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LETTER TO THE EDITOR

# Melanotic Schwannoma: Two Cases of Rare Lesions

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**To the editor**

Melanotic schwannomas (MS) arise from sympathetic nervous system commonly from the spinal or autonomic nerves near the midline [1]. Hundreds of cases of MS have been reported in the English medical literature. However, intracranial MS and intramedullary MS were rare, with respectively 20 and 10 cases being reported to date [2, 3]. We report two cases of MS occurring in the uncommon locations. Both the uncommon locations and the melanotic component of the reported lesions make them difficult to distinguish from malignant melanomas or meningeal melanocytomas.

A 33-year-old woman, she complained of episodic headaches, with numbness in the left face, developed over half month. No abnormality was in physical examination. She preoperative magnetic resonance imaging (MRI) detected a 4.2 cm*5.2 cm*6.0 cm mass in the left middle cranial fossa invading the left temporal lobe and compressing the left brain ventricle. The tumor was homogeneously hypointense on T1-weighted images and hyperintense on T2-weighted images and showed homogeneous enhancement on contrast- enhanced T1-weighted images. Since a gross total resection was performed. Microscopically, the neoplasm was composed of uniform cells, mostly round to oval, with prominent nucleoli, smooth nuclear contours, finely clumped chromatin, and fibrillary eosinophilic cytoplasm. Scattered pigmented cells were present throughout the specimen (Fig. 2a). Psammoma bodies were not presented. Mitotic activity showed 0–1 mitotic figures/10 HPF. The tumor cells were positive for S-100, Vimentin and melanocytic markers (HMB45, Melan-A) and negative for PR and EMA. The Ki-67 index was less than 5%. Combing imaging features with histological characteristics, we finally diagnosed the patient as a intracranial MS.

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A 61-year-old, previously healthy woman experienced progressive weakness of the lower limbs and increasing pain and numbness around the left lower extremity 3 years prior to admission. Neurologic examination revealed the lower extremities weakness of 3–4 without hyperreflexia. She preoperative MRI revealed a cystic and solid mass measuring 1.6 cm*4.5 cm in spinal canal at the level of L1. The cystic component was hypointense on T1-weighted images and hyperintense on T2-weighted images. The solid component was isointense on the T1-weighted images and T2-weighted images and showed heterogeneous enhancement on contrast-enhanced T1-weighted images (Fig. 1). The patient underwent removal of the mass. The tumor was mainly composed of spindled to epithelioid cells growing in short fascicles and sheets, variably abundant melanin pigment and inconspicuous vasculature (Fig. 2b). The nuclear showed relatively monomorphous, elongated nuclei with small nucleoli. Psammoma bodies and mitotic activity were not presented. The tumor cells were strong positive for Vimentin and Melan-A and local positive for S-100, and negative for PR and EMA. The Ki-67 index was 1%. Thus, the final diagnosis of the patient was identified as intramedullary MS.

Melanotic schwannoma (MS) is a rare pathologic variant of schwannoma, of which peak incidence is a decade earlier than that of conventional schwannoma [1]. Intracranial or intramedullary MS is more rare. Only 20 intracranial MS and intramedullary MS have been reported. Their clinical findings are separately summarised in Tables 1 and 2.

According to our data and analysis, intracranial MS showed a male prevalence with a mean age of 38.6 years, while intramedullary MS most were female and the average of them was 52.3 years. We performed a Student t test and $\chi^2$ test to determine if various locations presented age or gender differences. Statistical analysis showed no age ($P = 0.061$) and gender ($P = 0.045$) differences between intracranial and intramedullary MS.

The clinical manifestation of the patients were various. Neuropsychiatric symptoms, bowel and bladder dysfunction, and sensory and motor disturbances are common [4]. The symptoms were mainly related to compression of the spinal

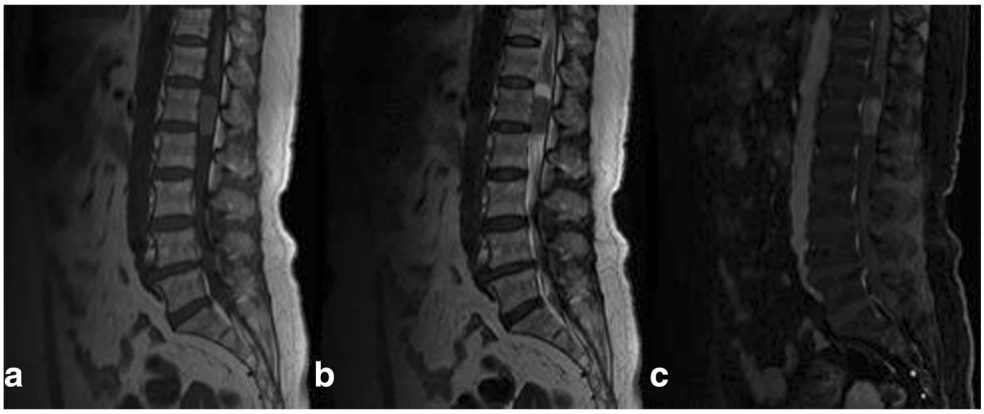


**Fig. 1** Case 2, hypointense and isointense on the T1-weighted images (**a**), hyperintense and isointense on the T2-weighted images (**b**), partial enhancement on contrast-enhanced T1–weighted images (**c**)

cord, cerebellum or cerebrum by an extra-axial mass, with focal neurological signs arising secondary to either hydrocephalus or local effects on the CNS parenchyma. These non-specific manifestation are difficult to distinguish from other space-occupying diseases of the nervous system.

The imaging findings, especially MRI, are very useful to distinguish intracranial or intramedullary MS from meningeal melanocytoma. MS usually shows a isolated mass, sometimes being partly cystic and showing peripheral edema and local compression. MRI shows homogeneously hypointense to isointense signals on T1-weighted images and isointense to hyperintense signals on T2-weighted images [3]. In contrast, meningeal melanocytoma exhibits isointensity to hyperintensity on T1-weighted images, hypointensity to isointensity on T2-weighted images, homogeneous enhancement on enhanced MRI [5].

The pathological findings play an important in distinguishing intracranial or intramedullary MS from malignant melanoma. MS is mainly composed by spindled to epithelioid cells growing in short fascicles and sheets and

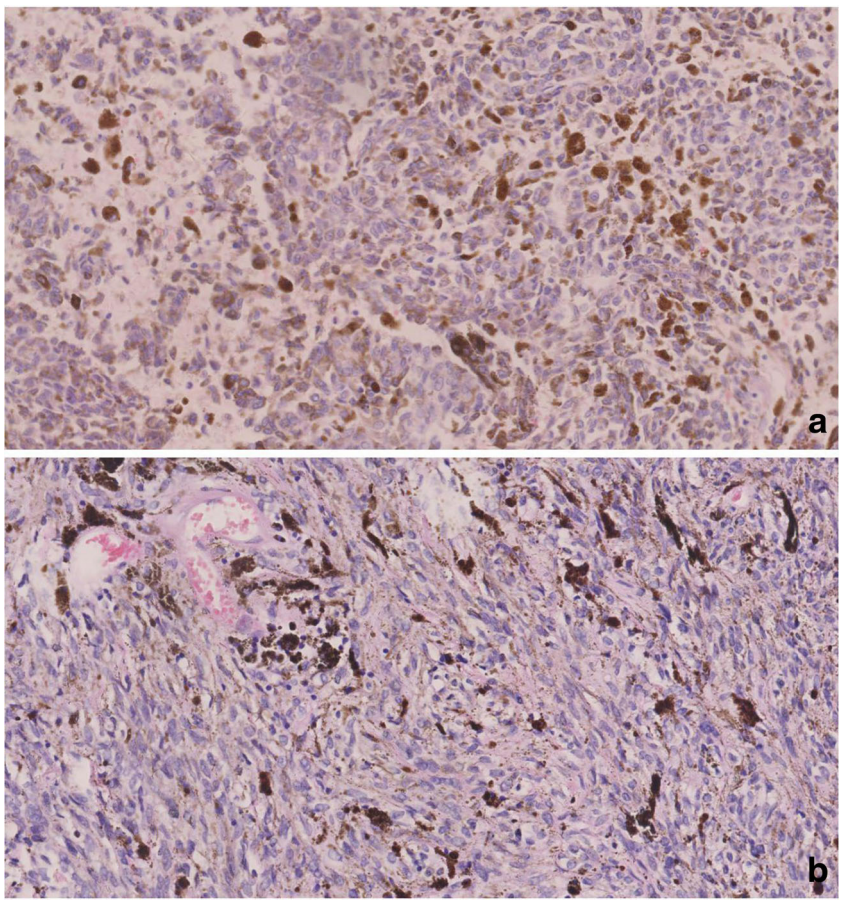


**Fig. 2** **a** Case 1, uniform cells, mostly round to oval, with prominent nucleoli, and scattered pigmented cells were present throughout the specimen(bar = 100 μm). **b** Case 2, the tumor showed epithelioid cells growing in short fascicles and sheets, abundant melanin pigment and vasculature(bar = 100 μm)

**Table 1** All intracranial MS cases available in the literature (including the present case)

| Case | Age | Sex | Location | Resection | Adjuvant treatment | Recurrence |
|---|---|---|---|---|---|---|
| 1 | 34 | M | Acoustic nerve | STR | None | Yes |
| 2 | 22 | M | Trigeminal nerve | GTR | None | No |
| 3 | 74 | M | CPA | GTR | None | No |
| 4 | 12 | M | Meckel's cave into CPA | NA | Radiotherapy | Yes |
| 5 | NA | NA | Trigeminal nerve | NA | NA | NA |
| 6 | NA | NA | Trigeminal nerve | NA | NA | NA |
| 7 | 22 | M | Orbit | GTR | None | Yes |
| 8 | 77 | M | CPA | STR | NA | NA |
| 9 | 56 | F | CPA | GTR | None | No |
| 10 | 28 | M | CN V | GTR | NA | NA |
| 11 | 11 | M | CN III | STR | Radiotherapy | No |
| 12 | 34 | F | CN V | STR | None | Yes |
| 13 | 22 | M | Orbit | GTR | Radiotherapy | Yes |
| 14 | 54 | M | Foramen magnum | GTR | None | No |
| 15 | 52 | F | CPA | STR | None | NA |
| 16 | 41 | F | Posterior fossa | STR | Radiotherapy | NA |
| 17 | 60 | M | Orbit | GTR | None | NA |
| 18 | 12 | F | Orbit | STR | None | Yes |
| 29 | 47 | F | CPA | STR | Radiotherapy | NA |
| 20 | 43 | F | Left middle cerebellar peduncle | STR | Radiotherapy | NA |
| 21 | 33 | F | Left middle cranial fossa | GTR | None | NA |

*STR*, subtotal resection; *GTR*, gross total resection; *NA*, not available; *M*, male; *F*, female; *CPA*, cerebello-pontine angle

variably abundant melanin pigment [1, 4]. The nuclear morphology varied significantly from case to case, with some cases showing relatively monomorphous, elongated nuclei with small nucleoli, and others showing striking pleomorphic, hyperchromatic nuclei with prominent macronucleoli [4]. Unlike malignant melanoma which usually has frequent mitosis, MS has rare mitosis. Melanin pigment was present in all cases but showed similar variability, with some cases containing only small amounts of intracytoplasmic melanin and other cases showing massive melanin deposition, at times obscuring cellular detail [4]. Sometime, psammoma bodies were presented. Sometime, psammoma bodies, calcification, or mature fat areas can be presented in tumor and the structures are more helpful for the diagnosis of MS [1]. Immunostaining analyses are important, but the differentiation between MS and malignant melanoma can be particularly challenging because both can be inmunoreactive for HMB-45 and S-100 protein antibodies. Unlike malignant melanoma, the

**Table 2** All intramedullary MS cases available in the literature (including the present case)

| Case | Age | Sex | Location | Resection | Adjuvant treatment | Recurrence |
|---|---|---|---|---|---|---|
| 1 | 69 | M | Caudal medulla-C3 | GTR | None | No |
| 2 | 72 | F | C4–C6 | GTR | None | No |
| 3 | 63 | F | C4–C6 | NA | NA | NA |
| 4 | 44 | F | T2–T3 | GTR | None | No |
| 5 | 35 | M | C4–C6 | GTR | Radiotherapy,Chemotherapy | Yes |
| 6 | 56 | F | T12–L1 | GTR | None | No |
| 7 | 62 | F | T11 | GTR | None | No |
| 8 | 23 | F | T4–T5 | GTR | None | No |
| 9 | 43 | M | T9–T10 | GTR | None | No |
| 10 | 47 | M | T4–T5 | STR | None | Yes |
| 11 | 61 | F | L1 | GTR | None | NA |

*STR*, subtotal resection; *GTR*, gross total resection; *NA*, not available; *M*, male; *F*, female

Ki-67 index is very low and usually less than 5% [4]. In a word, histologic features of ample cytoplasm, cytoplasmic process, and indiscernable cell border as well as low proliferative index contribute to the diagnosis of MS rather than malignant melanoma.

While there are no guidelines, review of the literature suggests that radiotherapy should be pursued especially when there is subtotal resection [2, 3]. Given the recurrence rate and the metastatic potential, radiation has been strongly advocated by some authors especially if they display any features of malignancy.

To conclusion, combing imaging features with histological characteristics is a key to the final diagnosis. In view of its unpredictable behavior, especially in the absence of overt malignant features, long terms follow-up with or without radiotherapy is recommended.

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**Author Contributions** Xiao-li Li designed the study, managed the data analyses and drafted the article. Shun-dong Dai evaluated of immunohistochemical stains, gave histopathological diagnosis and revised the article.

## Compliance with Ethical Standards

**Conflict of Interest** The authors declare that there is no conflict of interest.

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