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Case Report

Meningioangiomatosis without neurofibromatosis simulating encephalitis in neuroimaging

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Abstract

Background: Meningioangiomatosis (MA) is a rare entity characterized by a focal lesion that affects the leptomeninges and the cerebral cortex.

Case Description: We describe a case of a 32-year-old man diagnosed with MA not associated with hamartomatous lesions or with type 2 neurofibromatosis. Magnetic resonance images (MRI) showed an extensive parieto-occipital lesion and another right frontal lesion, initially suggestive of encephalitis. A biopsy of the meninges and brain was performed via a right parieto-occipital craniotomy. The histopathologic diagnosis, complemented by immunohistochemical studies, was MA.

Conclusion: Diagnosis of MA is very difficult based only on images, therefore lesions compromising the brain cortex, associated or not with calcifications, should be further examined through biopsy so as to have a precise diagnosis.

Key Words: Cerebral cortex, hamartoma, leptomeninges, meningioangiomatosis

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INTRODUCTION

Meningioangiomatosis (MA) is a rare entity characterized by a focal lesion that affects the leptomeninges and the cerebral cortex. It can be misdiagnosed as a brain tumor both clinically and radiologically and is usually associated with hamartomatous focal lesions. MA is considered benign, with no reports of malignancy until now, but its histogenesis remains uncertain.[6]

MA was initially described in patients presenting type 2 neurofibromatosis (NF2) in whom it is clinically silent and often diagnosed at autopsy.[4,12] A few cases have been published on patients without NF2, usually presenting with medically refractory seizures, which usually improve after total resection of the lesion. Most of those patients had a single lesion characterized by meningovascular proliferation and leptomeningeal calcifications.[4]

The present paper reports a patient diagnosed with MA not associated with hamartomatous lesions or NF2.

CASE REPORT

A 32-year-old man was admitted with simple partial seizures described as left hemifacial and left arm

automatisms, which he reported having had for 1 week. Clinical and neurological examinations were normal and he presented no signs or symptoms of NF2.

The patient developed status epilepticus, being initially treated with endovenous phenytoin and diazepam. He was transferred to the intensive care unit (ICU), so as to better control his seizures. Computerized tomography (CT) scan showed a right frontal nonenhancing hypodense lesion. Magnetic resonance images (MRI) revealed an extensive parieto-occipital lesion and another right frontal lesion, initially suggestive of encephalitis [Figure 1].

Liquor lab tests were normal. An open biopsy of the meninges and brain was performed via a right parieto-occipital craniotomy. The histopathologic diagnosis, complemented by immunohistochemical studies, was MA, characterized by diffuse proliferation of elongated perivascular, fibroblast-like cells in the cerebral cortex [Figure 2]. Immunostaining of smooth muscle actin or epithelial membrane antigen in the perivascular cells was not observed; however, immunostaining of CD34, a marker for endothelial cells, was positive only in the endothelial cells that normally cover the intima [Figure 3]. The cytoarchitecture of the cerebral cortex was normal, but selective neuronal ischemic necrosis of isolated cells and reactive fibrillary astrocytosis were observed. The meninges sample was also normal.

Continuous sedation was necessary and kept for 28 days so as to control the seizures. After this period, the patient left the ICU, being medicated with phenytoin (500 mg/day), clonazepam (2 mg/day), and valproic acid (1500 mg/day). He had important neurological sequelae caused by ischemic lesions due to the status epilepticus. The patient was seizure-free for 3 months. Thereafter, seizures recurred, remaining refractory to antiepileptic drugs and the patient died.

DISCUSSION

MA was originally described in 1915 as an incidental finding during autopsy, associated with NF2. However, the nomenclature was only established in 1937.[2,18] MA most commonly affects children and young adults and the most usual clinical manifestations are refractory seizures and headache.

Even though its histology is unique, its histogenesis and pathogenesis remain controversial.[14] Three hypotheses have been proposed: a) the lesion is a hamartoma that results in degenerative alterations in the encephalic parenchyma; b) the lesion is caused by the invasion of the cerebral cortex by a leptomeningeal meningioma; c) the lesion is a vascular malformation involving secondary proliferation of meningeal cells.[3,7-9]

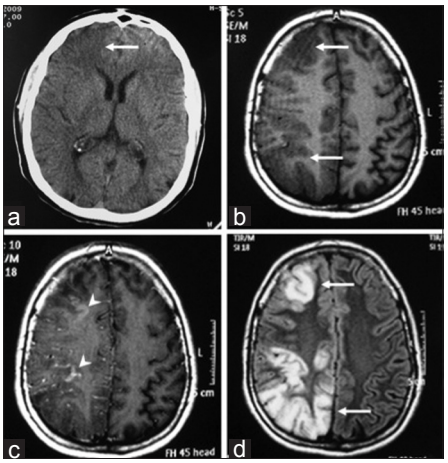

Figure 1: (a) CT scan. Axial slice shows right frontal hypodense lesion without calcifications (white arrow). (b) Axial MRI. T1-weighted image reveals an isointense lesion (white arrows). (c) Axial MRI. T1-weighted image with contrast agent shows gyriform enhancing (arrowheads). (d) Axial MRI. FLAIR-weighted image shows a hyperintense signal

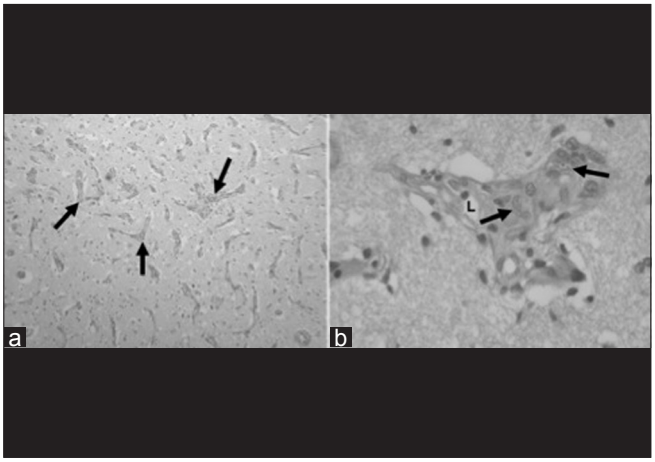

Figure 2: Cerebral cortex. (a) Diffuse perivascular proliferation (arrows). HE, Objective ×4. (b) Thickening of vascular wall and reduction of the lumen (L) by proliferation of perivascular, elongated, fibroblast-like cells (arrows). HE, Objective ×40

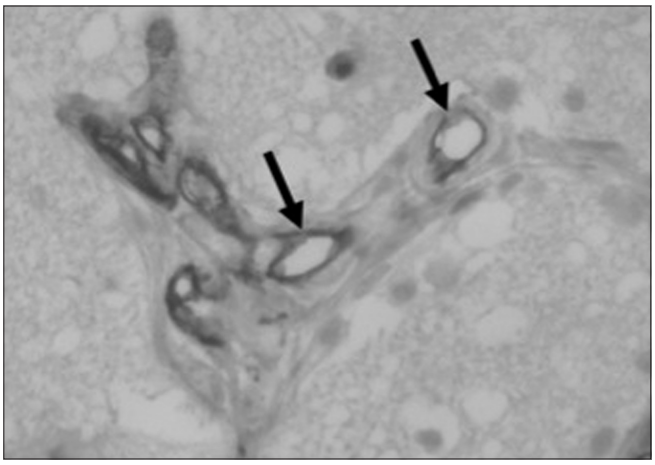

Figure 3: Cerebral cortex. Reactivity of the endothelium by anti-CD34, a marker for endothelial cells (arrows). Objective ×40

MA affects the cerebral cortex of the frontal and temporal lobes in approximately 90% of all cases, though it is occasionally observed in deep encephalic structures 4. In patients without NF2, the lesion is usually a single, solid, well-defined mass that presents slow growth.[11]

Multiple lesions tend to occur in patients with NF2 and can vary from microscopic lesions to large, macroscopically visible nodules. Radiological findings are nonspecific, while computed tomography (CT) scans show different characteristics from isodense to slightly hyperdense images, with or without calcifications.[1,15] Magnetic resonance imaging (MRI) also reveals different characteristics. This wide spectrum in the imaging exams makes clear clinical diagnosis difficult.[16] In the present case, MRI showed an extensive parieto-occipital lesion and another frontal right lesion, giving the initial impression of a patient with diffuse encephalic compromise suggestive of encephalitis.

The main histological characteristic of MA is meningovascular proliferation interlaced with layers of fibrous tissue associated with leptomeningeal calcification.[10] The histological spectrum can vary expressively, from cellular to vascular predominance, which may correspond to the evolutive stages of MA.[13,17]

In the present case, only angiomatoid microvascular proliferation was observed associated with diffuse perivascular proliferation of elongated, fibroblast-like cells that affected the cerebral cortex, with no leptomeningeal calcifications. Immunohistochemistry has limited diagnostic value in MA due to variable staining patterns.[17] In the case presented, immunostaining for epithelial membrane antigen, a meningothelial cell marker, was negative, in agreement with MA cases constituted only of elongated, fibroblast-like cells (Burger and Scheithauer, 2007). Similarly, positive immunostaining restricted to intima endothelial cells excludes the participation of endothelial hyperplasia in the genesis of perivascular proliferation.

The maintenance of the cortical cytoarchitecture in MA depends on the ratio of meningothelial/fibroblast-like perivascular proliferation to brain parenchyma, which is obscured proportionally by downgrowth of the lesion. The maintenance of the cortical cytoarchitecture associated with the proliferation of elongated, fibroblast-like cells in the present case, without the presence of meningothelial cells, suggests that it may correspond to an initial evolutive stage of MA (Suarez *et al*., 2006; Wiebe *et al*., 1999). On the other hand, the ischemic neuronal lesions observed could be explained by the status epilepticus presented by the patient.[5]

In most cases, treatment is surgical, involving resection of the lesion to control the seizures. However, seizures can persist in many patients, with control achieved by administration of antiepileptic drugs, a fact that suggests an extralesional epileptogenic area.[17] Surgical resection was not possible in the present case due to the extensive cortical compromise.

In conclusion, diagnosis of MA is difficult based only on imaging exams, but it should be considered in lesions affecting the brain cortex, associated with calcifications or not. Differential diagnoses of MA include vascular malformations, meningioma, gliomas, and granulomatous process.[6] Whenever possible, the treatment of choice is the surgical approach, often in association with antiepileptic drugs. In cases involving extensive multiple lesions without calcifications, alterations in the imaging exams can be suggestive of encephalitis.

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