaborate the clinical course, radiological and immuno-histochemistry features, focusing on the importance of surgical outcome.

Case Report

A 4-year-old female child presented to our department following seizure activity. She had been experiencing seizures for over a year. Seizures were accompanied by convulsions, repeated vomiting and loss of consciousness for which she was medically managed with Levetiracetam. Initially, she presented with partial-onset seizures which over time converted into generalized tonic-clonic seizures. MRI of the brain revealed a cortically based mass within the left frontal lobe with surrounding oedema. The mass was of mixed-intensity, but mainly hyperintense on T2- weighted imaging and hypointense on T1- weighted imaging. Post-contrast images showed irregular enhancement of lesion with the overall appearances of a primary brain tumor. An EEG was also done which suggested epileptiform activity arising from the region where the mass was located.

The patient underwent a lateral supraorbital craniotomy and complete excision of tumor was done which was then confirmed by an ultra-sound intra-operatively and MRI post-operatively. Histopathology reported central nervous tissue with two distinct meningiothelial components. One component was a *transitional meningioma* (WHO grade 1), the other component had features of *MA*.

Follow-up imaging with MRI was performed which showed no signs of re-growth. After one year of follow-up, the patient with respect to seizures after surgery was classified as Outcome Class 1 (free of disabling seizure), according to the Engel Epilepsy Surgery Outcome Scale.

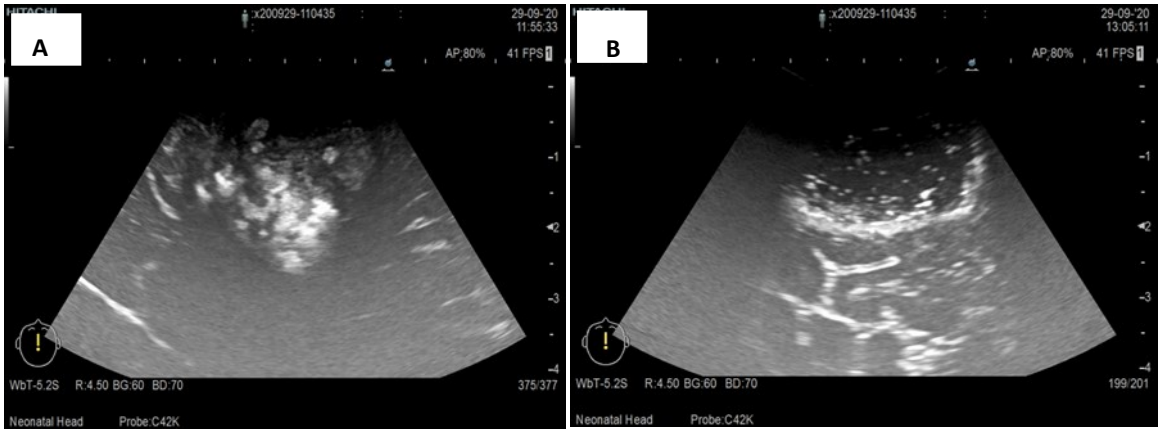

Figure 1: *USG findings corroborating to our case pre and post-operatively*

A: Intra-operative Ultra-sound image pre-resection of tumor

B: Intra-operative Ultra-sound image after resection of tumor

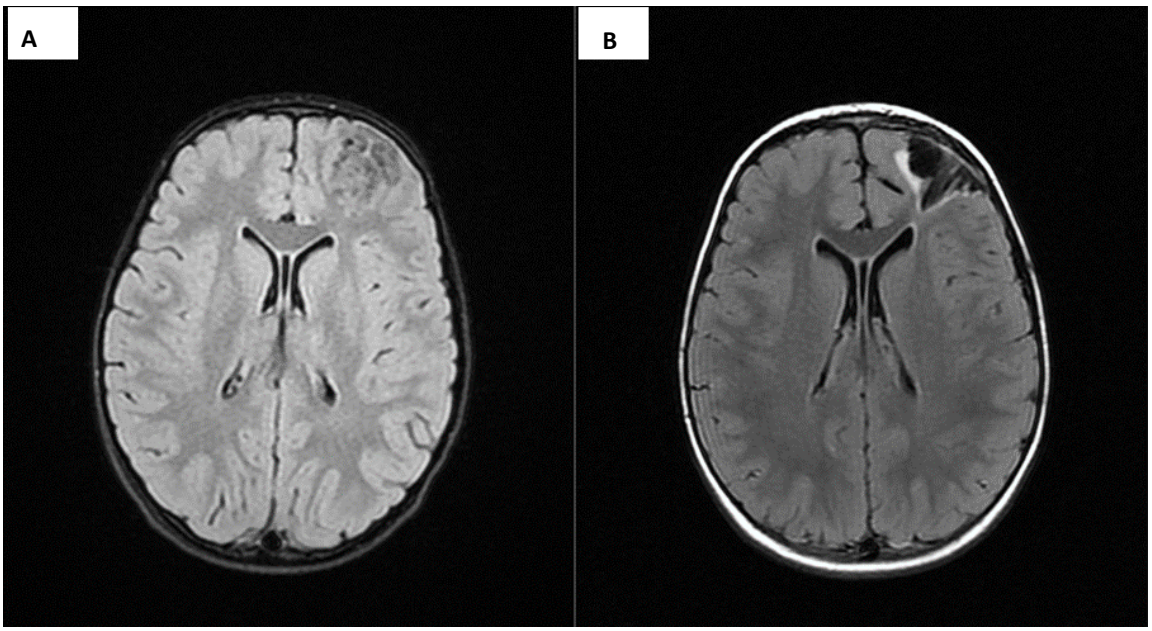

Figure 2: *Representative MRI findings pre and post-operatively in our case of MA-M*

A: Pre-operative MRI of the brain revealed a cortically based mass within the left frontal lobe with little surrounding oedema
B: MRI of the brain post-operatively, with complete resection of the tumor

Immuno-Histochemistry

Histology of paraffin section showed central nervous tissue with two distinct meningiothelial components. One component showed compact and syncytial growth pattern, tumor cell with small oval shaped nuclei and loosened chromatin structure. There was increased mitotic activity, and it also showed several interspersed round calcifications which could be identified as psammoma bodies. The other meningiothelial component showed a focally vascular association within central nervous tissue with gliotic changes. There was no necrosis or vascular proliferation. Immunohistochemistry of both components showed an expression of Vimentin while EMA and SSTR2 were only expressed in the compact meningioma component and not in the MA component. Interspersed central nervous tissue with immunoreactivity for Glial Fibrillary Acidic Protein (GFAP), Microtubule-associated protein 2 (Map2) and synaptophysin.

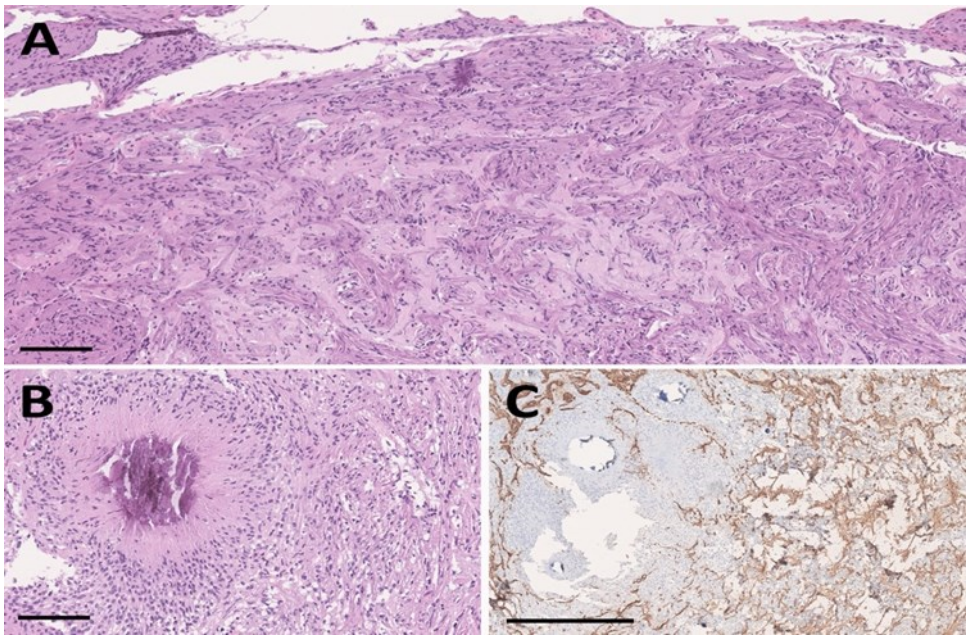

Figure 3: *Representative histopathology findings in our case with sporadic MA-M.*

A: Leptomeningeal and intracortical growth of MA. Hematoxylin-eosin staining. Scale bar=250μm. B: Meningioma component with psammoma body on the left and transition zone to MA. Hematoxylin-eosin staining. Scale bar=250μm. C: GFAP-immunohistochemistry capturing the MA-M aspects of this rare tumor at low power magnification. Hematoxylin-eosin staining. Scale bar=500μm.

Discussion

MA is a focal intra-cranial lesion which was elucidated as an incidental autopsy finding in a patient with neurofibromatosis-2 by Bassoe and Nuzum in 1915 [8]. Most studies conclude that MA commonly occurs in children, adolescents and young adults who suffer from intractable seizures and headaches [2,6]. The clinical hallmarks of MA can be neurofibromatosis, intracranial calcification or pathological vessels at angiography.

Although still a rare occurrence MA-M is the most frequently observed combination, with approximately 40 cases previously reported [16]. It has been studied that seventy percent of all MA-M lesions occur in the fronto-temporal region, and the sole most common site being the temporal lobe. Histopathologically, meningiomas occurring with MA are mostly of the transitional subtypes, though they can be of fibroblastic , meningiothelial, atypical, microcystic, and sclerosing subtypes [18].

It is difficult to determine the histogenesis of MA-M, but Kasatikul and Brown have suggested three possibilities; direct invasion of meningioma into the brain, a hamartomatous origin and malformative-angiomatous tissue developing perivascular meningiomatous components [3].

The differential diagnosis of MA associated with meningioma includes cortical invasion by meningioma and intracerebral schwannoma [16,18,19]. These two kinds of lesions can be easily distinguished by immunohistochemistry staining.

Recent studies suggest that loss of 22q12 (NF2 gene) and loss of heterozygosity have been found in pure MA and MA-M, aiming at it's pathogenesis [9,19]. This pattern of spread may be facilitated by meningiomas that are predominantly leptomeningeal or intracerebral in origin [6,19]. However, this needs further study.

Total removal of the MA-M could achieve reduction in epileptic seizure, so it was the focus in this case. The seizure outcome post-surgery after 1 year of follow-up was established as Outcome Class 1 (free of disabling seizure) according to Engel Epilepsy Surgery Outcome Scale. The epileptogenicity of tumor was documented during the EEG, and therefore, its complete resection was important for the proper control of seizures and for better prognosis.

Conclusion

Surgical resection is an adequate therapy strategy for MA associated with meningioma. The prognosis of patients with MA associated with meningioma depends on the histopathological grade of meningioma and the radicality of the operation. Grade I MA-M should be operated on taking into consideration its neoplastic origin, epileptogenicity and time of detection. Especially in cases where they are epileptogenic in origin, they should be operated on. Gross total resection is possible, and once achieved can lead to better prognosis and improved quality of life.

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