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Case Report

Angiomatous meningioma: A rare case report

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ABSTRACT

Angiomatous meningioma is a rare subtype of meningioma that accounts for 2.1-2.59% of all meningiomas. Microscopically, it shows numerous vascular spaces in addition to the normal meningotheial elements. Here, we are presenting a case of 41 years old female who presented with history of bilateral loss of vision, headache, loss of consciousness, seizures, giddiness and vomiting and was diagnosed as angiomatous meningioma on histopathological examination. Despite the availability of neuroimaging diagnostic modalities, histological examination and immunohistochemistry remain the gold standard for final diagnosis.

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1. Introduction

Meningiomas are the most common primary tumor of the central nervous system in adults.¹ Angiomatous meningioma is an out of the common WHO Grade I histological subtype of meningioma which accounts for 2.1-2.59% of all meningiomas.² Microscopically, haematoxylin & eosin stained sections pertaining to angiomatous meningioma show numerous blood vessels and classic meningioma morphology. The vascular channels may be small or medium sized, thin-walled or thick, and most of them are small with markedly hyalinized walls. This subtype therefore requires further evaluation to differentiate it from hemangioblastomas and hemangiopericytomas.³ Angiomatous meningioma is usually characterized by the onset of slow progressive symptoms and the main symptoms result from compression of the adjacent structures. Headache and epilepsy are the initial clinical manifestations. Due to its rich vascular supply, its surgical removal becomes quite challenging for neurosurgeons due to the risk of intraoperative

haemorrhage.

2. Case Report

A 41 years old female presented to the neurology OPD with clinical history of bilateral loss of vision since 20 days, headache, loss of consciousness, seizure, giddiness and episodes of vomiting since one month. According to patient's husband, the patient was in her usual state of health one month back. Her general physical examination and systemic examination were normal. All the baseline hematological and serological investigations were normal. Contrast Enhanced Magnetic Resonance Imaging (CEMRI) of brain showed meningioma in left frontoparietal convexity region, significant vasogenic edema and midline shift. The patient was then shifted to neurosurgery department where she underwent surgery, tumor was excised & sample was sent to the histopathology section of the department of pathology.

The surgery went uneventful & the patient did not develop any post-operative complications. One week post-surgery, patient was assessed for the above mentioned symptoms, which had resolved completely. Eventually, the

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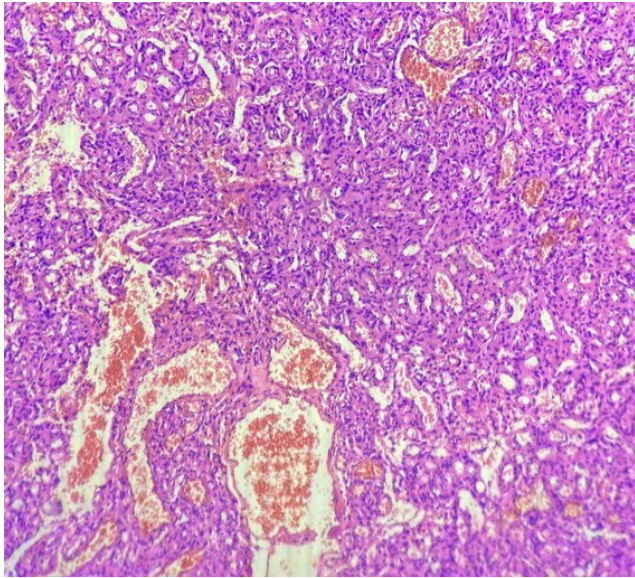


Fig. 1: Micropictograph depicting tumor composed of meningothelial cells and vascular spaces (400X, H&E stain)

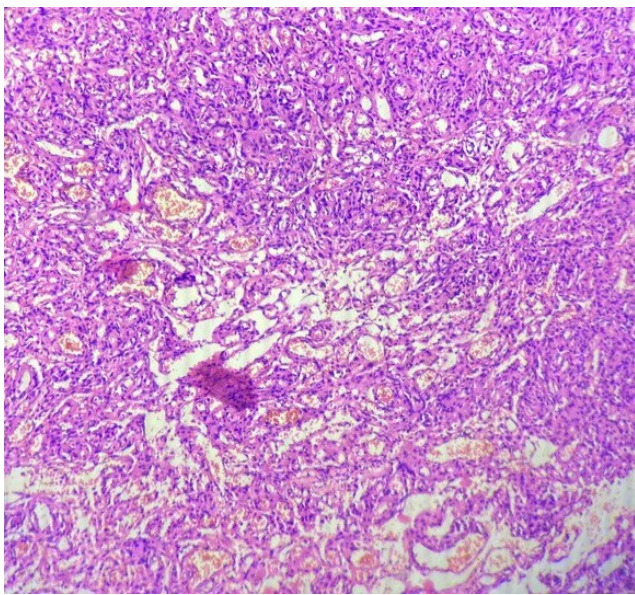


Fig. 2: Micropictograph depicting angiomatous meningioma (400X, H&E stain)

patient was discharged. Gross examination showed four fragments of tan, hemorrhagic tissue, firm in consistency with the largest tissue piece measuring 3.8x3.5x2.0cms and smallest measuring 1.0x0.7x0.5cm. Tumor tissue was fixed in 10% neutral buffered formalin. The tissue was then subjected to routine histopathological processing. Sections from the paraffin block were cut with thickness of 4 micron and staining with hematoxylin & eosin (H&E) was done. Microscopic examination of the stained sections depicted the morphology of angiomatous meningioma, showing a

tumor composed of meningothelial cells and numerous vascular spaces. The meningothelial areas were composed of spindle and oval cells with eosinophilic cytoplasm and round to oval nuclei.

3. Discussion

Meningiomas are benign brain tumors of adults and arise from the meningothelial cells of the arachnoid & are usually attached to the dura. Meningiomas account for 20% of all intracranial neoplasms. According to WHO classification, CNS tumours including meningioma are classified into various grades. Meningiomas have been categorized into grades I, II & III based on increased cellularity, high N:C ratio, prominent nucleoli, mitosis, patternless sheets & geographic or spontaneous necrosis. Grade I meningiomas are classified as the most frequent subtype, according to WHO. Angiomatous meningioma (AM) is a rare subtype of all meningiomas & constitutes 2.1-2.59% of all meningiomas. Angiomatous meningiomas exhibit some distinct features as compared to other benign meningiomas e.g. epidemiologically, these display slightly higher male preponderance. Anatomically, angiomatous meningioma is located commonly over cerebral convexity and are dura based lesions.⁴ Clinically, these tumors most commonly present as seizures and neurological deficits.⁵ MRI & CT are the most frequently used imaging modalities. Radiologically, angiomatous meningiomas show perilesional edema in addition to the characteristic features. Perilesional edema is attributed to VEGF secretion by the hypervascular tumour.⁶ Microscopic examination of the stained sections show tumor composed of meningothelial cells and numerous vascular spaces. The meningothelial areas are composed of spindle and oval cells with eosinophilic cytoplasm and round to oval nuclei. Subtyping of meningiomas primarily depends on its histopathologic features and immunohistochemistry which remain the gold standard for its diagnosis.⁷ Based on the diameter of vascular channels, Martin et al., classified angiomatous meningiomas into two subtypes: a) macrovascular with 50% of vessels having larger than 30um in diameter and b) microvascular subtype in which 50% of vessels were smaller than 30um in diameter.⁸ Histologically, these tumors show lesser degree of calcification and greater degree of peritumoral brain edema as compared to other benign meningiomas.^{9,10} The diagnosis of angiomatous meningioma can be confirmed by immunohistochemistry. The tumor cells show positivity for epithelial membrane antigen (EMA), cytokeratin, progesterone; and negativity is obtained with CD34.

Two close differential diagnoses of angiomatous meningioma include hemangioblastoma, and hemangiopericytoma. Capillary hemangioblastoma accounts for <1-2% of all CNS tumors and are mostly found in third to fifth decade. It occurs more commonly in

cerebellum, it can occur in brain stem and spinal cord also but a supratentorial location is rare. Computed tomography (CT) and MRI scans show a cystic lesion with an enhancing mural nodule. Microscopically, it is characterized by thin-walled blood vessels lined by plump endothelial cells and separate groups of polygonal stromal cells. These polygonal neoplastic cells contain vacuolated or foamy cytoplasm and nuclei may or may not show hyperchromasia and pleomorphism. But the tumor does not show any features of anaplasia. Hemangiopericytomas are dural-based lesions which can clinically mimic meningioma. Meningeal hemangiopericytoma was earlier described as angioblastic variant of meningioma, but Begg and Garret recognized that it was hemangiopericytoma arising within meninges.¹¹ Microscopic examination shows a highly vascular lesion composed of irregular cells with ill-defined cytoplasmic margins and staghorn vascular pattern. Reticulin stain is used to delineate a dense meshwork in the pericellular region. Immunohistochemical stains play a vital role in differentiating hemangiopericytoma from angiomatous meningioma and hemangioblastoma. Hemangiopericytoma are immunoreactive to vimentin and endothelial antigen CD34 but stain negatively with 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. Kuruvilla and Madhavan have reported positivity with vascular endothelial markers, factor VIII R Ag and Ulex Europaeus A lectin 1 antibody (UEA-1) in both endothelial and stromal components of the tumor whereas a negative/faint reactivity in flattened endothelial lining of blood vessels in hemangiopericytomas.¹² It is important to differentiate these tumors as they have different prognostic implications. Meningeal hemangiopericytomas are locally invasive lesions with recurrence rate higher than meningiomas. Total excision of tumor is the treatment of choice and the same was followed in our case.

4. Conclusion

We have presented a rare subtype of meningioma i.e. Angiomatous meningioma which needs to be distinguished from other vascular lesions in the CNS as they have different prognostic implications.

5. Conflict of Interest

The authors declare that there are no conflicts of interest pertaining to the publication of this paper.

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