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(CASE REPORT)

A case of myxopapillary ependymoma of filum terminale with atypical findings on MRI

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Abstract

Myxopapillary ependymoma is comparatively uncommon lesion of spinal canal with relatively characteristic location and MR imaging features. These lesions are a unique subtype of glioma that develops in the area of the filum terminale and conus medullaris, was first reported by Kernohan in 1932. Despite being slow-growing and generally benign, MPE has a propensity for local recurrence and can spread across the central nervous system. Atypical imaging features of the lesions may include long segment involvement, T1 hyperintensity etc. We are going to present one such atypical imaging presentation of myxopapillary ependymoma

Keywords: Filum Terminal; Ependymoma; Spinal Tumours; MRI; Conus Medullaris

1. Introduction

Only 5–10% of tumors in the central nervous system (CNS) are spinal tumors, making them comparatively uncommon. Seventy to eighty percent of spinal tumors are intradural extramedullary in site, with meningiomas and schwannomas being the most prevalent. The less frequent intradural extramedullary lesion known as myxopapillary ependymoma (MPE), a unique subtype of glioma that develops in the area of the filum terminale and conus medullaris, was first reported by Kernohan in 1932 (1). Although it makes up 13% of all spine ependymomas, it is the most prevalent tumor in the conus and cauda equina, making up 83% of all tumors in this area. Despite being slow-growing and generally benign, MPE has a propensity for local recurrence and can spread across the central nervous system, which could have negative consequences, particularly in young patients. Furthermore, a diagnosis might not be made until the tumor is fairly large and there is little chance of a cure due to its typical slow growth and subtle clinical signs. Early diagnosis, which is essential for a successful outcome, can be obtained by MR imaging (2).

2. Case report

A 33-year-old man with a 6-month history of aggravating low back and leg pain was presented. On physical examination, there was no significant neurological abnormality. MR images showed Small well-defined, space occupying lesion of approximate size 12 x 14 x 18 mm (AP x TR x CC) with internal solid and cystic components at distal end of the conus medullaris/filum terminale at L1 level, causing its expansion. Adjacent cauda equina roots appears peripherally displaced. Lesion was intradural extramedullary. Lesion showed mild T1 hyperintensity with non-suppression on STIR and T2 FS images- likely due to proteinaceous/mucinous component. On T2, hyperintense lesion appears along with heterogeneous contrast enhancement (Fig 1 & 2).

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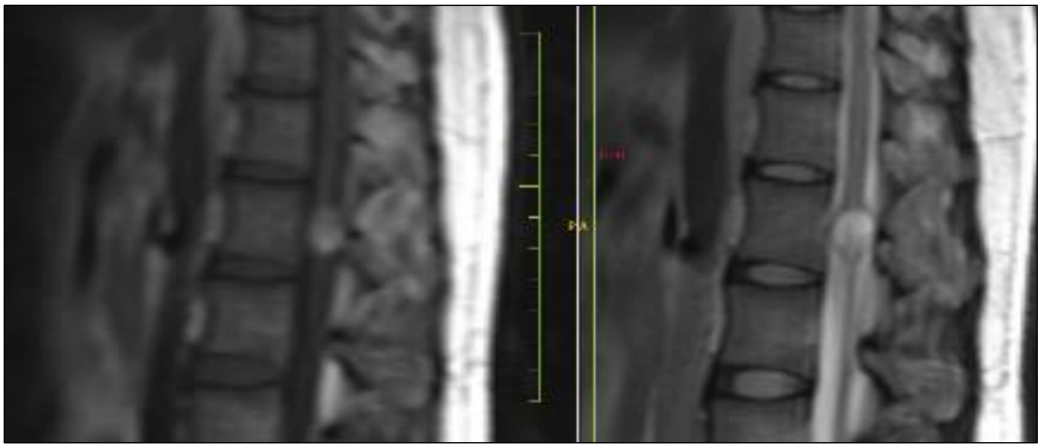

Figure 1 Sagittal T1 and T2 images of dorsolumbar spine showing a well defined oval shaped intradural lesion with hyperintense signal on both T1 and T2 images- Raising the suspicion of lipomatous lesion, however, no fat suppression was noted on fat saturated images

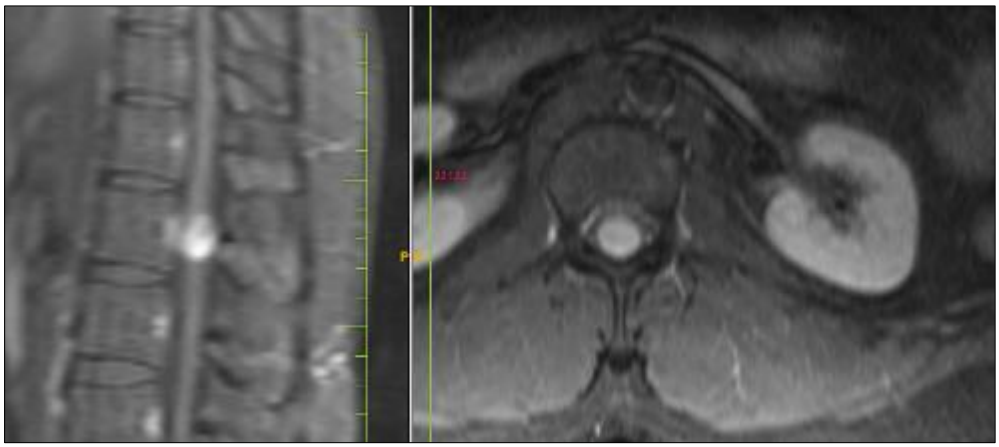

Figure 2 Sagittal and axial fat saturated images of the lesion showing non-suppression of T1 signal, raising the suspicion of proteinaceous content within the intradural lesion

Patient underwent excision biopsy which showed cuboidal to elongated tumor cells radially arranged in a papillary manner surrounding the vascularized stromal cores in a mucoid background, confirming the diagnosis of myxopapillary ependymoma

3. Discussion

In cauda equina and conus medullaris, myxopapillary ependymoma is usually extramedullary and intradural. The tumor typically starts in the filum terminale and often spreads to the conus medullaris and/or cauda equina nerve roots. L2 is most common site for its occurrence, followed by L1 and L3. MPE is typically solitary, but up to 43% of individuals have numerous lesions, which could be drop metastases (3).

The filum terminale myxopapillary ependymoma has a lengthy clinical history and is a histologically benign tumor. The most common symptoms include sciatica and/or low back pain. Additionally, the tumor manifests as gastrointestinal dysfunction, gait abnormalities, and saddle sensory loss. The surface of the sausage-shaped bulk is lobulated and enclosed. These tumors can occasionally develop in the extradural area (4).

About 14–43% of individuals have been observed to have multiple lesions. When they first appear, osseous alterations are seen in 63% of spinal ependymomas. The posterior vertebral bodies frequently scallop or the medial side of the pedicle erodes (5).

An intradural extramedullary, sausage-shaped, lobulated, and frequently encapsulated thoracolumbar mass that extends for multiple vertebral levels and may enlarge the spinal canal or, less frequently, a neural foramen is the classic presentation of MPE (6). While large tumors may enclose the lumbosacral nerve roots, small tumors tend to displace them. MPE is typically T1 isointense to the cord and T2 hyperintense; it is less frequently T1 hypointense or hyperintense, with the latter occurring as a result of bleeding or mucus. T1 hyperintensity aids in distinguishing ependymoma from other subtypes (7). Reduced signal intensity due to superficial siderosis may happen, indicating the tumor's vascularity (8).

Sometimes, myxopapillary ependymoma lesions may have proteinaceous content in cystic component, leading to intrinsic T1 hyperintensity, thus mimicking to the intradural lipoma. However, in such cases, no fat suppression is seen on fat saturated images (9).

4. Conclusion

Myxopapillary ependymoma is the most prevalent tumor in the conus and cauda equina region. It is a kind of ependymoma that usually occurs intradural extramedullary. Although it has a propensity for local recurrence and the potential for intrathecal metastases, it is often less aggressive than other forms of ependymoma. The clinical appearance frequently resembles discogenic illness, which could cause a delay in diagnosis. The ability to detect tumors when they are tiny is critical for a possible cure, and MR imaging is helpful in assessing the size of tumors and their link to intraspinal structures.

List of abbreviations

- MPE- Myxopapillary ependymoma
- MRI- Magnetic resonance imaging

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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