Case Report

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Transparsinterarticularis approach for resection of a malignant melanotic nerve sheath tumour of the dorsal spine: A case report

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Introduction: Malignant melanotic nerve sheath tumours are extremely rare central nervous system neoplasms. Initially termed as Melanotic schwannoma the nomenclature was revised in 2020 WHO classification to malignant melanotic nerve sheath tumour (MMNST). They are rare aggressive peripheral nerve sheath tumours. In spine, they more commonly occur in the lumbosacral region (47.2%), followed by the thoracic (30.5%) and cervical (22.2%) segments. Most common age group affected is between 20-50 years. MMNSTs often tend to metastasize early and have poor prognosis. Surgical excision is the mainstay of treatment followed by radiotherapy and/or chemotherapy.

Case report: Here we present the case of a 54-year- old male who presented with gradually progressive lower limb weakness and hypertonia with bowel and bladder involvement. Magnetic resonance study of the spine suggested an intradural extramedullary melanoma at D4 level of spine. The rest of the physical examination and metastatic workup were unremarkable. The patient subsequently underwent tumour excision via trans-parsinterarticularis approach. Histopathological examination was suggestive of malignant melanotic nerve sheath tumour. Following surgery the patient's lower limb weakness improved significantly. At six-month follow-up patient did not show any signs of recurrence.

Conclusion: Malignant melanotic nerve sheath tumours (MMNSTs) are extremely rare highly aggressive lesions that are often misdiagnosed on neuroimaging. When a spinal tumour arising from a nerve root demonstrating the characteristic of T1 hyperintensity and T2 hypointensity, MMNST should always be included in differential diagnosis and metastatic workup, Clinical and radiological evaluation should be done to rule out other associated syndromes. Complete surgical excision followed by vigilant follow-up for early detection of recurrence is recommended.

Keywords: *spinal tumour; malignant melanotic nerve sheath tumour; immunohistochemistry; spine surgery*

Introduction

Malignant melanotic nerve sheath tumours (MMNSTs) are extremely rare tumours of the central nervous system [1]. Initially termed as melanotic schwannoma the nomenclature was revised in 2020 WHO classification to malignant melanotic nerve sheath tumour (MMNST) [2]. These tumours are rare aggressive peripheral nerve sheath neoplasms [3]. In the spine they most commonly occur in the lumbrosacral region (47.2%), followed by thoracic (30.5%) and cervical (22.2%) regions [4]. Most common age group affected is between 20-50 years. MMNSTs often tend to metastasize early and have poor prognosis [5]. Surgical excision is the mainstay of treatment followed by radiotherapy and chemotherapy [6]. Through this case report we aim to highlight the presentation and management of a thoracic MMNST.

Case Report

Here we present a 54-year-old male with two-month history of gradually progressive weakness and tightness in both lower limbs. On examination the power of both lower limbs was assessed as grade 2/5 according to Medical research council (MRC). Hypertonia of grade 3 was noted bilaterally in the lower limbs, and sensory hypoaesthesia was present in approximately 80% of the area below the nipple line.

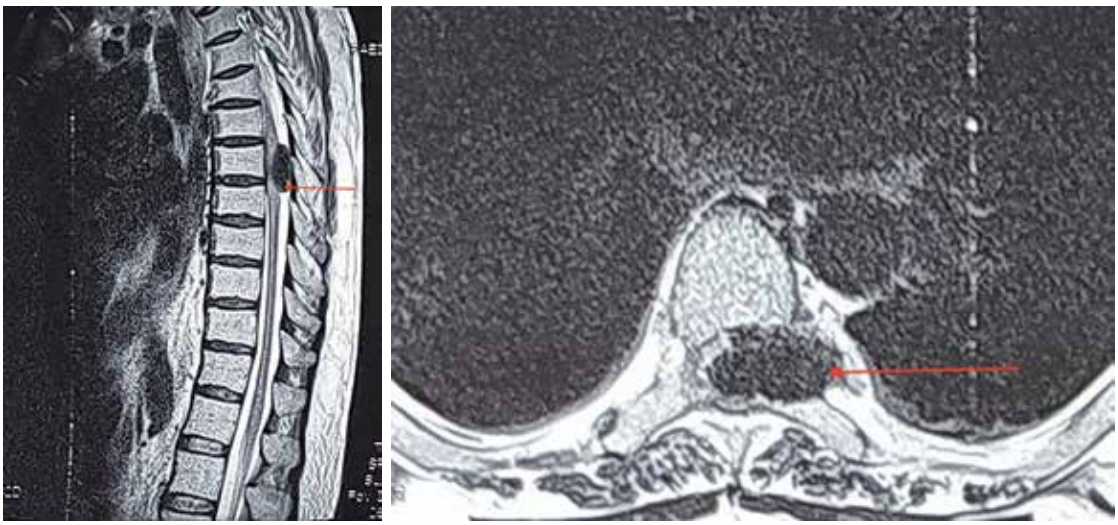
He had bladder and bowel incontinence. The remainder of his physical examination was within normal limits. Magnetic resonance imaging (MRI) of his spine revealed a T1 hyperintense and T2 hypointense lesion at the level of D4 vertebra. (***Figures 1A, 1B, 2A*** and ***2B***). The lesion was homogeneously enhancing on contrast and was located intradurally and extramedullary on the left side. A differential diagnosis of metastatic lesion was made and thorough metastatic workup done, which however failed to show any primary tumour elsewhere.

The patient underwent left transparsinterarticularis approach under general anesthesia and tumour was identified. Grossly, the tumour was blackish in colour, highly vascular and adherent to adjacent structures and the dura arising from the exiting dorsal nerve root

(***Fig.3***). The tumour was meticulously dissected from the dura and surrounding tissues; the involved dorsal nerve root was sacrificed (***Fig. 4***). The lateral portion of the tumour was found adherent to pleura and was excised (***Fig. 5***). The total duration of surgery was 120 minutes, with an estimated blood loss of approximately 150 ml.

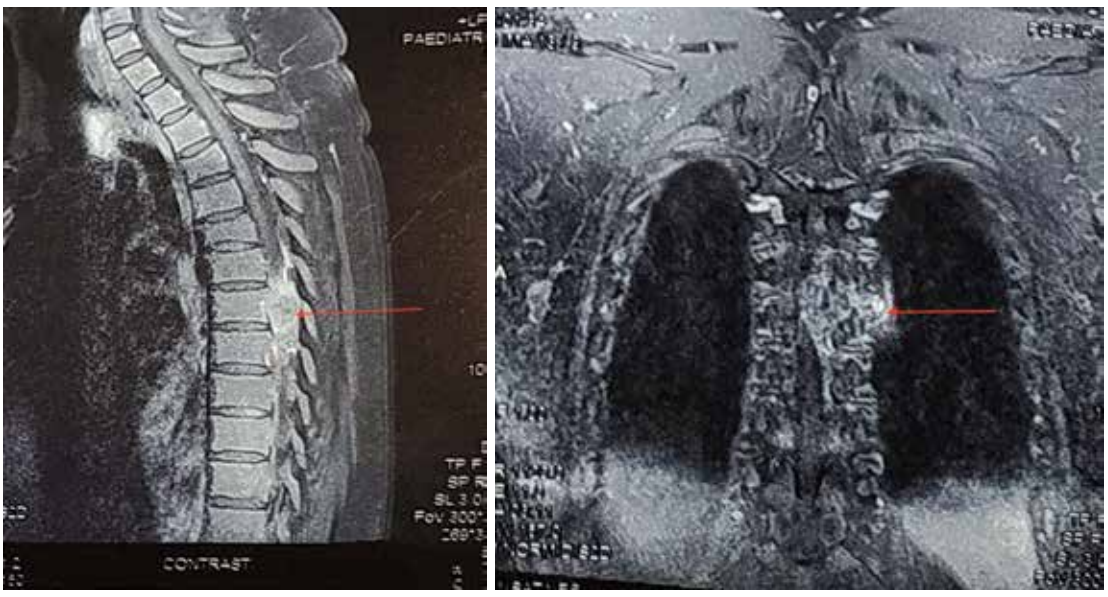
The patient was mobilized with support the very next day of surgery, followed by aggressive passive and active lower limb physiotherapy. Postoperatively, patient's lower limb power improved to MRC grade 4/5 and sensory hypoaesthesia reduced by 50%. The patient was able to walk with support. Histopathological examination revealed the tumour as malignant melanotic nerve sheath tumour (***Fig. 6A*** and ***6B***). IHC panel showed HMB45, S100, SOX10 positive, Ki67 index was 3%. GFAF, BRAF and V600E were negative (***Fig.7A*** and ***7B***).

The patient was followed up after 6 months demonstrating substantial improvement in neurological status and no evidence of recurrence. In the absence of recurrence at thesix-month months follow-up, no adjuvant therapy was recommended on multidisciplinary team meeting.



A **B**

Fig. 1A. Saggital T2-weighted MRI showing a hypointense lesion compressing the spinal cord at D4 level (red arrow)
Fig. 1B. Axial T2-weighted image showing a hypointense dumbbell-shaped lesion on left side with significant cord compression extending transforaminally (red arrow)



A **B**

Fig. 2A. Contrast-enhancing lesion at D4 (red arrow)
Fig. 2B. Coronal section image showing spinal lesion extending to the thorax (red arrow)

This article contains some figures that are displayed in color online but in black and white in the print edition.

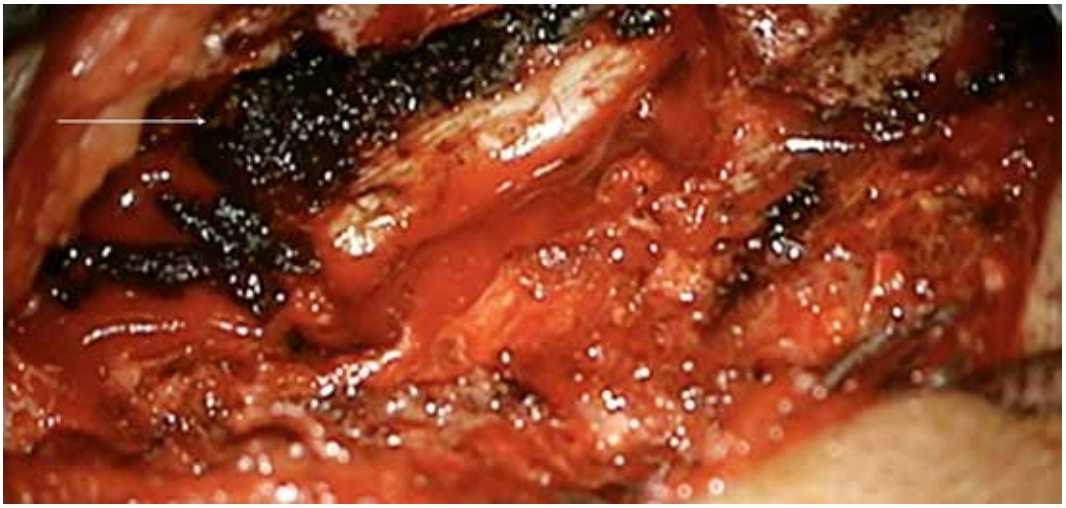


Fig. 3. Melanotic lesion observed lateral to thecal sac extending transforaminally (white arrow)

Fig.4. Complete excision of the lesion

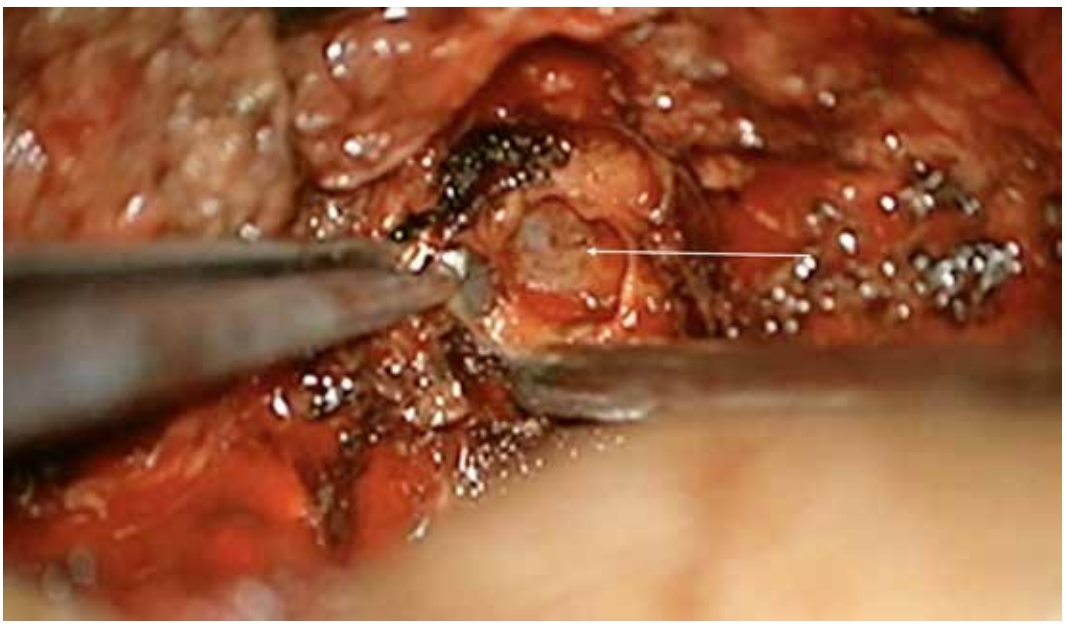
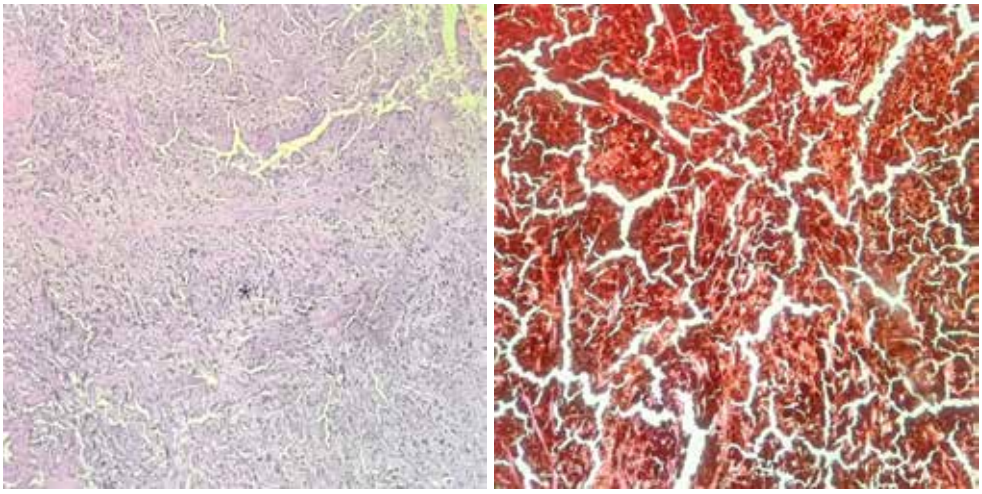
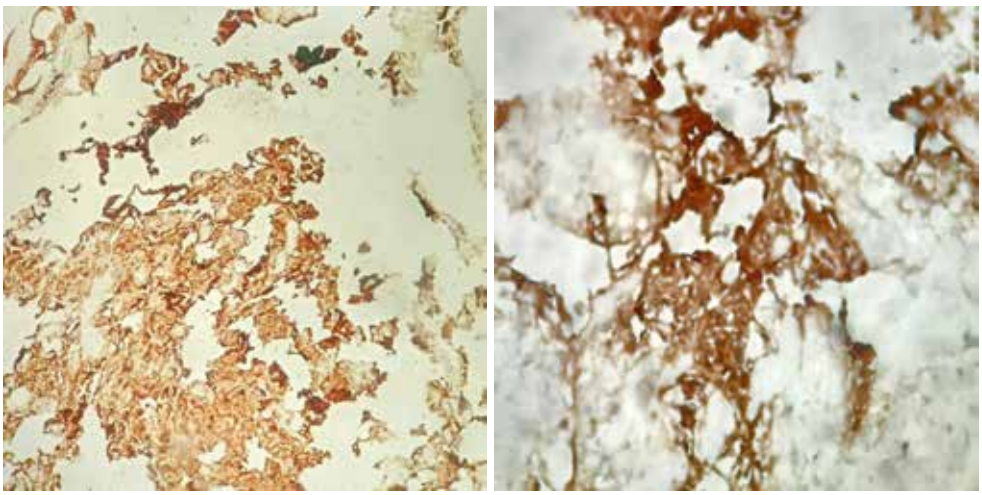


Fig.5. Careful dissection of the lateral portion of the tumour from the parietal pleura (white arrow)



A B

Fig. 6A. Tumour composed of plump spindle cells arranged in interlacing fascicles. (H&E, 100X), marked with asterisks
Fig. 6B. Tumour cells obscured by melanin pigment (H&E, 100X)



A B

Fig. 7A. Tumour cells positive for HMB45 (IHC, 400X)
Fig. 7B. Tumour cells positive for S100 (IHC 400X)

Discussion

Malignant melanotic nerve sheath tumour (MMNST) occurs due to melanogenesis of Schwann cells [7]. The most common syndromic association is Carney complex in 50% of cases [5]. Carney complex includes autosomal dominant disorder comprising of myxomas such as cardiac, cutaneous and mammary, spotty pigmentation and endocrine overactivity like Cushing's syndrome and acromegaly [8]. Our case had none of the above features in preoperative clinical and radiological evaluation. Extramedullary spinal melanotic schwannomas typically present in the 30-40 years age group [9]. However, in our case the age of the patient was 54 years old. According to the literature, about 50% of MMNST cases has local recurrence or distant metastasis at the time of detection [9]. Metastatic workup of our case did not reveal any metastasis. MRI findings includes T1 hyperintensity and T2 hypointensity in a dumbell shaped tumour arising from nerve root [10, 11] which was consistent with our case. However, signal intensity may vary depending upon the concentration of melanin [12]. Patients undergoing surgery for MMNST should be subjected to rigorous follow-up. The differential diagnosis of MMNST includes leptomeningeal melanocytoma, ancient schwannoma, pigmented neurofibroma, biphasic sarcoma, neurilemmoma and melanoma [5].

Conclusion

Malignant melanotic nerve sheath tumours (MMNSTs) are extremely rare and highly aggressive neoplasms that are often misdiagnosed in neuroimaging. Tumours arising from nerve root having characteristic of T1 hyperintensity and T2 hypointensity, MMNST should always be included in differential diagnosis. Metastatic

workup along with clinical and radiological evaluation should be done to rule out other associated syndromes. Surgical excision followed by vigilant follow-up for early detection of recurrence is recommended.

Disclosure

Conflict of Interest

The authors declare no conflict of interest.

Informed consent

Informed consent for data disclosure was obtained from the patient.

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