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CONUS SCHWANNOMA MIMICKING MYXOPAPILLARY EPENDYMOMA - A RARE CASE REPORT

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ABSTRACT Schwannoma and Myxopapillary ependymoma (MPE) can be difficult to differentiate on imaging, especially as both are intradural extramedullary tumours often found in the conus medullaris and cauda equina regions. MPEs, while generally benign, can exhibit a tendency for local recurrence and potential for dissemination, which is why complete surgical resection is crucial. We report a 47 years old male patient presented with low back pain with radiating pain to the left lower limb, radiologically diagnosed with Myxopapillary ependymoma underwent near total excision and histopathological examination reported as Schwannoma.

KEYWORDS : Schwannoma, Myxopapillary ependymoma(MPE), Conus medullaris, Cauda equina

INTRODUCTION

Myxopapillary ependymoma is a distinct subtype of spinal cord ependymomas with a predilection for the lumbosacral region.[1] It represents 13% of all spinal ependymomas and accounts for 90% of all tumours in the conus medullaris.[2][3] Myxopapillary ependymoma is a benign and slow-growing neoplasm that has been histologically classified as grade 2, according to the World Health Organization (WHO) classification. Schwannoma is a benign, slow growing, encapsulated tumour arising from Schwann cells of the nerve sheath[4][5]. Although schwannomas account for 30% of all spinal neoplasms, the conus medullaris is an unusual location for these tumours[6][7].

Case Report

We report a 47 years old male patient presented with low back pain with radiating pain to the left lower limb, minimal sensory symptoms and no focal neurological deficits. Patient underwent MRI of whole spine with contrast and brain screening.MRI spine showing fairly circumscribed heterogeneously enhancing ovoid intradural mass lesion is observed extending from superior third of L1 vertebra to L3-L4 disc level. The lesion measures 10.4 x 2 x 1.5 cm (CC x AP x W) . The lesion demonstrates heterogeneous signal on the T2-weighted images with hyper intense areas interspersed with intermediate signal solid components. Post contrast study shows intense enhancement of solid component of the lesion and non-enhancing cystic components. The lesion is ventral to the conus medullaris at the level of L1 vertebra. Conus medullaris is compressed and displaced posteriorly and slightly towards the right side. Cauda equina roots are compressed and displaced to the periphery of thecal sac. No abnormal enhancement of cauda equina roots. No extension of the lesion into the neural foramen suggestive of Myxopapillary ependymoma. Case was discussed in multidisciplinary tumour board and planned for laminectomy and safe maximal excision under Intra operative neuro monitoring(IONM).Near total excision was achieved with intact IONM at the end of surgery and no post operative deficits. Patient's symptoms was completely relieved. Histopathological examination of the specimen was done which showing fragments of tumour tissue composed of variable cellularity with hypocellular and hypercellular area. Cell showed elongated spindle shaped wavy nucleus with bland chromatin. Hyalinised blood vessels are noted. No significant atypia seen. No increase in mitotic activity seen. No necrosis seen. Immunohistochemistry [Table 1] showed S100-clone: EP32 and SOX-clone:EP268 marker were positive suggestive of Benign nerve sheath tumour - Schwannoma.

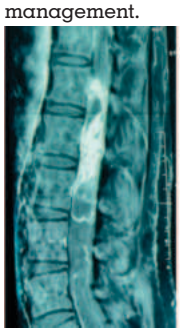
DISCUSSION

Schwannoma is a benign tumour of the peripheral nervous system that arises from Schwann cells and can occur in any body location where peripheral or cranial nerves are present. Schwannomas account for approximately 0.3% of primary intraspinal neoplasms. They tend to arise from the dorsal roots in the spinal region. The lumbar region is the most common location for spinal schwannomas[8][9]. Tumours arising in the cauda equina or around the conus medullaris can become larger than other spinal tumors because of the relative mobility of the roots and the wide diameter of the spinal canal. They may be well circumscribed, intradural or extradural, or combined intra-extradurally[10]. Schwannomas, together with meningiomas, are the most common intradural tumours. Generally, they occur between the fourth and sixth decades of life, and women and men are equally affected by this disease. Schwannoma can occasionally display degenerative changes like cyst formation, calcification, haemorrhage, and hyalinization. Myxopapillary ependymomas account for 1% to 5% of all spinal neoplasms and approximately 13%. Are a distinct subtype of glioma, arising from the ependyma of the filum terminale The age at onset varies between 30 and 50 years (mean age, 35 years). The most common location of myxopapillary ependymoma is the lumbosacral spine segment, mainly in the conus medullaris and cauda equina regions. Other rare locations include the cerebral ventricles and the brain.[11] Both MPEs and schwannomas are frequently found in the intradural extramedullary space, specifically around the conus medullaris and cauda equina. Their imaging features, especially on MRI, can be similar, making it difficult to distinguish them preoperatively. Both can cause similar symptoms, such as low back pain, leg pain, and neurological deficits. It's crucial to differentiate MPEs from schwannomas because MPEs can disseminate through the cerebrospinal fluid (CSF) during surgery, potentially leading to recurrence or metastasis. Radiologically MPEs may show a large, intensely enhancing, intradural extramedullary mass that extends for several vertebral levels and also have a high-T2, iso or low-T1 signal intensity masses with strong but inhomogeneous enhancement. Pathologically MPEs are characterized by perivascular degeneration and stromal mucin secretion and Schwannomas are composed of spindle-shaped cells arranged in a characteristic pattern. Surgical Considerations for MPEs, en bloc removal is often preferred to minimize the risk of CSF spread, incomplete resection or capsular violation can lead to increased recurrence

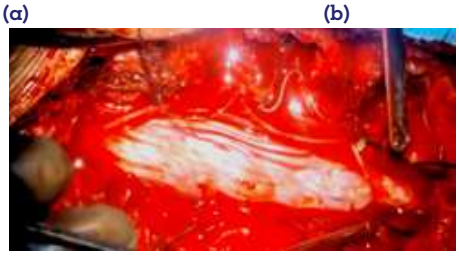
and neurological deficits. Schwannomas can be removed completely.

CONCLUSION

While MPEs and schwannomas can mimic each other on imaging, understanding their distinct features, including location, pathology, and potential for dissemination, is crucial for accurate diagnosis and appropriate surgical management.



1 (a) (b)



2(a)

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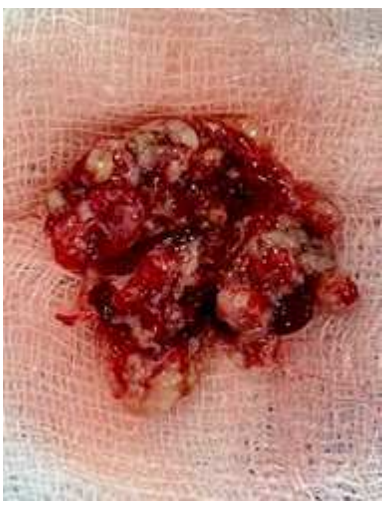

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1(a). MRI spine sagittal T1W1 contrast showing Fairly circumscribed heterogeneously enhancing ovoid intradural lesion with solid and cystic components extending from superior third of L1 vertebra to L3-L4 disc level

1(b). MRI spine axial T1W1 contrast showing the lesion is ventral to the conus medullaris which is compressed and displaced towards the right side

2(a). Intra operative image showing bulging of spinal nerves after durotomy

2(b). Intra operative image showing vascular tumour

2(c). Intra operative image showing complete decompression of spinal nerves after removing tumour

3. Tumour Specimen

Table 1 Immunohistochemistry

S.No	IHC marker	Result
1.	S100 - Clone : EP32	Positive
2.	SOX10 - Clone : EP268	Positive
3.	GFAP - Clone : EP13	Negative
4.	PR - Clone : 1E2	Negative
5.	SSTR2A - Clone :ZR233	Negative
6.	Ki67 - Clone : MIB - 1	6 - 8 %

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