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Surgical Neurology International

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Case Report

Spinal malignant melanotic nerve sheath tumor with atypical magnetic resonance imaging findings: A case report

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Received: 09 June 2024
Accepted: 28 June 2024
Published: 19 July 2024

DOI
10.25259/SNI_447_2024

Quick Response Code:

ABSTRACT

Background: Malignant melanotic nerve sheath tumors (MMNSTs) are relatively rare, comprising <1% of all neoplastic peripheral nerve lesions. Here, we describe a 79-year-old male who presented with atypical magnetic resonance imaging (MRI) findings of an MMNST.

Case Description: A 79-year-old male presented with lower back pain, paraparesis, and bladder/bowel dysfunction. The MRI showed an intradural extramedullary (IE) lesion at the T9–T10 level with low-signal intensity on T1-weighted images (WI) and high intensity on T2-WI, which markedly enhanced with contrast. The IE nerve root involved with the tumor was completely removed surgically. The lesion was confirmed to be an MMNST. In the absence of metastases, adjuvant therapy was deemed unnecessary. One year later, the lesion has not recurred.

Conclusion: A 79-year-old male patient presented with a T9–T10 MR intradural lesion that was pathologically proved to be an MMNST, which was treated with gross total surgical resection (i.e., removal of the involved nerve root alone).

Keywords: Malignant melanotic nerve sheath tumor, Schwannoma, Spinal tumor

INTRODUCTION

Malignant melanotic nerve sheath tumors (MMNSTs) are rare (comprising <1% of neoplastic lesions of the peripheral nerve).[14] Due to the melanin content in MMNST, their characteristic magnetic resonance imaging (MRI) findings include high signal intensity on T1-weighted images (WI), low signal intensity on T2-WI, and marked enhancement with contrast.[15] Here, a 79-year-old male presented with an isolated MR-documented T9–T10 MMNST that was treated with gross total excision without adjunctive treatment (i.e., in the absence of metastatic disease).

CASE DESCRIPTION

A 79-year-old male presented with a 1-month history of severe back pain, paraparesis (i.e., 2/5 proximal-distal motor function), a T10 sensory level, and bladder/bowel dysfunction. The thoracic MRI revealed an intradural extramedullary cephalad/caudad 15 mm well-defined T9–T10 lesion that markedly compressed to the cord to the left side of the spinal canal; it was hypointense on T1-WI, hyperintense on T2-WI, and markedly enhanced with contrast [Figure 1].

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The accompanying non-contrast computed tomography (CT) showed no tumor calcification [Figure 1]. Surgery, performed a day following admission, included T8–T10 laminectomy. On opening the dura, an extramedullary lesion was found compressing the spinal cord from the right to the left. The lesion was elastic, soft, hemorrhagic, and slightly brownish in coloration and was densely adherent to a single nerve root; once that root was removed, the tumor was removed in Toto.

Pathological findings

The histopathological diagnosis was consistent with MMNST [Figure 2]. There were spindle-shaped cells proliferating in indistinct, short, bundle-like arrays, accompanied by moderate cell density and medium to slightly large nuclei with mild increases in chromatin but without malformed nuclei. Cell division occurred in <1 cell per 10 views, and there was no evidence of necrosis. Small scattered vessels with vitreous-like walls and no psammomatous bodies were observed. The spindle-shaped cells had a brown substance in their cytoplasm; however, the majority did not contain hemosiderin. Immunostaining of spindle-shaped cells was positive for S100, HMB45, and Melan A and negative for PgR; it was difficult to determine EMA. The MIB1 labeling index was 1–5%. Intermediate to high-intensity positive cells were approximately "30–40% for p53." The majority of the brown material was melanin rather than hemosiderin, as confirmed by staining with Fontana-Masson's solution and a negative reaction to Berlin blue staining.

Postoperative course

Postoperatively, the patient had a residual paraparesis with sphincter dysfunction. Following gross total T9/T10 tumor excision, there was no further evidence of residual and/or metastatic disease on subsequent non-contrast and enhanced MRI brain, cervical, thoracic, and lumbar MR studies. As the systemic metastatic work-up was negative, no further adjuvant therapy was warranted [Figure 3]. Notably, however, the patient continues to be closely followed for the risk of tumor recurrence [Figure 3].

DISCUSSION

MMNSTs are rare lesions, accounting for <1% of peripheral nerve tumors.[14] Since 2020, these lesions have been renamed MMNST, indicating their potential for malignancy.[14] Patients developing MMNST are, on average, 33.2 years of age (i.e., for sporadic cases) but

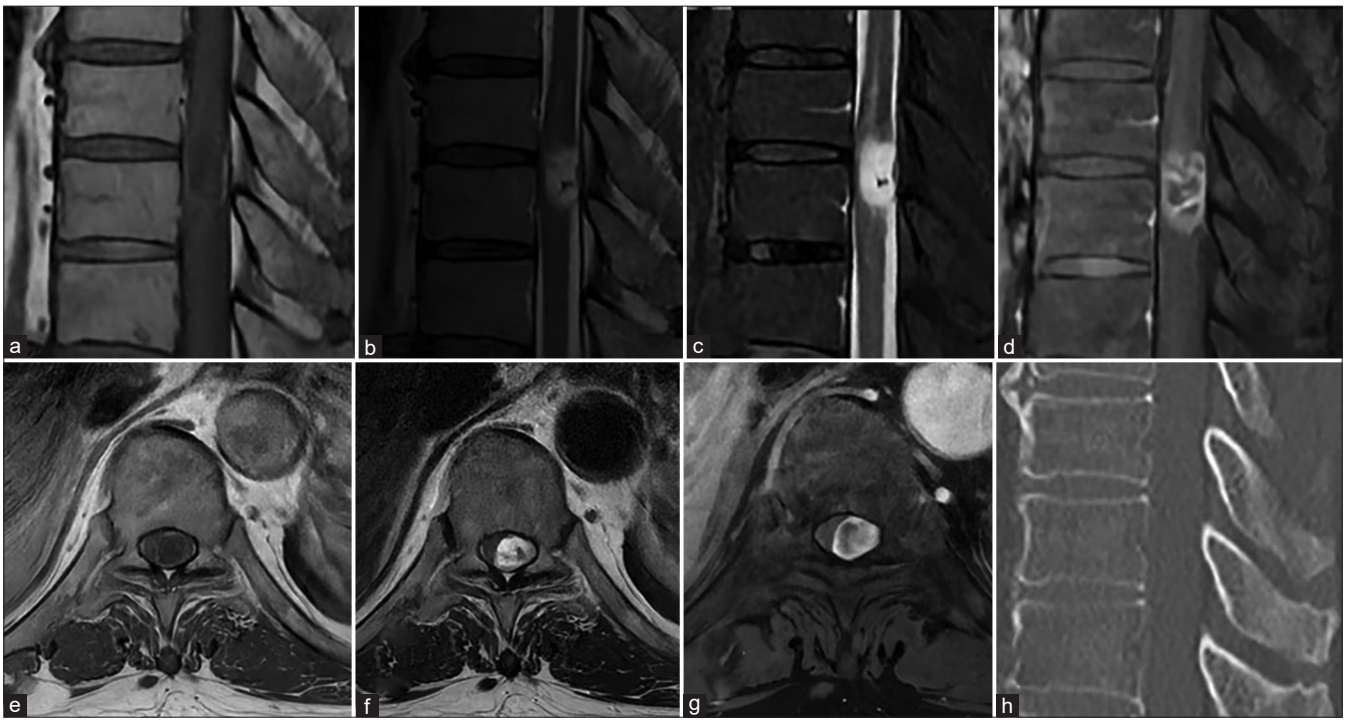

Figure 1: Thoracic spine images on admission. (a-d) Magnetic resonance imaging of sagittal section of the thoracic spine (a) T1-weighted image, (b) T2-weighted image, (c) short-tau inversion recovery [STIR] image, and (d) T1-weighted image with contrast enhancement. An intradural extramedullary mass is seen at the T9–T10 level. (e-g) MRI of the axial section of the thoracic spine (e) T1-weighted image, (f) T2-weighted image, and (g) T1-weighted image with contrast enhancement). The mass can be seen to compress the spinal cord to the right. (h) Thoracic computed tomography scan. The mass does not have any calcification.

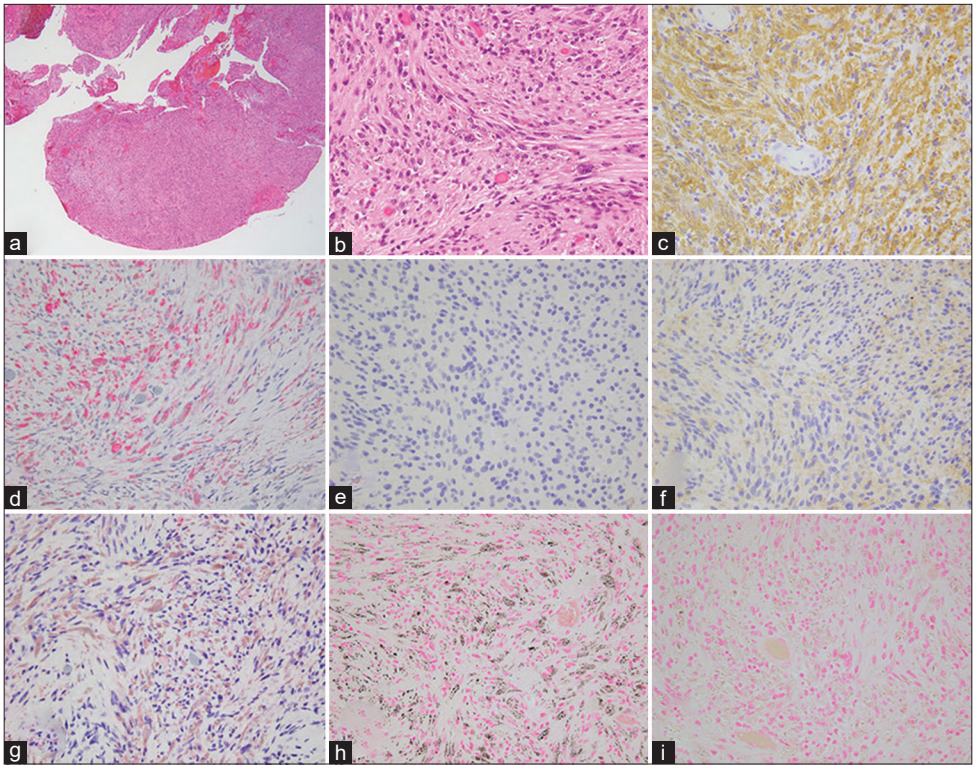

Figure 2: Pathological examinations. (a) Hematoxylin-Eosin (HE) staining weakly magnified (× 40). (b) HE staining strongly magnified (×400). c-g: Immunohistology staining (c) S100 [positive], (d) HMB45 [positive], (e) PgR [negative], (f) EMA [difficult to determine], (g) Melan A [positive], (h) Fontana Masson, and (i) Berlin blue.

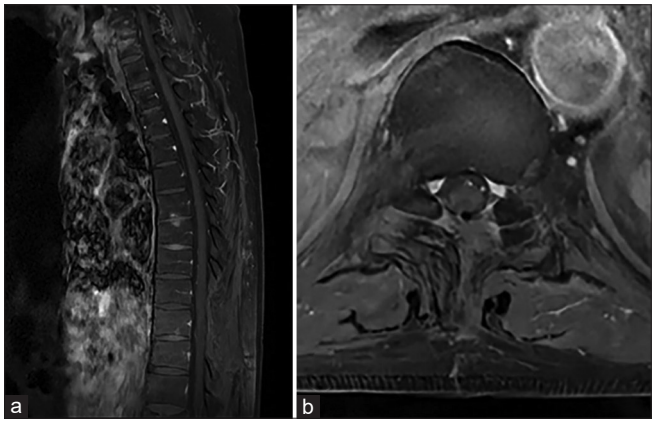

Figure 3: Contrast-enhanced thoracic magnetic resonance imaging 1 year after surgery. (a) Sagittal section and (b) axial section. No tumor recurrence can be observed.

average 22.5 years of age for cases associated with the Carney complex.[17] MMNSTs are melanogenic Schwann cells. Spinal MMNSTs occur most often in the nerve roots of the lumbosacral spine (47.2%), followed by the thoracic spine (30.5%) and cervical spine, with rare intramedullary occurrences (22.2%).[9,13] They originate from dorsal nerve roots in 30.5% of cases, and the spinal cord, sympathetic trunk, cranial nerves, peripheral nerves, and intestinal tract can also be involved.[15]

MR Findings

MRI typically shows high-signal intensity on T1-WI and low-signal intensity on T2-WI due to the free radical stabilization by melanin and a marked contrast effect. Solomou *et al.* found that 64.7% of MMNSTs had a T1 high signal, and 70.6% had a T2 low signal.[15] In the present case, the patient's MR showed a low T1 signal and a high T2 signal, which are atypical findings for MMNST. Solomou *et al.* reported similar atypical signal changes seen in 11.7% of patients (i.e., two of 17 cases); nevertheless, the two patients with MRI findings similar to ours did not develop tumor recurrence [Table 1].[15]

Treatment with gross total excision for isolated lesions without adjunctive treatment

Treatment often involves gross total resection or irradiation after subtotal resection.[8] Notably, to date, there are no established adjuvant therapies (i.e., including radiotherapy and chemotherapy) recommended for MMNST despite the high risk for recurrence (i.e., local tumor recurrence (42%), distant metastasis (27%), and mortality (26%), and/

Table 1: Magnetic resonance imaging findings and metastases in reported malignant melanotic nerve sheath tumors cases.

	Level	Sex	Age	Metastasis	T1-WI	T2-WI
Er *et al.*[6]	C1	M	54	No	High	Low
Faria *et al.*[7]	C4–5	F	32	Yes	High	Low
Kuchelmeister *et al.*[11]	C5–6	F	53	No	Iso	High
Yokota *et al.*[19]	C7	M	64	Yes	High	Low
Bendszus *et al.*[2]	T1	F	17	No	High	Low
Chen and Gu[4]	T2–T4	M	47	No	High	Low
Tawk *et al.*[16]	T7	M	25	Yes	High	Low
Mahesh *et al.*[12]	T8–T12	M	67	No	High	Low
De Cerchio *et al.*[5]	T9–T10	M	53	No	High	Low
Bosman *et al.*[3]	L1–2	M	43	No	Low	High
Keskin *et al.*[9]	L1–2	M	22	No	Low	High
Khoo *et al.*[10]	L3	F	46	Yes	High	High
Aprile *et al.*[1]	L4–5	M	60	–	High	Low
Khoo *et al.*[10]	L5–S1	F	36	Yes	High	Mixed
Welling *et al.*[18]	N/A	M	–	–	Mixed	Low
Welling *et al.*[18]	N/A	M	–	–	Mixed	Low
Welling *et al.*[18]	N/A	M	–	–	Mixed	Low
Present Case	T9–T10	M	79	No	Low	High

N/A: Not available, T1-W: T1-weighted images, T2-WI: T2-weighted images

or metastasis).[8] Ghaith *et al.* reported that the mean time to recurrence in eight cases was 3.5 years, with some recurring after >5 years.[8]

CONCLUSION

We successfully treated a 79-year-old male who presented with paraparesis and loss of sphincter function attributed to an isolated atypical MR-documented T9–T10 MMNST with a decompressive laminectomy alone.

Ethical approval

The Institutional Review Board approval is not required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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How to cite this article: Kageyama H. Spinal malignant melanotic nerve sheath tumor with atypical magnetic resonance imaging findings: A case report. Surg Neurol Int. 2024;15:250. doi: 10.25259/SNI_447_2024

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