Case Lesson 46-2026

Primary leptomeningeal melanocytic tumor of intermediate grade of the thoracic spine: An extremely rare tumor

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Introduction

We report a case of a primary leptomeningeal melanocytic tumor of intermediate grade presenting as an intramedullary thoracic spinal cord lesion with progressive neurological deterioration, highlighting the diagnostic challenges and the importance of surgical intervention for both decompression and definitive histopathological diagnosis. These tumors are very rare, with an estimated incidence of 1 per 10 million individuals[4].

Case presentation

A 76 years old man, presented with a one-year history of thoracolumbar pain associated with urinary disturbances. The symptoms progressed, with the patient reporting worsening thoracolumbar pain, new-onset weakness of the right lower limb, and increasing difficulty with ambulation. A spinal MRI showed an intradural lesion at the T8–T9 level, isointense on T1-weighted images, heterogeneously iso- to hypointense on T2/FLAIR sequences, and demonstrating contrast enhancement after gadolinium administration. The radiological differential diagnosis included probably meningioma versus cavernous malformation.

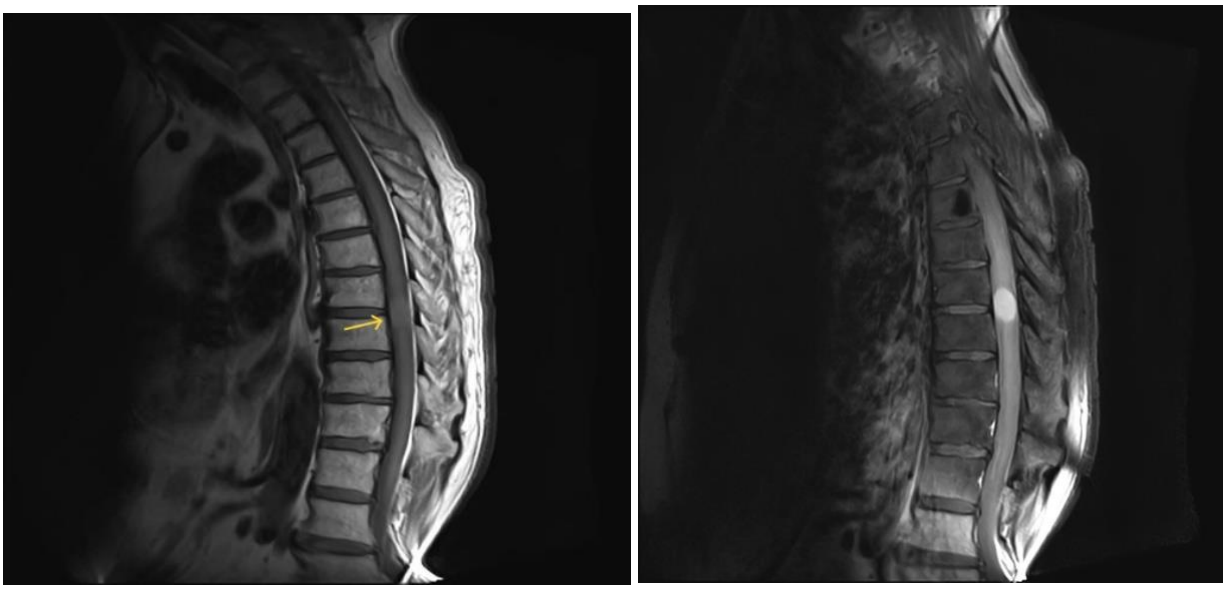

Fig1. *Sagittal T1 weighted MRI sequences with and without gadolinum: Intradural hyperintense lesion at T8-T9 level, and contrast enhancement after gadolinium administration.*

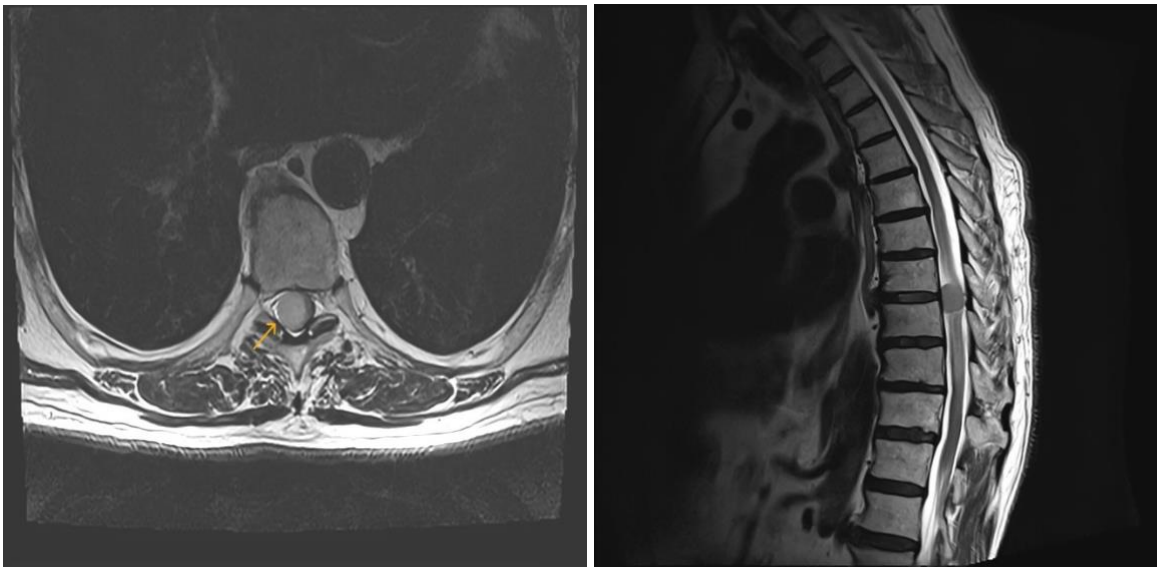

Fig2. *Axial and Sagittal T2 weighted MRI sequences: Intradural iso to hypointense lesion at T8-T9 level*

Neurological examination demonstrated right lower limb monoparesis with muscle strength graded at 2/5, a sensory level at T6, and hyperreflexia predominantly on the right side compared to the left. There was also a reduction in tactile and thermoalgesic sensation on the right side.

The patient underwent surgical excision via T8-T9 laminectomy. Intraoperatively, the lesion was intradural, attached to the posterior radix, expanded subduraly brown to dark mater without clear

pial cleavage to the medulla. It was possible through aspiration to consider GTR. Intraoperative impression may be cavernoma or other origin.

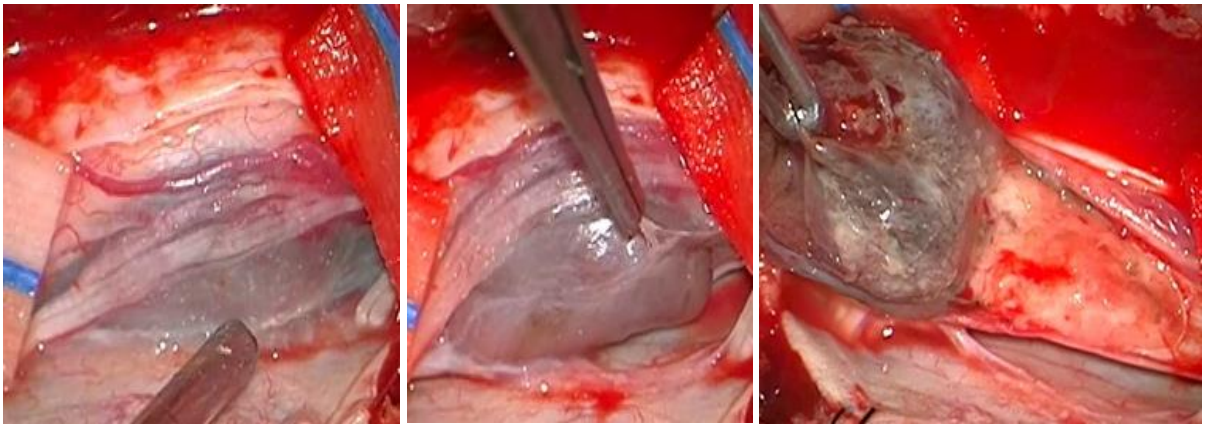

Fig3. *Intraoperative view of the lesion.*

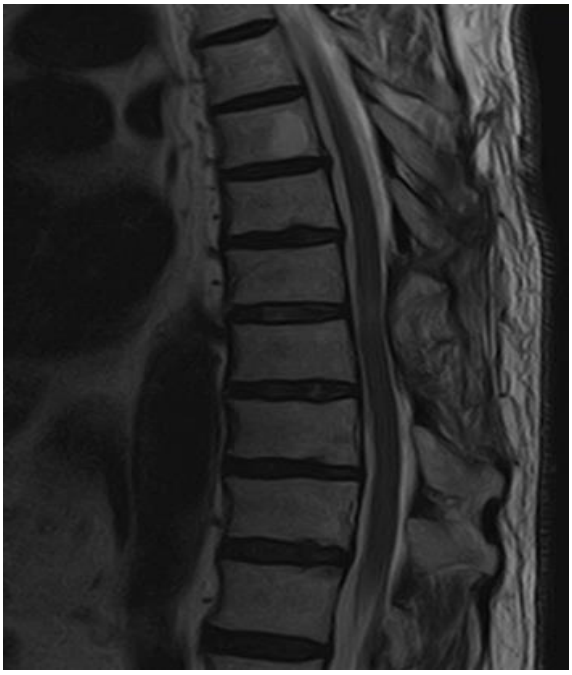

Fig4*. Immediate post-operative Sagittal T2 weighted MRI*

Histopathological comments of Dr. B.Latifaj Primary leptomeningeal melanocytic tumor of intermediate grade (also known as intermediate-grade meningeal melanocytoma or intermediate-grade melanocytic neoplasm per WHO 2021 classification of CNS tumors).

This ultra-rare tumor (incidence ~1 per 10 million/year for melanocytomas overall; intermediate-grade even rarer) originates from leptomeningeal melanocytes and fits the spectrum of circumscribed primary meningeal melanocytic tumors. It is distinct from metastatic melanoma,

primary leptomeningeal melanoma (malignant, with marked atypia, higher mitoses/necrosis/Ki-67 >10%, and poorer prognosis), and benign/low-grade melanocytoma (typically <0.5–1 mitosis/mm², Ki-67 <5%). The intermediate grading here is driven by hypercellularity, mitotic activity (~2/mm²), and elevated Ki-67 (~8%), despite lacking necrosis, significant atypia, or parenchymal invasion.

The grey-brown intraoperative appearance reflects variable pigment content align with reported features of these vascular/pigmented lesions. MRI variability (here T1 isointense rather than classic T1 hyperintense) occurs with lower melanin/hemorrhage amounts, explaining the preoperative meningioma/cavernoma differential.

Gross total resection is the cornerstone of management and was successfully achieved, offering the best chance for long-term control. These tumors generally carry a better prognosis than malignant melanoma but higher recurrence risk than benign melanocytoma recurrence rates after GTR range ~20–40% in series (higher with subtotal resection), with potential for local progression or (rarely) malignant transformation.

Given the recent surgery (only ~3 weeks prior) the patient is in the early postoperative period. Expected neurological recovery depends on the degree and duration of preoperative cord compression—many experience stabilization or gradual improvement in paraparesis and sensory symptoms with rehabilitation, though full recovery is not guaranteed if deficits were advanced.

Close clinical and radiological follow-up is strongly recommended (e.g., serial spinal MRI every 3–6 months initially, then annually if stable) to monitor for residual/recurrent disease. Adjuvant radiotherapy is not routinely indicated after GTR for intermediate-grade lesions but may be considered if recurrence develops. No role for systemic therapy unless progression or transformation occurs.

This diagnosis explains the unusual pigmentation and intraoperative findings that mimicked a vascular malformation, highlighting the importance of histopathology in resolving such differentials

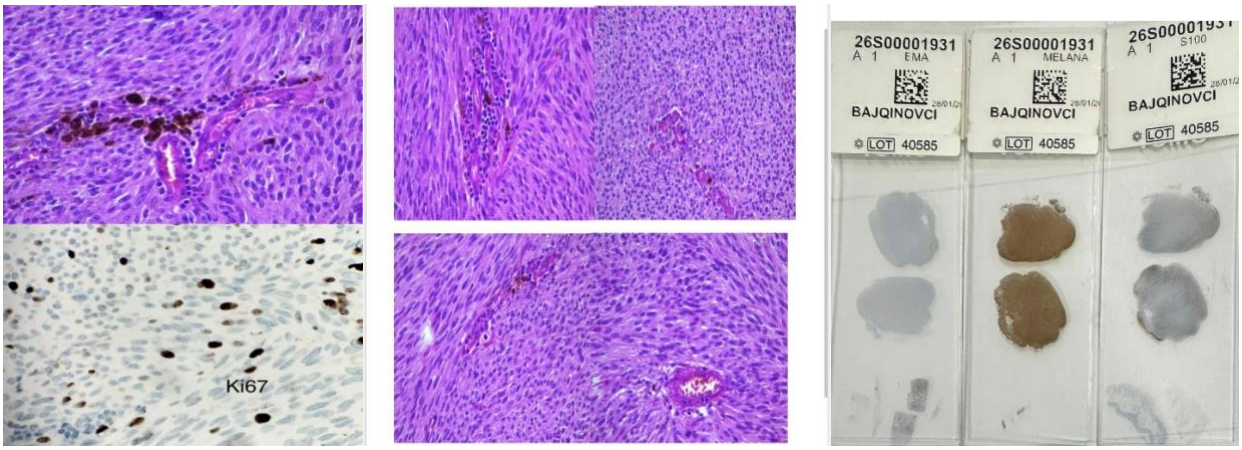

Fig4. *Immunohistochemistry: melanocytic differentiation; Melan-A positive, S100 positive, EMA negative (excluding meningioma). The Ki-67 proliferation index was approximately 8%.*

Discussion

Primary leptomeningeal melanocytic tumors arise from neural crest–derived melanocytes normally present in the leptomeninges[2]. These tumors were first reported in 1972, by Limas and Tio[1,3]. Since then 100 cases have been reported in the literature[5]. Spinal involvement is less frequent than intracranial. In 1999. Brat et al[6] introduced a new pathological diagnosis: intermediate-grade melanocytic tumors which distinguished itself from well differentiated melanocytomas and the malignant melanomas. They represent a particularly uncommon and poorly characterized subgroup. Literature reviews up to 2024 identified 19 cases of intermediate grade melanocytoma[7].

In addition to imaging and histopathological evaluation, molecular findings, particularly mutations in GNAQ or GNA11, further aid in differentiating primary leptomeningeal melanocytic tumors from metastatic melanoma, which more commonly have BRAF or NRAS mutations[1].

Conclusion

This case contributes to the limited series of literature on intermediate-grade primary leptomeningeal melanocytic tumors of the spine and highlights the importance of histopathology in resolving such differentials.

Reference:

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