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A RARE CASE OF ANGIOMATOUS MENINGIOMA AT A TERTIARY CARE HOSPITAL IN A RURAL SETUP

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Abstract:

Meningiomas are cancerous tumours that develop from meningothelial cells. The majority of these tumours are intracranial, with a few being intraspinal and a few being extracranial. Many histological abnormalities are divided into three classes based on clinical characteristics. Classification is essential for defining the therapy method. Meningiomas are usually slow-growing, benign neoplasms originating from the arachnoidal cap cells in the meningeal coverings of brain and spinal cord. They are the most frequently occurring benign intracranial tumours which account upto 34% of these neoplasms, peak incidence being in middle aged patients; with female:male ratio 2:1. Due to compression of the adjacent structures, they cause neurological signs and symptoms, having fickle clinical presentations, heterogeneous histological picture and an inherent trend to recur. The rate of recurrence is determined by the degree of surgery and histopathological grade. Angiomatous meningioma comprises 2.1% of all the meningiomas, being classified as WHO grade 1 tumour histologically, presenting just like a benign meningioma in which blood vessels exceed upto 50 percent. Hereby, presenting a 61 years old female patient diagnosed histologically as angiomatous meningioma delineating its clinical, radiological and histopathological characteristics. The primary goal of presenting this case is to showcase the histomorphological characteristics of this rare variant of meningioma that may aid in distinguishing it from Hemangioblastoma and Hemangiopericytoma. To differentiate this sub-type of meningioma from all the other vascular lesions in the central nervous system, a thorough assessment and interdisciplinary approach are required. Histopathological investigation and immunohistochemistry are important in verifying the diagnosis.

Key Words: Angiomatous meningioma, rare variant, hemangiopericytoma.

Background:

Meningiomas are cancerous tumours that develop from meningothelial cells. Meningioma is a benign, slowly progressing neoplasm, most frequently arising from the arachnoidnal cap cells of the cerebrum and spinal cord; constituting 15 percent of all intracranial tumors.[1] It produces neurological signs and symptoms

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because of compression of the adjacent structures, having fickle clinical presentations, heterogeneous histological picture and inherent trend to recur. The peak incidence of meningiomas is in middle aged patients; with females being twice as affected as males. In the elderly or young, there is no sex predilection. The majority of these tumours are intracranial, with a few being intraspinal and a few being extracranial. Many histological abnormalities are divided into three classes based on clinical characteristics. Classification is essential for defining the therapy method. The variants of meningioma are further sub-divided into the following three grades as per to World Health Organization:- Grade I - benign, grade II - atypical and grade III - malignant with metastatic potential grounded on cellularity, NC ratio, mitosis as well as necrosis. Angiomatous meningioma, which is a rare sub-type is classified as WHO Grade I meningioma, is characterized by an extensive vascular component, accounts for approximately 2.1% of all the meningiomas.[2]

In Magnetic Resonance Imaging, meningiomas are isointense or hyperintense as compared with the cerebral cortex. The presence of a short extension of contrast enhancing tissue along the duramater (dura-tail) is a useful diagnostic characteristic.

There are no other radiographic features that aid in the categorization of meningiomas, albeit angiomatous meningioma (despite being classified as WHO grade I) exhibits perilesional oedema. Glioma, hemangioblastoma, and normal meningioma all have contrast enhancement. Peri-lesional oedema is common in atypical meningioma, however it is not a marker of atypia or carcinoma in this type. The immunohistochemistry pattern of angiomatous meningioma is identical to that of a normal meningioma. It displays positive staining with antibodies to vimentin, desmoplakin, and EMA, as well as a localised positive reactivity to progesterone antibodies. Electron microscopy can be used to validate the findings. Neoplastic meningeal cells feature prominent cytoplasmic processes and well-defined synapses on the ultrastructure. Foamy cells in angiomatous meningioma are formed by plasma lipid leaking over the thin artery wall. A complete hemangioblastic meningioma can't be recognised from a hemangioblastoma unless by its connection to the dura, immunohistochemical markers and microscopy.

Hemangiopericytomas are dural-based tumours that are frequently misdiagnosed as meningiomas. Meningeal hemangiopericytoma was previously characterised as an angioblastic variety of meningioma, but it was Begg and Garret that identified it as a hemangiopericytoma developing inside the meninges. They account for two to five percent of meningeal lesions, are more prevalent in adults, and are more common in men. Hemangiopericytoma imaging results are comparable to meningiomas. Immunohistochemical stains are quite useful in distinguishing hemangiopericytoma from angiomatous meningioma and hemangioblastoma. Because of the diagnostic differences, it is critical to differentiate these malignancies. Meningeal hemangiopericytomas are locally invasive lesions with a high rate of recurrence than meningiomas.

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Meningiomas of grade 1 have a high survival rate. As angiomatous meningioma is a member of this category, its behaviour is similar. In the patient investigated, postoperative CT scans revealed no residual tumour, and the postoperative phase was uncomplicated. Angiomatous meningioma is an uncommon kind of meningioma with few distinguishing clinical, radiological, histological, and immunohistochemical characteristics. It may, however, mirror other vascular neoplasms such as hemangioblastoma or hemangiopericytoma, posing a diagnostic quandary.

Case History:

A 61 yr old female patient presented to neurosurgery out-patient department of tertiary care hospital in central India with complaints of difficulty in swallowing which was insidious in onset and gradually progressive in nature, headache which was gradually progressive in nature, gait disturbance and vomiting since 7 months. Patient is a known case of acoustic schwanoma since 2017. There was no other significant personal history or family history as well. Patient was conscious and oriented and his general examination was normal. On central nervous system examination, Glasgow Coma Scale was E4 V5 M6. Post-contrast magnetic resonance imaging (MRI) indicated a broad-based solid enhancing tumour at the cerebellopontine angle. A preoperative diagnosis of hemangioblastoma in cerebellopontine angle was obtained formulated on the basis of the symptoms, signs, and Gadolinium-enhanced Magnetic Resonance Imaging. Since the patient refused surgery, a wait-and-see approach with close monitoring was employed. He developed a worsening headache and gait disturbance two years later. Subsequently, craniotomy along with complete resection of the solid mass was performed. The specimen of the tumour mass was sent to the Department of Pathology. Postoperatively patient was good on regular follow-up.

Grossly, we received multiple, irregular, bluish brown tissue pieces aggregating 4 x 2.5 x 1.5 cm. Tumor tissue was routinely processed after being fixed in a 10 percent of the formalin solution. Paraffin blocks were made and then the sections were cut with a thickness of 5 microns. Hematoxylin and eosin stain was used to stain the sections and then the slides were viewed under a light microscope.

Microscopically, the tumour tissue seemed to be highly vascularized, with meningeal cells assembled in nests and a palisading pattern. The nuclei of the cells were delicate round to oval, and the cytoplasm looked eosinophilic. There were numerous tiny capillary-like blood capillaries with thin walls. The vessel wall was also found to be hyalinised with no indication of atypia or malignancy. The concluding diagnosis was established as Angiomatous Meningioma – WHO grade I.

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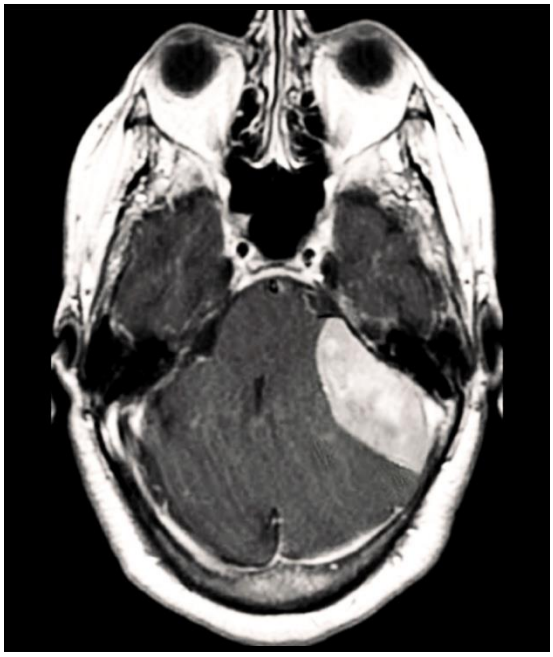
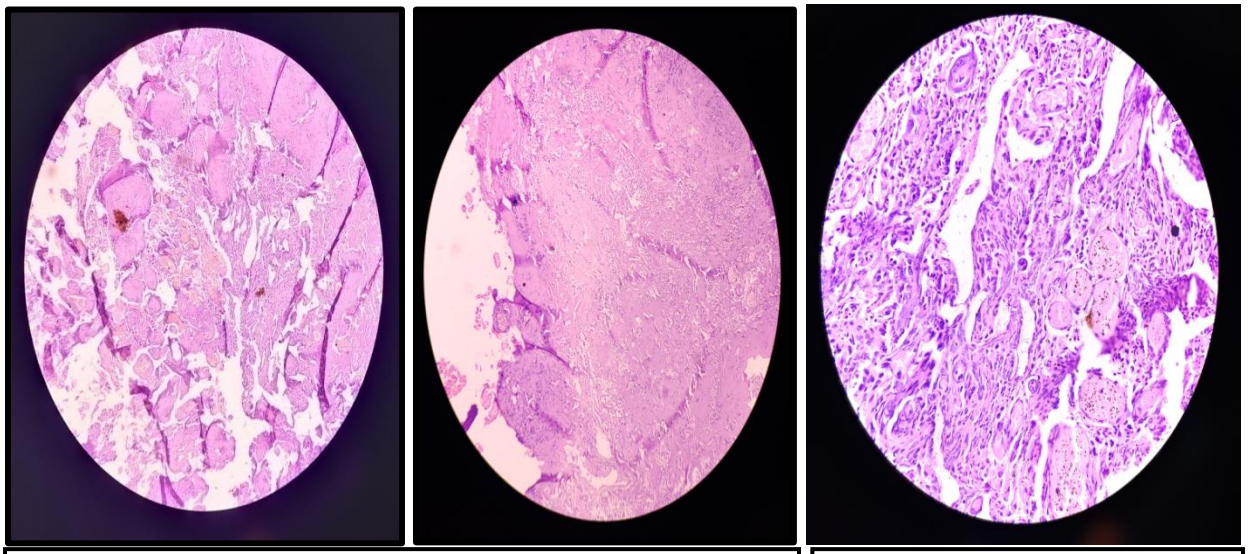


A fairly defined left C/P angle isodense mass lesion is seen. It shows homogenous enhancement in post contrast study. It is surrounded by significant low attenuation vasogenic edema. Both the mass and surrounding edema exerts mass effect in the form of compression of the 4th ventricle & compression of the nearby brain stem and vital structures.

The current CT findings are impressive of Left Cerebellopontine angle extra-axial large SOL most likely meningioma.



Angiomatous Meningioma as seen on 10x magnification with hematoxylin and eosin stain.

Microscopic H and E staining of angiomatous meningioma 40x magnification

Discussion:

Meningiomas account for about 20 percent among all of the intracranial neoplasms, 90 percent of which are usually benign.[3] Grade I being the most common subtype among the meningiomas, according to the WHO. Atypical meningiomas (Grade II-WHO) and anaplastic (Grade III-WHO) are rare, aggressive tumours with a much greater tendancy to recur after being treated.[4] Angiomatous meningiomas have a

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vascular component surpassing 50 percent of the tumour area which comprises barely about 2.21 percent of all of the meningiomas.[5] This form of meningioma has clinical characteristics that are comparable to benign type of meningiomas. The average age of commencement was 51.8 yrs, with males experiencing a greater rate of occurrence. Angiomatous meningioma is a type of dural-based lesion that is most typically found over the cerebral convexity.[6] As in our case, the most common presenting symptoms were headache and dizziness, tailed by transient loss of consciousness, seizures, nausea and vomiting. Since headache isn't a distinctive sign of meningiomas, imaging modality is essential for the assessment of such patients. The most widely used imaging diagnostic modalities are MRI and CT. Typical meningiomas have a distinct dural tail and mottling, signifying that the tumour is highly vascular.[7,8] Angiography may be used to rule out other vascular disorders like aneurysms and cerebrovascular conditions.[9] Based on radiology, meningiomas have no further subtyping features; however, angiomatous meningiomas have peri-lesional edema due to the hypervascular tumor's enhanced capillary permeability and VEGF release.[10]

Due to the constraints of merely imaging studies, meningioma subtyping is primarily based on histopathologic features such as immunohistochemistry and electron microscopy.[11] Highly vascular meningiomas must be evaluated for angiomatous meningioma, hemangiopericytoma as well as hemangioblastoma.[12] Martin et al. categorized angiomatous meningiomas into two subgroups established on the vascular channel diameter, namely macrovascular having >50 percent of vessels wider than 30m and microvascular having >50 percent of vessels narrower than 30m in diameter.[13] Microvascular angiomatous meningiomas and hemangioblastomas are difficult to distinguish, necessitating immunohistochemistry and electron microscopy for confirmation. EMA, vimentin, S100, and PR are all immunoreactive in angiomatous meningiomas. EMA was positive in our case.[14] Surgery i.e. total excision of the tumour will remain the choice of treatment as being followed in our case.

Conclusion:

Angiomatous meningioma is a rare form of WHO grade I meningiomas with a few distinguishing clinical, radiological, histological, and immunohistochemical characteristics. To differentiate this sub-type of meningioma from all the other vascular lesions in the central nervous system, a thorough assessment and interdisciplinary approach are required. Histopathological investigation and immunohistochemistry are important in verifying the diagnosis. Gross total resection is still the treatment of choice. Patients with residual tumor after surgery can benefit from radiation therapy. The meningiomas of grade 1 have a high survival rate. As angiomatous meningioma is a member of this category, its behaviour is similar. In the patient investigated, postoperative CT scans revealed no residual tumour, and the postoperative phase was uncomplicated. To summarize, angiomatous meningioma is an uncommon kind of meningioma with a few distinguishing clinical, radiological, histological, and immunohistochemical characteristics. It may,

however, mirror other vascular neoplasms such as hemangioblastoma or hemangiopericytoma, posing a diagnostic quandary.

Conflicts of Interest: None.

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