Angiomatous Meningioma – a rare variant

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ABSTRACT

Angiomatous meningioma is a rare WHO grade 1 histological subtype of meningioma, comprising of 2.1% of all meningiomas. Metastasis from a histologically “benign” meningioma is a rare but well documented event. In 60% of the cases the metastases are in the lungs. They have the histological and clinical features of benign meningioma in which the blood vessels components exceed upto 50%. They are rarely reported in the literature and till now their clinico-pathological characteristics have not been systematically analyzed and studied. The main aim of presenting this case is to highlight the histomorphological features of this uncommon variant of meningioma that could help in distinguishing it from Hemangioblastoma and Hemangiopericytoma.

Keywords: Angiomatous meningioma, uncommon variant, hemangiopericytoma.

INTRODUCTION

Meningioma is regarded as a benign neoplasm, which commonly arises from the arachnoid cap cells of the cerebrum and the spinal cord and constitutes 15% of all intracranial tumors1. It is known, however, that in 0.1% of the cases the meningioma metastases. It is also known that in 60% of the cases the metastases are in the lungs2,3. Primary intraventricular meningiomas are very rare and account for 1-2% of all cases3. In nearly 78% of the cases they are localized in the lateral ventricle, in 15% in the third and in 6% in the fourth ventricles4. Angiomatous meningioma is a rare subgroup of meningiomas in which numerous vascular channels prevail and constitute 2.1% of all meningiomas5.

CASE REPORT

A 50yr old male presented with headache and difficulty in swallowing since one month. Contrast magnetic resonance imaging (MRI) study showed extra axial lesion with dural tail heterogenous enhancing lesion noted in right fronto temporal region with moderate perilesional vasogenic edema causing compression of ipsilateral lateral ventricle and midline shift of 6 mm towards left side(Figure 1). Suggestive of either an Angioblastic meningioma with internal bleed or High grade Glioma in view of dural tail. Surgical excision of tumor from right sphenoid wing was done and specimen was sent for histopathological examination. Microscopy showed highly vascular tumor consisting predominantly small capillaries constituting more than 50% of lesion and also showing meningiothelial component in the background and tumor did not show any endothelial proliferation or mitotic activity and it was diagnosed as Angiomatous meningioma. (Figure 2,3 & 4)

DISCUSSION

Angiomatous meningioma is a rare subgroup of meningioma’s in which numerous vascular channels prevail on the background of an otherwise typical meningioma6.
Meningiomas show isointensity or hyperintensity to the cerebral cortex in MRI. Short extension of contrast enhancing tissue along the dura (dura tail) is a valuable diagnostic feature. 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

The clinical data, radiological, and histopathological features were suggestive of either of the three lesions with a similar pattern; these were angiomatous meningioma, Hemangioblastoma, and Hemangiopericytoma. Angiomatous meningioma shows similar pattern of immunoreactivity as that of a typical meningioma. It shows positive staining with antibodies to vimentin, desmoplakin, and EMA, and focal positive reaction to antibodies to progesterone. Further confirmation can be done by electron microscopic study. Ultrastructurally, neoplastic meningeal cells have prominent cytoplasmic processes and well-defined junctions. Angiomatous meningioma can have foamy cells which are related to leakage of plasma lipids across thin vessel wall. A meningioma that is entirely hemangioblastic cannot be distinguished from hemangioblastoma except by its attachment to dura, immunohistochemical markers, and electron microscopy.

WHO has subclassified all 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 immunostaining on tissue sections. The present case has no proliferative activity confirming them to be WHO grade I.

Hemangiopericytomas are dural-based lesions which are often confused clinically with meningioma. Meningeal hemangiopericytoma was earlier described as angioblastic variant of meningioma, but it was Begg and Garret who recognized that it was in fact a hemangiopericytoma arising within meninges. They account for 2-5% of meningeal lesions, occur commonly in adulthood and found more commonly in males. Image findings of hemangiopericytoma are similar to meningiomas. Immunohistochemical stains play a vital role in differentiating hemangiopericytoma from angiomatous meningioma and hemangioblastoma. The importance of differentiating these tumors needs to be emphasized due to the prognostic differences. Meningeal hemangiopericytomas are locally invasive lesions with recurrence rate higher than meningiomas.

Grade 1 meningiomas have a favorable prognosis. Since angiomatous meningioma belongs to this group, it has a similar behavior. Postoperative CT scans showed no residual tumor and the postoperative period was uneventful in the case studied. To conclude we would like to summarize that angiomatous meningioma is a rare variant of meningioma with few distinctive clinical, radiological, histopathological, and immunohistochemical features. However, it may mimic other vascular neoplasms like hemangioblastoma or hemangiopericytoma creating a diagnostic dilemma.

REFERENCES


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