

# Intramedullary melanotic schwannoma of the cervical spine: A case report and literature review

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Received September 15, 2017; Accepted December 6, 2017

DOI: 10.3892/mco.2018.1584

**Abstract.** Intramedullary melanotic schwannomas (IMS) are rare lesions, with only 8 cases reported in the literature to date. We herein describe the case of a 40-year-old male patient with an IMS of the cervical cord, which, to the best of our knowledge, is the ninth reported case of IMS, and review the relevant literature. The patient presented with numbness of the left arm that gradually worsened over a period of 4 months. Magnetic resonance imaging (MRI) of the cervical spine with gadolinium enhancement revealed an intramedullary lesion at the level of C1-C2. The mass was T1 hyperintense, T2 hypointense and homogeneously enhanced. During surgery, an intramedullary dark gray lesion was identified and was partially removed. The left arm numbness partially subsided 2 weeks after surgery. Although rare, IMS is associated with characteristic MRI findings (T1 hyperintensity, T2 hypointensity and homogeneous enhancement) and MRI is the preferred method for evaluating lesions of the spinal cord. Correct diagnosis is crucial for management planning; therefore, immunohistochemical examination is required. In addition, IMS cases must be closely followed up, particularly when the mass cannot be completely resected.

## Introduction

Schwannomas are benign, slowly growing neoplasms composed of neoplastic Schwann cells (1). Melanocytic schwannoma (MS) is a rare schwannoma variant composed of melanin-producing cells with the ultrastructural characteristics of Schwann cells (2), accounting for ~1% of primary peripheral nerve sheath tumors (1,2). MS is primarily considered to be a benign tumor, with a relatively rarely reported propensity to metastasize (2,3); however, recent published literature suggests that MS must be reconsidered as a malignant neoplasm (4) with a greater potential to metastasize. The most common location of the tumor is in the nerve roots (5-7). MS is also encountered

in extramedullary sites and the peripheral nervous system, but is particularly rare in intramedullary sites (8,9). There are only 8 reported cases of intramedullary MS (IMS) (1,8-14); we herein report the ninth IMS case in a 40-year-old man with a lesion located in the cervical cord, which was diagnosed based on the magnetic resonance imaging (MRI) and histopathological findings and treated with surgery.

## Case report

A 40-year-old man presented in the Second Affiliated Hospital of Zhejiang University School of Medicine in March 2017 with left arm numbness that gradually worsened over a period of 4 months. The patient did not have any other neurological symptoms. Upon physical examination, no obvious deposition of pigment was found in the skin and mucosa. There was no previous history of a surgical procedure for the removal of MS. Upon neurological examination, the distal pinprick sensation in the left upper limb was slightly decreased. The muscle strength in all upper and lower limbs was unaffected (5/5) and the muscle tone was normal. All limb tendon reflexes were normal. No positive pathological reflexes were present bilaterally.

An MRI of the cervical spine revealed an intramedullary mass, sized 1.5x1.0 cm, within the spinal canal. The mass occupied 60% of the spinal canal at the level of C1-C2, with displacement of the spinal cord to the right side (Fig. 1A-D). The mass was T1 hyperintense (Fig. 1A) and T2 hypointense (Fig. 1B). Following enhancement, the mass was homogeneously enhanced (Fig. 1C and D). There was spinal cord expansion and edema above and below the mass, as evidenced by T2 hyperintensity (Fig. 1B). The radiological and clinical findings were consistent with melanoma, including primarily intramedullary melanoma or metastatic malignant melanoma. As primary tumors were not found anywhere on the skin of the limbs or the trunk, metastatic malignant melanoma was not considered in the initial diagnosis. A cavernous malformation with subacute hematoma was considered in the differential diagnosis; however, the history did not include a sudden onset, and the patient's symptoms gradually worsened over a relatively long period of time, which is not consistent with haemorrhagic manifestations. Finally, the preoperative diagnosis was a primarily intramedullary melanoma.

A C1 laminectomy was performed. The dura mater and arachnoid sheath were opened longitudinally and dark

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**Key words:** melanoma, schwannoma, diagnosis, spinal

Table I. Summary of the intramedullary melanotic schwannoma cases reported in the literature.

Case no.	Authors	Sex	Age, years	Location	Treatment	Outcome	Follow-up	(Refs.)
1	Solomon <i>et al</i>	Male	69	Caudal medulla and C3	Gross total removal	-	-	(11)
2	Marchese <i>et al</i>	Female	72	C4 -C6	Partial removal	Functional recovery	-	(12)
3	Sola-Pérez <i>et al</i>	Female	63	C7-T1	Needle aspiration	-	-	(13)
4	Acciarri <i>et al</i>	Female	44	T2 -T3	Gross total removal	Partial neurological recovery	-	(14)
5	Santaguida <i>et al</i>	Male	35	C4-C5	Gross total removal	Partial neurological recovery	Recurrence at 2 years, radiotherapy, and repeatresection at 4 years	(1)
6	Mouchaty <i>et al</i>	Female	56	Conus	Gross total removal	Partial neurological recovery	No recurrence at 12 months	(9)
7	Hoover <i>et al</i>	Female	62	T11	Gross total removal	Good neurological recovery	No recurrence at 10 months	(8)
8	Mohamed <i>et al</i>	Male	43	T9-T10	Gross total removal	Good neurological recovery	No recurrence at 12 months	(10)
9	Present case	Male	40	C1-C2	Partial removal	Partial recovery	-	

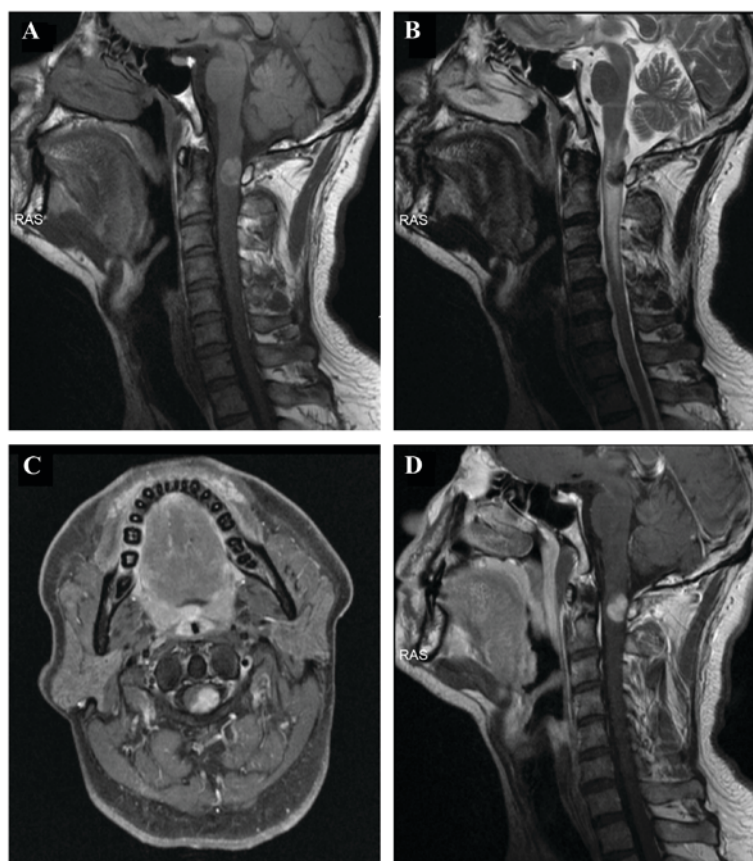


Figure 1. The mass was pre-contrast (A) T1 hyperintense and (B) T2 hypointense. (C and D) The mass lesion, which appeared to be intramedullary at the level of C1-C2, exhibited homogenous enhancement.

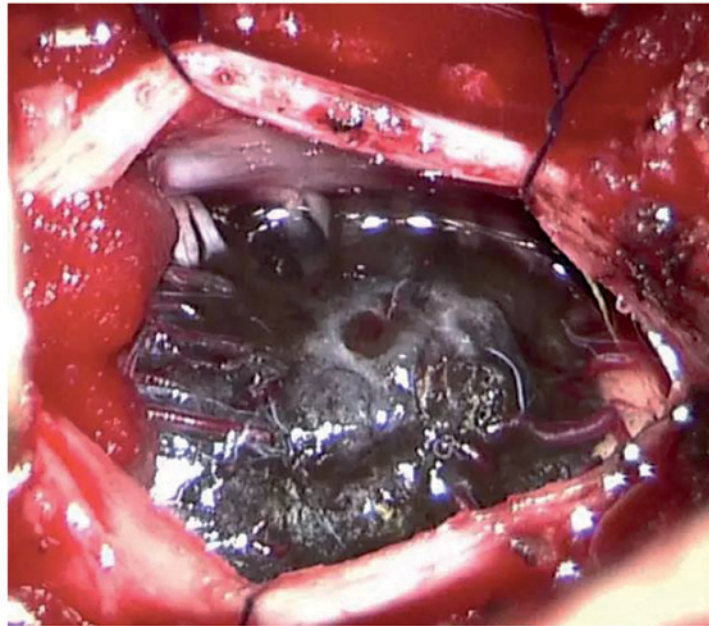


Figure 2. Intraoperative photograph showing an intramedullary lesion following opening of the dura mater.

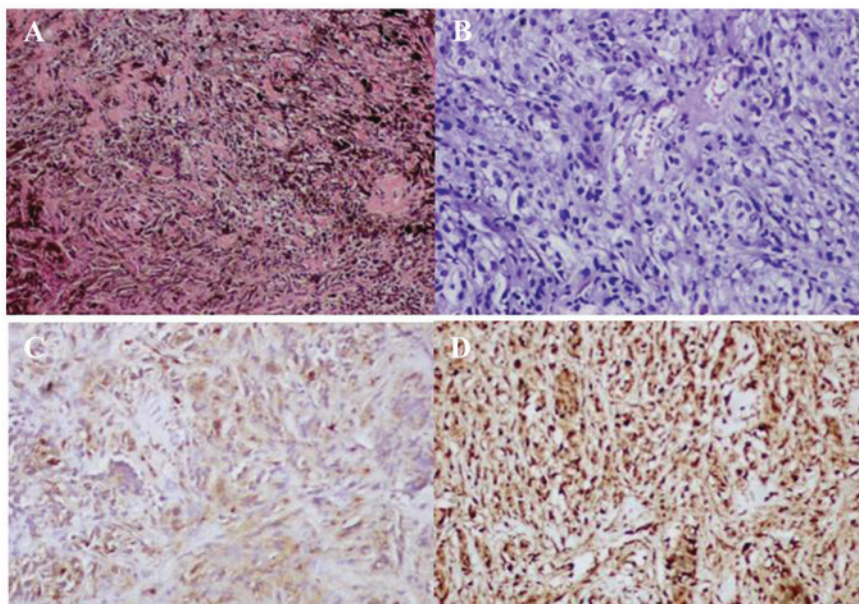


Figure 3. (A-D) Light microscopy findings of melanotic schwannoma. (A) The tumor cells had abundant cytoplasm containing melanin granules and spindle-shaped cell nuclei; hematoxylin and eosin staining; magnification, x40. (B) Following depigmentation, no obvious atypia of the nuclei was observed; magnification, x10. Immunocytochemical examination revealed that the tumor cells were (C) human melanoma black 45-positive and (D) S-100-positive; magnification, x10.

pigmentation was visible through the dura (Fig. 2). The dura was opened in the midline to expose an intramedullary tumor, with neurophysiological monitoring during surgery. The tumor invaded deeply into the spinal cord and had an unclear boundary. Spinal nerve roots were also invaded by the tumor. Due to the difficulty of complete removal and an intraoperative diagnosis of metastatic malignant melanoma by frozen section biopsy, the lesion was partially resected. The left arm numbness partially subsided 2 weeks later after the surgery.

Sections stained with hematoxylin-eosin revealed that the tumor was composed of polygonal epithelioid and spindle-shaped cells, with abundant cytoplasm containing

melanin granules (Fig. 3A). Depigmented sections revealed no obvious atypia of the tumor cell nuclei (Fig. 3B). Immunocytochemistry for human melanoma black 45 (Fig. 3C), p53, vimentin and S-100 (Fig. 3D) was positive, whereas the tumor cells were negative for melan-A and epithelial membrane antigen. The Ki-67 proliferative index was <1%. The pathological diagnosis was IMS. During the follow-up on May 2017, the patient status was stable with the left arm numbness.

Written informed consent was obtained from the patient regarding the publication of the case details and associated images.



## Discussion

MS is a tumor derived from progenitor neural crest cells that can differentiate into both Schwann cells and melanocytes, which is characterized by deposition of melanin in the Schwann cell cytoplasm (2). Theories for the production of melanin by these cells include neoplastic differentiation of neural crest cells into Schwann cells with melanogenetic properties, and melanocytic transformation of previously normal Schwann cells (1). The presence of psammoma bodies is typical of the psammomatous variant of MS. Approximately 50% of psammomatous MS are part of the Carney syndrome (along with myxomas, skin pigmentation and endocrine tumors or overactivity) (15). The most frequent sites of MS are the dorsal spinal nerve roots, sympathetic chain, acoustic nerve, cerebellum and orbit (10). IMS is particularly rare and, to the best of our knowledge, this is the ninth case reported to date (1,8-14) (Table I).

MRI is currently the optimal diagnostic modality for evaluating lesions of the spinal cord. Melanin may be associated with shortened T1 and T2 relaxation times due to its content of paramagnetic free radicals; thus, melanotic lesions appear hyperintense on T1-weighted and hypointense on T2-weighted images (16). By contrast, non-melanotic tumors are hypointense on T1-weighted and hyperintense on T2-weighted images, which helps differentiate melanotic from non-melanotic lesions. However, subacute hematoma is difficult to differentiate from melanotic lesions, as subacute hematoma may display similar characteristics on MRI (17). In the present case, a cavernous malformation with subacute hematoma was also initially considered on T1- and T2-weighted images. However, considering the patient's medical history, the onset of the symptoms was not sudden but rather a chronic process; thus, subacute hematoma was not considered in the initial preoperative diagnosis. Following administration of gadolinium, the tumor typically exhibited homogeneous enhancement; however, enhancement may be heterogeneous in the presence of haemorrhage within the lesion (10). A diagnosis of intramedullary schwannoma may be confidently made when there is continuity of the intramedullary lesion with a contrast-enhanced thickened spinal root (18); however, no obvious enhancing thickened spinal root involvement was observed in the present IMS case.

From the 8 previously reported cases (Table I), the main treatment for IMS is gross total removal. Approximately 10% of MS cases are reported to exhibit an aggressive clinical course, with local recurrence and metastasis (1,16). Due to the rarity of IMS, the role of radiotherapy has not been established. The fifth documented case (Table I) of a patient with IMS recurred 2 years after initial gross total removal. Subsequently, the patient received radiotherapy, but the tumor progressed 2 years after radiotherapy and the patient was again treated with surgery. Considering the recurrence potential of IMS, annual follow-up of patients with IMS with MRI is required (8). In the present case, the lesion was partially resected, due to the difficulty of complete removal, and an intraoperative diagnosis of metastatic malignant melanoma was made by frozen section biopsy. Due to the rarity of IMS, intraoperative frozen section diagnosis is challenging without immunohistochemical examination.

In conclusion, MRI is the preferred method for evaluating lesions of the spinal cord. A standard IMS would typically

be T1 hyperintense, T2 hypointense and homogeneously enhanced. However, although IMS has these characteristic MRI features, preoperative diagnosis as well as intraoperative frozen section diagnosis are challenging due to the rarity of this tumor. Correct diagnosis is crucial for management planning; therefore, immunohistochemical examination is warranted. In addition, careful follow-up is required for all IMS patients, particularly when the mass cannot be completely resected.

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