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Case Report

Myxopapillary ependymoma with anaplastic features: A rare case report

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Abstract

Myxopapillary ependymoma (MPE) is a typically encountered neoplasm in the filum terminale/lumbosacral region of the spinal cord, categorised as a grade 2 tumour in the latest 2021 WHO classification of tumours of the CNS. Despite its usual indolent behaviour, cases in extradural locations may exhibit aggressive clinical tendencies. Anaplastic transformation in MPE is exceptionally uncommon, with fewer cases documented in literature. Diagnostic criteria and definitive grading for such cases remain uncertain. This report presents a unique case of MPE demonstrating anaplastic features, characterized by histology consistent with conventional MPE alongside areas displaying significant atypia, frequent mitotic figures, and elevated Ki-67 proliferation indices (10–12%). The review of literature included discusses common histologic and molecular findings associated with anaplastic features in MPE.

Keywords: Anaplastic, Myxopapillary, Ependymoma.

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1. Introduction

Ependymomas are subset of gliomas that exhibit prominent ependymal differentiation, both morphologically and histologically. They account for just 10% of all neuroepithelial tumours and are the most common primary spinal cord tumours. Additionally, they are third most prevalent paediatric CNS tumour, making up to 30% of intracranial tumours in children under 3 years old.[1] Ependymomas show a bimodal age distribution, with peak incidences occurring at ages 6 and between 30 to 40 years. While paediatric cases mostly arise intracranially, over 60% of adult ependymomas are centered in the spinal cord.[2]

Myxopapillary ependymomas are distinguished by their well-defined borders in regions such as the conus medullaris, cauda equina, and filum terminale and they differ from conventional ependymomas. They appear hyperintense on T1-weighted MRI due to their mucin content.[3] Anaplasia in ependymomas is generally characterized by features like hypercellularity, frequent mitotic activity, pseudopalisading necrosis, vascular proliferation, and cellular and nuclear pleomorphism.[4] However, the histologic criteria for identifying and grading anaplasia in myxopapillary ependymomas remain a topic of debate, which complicates both diagnosis and prognosis. These tumours are known for their local invasiveness, tendency to spread through cerebrospinal fluid pathways, and high rates of recurrence, all of which present significant challenges in treatment and survival outcomes.[2,4]

In this article, we present a rare case of myxopapillary ependymoma with anaplastic features in an 8-year-old detailing clinical, histopathological, and immunohistochemical findings.

2. Case Report

An eight-year-old boy presented to neurology outpatient clinic with history of walking on his toes for duration of one year. He also experienced on-and-off stiffness and numbness in both limbs. His clinical examination and blood investigations were within normal limits. MRI spine showed a large T2 hyperintense T1 isointense heterogeneously

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enhancing well defined intradural mass lesion seen within the spinal canal. The lesion is seen extending from lower L3 level to S2 level. Lesion is seen completely filling the spinal canal. Lesion approximately measures 82 (CC) x 16.8(AP) x 24.1 (TR) mm. No significant extension into the neural foramina is noted. In Diffusion-weighted imaging, lesion shows peripheral rim of diffusion restriction and causes mild scalloping of the posterior part of L4 L5 S1 vertebral body as seen in (**Figure 1** a-c). Pre operative diagnosis made for this case was ependymoma. Radiological findings favoured possiblity of neurogenic tumor and a diffrential diagnosis of ependymoma was given by neuroradiologist.

Following all the investigtions patient was taken up for Posterior lumbar approach surgery, Laminectomy of L3 to upper sacral level with gross total resection of tumor was performed.

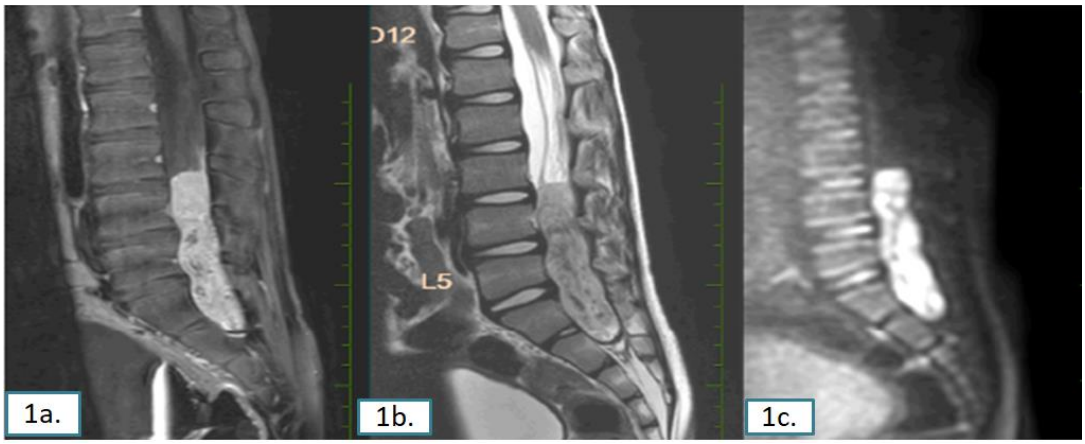

Figure 1: **a):** A large T2 Hyperintense heterogeneously enhancing well defined intradural mass lesion seen within the spinal canal. **b):** Lesion is extending from lower L3 Level to mid S2 level. **c):** Lesion shows peripheral rim of diffusion restriction, lesion is seen completely filling the spinal canal

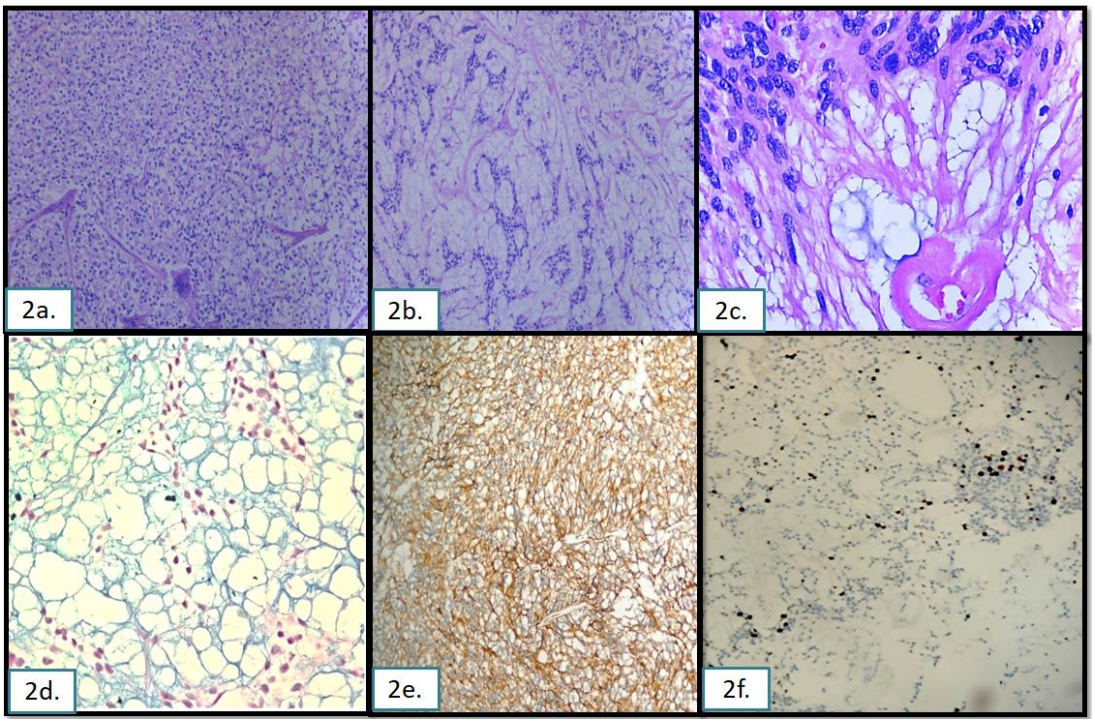

Figure 2: **a)**: Cellular tumour in sheets with increased mitotic activity (arrow), 10x H&E.(Hot spot); **b)**: Papillary,myxoid and microcystic areas with tumor cells around hyalinized fibrovascular cores,10 x. H&E. **c):** Rare eosinophilic globules and myxoid areas highlighted PAS stain 40x.(arrow). **d**); Alcian blue stain highlighting myxoid areas, 40x. **e)**: Tumour cells are diffuse and strongly positive for GFAP, 20x. **f)**: Increased MIB-1 Index (Hot spot region) 10x

3. Histopathology

Grossly tumour was received in multiple fragments altogether aggregating to 2 x 2 cm. serial sectioning through all the fragments shows grey white, tan and soft areas. Grossly no necrosis or haemorrhage was identified. Microscopic examination showed moderately cellular neoplasm arranged predominantly in solid sheets. Individual tumour cells are round to oval in shape with abundant clear cytoplasm and speckled chromatin. Focal perivascular myxoid areas with papillary arrangement around hyalinised fibrovascular cores are noted (**Figure 2** a,b). Microcystic myxoid areas surrounded by tumour cells, along with rare eosinophilic globules, are observed (**Figure 2** c). Scattered mitosis noted focally, reaching up to 5 per 10 high-power fields. There is no significant nuclear anaplasia, microvascular proliferation, or tumour necrosis noted.

Special stains such as Alcian Blue (**Figure 2** d) highlight myxoid areas. Immunohistochemistry reveals diffuse positivity for GFAP (**Figure 2** e), Vimentin, CD99, and focal positivity for D2-40. Tumour cells are negative for Pancytokeratin, Olig-2, S100, EMA, Synaptophysin, NeuN, and neurofilament protein. The MIB-1 proliferation index is focally increased up to 10- 12% (**Figure 2** f).

4. Discussion

Conventional myxopapillary ependymomas (MPE) usually has good overall survival, however, many of the cases will present with recurrence even with adequate excision. The recurrences are usually noted 2 to 15 years after post-surgery.[4-6] Recurrences usually occur in same site within the spine and very rare instances of metastasis to the cranium are also observed.[7] All the conventional myxopapillary ependymomas (MPE) patients have to be kept in follow up to prevent recurrences and also radiation therapy is recommended in many of the cases.[7]

Myxopapillary ependymomas usually arises in the filum terminal region but can also originate in other spinal cord levels such as intracranial sites which includes intraventricular, intraparenchymal and also occasionally in sacrococcygeal areas.

This entity was first reported in 1932 by Kernohan demonstrating mucinous changes and fibrovascular connective tissue.[8,9] Multiple genetic analyses show that myxopapillary ependymoma has got genome wide polyploidy occurring in between several chromosomes.[10] MPE shows specific loss of chromosome 16 and chromosome 12, also noted changes are increases of chromosome 4, chromosome 9 and chromosome 18, while the classical grade 2 ependymoma shows a specific loss of chromosome 16 and an increase of chromosome 12.[11] However, specific familial echogenic and environmental causes which predisposes to the malignant transformation or anaplastic transformation of myxopapillary ependymoma is not been identified or not been well understood.

Myxopapillary ependymoma may also present like disseminated disease with multiple spinal and intracranial metastasis. Same findings could be observed in both paediatric and adult patients.

The grading of MPE was revised from WHO grade 1 to CNS WHO grade 2 in the current version of the WHO classification, in response to the recognition of its partially aggressive clinical behaviour, recurrence, and dissemination.[13]

The potential for malignant behaviour in MPE was first suggested by Davis and Barnard in 1985, who reported cases of intracranial metastasis without distinct anaplastic features.[13,14] Subsequent reports have highlighted potential anaplastic features in myxopapillary ependymomas (MPE), though consensus on diagnostic criteria remains unclear. Anaplastic MPEs are known for frequent recurrence and reduced survival time. These tumours also tend to spread to other areas of the central nervous system via cerebrospinal fluid (CSF) and often show local invasiveness. It was proposed by WHO ‘typical myxopapillary ependymoma shows low level Mitotic activity typically with MIB-1 proliferation index not exceeding 2-3%. Exceptional examples are termed as anaplastic myxopapillary ependymoma which manifest regional hypercellularity, reduced mucin and also associated with at least 2 of the following features: ≥ 2 mitosis per mm^2, Ki-67 labelling index ≥ 10%, microvascular proliferation and spontaneous necrosis.[2,14]

In our case we had both findings of increased mitosis and also increased MIB- 1 proliferation index. With all the above-mentioned findings along with immunohistochemistry, diagnosis of anaplastic myxopapillary ependymoma is made.

Kanno H et al. conducted an extensive review of the literature and analysed 20 reported cases of anaplastic meningeal pattern epilepsy (MPE). Their findings revealed that the age of patients with anaplastic MPE ranged from 0.9 to 57 years, with an average age of 24.7 years. The majority of these patients were under 20 years old. They also observed that 50% of anaplastic MPE cases involved cerebrospinal fluid (CSF) dissemination and adjacent tissue involvement. Additionally, distant metastases to the spinal cord were noted in 9.3% of cases, while brain metastases were present in 6%.[13]

Malignant transformation of MPE can occur in both paediatric and adult patients, and is often linked to relapse, local invasion, CSF spread, or metastatic disease. These anaplastic clinical features suggest a more aggressive biological potential behaviour compared to classic MPE. As a result, close monitoring is essential. Additional studies are needed to refine the proposed assessment criteria for

anaplastic MPE and to identify genetic biomarkers associated with tumorigenesis and the malignant transformation of MPE.[15]

Gross total resection should be the primary goal of treatment as it tends to offer superior outcome. Adjuvant radiotherapy has also shown to aid in reducing recurrence.

5. Outcome

Post treatment patient was symptom free and kept on regular follow up. Repeat MRI performed after three months showed Post-surgical changes with no abnormal residual or recurrent mass lesion in the spinal canal with focal areas of fibrosis and adhesions seen in the subarachnoid space of lower lumbar spine.

6. Conclusion

In conclusion, while anaplastic MPE is rare in the literature, it should be considered in the differential diagnosis of spinal tumours, particularly in the filum terminale or lumbosacral regions. Due to the lack of a definitive histopathological classification, diagnosing this condition is challenging and requires specific microscopic or molecular criteria. Identifying MPE with anaplastic features is vital for accurate grading and management, given its aggressive nature and higher likelihood of recurrence.

7. Source of Funding

None.

8. Conflict of Interest

None.

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