NEUROSURGICAL TREATMENT OF SPORADIC MENINGIOANGIOMATOSIS

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Received: November 21, 2009
Accepted: December 11, 2009

Pedijatrija danas 2010;6(1):82-86

Intracranial meningioangiomatosis is a rare, benign, focal cortical disorder characterised by a proliferative process. There is often diagnostic confusion with meningiomas or low grade glioma. Potentially amenable to surgical cure, early recognition of this condition is key to avoiding unnecessary alternative therapies. We present here the first case of meningioangiomatosis diagnosed and treated microsurgically in Bosnia and Herzegovina. A 13-year-old boy, previously well and having no family history of note, presented with a 2-year history of intractable seizures unresponsive to medical therapy. There was associated headache but no focal neurological deficit. Brain MRI showed a cortical lesion located on the medial side of the dominant frontal lobe. An interhemispheric frontal approach through a key-hole craniotomy was used and total resection of the lesion was performed. Intraoperatively, the specimen was noted to be of firm consistency with well-demarcated edges and minimal vascularity. Light microscopy showed spindle-shaped cells proliferating in a fascicular pattern. Reticulin was abundant in these areas. In the background, multiple foci of sammoma bodies were noted. Conclusion The boy remained seizure-free and off antiepileptic medications postoperatively. There was no recurrence radiologically or clinically in the 20-month follow up period.

Key words: Meningioangiomatosis ▪ Sporadic ▪ Neurosurgery

Introduction

Meningioangiomatosis (MA) is a rare, focal cerebral benign disorder of a hamartomatous nature characterised by a proliferative process. MA affects children and young adults, with a preponderance in males. There is often diagnostic

confusion with invasive meningiomas (increase in meningothelial cells within the leptomeninges) or low-grade astrocytoma (astrocytic proliferation). The lack of cellular atypia, nuclear pleomorphism and mitotic activity serve to establish MA as a vascular hamartomatous disorder. Sporadic cases tend to present with solitary lesions in the frontal or temporal lobe and these are more common. The multifocal disease is particularly closely associated with neurofibromatosis type 2. The aetiology and pathogenesis remain unclear. There are three types of cells that have been proposed individually as possible cells of origin: meningothelial, fibrobast and perivascular connective tissue cells. Potentially amenable to surgical cure, early recognition of this condition is key to avoiding unnecessary alternative therapies. The first case of sporadic intracranial meningioangiomatosis diagnosed and treated microsurgically in Bosnia and Herzegovina is presented.

Case presentation

A 13-year-old boy, previously fit and well and having no family history of note, presented with a 2-year history of intractable seizures unresponsive to medical therapy. There was associated headache but no focal neurological deficit. Brain MRI showed a cortical lesion located on the medial side of the dominant frontal lobe. An interhemispheric frontal microsurgical approach through a key-hole craniotomy was used and total resection of the lesion was performed by the author (Figure 1).

Intraoperatively, the specimen was noted to be of firm consistency with well-demarcated edges and minimal vascularity. The intraoperative mesoscopic impression of the lesion was that of a low-grade astrocytoma. The lesionectomy specimen, measuring 4 x 2.5 x 2 cm was of whitish color and firm consistence. Histologically, the lesion was confined to the cortex, with focal involvement of the overlying leptomeninges. Its central part was moderately to highly cellular (Figure. 2a) with focal storiform pattern, and rare whorl formations, (Figure 2b) and psammoma bodies (Fig. 2c).

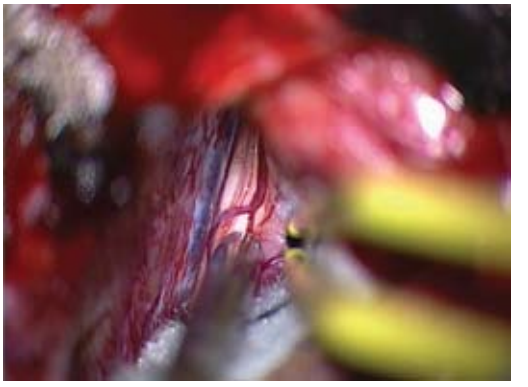

Figure 1 *Intraoperative image. An interhemispheric microsurgical frontal key-hole approach. Operation performed by the author*
Slika 1 *Intraoperativna slika. Interhemisferični mikrohirurški frontalni minimalno invazivni pristup. Operacija izvedena od strane autora*

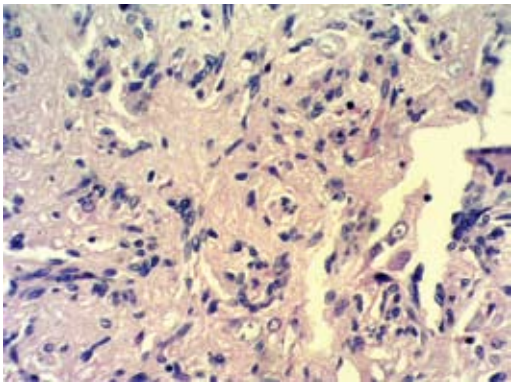

Figure 2a *Highly cellular areas showing storiform growth pattern. There is complete effacement of the cortical architecture. HE x 40*
Slika 2a *Područja bogata ćelijama koja pokazuju storiformni obrazac rasta. Postoji kompletno zbrisana kortikalna arhitektura. HE x 40*

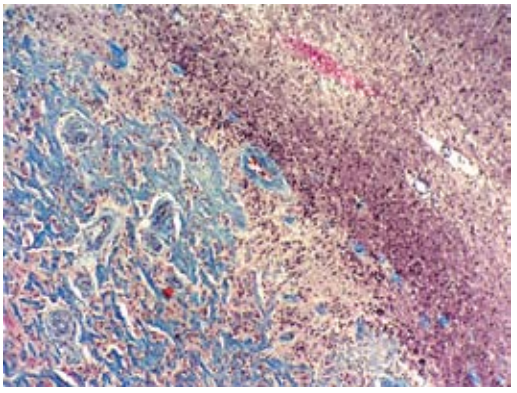

Figure 2b *Vague storiform growth pattern with occasional cellular whorl formation (Masson Trichrom x 20)*
Slika 2b *Neodređeni storiformni obrazac rasta sa mjestimičnim celularnim vrtlozima (Masson Trichrom x 20)*

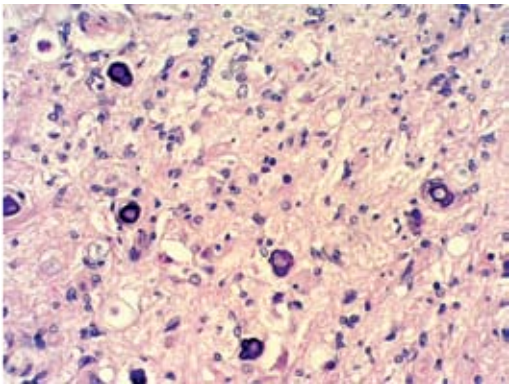

Figure 2c *Spindle-shaped cells proliferating in a fascicular or storiform pattern. In the background, multiple foci of psammoma bodies were noted (hematoxylin and eosin ×40)*
Slika 2c *Vretenaste ćelije koje proliferiraju u fascikularnom ili storiformnom obrascu. U pozadini se vide multipli fokusi psamoznih tijela (hematoksilin i eosin ×40)*

The perivascular arrangement of cells was more evident in the periphery of the lesion (Figure 2a). Blood vessels were increased in number; some had slit-like lumina, whereas others possessed a thickened, hyalinized wall. (Figures 2a and 2b) They were surrounded by cuffs of cells, the appearance of which ranged from meningothelial to spindled, fibroblastic (Fig 2a and 2b). Immunohistochemically, there was positivity for Vimentin. The patient remained seizure-free and off antiepileptic medications postoperatively. Postoperative MR images of the brain showed complete resection of the lesion (Figure 3). There was no recurrence radiologically or clinically in the 20-month follow-up period.

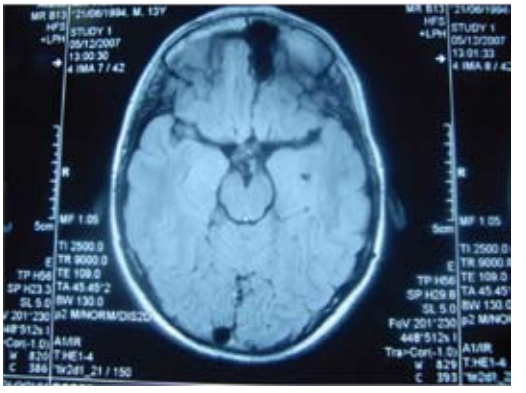

Figure 3 *Postoperative MRI of the brain*
Slika 3 *Postoperativna MRI slika mozga*

Discussion

Meningioangiomatosis is a rare cerebral disorder of a hamartomatous nature. Since its first description in 1915 and subsequent naming by Worster- Drought et al., there have been less than a hundred reported cases in literature. This is a non-neoplastic lesion that is histologically classed into two types: the predominantly vascular and the predominantly cellular forms. The association with neurofibromatosis 2 (NF 2) forms the basis for one theory on the underlying pathogenesis. The gene for NF 2, a tumour suppressor located on chromosome 22, is believed to give rise to the other neoplasms that characterise the disorder – schwannomas, meningiomas and gliomas. The characteristic histology forms the mainstay of diagnosis. Radiology and intraoperative findings vary widely and differential diagnosis includes invasive meningioma, astrocytoma or even vascular malformation.

The marked increase in meningothelial cells within the leptomeninges can be mistaken for that of an invasive meningioma. Astrocytic proliferation can lead to misdiagnosis of astrocytoma. However, the presence of neuronal networks and the lack of mitotic activity should preclude such errors in diagnosis. Immunohistochemical staining is largely non-beneficial in establishing the diagnosis. Vimentin is one stain that has been reproduced in most patient samples. Only two additional markers, EMA and CD34, can be focally positive. The Ki-67 proliferation index is always below 0.1% . The main clinical presentation is recurrent seizures. Definitive treatment for MA is radical surgical resection of the lesion, which was achieved in our patient. Recurrence is only reported where there has been incomplete resection. Avoidance of alternative treatment modalities, as well as patient reassurance (given the benign, slow-growing natural history) have obvious benefits.

Conclusion

Meningioangiomatosis is a rare hamartomatous disorder that combines the histological features of a meningioma with those of a vascular proliferation. Immunohistochemical staining patterns vary between cases showing positivity only for Vimentin. Microsurgical total resection is the treatment of choice. Our patient remained symptom-free with the cure of seizures postoperatively

Conflict of Interest: The authors declare that they have no conflict of interest. This study was not sponsored by any external organisation.
Sukob interesa: autori su se izjasnili da nisu u sukobu interesa. Studija nije bila sponzorirana od neke vanjske institucije.

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Sažetak

NEUROHIRURŠKI TRETMAN SPORADIČNE MENINGIOANGIOMATOZE

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Intrakranijalna meningioangiomatoza je rijetki, benigni, fokalni, kortikalni poremećaj karakteriziran proliferativnim procesom. Često se može zamijeniti sa meningeomom ili niskogradusnim gliomom. S obzirom da se hirurški može izliječiti bitno je rano prepoznati ovu kondiciju radi izbjegavanja nepotrebnog alternativnog tretmana. Mi smo prezentirali prvi slučaj meningioangiomatoze dijagnosticiran i mikrohirurški tretiran u BiH. Ranije zdrav trinaestogodišnji dječak javio se nakon refraktarnih epileptičkih napada koji su trajali dvije godine i nisu reagirali na medikamentoznu terapiju. Postojala je glavobolja ali bez neurološkog fokalnog deficita. Načinjeni MRI mozga je pokazao kortikalnu leziju lociranu u dominantnom frontalnom lobusu sa medijalne strane. Načinjeno je totalno odstranjenje lezije kroz interhemisferični frontalni key-hole pristup. Lezija se intraoperativno prezentirala kao dobro demarkirana, čvršće konzistencije i minimalne vaskularnosti. Svjetlosna mikroskopija je pokazala fascikularni obrazac proliferacije vretenastih ćelija. U ovim područjima je bila velika količina retikulina. U pozadini su uočeni multipli fokusi psamoznih tijela. **Zaključak** Postoperativno je dječak bez epileptičkih napada i bez antiepileptičkih medikamenata. Tokom 20 mjeseci praćenja nisu uočeni radiološki niti klinički znakovi rekurensa.

Ključne riječi: Meningioangiomatoza ▪ Sporadična ▪ Neurohirurgija

Primljeno: 21. 11. 2009.
Prihvaćeno: 11. 12. 2009.