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Seizures Revealing Familial Meningiomatosis in Two Half Brothers in Pointe-Noire, Congo

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ABSTRACT

Meningiomatosis is defined as the presence of more than two meningiomas at different intracranial sites, in the same time in the absence of neurofibromatosis. Family forms are extremely rare. We report two cases of a family meningiomatosis in half brothers. The youngest presented a partial motor seizure with secondarily generalization and headache. Physical examination was normal. Cerebral MRI showed numerous T1 isosignal formations and multiples foci with hyperoedematous flair signal in T2 hyposignal enhanced after injection of contrast medium, suggestive of meningiomatosis. They received only symptomatic treatment. The prognosis is generally good with a good quality of life despite the multiplicity of lesions, the potential number of recurrences or multiple surgeries.

Keywords

Seizure, Meningiomatosis, Neurofibromatosis.

Introduction

Meningiomas are the most common primary intracranial benign tumors, accounting for up to 35% of brain tumors in adults. They develop from the meningothelial cells of the arachnoid [1,2]. Multiple meningiomas (MM) or meningiomatosis are characterized by the presence of more than two meningiomas at different intracranial sites, occurring at the same time in a patient with no neurofibromatosis [2,3]. Family forms without neurofibromatosis are extremely rare. We report 2 familial cases of multiple meningioma in two half brother patients.

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These are two half brothers who's the youngest, the third of a family of 3 children was 21 years old at the time of the seizure onset. He has no significant medical history. He was admitted to the emergency room for seizure. It was a right-sided partial seizure with secondary generalized tonic-clonic seizure. The seizure lasted 5 minutes and was followed by a sphincter relaxation, tongue bite and amnesia. The parents reported 3 consecutive seizures in less than 24 hours. Physical examination at presentation was normal, as well as a comprehensive dilated eye exam. Cerebral MRI showed numerous T1 isosignal formations and multiples foci with hyperoedematous flair signal in T2 hyposignal enhanced after injection of contrast medium, suggestive of meningiomatosis, suggestive of multiples meningiomas (Figure 1). Others investigations, including

the electroencephalogram, looking for peak-type paroxysmal abnormalities or slow theta waves, was normal. Cardiac, renal ultrasounds showed no significant abnormalities. Symptomatic treatment was initiated with carbamazepine 15 mg/kg PO q12h. The clinical course was satisfactory with seizure disappearance and the patient presents was symptom free at 6-month follow up consultation.

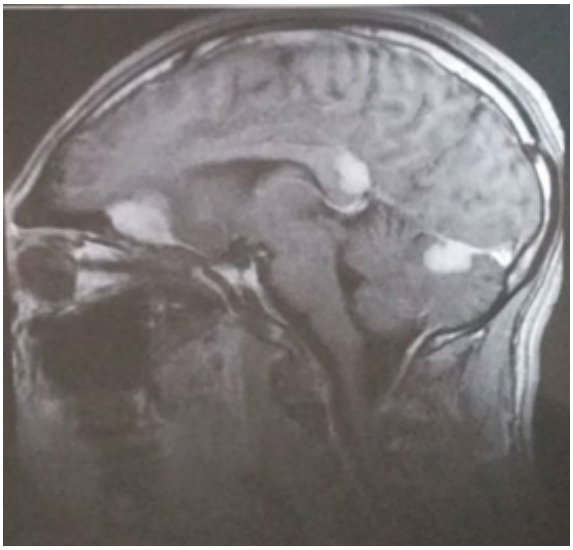

Figure 1: Cerebral MRI, sagittal section, T1 sequence with gadolium, evidence of hypersignals localized in vermix, corpus callosum and frontal lobe extra parenchymatous, evoking meningiomatosis.

His older half brother reported headache of moderate intensity with no associated symptoms, evolving in recent years. A history of a seizure one year ago but did not seek medical attention. The physical examination was normal. Cerebral MRI (Figure 2) demonstrated nodule formation in T1 signal and hyposignal flair, which enhanced after injection of contrast medium and a hyper signal flair, suggesting a meningiomatosis.

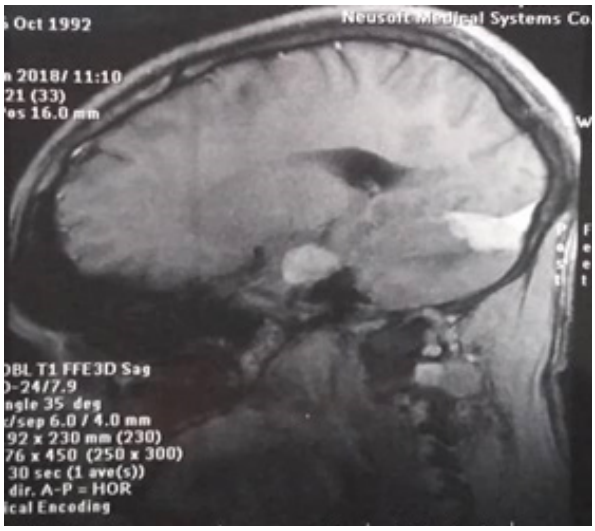

Figure 2: Brain MRI, axial section, T1-weighted sequence with gadolium, evidence of hypersignals at the hippocampal and cerebellar level resting on meninges suggestive of meningiomatosis.

The younger sister has no symptoms and investigations showed no abnormality.

Discussion

Multiple meningiomas or meningiomatoses have been described for the first time by Anfirmow and Blumenna [4]. It is important to differentiate between meningiomatosis related to neurofibromatosis (NF) and multiple meningiomas in the absence of a familial history [1,2,4]. Few studies have been found in literature. In Morocco, a series of 4 patients has been described [5]. In subsaharan Africa, no publication has been found. We describe the two familial cases of meningiomatosis in Pointe-Noire.

Multiples meningiomas show greater female preponderance and higher expression of progesterone receptors [6], both of our patients were male.

The mechanisms explaining the development of meningiomatosis are still poorly known, the mutation of the tumor suppressor gene NF2 plays an important role, both in sporadic forms and in multiple meningiomas associated with Type 2 neurofibromatosis. The Genetic factor is therefore certainly one of the most important causes for the development of multiple meningiomas. Numerous studies confirm the mutation of the NF2 gene and the deletion of chromosome 22 [7,8].

In the literature, familial forms of meningiomatosis without type 2 neurofibromatosis rare extremely rare. We describe the family case of two half brothers from the same father and different mothers. Aavikko et al. reported a case of familial meningiomatosis, affecting 5 children of the same parents secondary to a mutation in the gene SUFU on chromosome 10 [9].

The symptoms at presentation are similar to a solitary meningioma, and are usually represented by seizures, headaches, neurological deficits or visual disturbances. They can remain asymptomatic [3,7,8], this is the case of our patients whose seizures and headache were the telltale symptoms.

The localization of the tumors of our patients was bilateral, on the supratentorial level and at the posterior cerebral fossa. However this localization is not uncommon as showed the studies, although there is a predominance for the supratentorial localisation, cerebellar convexity or falx cerebri [10].

The natural history of multiple meningiomas is similar to that of meningiomas. The evolution curve, in terms of the volume of multiple meningiomas, does not appear to be greater than of isolated meningiomas, suggesting clinical and radiological follow up is sufficient for asymptomatic lesions [11]. Several authors have suggested that surgery is only indicated for symptomatic lesions, the rest should benefit from clinical and imaging follow-up every 6 to 12 months. Our patients have benefited from symptomatic treatment and they were symptom-free at 6 month follow up consultation.

For diagnosis confirmation pathological exam is needed. It is noted that in the majority of meningiomas are WHO grade I, suggesting a better prognosis despite the high number of tumors. Multiple cases of multiple meningiomas with different grades have been reported in the same patient [12].

Multiple meningiomas are benign tumors in the vast majority of cases. The prognosis is usually good and does not differ from solitary meningiomas, except in the case of radiation-induced meningiomatosis in children or when they are part of type 2 neurofibromatosis [13].

Conclusion

Familial meningiomatosis is a rare entity. Like solitary meningiomas, they are more common in women. Seizures are usually the the symptom at presentation. Their anatomical localization is variable, often unilateral and at supratentorial level even if posterior fossa localisations have been described. In the end, despite the multiplicity of tumors, the potential number of recurrences, or multiple surgeries, patients have a fairly good quality of life and prolonged survival.

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