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Review Article

Myxopapillary ependymoma of the spinal cord: A case with literature review

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ABSTRACT

Myxopapillary ependymoma is a rare spinal cord tumors. The intramedullary myxopapillary ependymomas are. Herewith case of a 41-year-old male complaint of a nonspecific low back pain, progressive inability to bend, right lower limb pain with numbness of six month duration. On clinical evaluation right lower limb weakness was noted while the sensations were normal. There was no any significant past history. MRI lumbo-sacral spine with whole spine screening was performed. It showed lobulated heterogenous intensity enhancing intra medullary mass lesion in thecal sac extending from L4-L5 to S1-S2 level. It measured about 52 x 28 x 21 mm. MRI features suggestive of neoplasm – ependymoma was made. The gross total surgical resection was performed. On histopathological findings reported as myxopapillary ependymoma grade 2. The tumor on immunohistochemistry showed positive for EMA, GFAP. While negative for CMYC. The Ki-67 proliferation index (MIB1) was 3%. On follow up there was no tumor recurrence. Rehabilitation therapy was initiated and follow up is advised.

1. Introduction

Myxopapillary Ependymoma (MPE), a rare distinct subtype of glioma. Commonly noted at filum terminale and conus medullaris, in the spinal cord tumor is at medullary conus at lumbar spinal level. Their extension to thorasic spine, and sacrum is observed.[1] In spinal cord tumors MPE accounts for 1–5%. The overall incidence is 0.0–0.08 cases per 100,000 persons annually.[2]

On histopathological examination MPE shows papillary or radial growth pattern. The neoplastic cells are cuboidal or epithelioid. Also, spindle cell morphology is noted. Perivascular tumoral arrangement in seen in many cases. Tumor background is usually showed myxoid change. The microcyst formation is frequently noted. As per recent WHO classification, are associated with a higher proliferative index and is considered as grade 2 categories. It carried clinical significance as Grade 2 tumors are potentially more aggressive behavior. MPE shows higher proliferative index.

2. Case Report

A 41-year-old male complained of nonspecific low back pain, progressive inability to bend, right lower limb pain with numbness of six month duration. Neurological examination revealed right lower limb weakness, sensations were normal. There was no any significant past history.

MRI lumbo-sacral spine with whole spine screening was performed. Screening of entire spine was also performed. Normal spine curvature and vertebral alignment is observed. Spondylosis çhanges noted in the form of perivertebral osteophytes and endplate marrow changes at places. The vertebral bodies are normal in height and signal intensity.

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Mild annular bulges of L2-3 and L3-4 discs noted effacing thecal sac with no significant nerve root compression or canal narrowing. Diffuse bulge of L4-5 disc with moderate left posterołateral protrusion of disc compressing intrathecal and exiting nerve roots. Mild to moderate canal stenosis and foraminal narrowing noted aggravated by perivertebral osteophytės. The facet joints appear normal. Ligamentum flava are not significantly thickened. The visualized ćord is normal in signal and morphology. There is no abnormal pre or paraspinal soft tissue. Visualized sacro-iliac joints shows, no significant abnormality. Screening of cervical spine showed loss of cervical lordosis with degenerative changes. Mild disc bulges are seen at C3-4 and C+-5 level indenting anterior subarachnoid space and abutting nerve roots. Mild canal and mild to moderate foraminal narrowing noted, aggravated by peridiscal osteophytes. Posterocentral mild protrusion of C5-6 disc noted. Impression given was lobulated heterogenous intensity enhancing intra medullary mass lesion in thecal sac extending from L4-L5 to S1-S2 level (Figure 1). It is hyperintense on T2WI and hypointense on TIWI Moderate heterogenous enhancement seen on contrast study. It measures about 52 x 28 x 21 mm. MRI features suggestive of neoplasm – ependymoma. Displacement of adjacent nerve roots noted. There is no abnormal pre or paraspinal soft tissue any significant abnormality. Patient treated with surgical excision of lesion with L5-S1 laminectomy. On microscopy showed radial and in areas fascicular growth pattern the tumor cells were cuboidal to epithelioid showed perivascular arrangement at places. In areas microcysts formation was noted (Figures 2, 3, 4 and 5). On histopathological findings reported as myxopapillary ependymoma grade 2. The tumor on immunohistochemistry showed positive for EMA dot-like cytoplasmic pattern (Figure 6) and positive for GFAP. While negative for CMYC. The Ki-67 proliferation index (MIB1) observed was 3%.

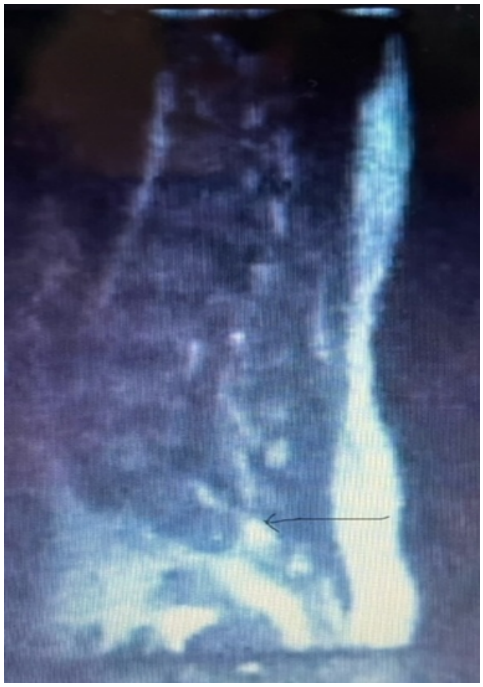

Figure 1: MRI lumbo-sacral spine showed lobulated heterogenous intensity enhancing intra medullary mass lesion in thecal sac extending from L4-L5 to S1-S2 level

3. Discussion

Myxopapillary ependymoma is rare neuroepithelial tumor, as distinct subtype of glioma. The origin of ependymomas is from ependymal cells lining the ventricular system, choroid plexus, central canal of the spinal cord and filum terminale within the central nervous system. Ependymomas are a relatively uncommon entity, accounting for approximately 1.8% of all primary central nervous system tumors and 6.8% of glial neoplasms. Kernohan in 1932 first defined MPE as a distinct subtype of ependymomas.[3]

The anatomical location for spinal ependymomas is at any level of cervical, thoracic, or lumbar regions. Spinal MPE is an uncommon primary spinal neoplasm. The spinal cord MPE is noted 1 to 5% of all spinal neoplasms and 13% of all spinal ependymomas. The incidence of MPEs is 0.05–0.08 per 100,000 individuals per year.[4]

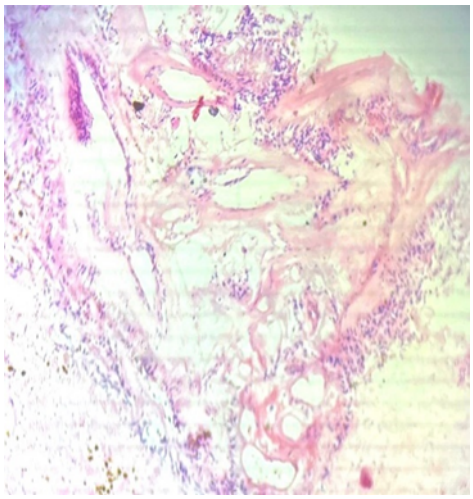

Figure 2: Microscopy shows tumor cells arranged in radial and in areas fascicular growth pattern.(Haematoxylin & Eosin stain, 40x)

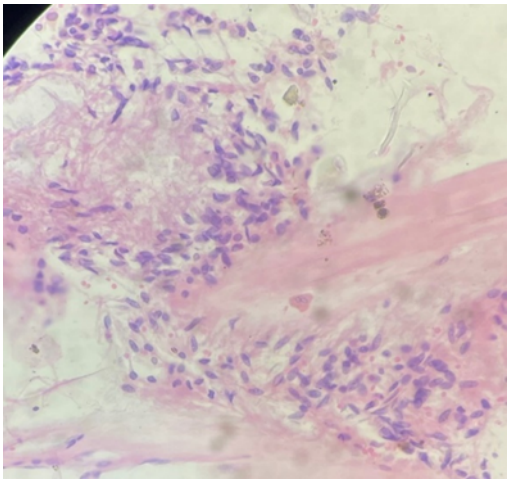

Figure 3: Microscopy shows cuboidal to epithelioid elongated glial tumor cells.(Haematoxylin & Eosin stain, 40x)

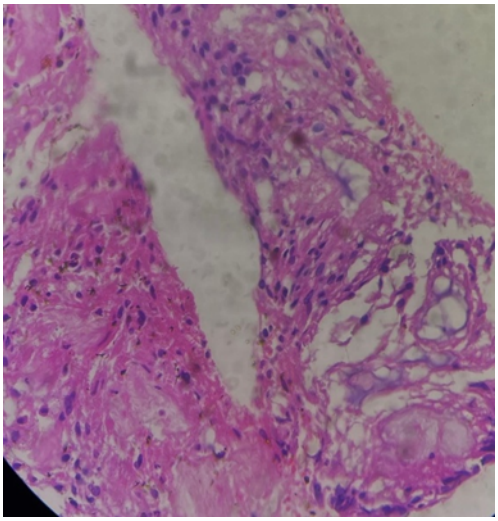

Figure 4: Photomicrograph showing tumor cells around hyalinized fibrovascular tissue with areas of microcysts formation.(Haematoxylin & Eosin stain, 40x)

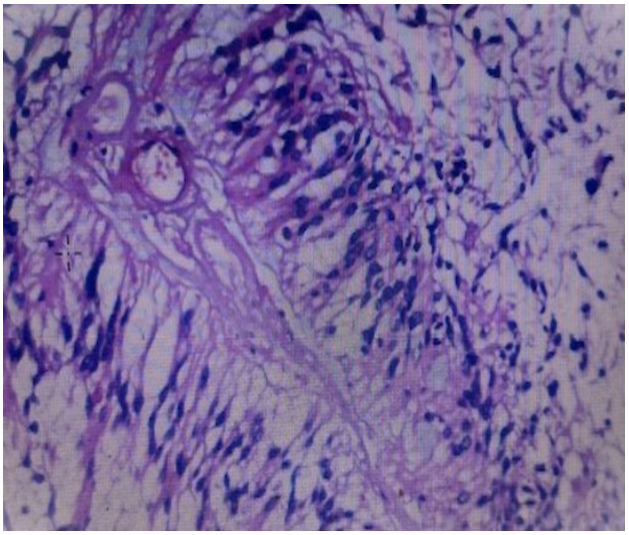

Figure 5: Photomicrograph showing tumor cells, fibrovascular tissue with areas of microcysts formation. (Haematoxylin & Eosin stain, 40x)

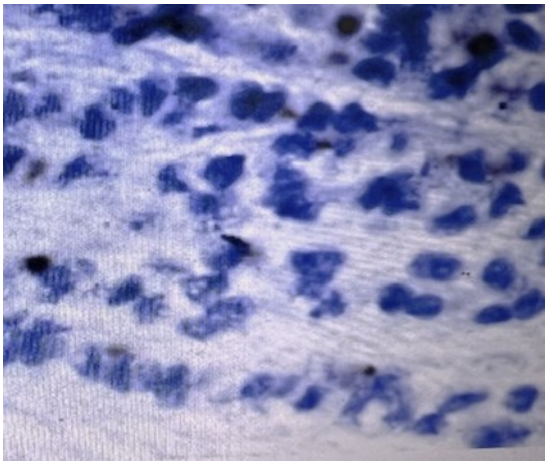

Figure 6: Tumor showed positive for EMA cytoplasmic pattern

It is considered that MPE is usually slow-growing tumor. The age group affected is at 30 and 50 years of age. In relation to proliferative index of tumor and occurrence in pediatric patients few of MPEs behave in an aggressive manner. Bagley CA et al., observed that the recurrence rate of 64% in paediatrics patients compared with 32% in adults.[5]

Clinically patients present with symptom of nonspecific back pain, weakness in legs, and sensory disturbances as a most common complaints. Other menifestations are gait ataxia, sexual problems, sphincter problems or bladder dysfunctions. These sign and symptoms are related to tumor size, site, and the local extent of the tumor.

Various risk factors linked to an increased of spinal ependymoma are hereditary, immunodeficient states, radiation, neurofibromatosis type II, post-transplantation, acquired immunodeficiency syndromes etc.

In the 2021 the World Health Organization classification of CNS tumors, ependymal tumors are classified into 3 grades as Grade 1: Subependymoma, Grade 2: Classic Ependymoma or myxopapillary ependymoma, Grade 3: Anaplastic ependymoma.[6] This is based on their outcome and recurrence rates.

The diagnosis of Spinal MPE is done with clinical, radiological and tissue biopsy, immunohistochemistry and molecular- unique DNA methylation profile study.

The radio imaging findings of MPE on MRI shows hyperintense contrast enhancement. The MPE is tumor

typically intradural and extramedullary which are well demarcated, ovoid mass lesion. Located at the cauda equina or conus or filum terminale region.[7]

4. Histopathological Features

On gross histopathological examination tumors are sausage-shaped, well-encapsulated, soft in consistency, and light reddish in color. The mucinous, gelatinous or hemorrhage or cystic changes may be seen in these tumors.

On microscopic morphology of MPE is composed of cellular rosettes and pseudorosettes. Tumor cells are arranged in papillary pattern or in areas fasicular type. Tumor contain vascular cores. In areas microcysts formation is noted. Accumulation of basophilic myxoid material is noted for which staining with PAS and Alcian blue is needed. Chen X et al. noted MPE is characterized by a papillary architecture with fibrovascular core, which contains both hyalinized blood vessels. The extracellular mucoid material was commonly observed.[8] Occasionally tumor may show pleomorphic tumor giant cells. Lee JC et al. observed, malignant behavior in a small subset of ependymomas.[9]

To study tumor behevior and progress of tumor, WHO grading criteria and genetic indicators are required. Lee JC, et al in this study observed that in anaplastic myxopapillary ependymomas cellularity is increased and reduced mucin in association with at least 2 of the following features: $\geq$ 5 mitoses / 10 high power field, Ki67 labeling index $\geq$ 10%, microvascular proliferation, spontaneous necrosis.[9]

The various differential for spinal MPE, which should be carefully looked for are schwannoma, fibrous meningioma, signet cell adenocarcinoma and metastatic carcinoma. For which morphological features and immunohistochemistry is required.[10]

5. Immumohistochemistry and Molecular Study

MPE shows diffuse expression of GFAP, tumor cells stained for S100, CD99, vimentin and CD56. Also, many of the shows COX-2 positivity. While tumor is negative for cytokeratin.

The new guidelines by the Consortium to Inform Molecular and Practical Approaches to CNS Tumor Taxonomy (cIMPACT-NOW) committee in 2020 have distinguished ependymal tumors according to methylome profiling data to indicate specific molecular groups based upon anatomic location.[11]

The histologic and molecular analysis of spinal ependymomas is given on N-MYC proto-oncogene amplified/ non amplified. The molecular markers of MPEs are receptor tyrosine kinase and HOXB13 gene. The HOXB13 is more specific for MPE while HOXA9 is more specific for ependymoma. Unique DNA methylation profile may also helpful for myxopapillary ependymomas.[12]

6. Treatment

The gross total resection is the treatment of choice for MPE. The total resection of may be difficult, as these tumors are well-vascularized, may noncapsulated, may adherent to nerve roots and the spinal cord or may be even infiltrated by the tumor. The presence of residual tumor due to infiltrative nature, local recurrence and metastasis, has been reported to occur in one-third of patients. The National Cancer Database was analyzed for patterns of care for adult MPE diagnosed between 2002 and 2016. Among the 753 qualifying cases, the majority of patients underwent resection (81.9%).[13]

Chemotherapy has been suggested as a potential treatment to prevent recurrence, but its efficacy has not been established in MPE. The patient followup is required.

7. Prognostic Factors

MPE are slow growing tumor. These patient shows long-term survival rates. However they have incidences of local recurrence.

The ependymoma are associated with relatively favorable prognosis with 10 year survival rates > 90.6%.[14]

Bandopadhayay P et al., studied the MEP in children and observed that recurrence is common. However the outcomes and survival is good.[15]

It is noted that radiotherapy improves progression free survival. In the cohort study by Daphne B Scarpelli et al on National trends in management of adult myxopapillary ependymoma showed the impact of on overall survival is indeterminate given the 1.6% death rate. In our case on follow up there was no tumor recurrence. Rehabilitation therapy was initiated and advised follow up.

8. Conclusion

Ependymal tumours are a heterogenous group of neoplasms. Myxopapillary ependymomas are a relatively uncommon entity, The morphological, radiological, immunohistochemical and molecular features will helpful to improve the prognostic relevance of the different diagnostic entities of spinal tumors.

9. Source of Funding

None.

10. Conflict of Interest

None.

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