



Case Report

Acute onset of paraplegia due to melanocytic cervicodorsal lesion

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ABSTRACT

Background: Melanocytomas are rare pigmented tumors of the central nervous system (CNS). They are more frequent in females in their fifties and are typically benign, but locally aggressive lesions that very rarely are found in the spine. They can occur in all the age groups.

Case Description: A 62-year-old male presented with acute paraplegia attributed to a C7-T1 intradural extramedullary melanocytoma. Urgent surgery, consisting of subtotal removal of the lesion, resulted in subtotal recovery of motor function with full resolution of his sensory deficit. Histologically, the lesion proved to be a melanocytoma.

Conclusion: Rarely, patients with intradural extramedullary melanocytomas of the spine may present with acute paralytic deficits warranting emergent surgical intervention.

Keywords: Melanocytoma, Paraplegia, Primary spinal tumor, Spinal cord

INTRODUCTION

Melanocytomas are rare pigmented tumors originating from leptomeningeal melanocytes. They are benign lesions that infrequently occur in the spine, where they may cause acute radiculopathy and/or myelopathy. In this case, a 62-year-old male acutely presented with paraplegia attributed to a C7-T1 intradural extramedullary anterior melanocytoma.

CASE PRESENTATION

Clinical and surgical history

A 62-years-old male, with a 2 months history of interscapular pain, presented with acute T2-level paraplegia. The MRI showed an intradural extramedullary well-defined mass at the C7-T1 level located anterior to the spinal cord. The T2-weighted image demonstrated an isointense lesion with a wide dural attachment while the enhanced study demonstrated heterogeneous enhancement with compression and posterior displacement of the cord [Figure 1].

An emergent decompressive C7-T1 laminectomy was performed and achieved subtotal tumor removal. Intraoperatively, the cervicodorsal dura was blackened, and the lesion was firm.

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Histopathology

The histopathologic examination revealed large cells, oval to polygonal in shape with prominent nucleoli, and eosinophilic cytoplasm filled with melanin pigment. There was no significant increase in mitosis (1/10 HPF and low proliferation index Ki 67: 2%). On immunohistochemistry tumor cells showed positivity with S100, Vimentin and HBM45 and negativity for EMA (which is an indicator of meningioma), thus confirming the diagnosis of melanocytoma [Figure 2].

Postoperative course

The 1 day post-operative MRI revealed removal of the 90% of the lesion and no residual compression of the cervicothoracic cord at the same level. After the 1st postoperative week, he demonstrated subtotal recovery of motor (4/5 BMRC grade) and total sensory recovery of function. He was discharged in a rehabilitation unit. Three months later, the patient's neurological status was improved (5/5 BMRC) and he is being referred for radiation therapy.

DISCUSSION

The World Health Organization's classification classifies primary melanocytic lesions into four categories; meningeal



Figure 1: Sagittal T2-weighted image showing an isointense extramedullary mass with dural attachment.

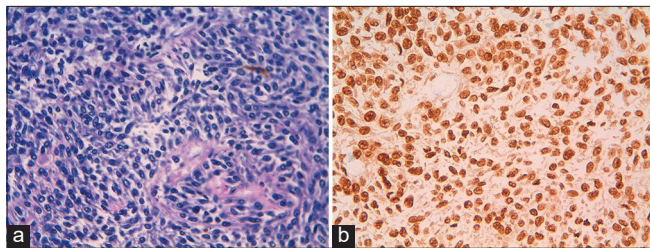


Figure 2: (a) Spindle-shaped cells with focal melanin pigmentation (b) immunohistochemical positivity for S100.

melanocytosis, meningeal melanocytoma, meningeal melanoma, and meningeal melanomatosis.^[3,6] Melanocytoma commonly presents in posterior fossa and foramen magnum, but only rarely in the spine.^[2] Due to the benign nature of these tumors, and their typical slow growth, spinal symptoms/signs have a slow onset, and reflect the location of the lesion; acute paraplegia is very rare.^[4] We treated this lesion as an acute spinal cord compression to grant the total neurological recovery of the patient, giving priority to clinical status with respect to complete removal of the tumor; although these are benign tumors, for gross total, subtotal or partial excision, the risk of recurrence is sufficiently high to warrant adjuvant radiation therapy.^[1,5]

CONCLUSION

In the presence of spinal cord compression due to intradural extramedullary lesion with acute neurological onset, the surgical decompression and the removal of the lesion, despite subtotal, are necessary to achieve a good neurological outcome.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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