

## Totally extradural spinal en plaque meningiomas – Diagnostic dilemmas and treatment strategies

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### Abstract

**Background:** Meningiomas are the second most common intraspinal tumors, constituting ~25% of all intraspinal tumors; however, in the context of extradural spinal lesions, the diagnosis of meningioma is an uncommon one. Purely extradural spinal meningiomas, especially of the en plaque variety, frequently mimic metastatic disease and may result in inadequate therapy.

**Case Description:** We report two cases of totally extradural en-plaque meningiomas of the spine, one each in the cervical and dorsal spine. We present the significant diagnostic dilemmas posed by these cases and discuss the pathogenesis, treatment strategies, and long-term behavior of these uncommon lesions.

**Conclusion:** Attention needs to be drawn to this dangerous preoperative and intraoperative misinterpretation. Intraoperative histopathology support for correct identification, gross total resection at surgery, inclusion of a durotomy to rule out intradural extension, and long-term follow-up are cornerstones for successful management of totally extradural en plaque spinal meningiomas.

**Key Words:** En plaque meningiomas, extradural spinal lesions, spinal meningiomas

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## INTRODUCTION

Meningiomas account for 25% of all intraspinal neoplasms and are the second most common primary intraspinal tumor.<sup>[2]</sup> The majority of spinal meningiomas are entirely intradural and <10% show an extradural component.<sup>[5]</sup> Meningiomas located purely in the extradural space are exceedingly rare and, therefore, are seldom considered in the differential diagnosis of spinal extradural lesions.<sup>[5]</sup> We present two cases of totally extradural en plaque meningiomas of the spine, which posed significant intraoperative dilemma as to the nature of the pathology and the treatment strategy.

## CASE REPORTS

### Case 1

#### *Clinical and radiological presentation*

A 35-year-old female presented with the chief complaints of neck pain, progressive spastic quadriparesis, and decreased sensations below the spinal level of C7. Magnetic resonance imaging (MRI) revealed an extradural en plaque lesion extending from C3 to C6 vertebral levels and invading the C4 and C5 lateral spinous processes, along with encasement of the vertebral artery traversing the C4 and C5 transverse (vertebral) foramina. The lesion was iso- to hypointense on T1-weighted as well as

T2-weighted MR images and showed intense contrast enhancement [Figure 1]. Pre-operative impression was an extradural malignant spinal tumor.

*Operative intervention*

The lesion was explored through C3-C6 hemilaminectomy. Intraoperative findings were that of a firm, fleshy, highly vascular tumor, and frozen section histopathology revealed meningothelial meningioma. In view of the benign pathology at hand, near-total excision of the tumor was achieved at surgery, leaving behind the part encasing the vertebral artery.

*Postoperative course*

The patient had complete resolution of symptoms and follow-up radiology at 1 year showed small residue in the region of the lateral process encircling the vertebral artery, but without any dural compression [Figure 1d and h]. At 16 months of follow-up, the patient is asymptomatic, and will be kept under regular clinical and radiological follow-up in view of the residue.

**Case 2**

*Clinical and radiological presentation*

A 23-year-old female presented with spastic paraplegia and bowel and bladder involvement of 7 days duration. This was preceded by pain in the dorsal spine, radiating to both lower limbs for 2 months, and progressive weakness and spasticity in the lower limbs for 2 weeks. On examination, she had spastic quadriplegia and loss of all sensory modalities below the D5 dermatome. An urgent MRI of the thoracic spine showed an extradural en plaque spinal lesion located posterolateral to the spinal cord, pushing the dura anteriorly [Figure 2]. The lesion was hypointense on T1-weighted MR, isointense on T2-weighted MR, and intensely enhancing on contrast administration.

It arose primarily from the left posterolateral corner of the spinal canal, and extended predominantly dorsally but also ventrally in relation to the cord [Figure 2a-c]. The lesion was traversing the D4-D5 intervertebral foramen up to its outer limit. In view of the extradural location in the spine, the differentials were metastases, lymphoma, or a granulomatous infection like tuberculosis.

*Operative intervention*

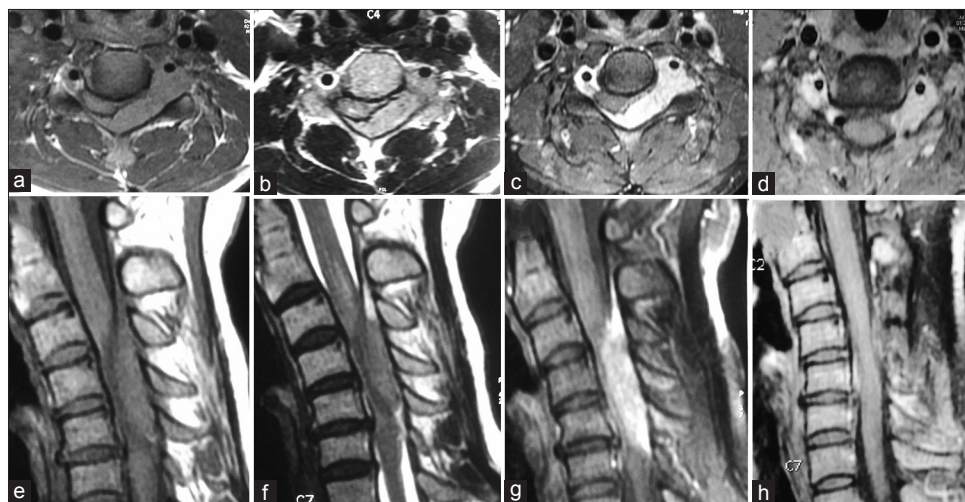
The patient underwent emergency D3-D5 laminectomy and decompression of the lesion. Intraoperative findings revealed a vascular and fleshy lesion present over the dura. Unfortunately, frozen section was not done at the first surgery, as this case was taken up as an emergency during the night. Histopathology was unequivocal for meningioma [Figure 3b].

*Postoperative course*

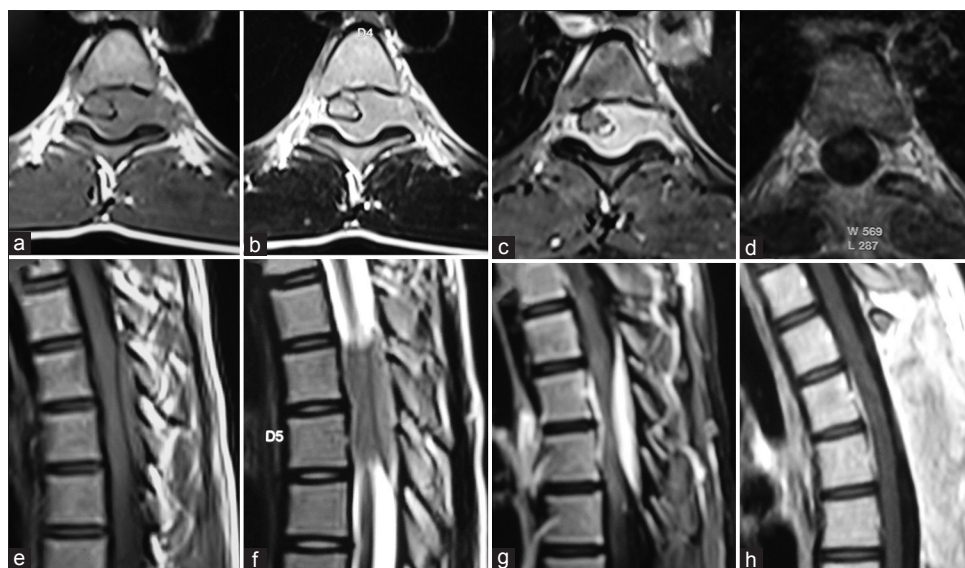
Although the patient improved after decompression, the benign nature of the pathology prompted us to re-explore with a wider left posterolateral approach and perform a total excision of the tumor. At surgery, the tumor could be peeled off the dura, enabling a total excision [Figure 3a]. The patient improved in the postoperative period and has regained grade 5/5 power in both lower limbs, sensations, and bowel and bladder function at 3 months follow-up. Follow-up MRI showed no evidence of residual or recurrent lesion [Figure 2d and h].

**DISCUSSION**

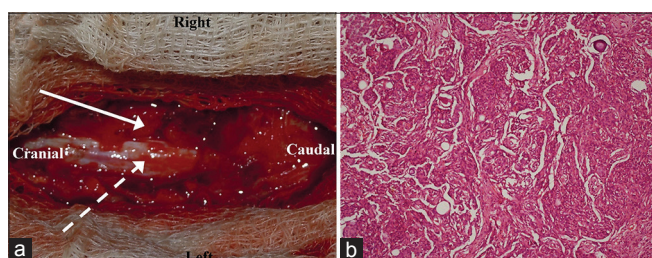
Meningiomas are the second most common intraspinal tumors. Approximately 80% occur in the thoracic spine, affecting women four times more often than men, with a peak incidence during the fifth to sixth decades of



**Figure 1: MRI findings in Case 1, showing an extradural lesion extending from C3 to C6 vertebral levels. The lesion appears isointense on T1-weighted images [(a) axial, (e) sagittal], hyperintense on T2-weighted images [(b) axial, (f) sagittal], and exhibits intense enhancement on contrast administration [(c) axial, (g) sagittal]. Postoperative contrast-enhanced MRI [(d) axial, (h) sagittal] at 1 year follow-up showing small residue laterally on the left side, encircling the vertebral artery without any compression on the dural sac**



**Figure 2:** MRI findings in Case 2, showing an extradural lesion extending from D3 to D6 vertebral levels. The lesion is isointense on T1-weighted images [(a) axial, (e) sagittal], hyperintense on T2-weighted images [(b) axial, (f) sagittal], and intensely contrast enhancing [(c) axial, (g) sagittal]. Postoperative contrast-enhanced MRI [(d) axial, (h) sagittal] at 3 months follow-up shows gross total excision of the tumor



**Figure 3:** (a) Intraoperative image of the fleshy extradural meningioma (arrow) on the right side of the thecal sac, with the shining white dura (dashed arrow) under the tumor. (b) Photomicrograph (HE stain) showing a meningothelial meningioma arranged in syncytium and whorling pattern. Few psammoma bodies are also present

life.<sup>[3]</sup> Spinal meningiomas are predominantly ventrally or ventrolaterally placed intradural extramedullary lesions.<sup>[2]</sup> They may have an extradural component in approximately 10% of cases; but an exclusively extradural meningioma is an uncommon entity.<sup>[5]</sup> This entity assumes significance in the context that extradural spinal lesions are most commonly metastatic neoplasms (including lymphoma), and hence, the correct intraoperative identification is indispensable for instituting the correct treatment strategy.

### Pathogenesis

Several theories have been postulated to explain the pathogenesis of purely extradural meningiomas. Most likely, they arise from the ectopic or separated arachnoid tissue around the periradicular nerve root sleeve, where the spinal leptomeninges merges directly into the dura.<sup>[5]</sup> It has also been suggested that the periradicular dura, being less thick, may contain vestigial remnants of the superficial layer of the embryonal arachnoid mater and

villi. This phenomenon might give rise to the extradural location and root proximity of some meningiomas.<sup>[3]</sup> Another theory suggests that islands of arachnoid tissue that may have migrated into the extradural space can be the source of these meningiomas.<sup>[3,5]</sup>

### Treatment strategies

Purely extradural spinal meningiomas, especially of the “en plaque” variety, may mimic metastatic disease.<sup>[1]</sup> Intraoperative histology is mandatory for optimal surgical decision-making; hence, frozen section histopathology should be available for these cases. As seen in Case 1, intraoperative frozen section enabled us to correctly identify this pathology and perform near-total resection. However, the absence of intraoperative frozen section in Case 2 (as the case was taken up as an emergency) proved detrimental as only decompression was achieved at the first setting. Attention needs to be drawn to this dangerous preoperative and intraoperative misinterpretation, which may easily entail an inadequate therapy.<sup>[1,2]</sup>

Once intraoperative diagnosis of meningioma is confirmed for an extradural spinal lesion, the surgeon should consider gross total resection of the tumor, including excision of the tumor extending into the bone or the paraspinal space. In order to gain access to the tumor on the ventral aspect of the spinal cord through the postero-lateral approach, wide lateral removal of the lamina, the pedicles and the facet joints may be necessary. Stabilization of the spine may be required, if the postero-lateral approach compromises the stability of the spine.<sup>[4]</sup> This strategy is likely to give the best postoperative outcome, as prognosis depends on the extent of resection.<sup>[1]</sup>

Another important aspect to be considered is the treatment strategy for the underlying dura. The pathogenesis of extradural spinal meningiomas suggests that these tumors arise from the dural root sleeve and not from the external surface of the spinal dura. Consequently, as seen in both our cases, these tumors can be stripped off from the spinal dura, without the need to excise the dura. Figure 3a shows the intraoperative image of Case 2, in which the tumor has been stripped off in the center to expose the shining-white dura underneath. Even though there is no need for the dura to be excised, it is imperative that the dura be opened to rule out intradural extension of the meningioma. The rationale for this maneuver comes from the observation made by Tuli *et al.* that out of the 47 reported cases of purely extradural spinal meningiomas, only 12 cases were subjected to a durotomy; and in three of these durotomies, an intradural extension was observed.<sup>[3]</sup>

### Long-term behavior

Conflicting reports exist for the long-term prognosis of patients with extradural spinal meningiomas. While some authors have suggested that these tumors have a locally malignant course, others have shown these meningiomas to be benign.<sup>[3]</sup> These meningiomas are most likely to behave as per their pathological (WHO) grade. The worse prognoses for patients with extradural meningiomas may be because of the difficulty in completely removing the tumor due to its bony involvement or paraspinal extent.<sup>[1]</sup> As seen in Case 1, the small residue has been static over 1 year without any adjuvant treatment being administered to the patient. The follow-up in our cases suggests that these tumors behave in a benign manner, but taking into consideration the literature review, it would be wise to keep these patients under long-term clinical and radiological follow-up.

The prevalence of extradural meningiomas has been described as ranging between 3.3% and 21.4% of all spinal

meningiomas.<sup>[3]</sup> However, in the context of extradural spinal lesions, the diagnosis of meningioma is an uncommon one. Tuli *et al.* have tabulated all such case reports in their extensive review of literature, and may have correctly inferred that the presence of extradural meningiomas is not as rare as once thought.<sup>[3]</sup> This highlights the fact that meningiomas should be rightfully included in the differential diagnosis of extradural intraspinal masses. The ubiquity of MR imaging, coupled with prolonged survival of patients with malignant disease in view of advanced oncology care, is likely to result in frequent diagnostic and therapeutic dilemmas for such intraspinal masses.<sup>[5]</sup> Intraoperative pathology support is essential in establishing the diagnosis of this uncommon entity. It assumes importance as total excision undertaken for this uncommon benign entity, vis-à-vis decompression undertaken for the common malignant pathologies, can significantly affect the patient outcome.

### Disclosures

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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