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Intracranial Angiomatous Meningioma: Case Report and literature review

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ABSTRACT

Angiomatous meningioma (AM) is a rare subtype of meningioma characterized by highly vascular tumor tissue comprising predominantly variable sized hyalinized blood vessels. This manuscript describes a case of intracranial Angiomatous meningioma and reports briefly its clinical, radiological, histopathological and therapeutic characteristics through a literature review.

Keywords: Angiomatous meningioma, intracranial tumor, case report.

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INTRODUCTION:

Angiomatous meningioma is a rare subgroup of meningiomas in which numerous vascular channels prevail on the background of an otherwise typical meningioma, it belongs to WHO grade I category and accounts for 2.1% of all meningiomas. Although angiomatous meningiomas share similar clinical features and prognosis with benign meningiomas, they have some unique characteristics such as, relative high male to female ratio compared with meningiomas in general, more frequent peritumoral edema, and rich blood vessels in the tumor. Gross total resection is still the treatment of choice. Patients with residual tumor after surgery can benefit from radiation therapy. Overall, the prognosis of angiomatous meningiomas is as good as other benign meningiomas.

PATIENT AND OBSERVATION:

37 years old male patient presented with headaches resistant to analgesic treatment since 6 months. His general and central nervous system (CNS) examination was normal. Magnetic Resonance Imaging (MRI) of brain showed extra axial median process at left parasagittal region well circumscribed of 43mm of great axis in hyposignal T1 in hypersignal T2 with intense and homogeneous enhancement

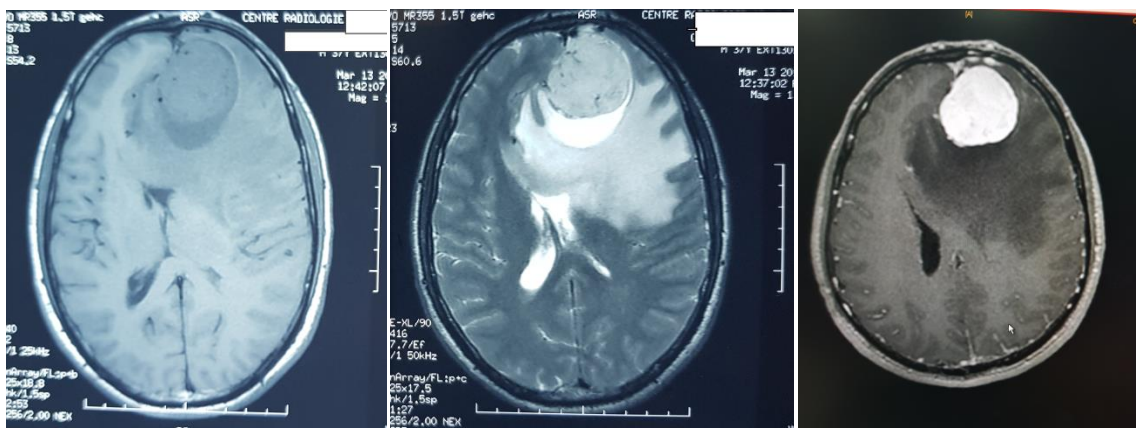
after injection of gadolinium, this lesion is in contact with the superior longitudinal sinus, it is accompanied by an important perilesional edema and important mass effect on the median structures (Figure 1). Patient underwent surgical biopsy. The tumor tissue was sent to histopathology department. Microscopic examination showed tumor proliferation made of fusiform cells arranged in layers or bundles with storiform clusters. these cells are provided with regular nuclei devoid of atypies and mitoses and eosinophilic cytoplasm, poorly limited; the tumor stroma is richly vascularized with numerous capillaries representing more than half of the tumor, sometimes with a hyalinized thick wall. Final impression was given as angiomatous meningioma (WHO grade I). The patient did not receive tumor resection because of risk of hemorrhage of the tumor, he benefited from radiation therapy at the total dose 50,4Gy in 28 fractions.

DISCUSSION

In adults, 15–18% of all intracranial tumors and 33% of all incidental intracranial neoplasms are meningiomas [1]. Angiomatous meningioma belongs to WHO grade I category and accounts for 2.1% of all meningiomas [2].

Among the population of benign meningioma, there was usually a female predominance [3].

Figure 1: extra axial process at left parasagittal region in hyposignal T1 (A) in hypersignal T2 (B) with intense and homogeneous enhancement after injection of gadolinium (C), with an important perilesional edema.



A: T1-weighted axial MRI

B: T2-weighted axial MRI

C: T1-weighted with Gadolinium

AM is usually characterized by the onset of slow progressive symptoms and the main symptoms result from compression of the adjacent structures. Few patients showed rapidly progressive symptoms, which may be a reason for the relatively long period between the onset of symptoms and admission time. Like other benign meningiomas, AM can locate in almost all intracranial areas as well as the spine, with cerebral convexity being the most common location [3].

Meningiomas show isointensity or hyperintensity to the cerebral cortex in magnetic resonance imaging (MRI). The most important diagnostic feature is short extension of contrast enhancing tissue along the dura which is also known as dura tail [1]. Angiomatous meningioma seems to have more obvious signal voids of vessels in the tumor on MRI [4].

Radiographically, there is no other additional feature to help in subclassification of meningiomas, though angiomatous meningioma (in spite of belonging to WHO grade I) shows perilesional edema. Contrast enhancement is a feature seen in glioma, hemangioblastoma, and typical meningioma. Perilesional edema is usually seen in atypical meningioma but when seen in this variant it is not a sign of atypia or malignancy [2]. Our case showed contrast enhancement and perilesional edema.

On histopathology, angiomatous meningioma is characterized by abundance of well-formed vascular channels, sinusoids or capillaries. The study by Martin et al.[5] showed two subtypes of angiomatous meningioma, viz., microvascular subtype (more than 50% contain vessels with diameter below 30 μ), and a macrovascular subtype. These tumors may also display microcystic change along with foamy cells (which are related to leakage of plasma lipids across thin vessel walls); in addition to solid areas with meningotheliomatous meningioma elements[1]

World health organization (WHO) has subclassified all central nervous system (CNS)

tumors including meningioma into various grades. Meningiomas have been categorized into grades I, II, and III based on increased cellularity, high nucleocytoplasmic ratio, large prominent nucleoli, patternless sheets, mitosis, and spontaneous or geographic necrosis. Hence, a workup would be incomplete without the assessment of grade. Counting the mitotic figures is quite subjective and an objective method of evaluating proliferative activity is by performing Ki-67/MIB-1 immuno-staining on tissue sections [2]. In our case, the tumor was reported as angiomatous haemangioma WHO grade I

The differential diagnosis of angiomatous meningioma includes vascular tumors like hemangiopericytoma and capillary hemangioblastoma [2]. But microscopically, haemangiopericytoma consisted of tightly packed spindle shaped cells surrounding ramified thin-walled endothelium-lined vascular channels with characteristic 'antler' or 'staghorn' configuration [1]. Capillary hemangioblastoma microscopically is characterized by thin-walled blood vessels lined by plump endothelial cells and separate groups of polygonal stromal cells. Immunohistochemical stains play a crucial role in differentiating hemangiopericytoma from angiomatous meningioma and hemangioblastoma. Hemangiopericytoma are immunoreactive to vimentin and endothelial antigen CD34 but stain negatively with Epithelial Membrane Antigen (EMA). Stromal cells of hemangioblastomas are immunoreactive to vimentin, neuron-specific enolase, S100, Glial fibrillary acidic protein, and calponin but fail to stain with EMA and prognosis is generally excellent. Angiomatous meningioma is immunoreactive for EMA, vimentin and S-100 protein, which confirm a diagnosis of angiomatous meningioma [1].

Abedalthagafi et al. [6] performed high-resolution array comparative genomic hybridization on 16 AMs and found that AMs had distinct genomic features, with polysomies

of at least 1 chromosome. This factor may apply to clinical diagnosis in the future⁸ ; however, it will not change the current treatment strategy [3].

Gross total resection is the treatment of choice [1]. In our case the patient only benefited from a surgical biopsy.

For cases with residual tumor after surgery, radiation therapy can be used for treating these patients [4]. The molecular basis of the effectiveness of radiation therapy is that radiotherapy can inhibit the expression of vascular endothelial growth factor (VEGF) and somatostatin receptor, which causes blood vessel contraction to reduce blood supply and then shrink the tumor [7]. Nicolato A et al reported that, in the 122 cases of cavernous sinus meningiomas who received gamma-knife stereotactic radiosurgery, 118 (97%) cases had stable clinical symptoms or greatly improved [8]. Subach et al think that patients with residual tumor, recurrent tumors, or tumor progression after subtotal resection, and inoperable patients, can receive the radiosurgery [9]. In our case, the patient received postoperative radiotherapy at the total dose 50,4Gy in 28 fractions.

Grade 1 meningiomas have a favorable prognosis. Since angiomatous meningioma belongs to this group, it has a similar behavior.

CONCLUSION:

Angiomatous meningioma is a rare variant of meningioma with few distinctive clinical, radiological, histopathological, and immunohistochemical features. Histological examination and immunohistochemistry play principal role in confirming diagnosis. Gross total resection is still the treatment of choice. Patients with residual tumor after surgery can benefit from radiation therapy.

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