

## Intramedullary Melanocytoma of the Cervicothoracic Cord: Case Report and Review of Literature

### Abstract

Melanocytoma is rare pigmented tumor of the leptomeninges which arise from the neural crest. Intramedullary location of the tumor is extremely rare, and only a few case reports are available in the literature. We report a case of 35-year-old female with the entity who had a near total removal of the intramedullary tumor with good postoperative outcome. The available literature is reviewed.

**Keywords:** Cervicothoracic tumor, intramedullary tumor, melanocytoma

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### Introduction

Melanocytoma is a rare pigmented neoplasm of the leptomeninges first described in 1972 by Limas and Tio.<sup>[1]</sup> Melanocytoma is derived from scattered melanocytes that are present in the leptomeninges. They usually have an intracranial localization but also involve the spinal column, where they are most often detected as an intradural extramedullary lesion.<sup>[2-4]</sup> Intramedullary lesion of the spine is an extremely rare entity, and after searching PubMed database and Google search, we were able to find only 24 cases having this entity [Table 1]. We report a case of 35-year-old female with this entity and the available literature is reviewed.

### Case Report

A 35-year-old female presented with complaints of pain over lower cervical and upper thoracic region and progressive spastic paraparesis for the last 1½ years. On neurological examination, there was spastic paraparesis with power of grade 3/5 at bilateral hips and 1/5 at B/L ankle joints. There was no bladder or bowel involvement.

On magnetic resonance imaging (MRI), there was an intramedullary mass present from C7 to D6 level. It was heterogeneously hyperintense on T1-weighted image and hypointense on T2-weighted image. There were syrinx formation and edema of cord both cranial and caudal to the lesion. MRI

contrast images showed heterogeneous enhancement of the lesion [Figure 1].

A C6 to D7 laminectomy was performed. Dura opened in the midline and tented laterally. Arachnoid opened. A standard midline myelotomy was performed through the posterior median septum. Intraoperatively, a dark black intramedullary mass was found having intratumoral tangle of vessels. Part of the tumor was demarcated from surrounding spinal cord tissue, but the tumor was infiltrating anteriorly to the spinal cord tissue at some places. A near total excision was performed. Histopathological examination [Figure 2] revealed heavy brown pigment in the cell cytoplasm which was consistent with melanocytoma. Postoperatively, patient's condition gradually improved. There was improvement in the power of lower limbs. After 6 months, the power in bilateral lower limb was of grade 4/5, and she was able to stand and walk without support.

Six months follow-up MRI contrast showed postoperative changes from C6 to D7 level with a small area of minimal contrast uptake suggestive of near total excision [Figure 3].

Table 1 shows the comparison of previous case reports with the present case.<sup>[5]</sup>

### Discussion

Primary melanocytic tumors of the spine are part of a spectrum of rare neoplasm derived from scattered melanocytes located in the leptomeninges. The World Health Organization has classified melanocytic

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**Table 1: The comparison of previous case reports with present case**

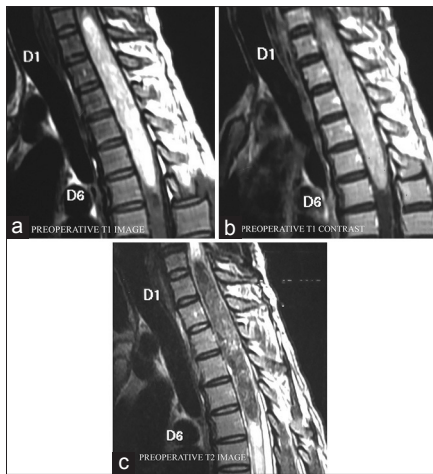
Author, year	Age	Sex	Location	Resection	Follow-up	Recurrence	Radio-therapy
Barth <i>et al.</i> 1993	49	Female	T10-T12	Subtotal	48	Yes	No
Glick <i>et al.</i> 1997	69	Male	C1-C2	Total	60	No	No
	39	Female	T8-T9	Subtotal	12	No	Yes
	27	Female	T1-T6	Total	24	No	No
	56	Female	T12	Total	48	No	No
	74	Female	T11-T12	Total	12	No	No
	70	Male	C1	Total	Nil	-	-
	24	Female	T12 to L1	Total	24	No	No
Delhay <i>et al.</i> 2001	38	Female	T6-T9	Subtotal	48	Yes	No
Iida <i>et al.</i> 2002	42	Male	T10	Not described	4	No	no
Turhan <i>et al.</i> 2004	19	Female	T8	Total	36	No	No
Van Paesschen <i>et al.</i> 2004	51	Male	C1-C2	Total	Nil	-	-
Horn <i>et al.</i> 2008	37	Female	C1-C3	Total	38	Yes	No
	37	Female	T9-T10	Total	16	Yes	No
	48	Male	T12	Total	185	Yes	No
Chacko <i>et al.</i> 2008	22	Male	T6-T11	Total	96	No	No
Karikari <i>et al.</i> 2009	32	Female	T10	Total	3	No	No
	20	Male	T12	Total	2	No	No
Caruso <i>et al.</i> 2009	62	Male	T11-T12	Total	24	No	No
Perrini <i>et al.</i> 2009	79	Female	T10-T11	Subtotal	30	Yes	No
Eskandari <i>et al.</i> 2010	45	Male	T11	Subtotal	36	Yes	Yes
Muthappan <i>et al.</i> 2012	61	Female	C3-C4	Total	36	No	No
Kahilogullari 2012	28	Female	Thoracic	Total	Nil	-	-
Wagner 2015	63	Male	C2-C3	Total	18	Yes	Yes
Present case	35	Female	C6-T6	Near total	6	No	No

lesions of the central nervous system (CNS) into<sup>[1]</sup> diffuse melanocytosis and melanomatosis,<sup>[2]</sup> melanocytoma, and<sup>[3]</sup> malignant melanoma. The melanocytic lesions of CNS have been classified as low, intermediate, and high grades by Brat *et al.*<sup>[2]</sup> These melanocytes are most frequently encountered at the posterior fossa and upper part of the cervical spinal cord.<sup>[1,6]</sup>

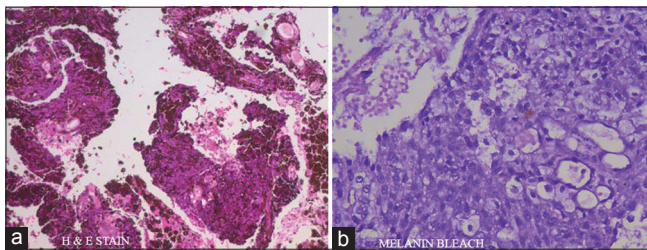
These tumors were first described as meningeal melanocytoma by Limas and Tio in 1972.<sup>[1]</sup> Melanocytoma is commonly solitary, low-grade neoplasms that do not invade surrounding structures. In the spine, they usually present as intradural extramedullary lesions.<sup>[2-4]</sup> In cases

with intraparenchymal localization, the melanocytes most probably originate from the Virchow–Robin spaces.<sup>[5,6]</sup> Intramedullary melanocytoma is extremely rare and very little data exist to guide the clinicians about the management of the patients with this entity.

MRI with gadolinium is the preferred imaging modality for the diagnosis of melanocytoma. Intramedullary melanocytoma lacks characteristic imaging features. Typical MRI findings are that of an intramedullary lesion iso- to hyper-intense on T1-weighted sequences, hypointense on T2-weighted sequences, and with homogeneous enhancement.<sup>[5,7]</sup> In our case, mass was hyperintense on



**Figure 1:** (a) Preoperative T1-weighted magnetic resonance imaging, (b) preoperative magnetic resonance imaging T1 with gadolinium, (c) preoperative magnetic resonance imaging T2-weighted image



**Figure 2:** (a) H and E stain, (b) melanin bleach stain



**Figure 3:** (a) Postoperative magnetic resonance imaging T1-weighted image, (b) Postoperative magnetic resonance imaging T1 with gadolinium, (c) postoperative magnetic resonance imaging T2-weighted image

T1-weighted images and hypointense on T2-weighted images, and it showed heterogeneous enhancement. These signal features are biased by variable degrees of tumor melanization that affects the signal characteristics on MRI.<sup>[5]</sup> Diagnosis on the basis of MRI findings is still challenging as individual appearances may vary widely.

Enough clinical evidence exists, based on multiple small case reports and series, to show that gross total resection

is the initial treatment of choice for the management of these tumors; however, the question remains about the best possible follow-up method for postsurgical patients and how residual tumor or recurrences should be managed. Local control rates have been shown to be four times higher if complete resection is achieved and morbidity and mortality rates for incomplete resection now mandate the use of adjuvant radiation therapy.<sup>[3,8-12]</sup> Although they are considered to be a benign tumor, melanocytomas have been reported to recur even after gross total resection.<sup>[11]</sup> Rades and Schild<sup>[10,11]</sup> have proposed high-dose local radiation, even in cases of complete tumor resection, to prevent local recurrence. More aggressive approaches have also been advocated including re-resection of recurring tumor and reservation of adjuvant radiotherapy for those lesions that cannot be completely resected.<sup>[8]</sup>

Close postoperative monitoring and diligent follow-up, even in the case of gross total resection, is extremely important. Although these tumors are histopathologically benign, they do have an aggressive clinical course secondary to their location and the mass effect they impose within the spinal cord. It is therefore crucial that recurrences should be caught early as this allows for early adjuvant therapy or re-resection. Hopefully, more reports and longer follow-up periods after surgical resection, recurrence, and subsequent treatment will enable clinicians to render more accurate information as to the best treatment for melanocytomas.

## Conclusion

Melanocytoma is an extremely rare entity. The lack of distinctive imaging characteristics increases the challenge during preoperative diagnosis. Complete surgical excision should be advocated as optimal treatment. The question remains whether more aggressive early adjuvant therapy should play a role in treating patients with these benign, yet locally aggressive intramedullary tumors.

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## Conflicts of interest

There are no conflicts of interest.

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