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Review Article

Update on atypical meningioma clinicopathological profile

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Abstract

Atypical meningioma is a more aggressive form of meningioma and denotes a WHO grade 2 tumor. Case report: A 45-year-old female presented with a sudden onset of persistent generalized headache with dizziness. It was aggressive in nature with associated nausea, vomiting of five days duration. On presentation, the patient was alert, oriented, and cooperative. She did not have any motor or sensory defects. There was no significant any past illness. CT scan of brain showed a large well defined iso to hyperdense lesion in the right lobe of cerebellum, measuring 4 x 3.6 cm. It was causing infiltration of the adjacent brain parenchyma, fourth ventricle, quadrigeminal cistern with effacement. Patient underwent midline suboccipital craniotomy with excision of tumor from the right lobe of cerebellum. On histopathology reported as Atypical meningioma intermediate grade tumor, WHO 2 right cerebellar region. Postoperatively she had no evidence of recurrence on follow-up imaging. Conclusion: Atypical meningioma exhibits characteristic histological features. Patients with an atypical meningioma may shows aggressive nature and requires regular follow up. We present this update with review of atypical meningioma WHO grade 2 tumor for its clinical, radioimaging and histopatholgical findings.

Keywords: Atypical meningioma, Histopathology, CNS WHO grade 2 tumors

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1. Introduction

Meningioma is a primary CNS tumor, arising from arachnoid cap cells associated with dura mater or choroid plexus. Atypical meningioma refers to a more aggressive form of meningioma and denotes a WHO grade 2 tumor.[1] The term "atypical meningioma" is considered when a tumor showing borderline histologic and clinical features between benign and malignant meningioma. The 2021 WHO classification of tumors of the central nervous system for atypical meningioma is considered when the tumor fulfilling either 1 of 2 major criteria or 3 of 5 minor criteria. The major criteria are: 4 - 19 mitotic figures/10 high power fields. Mitotic rate is defined as the highest count over 10 consecutive high power fields (1 high power field = 0.16 mm²) and brain invasion. The minor criteria are: patternless or sheet-like growth, increased cellularity, small cells with high nuclear-cytoplasmic ratio, macronucleoli, foci of spontaneous or geographic necrosis.[2] In this case tumor showed sudden clinical presentation with aggressive nature.

1.1. Instance

A 45-year-old female presented with a sudden onset persistent generalized headache with dizziness. It was aggressive in nature with associated nausea, vomiting of five days duration. On presentation, the patient was alert, oriented, and cooperative. She did not have any motor or sensory defects. The routine investigations were within normal limits. There was no significant any past illness. CT scan of brain showed a large well defined iso to hyperdense lesion in the right lobe of cerebellum, measuring 4 x 3.6 cm. (**Figure 1**). It was causing infiltration of the adjacent brain parenchyma, fourth ventricle, quadrigeminal cistern with effacement of it. There was cerebellar tonsillar heriation and

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descending tonsillar herniation. The conclusion was a large well defined iso to hyperdense lesion is seen in the right lobe of cerebellum suggestive of neoplastic etiology. Patient underwent midline suboccipital craniotomy with excision of tumor from the right lobe of cerebellum.

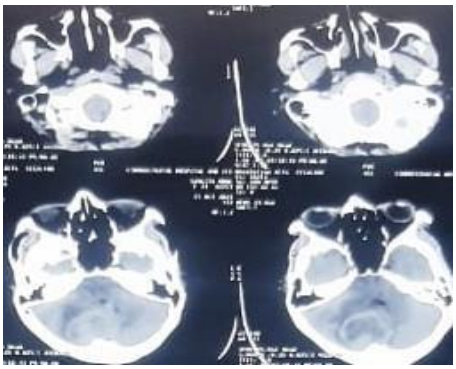

Figure 1: CT scan of brain shows a large well defined iso to hyperdense lesion in the right lobe of cerebellum.

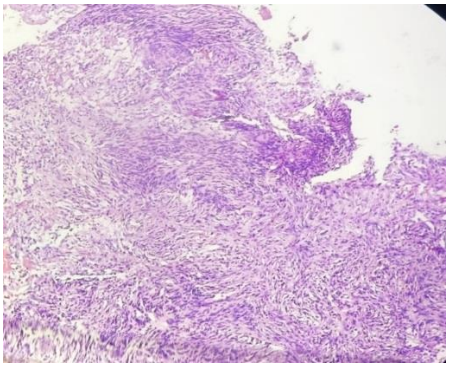

Figure 2: Histopathology show a meningothelial tumor arranged in sheets, whorls and diffuse pattern. (Haematoxylin and eosin stain, 40x)

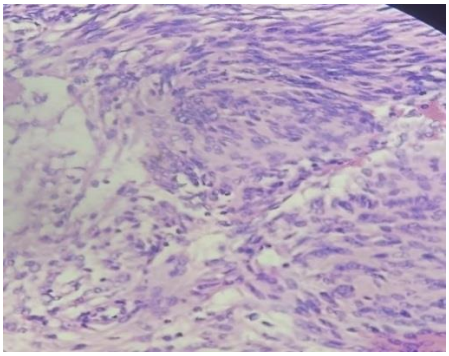

Figure 3: Phomicrograph shows hypercellular neoplastic cells with indistinct cell membranes, round nuclei with mild atypia, high N/C ratio and having eosinophilic cytoplasm. (Haematoxylin and eosin stain, 100x)

On histopathology showed a meningothelial tumor arranged in sheets, whorls and diffuse pattern with invasion of brain. The neoplastic cells were syncytial cells with indistinct cell membranes, round nuclei with mild atypia, high N/C ratio and having eosinophilic cytoplasm. Tumor showed hypercellurarity, intermediate mitotic rate 4 mitoses/10 HPF. The occasional tumor cell with macronucleoli were noted. In areas scattered psammoma bodies were noted (**Figure 2**, **Figure 3**, **Figure 4**, **Figure 5**).On histopathology reported as Atypical meningioma intermediate grade tumor, WHO 2 right cerebellar region.

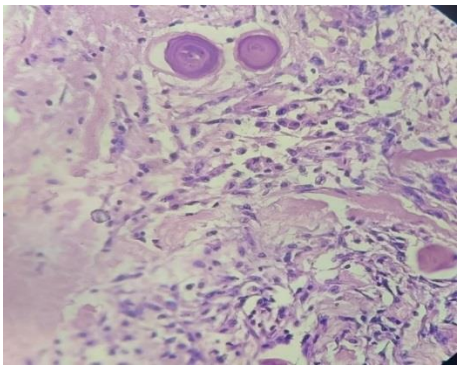

Figure 4: Phomicrograph shows meningothelial tumor with scattered psammoma bodies. (Haematoxylin and eosin stain, 100x)

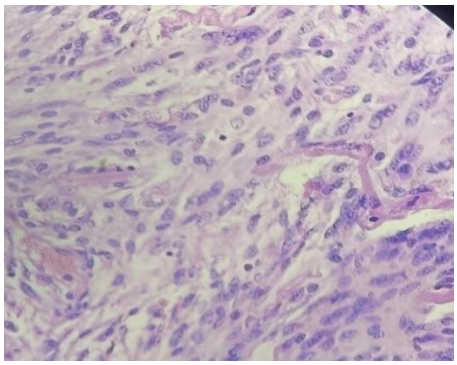

Figure 5: Phomicrograph shows tumor hypercellurarity, increased mitotic rate and macronucleoli. (Haematoxylin and eosin stain, 100x)

The patient experienced a significant improvement in her symptoms postoperatively and had no evidence of recurrence on follow-up imaging.

2. Discussion

Background: Meningiomas are common brain tumors that are classified as either benign, atypical, or malignant. The meningiomas constitute about 33% of all brain tumors.[3] Most of menningiomas are typical or grade 1(80%).[4] Most of menningiomas are slow growing tumor of WHO grade 1 and considered as benign. Grade 2 meningiomas are a) Brain-invasive meningioma b) Atypical meningioma and c) Clear cell or chordoid subtype. The next rare is anaplastic (WHO grade 3) meningiomas.[5,6] Due to rarity of atypical meningioma, the literature on the natural history, histomorphology and treatment is relatively *less to satisfy the need* . The study from Recker MJ et al , observed total of 4476 patients diagnosed with meningioma were identified from The National Cancer Institute's Surveillance, Epidemiology,

and End Results (SEER) database from 2004 to 2018, showed the incidence of atypical meningioma increased at an annual percent change of 5.6% cases.[7]

2.1. Atypical meningioma: Clinical presentations

Atypical meningioma, a primary CNS tumor, arising from arachnoid cap cells is a variant of meningioma. It comprising 5 - 15% of meningiomas. Their clinical presentation varies greatly, ranging from asymptomatic incidental tumor to fatal tumor. Clinically symptoms are of headache, vomiting, seizures neuropsychological decline, seizure, bulging mass and focal neurological deficit due to tumor compression.[8] Atypical meningioma generally grow faster with aggressive behaviour. A meningioma with brain invasion is now considered a grade 2 tumor.In this case also sowed aggressive behaviour with rapid growth and invasion in surrounding tissue. Their location may be Intracranial, intraspinal or intraorbital.

2.2. Radio imaging characteristics of atypical meningiomas

The radio imaging has an important role in detection of tumor, locations, size detecting tumour-related complications and in preoperative differential diagnosis for optimizing treatment strategies. Typically, meningiomas are sharply demarcated and hyperdense on CT. On radio imaging it is difficult to distinguish between benign from atypical and anaplastic meningiomas on there morphology. Magnetic resonance imaging (MRI) is considered better modality for evaluation of meningiomas, as it provides excellent contrast differentiation.[9] It also distinguish between intra- and extraaxial lesions. The most reliable feature is the presence of lower apparent diffusion coefficient values. Preoperative diffusion tensor imaging (DTI) showed that intratumoral microscopic water motion is less organized in classic than in atypical meningiomas. However DTI is not useful for differentiation. The mean reduced Apparent Diffusion Coefficient (ADC) values are a more reliable imaging feature to distinguish atypical or anaplastic meningiomas. These tumor shows extra-axial mass with dural tail having uniformly contrast enhancing mass lesions. In cases the extensive peritumoral edema is noted which is associated with brain invasion.[10] Tomura *et al.* observed that partial or complete disappearance of the peritumoral band had been seen in a majority of atypical meningiomas and a relatively large amount of perifocal edema.[11]

2.3. Histopathology of Atypical Meningiomas

On gross examination tumors are well circumscribed, rubbery soft to firm. These are firmly attached to the inner surface of the dura. In atypical meningiomas these are readily adherent to adjacent brain tissue.

On histopathological examination diagnostic criteria on microscopic features for Atypical meningioma, are as per the 2021 WHO classification of tumors of the central nervous system having major and minor criteria. Diagnostic criteria: fulfilling either 1 of 2 major criteria or 3 of 5 minor criteria. Major criteria are 4 - 19 mitotic figures/10 high power fields or brain invasion. While Minor criteria are patternless or sheet-like growth, hypercellularity, small cells with high N/C ratio, prominent nucleoli and foci of spontaneous or geographic necrosis. In 2007, the WHO definition of atypical meningioma added brain invasion as an alternative criterion.[12,13] The brain invasion is defined as irregular projections of tumor cells into adjacent CNS parenchyma without an intervening layer of leptomeninges at the tumor to brain interphase.

The differential diagnosis for meningioma from the others tumors can be challenging. The differential are schwannoma, solitary fibrous tumor/ hemangiopericytoma, anaplastic meningioma.[14,15,16] Meningioma with overtly malignant cytomorphology are seen in anaplastic meningioma which shows mitoses >20/10 high power field.

2.4. Immunohistochemistry and molecular study of meningiomas

Immunohistochemistry plays a role in meningioma variants. Immunohistochemical markers for meningiomas are epithelial membrane antigen (EMA) and progesterone receptor (PR), somatostatin receptor 2A (SSTR2A). For the diagnosis of meningioma, the single marker SSTR2A showed the best sensitivity (95.2%), specificity (92%).[17]

The assessment of the proliferative index of antibody MIB-1 if > 5% play important role and are associated with increased risk of recurrence in atypical meningiomas.[18]

On molecular study shows that the majority of atypical meningiomas have loss of *NF2* combined with either genome instability or loss of *SMARCB1*. While recurrent losses of chromosome 1p, 6q, 14q, 18q and gain of 1q are indicators of poor prognosis. Mutations in the NF2 gene and loss of chromosome 22q are the most common genetic alterations associated with the initiation of meningiomas.[19] The tumor recurrence and prognosis may better predict on DNA methylation profiling.[20]

2.5. Treatment and Prognosis in atypical meningiomas

The treatment of meningiomas is surgical gross total resection of tumor. Postsurgical radiation may required for atypical meningiomas. The radiotherapy as external beam or brachytherapy often added both to complete and incomplete resections. It has shown improve local control and prolongs overall survival.[18] Ros-Sanjuan A, et al, noted that the Atypical meningiomas have an intermediate recurrence rate between benign and malignant meningiomas.[21] For the treatment of atypical meningiomas there are no large published series that provide definitive guidelines. Toh CH, et al observed, that the five-year recurrence rate is significantly higher (41%) than that seen in grade 1 (benign) meningiomas (12%).[22]

Yip Chi-Man et al study on 27 consecutive patients of intracranial atypical meningiomas treated between January 2005 and December 2014, 33.33% showed tumor progression or recurrence during follow-up.[23]

Postoperatively in our patient showed significant symptomatic improvement and had no evidence of recurrence on follow-up imaging.

3. Conclusion

Atypical meningioma exhibits characteristic histological features. Patients with an atypical meningioma may develop a recurrent tumor, aggressive nature and requires regular follow up. We present this update with review of atypical meningioma WHO grade 2 tumor for its clinical, radioimaging and histopatholgical findings.

4. Conflict of Interest

None.

5. Sources of Funding

None.

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