

## Case Report

# Melanotic schwannoma of thoracic spinal root mimics metastatic melanoma: a potential pitfall for misdiagnosis

Baizhou Li<sup>1</sup>, Qin Chen<sup>2</sup>

<sup>1</sup>Department of Pathology, Second Affiliated Hospital of Zhejiang University, Medical school, Hangzhou 310009, China; <sup>2</sup>Department of Pathology, Women's Hospital, Zhejiang University, School of Medicine, Hangzhou 310006, China

Received May 7, 2015; Accepted June 23, 2015; Epub July 1, 2015; Published July 15, 2015

**Abstract:** Melanotic Schwannoma (MS) is a kind of rare subtypes of Schwannoma. In this tumor, amounts of melanin always mislead inexperienced pathologists to a diagnosis of primary or metastatic melanoma. Different from the ordinary Schwannoma, MS is considered as a low malignant nerve sheave tumor. Here we present a case of MS arising from the thoracic spinal and initially was misdiagnosed as metastatic melanoma. But the patient followed a benign course, without recurrent in 30 months follow-up.

**Keywords:** Melanotic tumor, melanoma, Schwannoma, diagnosis, differentiation

### Introduction

Melanotic Schwannoma (MS) is rare tumors that usually involve spinal nerve roots. Up to now, fewer than 200 cases MS have been reported in literatures. Different from the conventional Schwannoma, MS shows unique morphological and immune-phenotypic features. The most characteristic of MS is more or less melanin granules found in tumor cells. This feature raises a diagnostic problem whether a nerve sheath tumor or a melanoma, a primary or metastatic tumor? Here we present a case of MS arising from the 7th thoracic spinal with radiological, pathological and clinical follow-up information, for better understanding and manage this rare tumor.

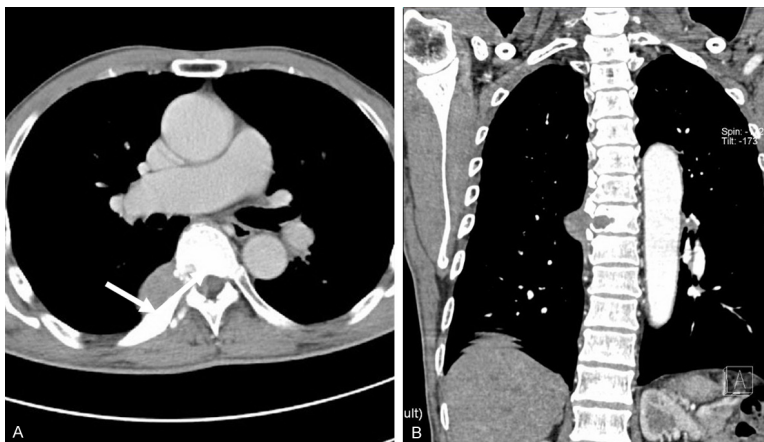
### Case report

#### *Clinical information*

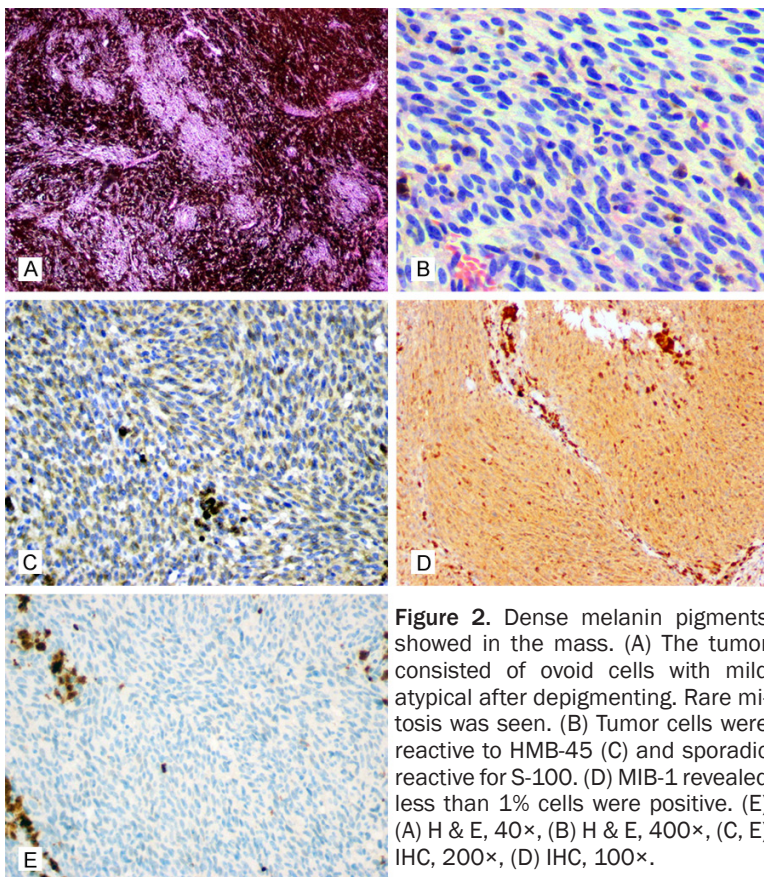
A 62 years old healthy male was found a tumor at 7th thoracic spine in a routine physical examination. He did not have any complaint, except for an erosive destruction in the 7th thoracic spinal body in CT scanning. Enhancing scanning showed a well circumscribed dumb-bell shape tumor from intraspinal to paravertebral

soft tissue, with obvious enhancing effect. The neighboring ribs were intact (**Figure 1A, 1B**). The imaging impression is a neurogenic tumor and metastatic tumor for further excluded (**Figure 1A, 1B**). And the surgical excision was executed. During the operation a black mass closely adhesive to the dura infiltrated into the soft tissue though T8 centrum. The tumor size is 5×3×2 cm. The frozen pathologic diagnosis was favor of metastatic melanoma. Under the gross examination, a pile of pieced black tissue with soft texture and sized 5×3×2 cm was seen. All tissue was embedded for microscopic review. Histological examination revealed a dense spindle cells lesion with amounts of melanin in the cytoplasm and covered nucleus. The uniform spindle cells were arranged in fascicular pattern with inconspicuous nucleoli, tumor cells were with medium chromatin, without necrosis and hardly found mitosis (**Figure 2A, 2B**). Immunostaining showed spindle cells were reactive for HMB45, focal and scattered stain for S-100 (**Figure 2C, 2D**). The tumor cells were negative for MBP, GFAP, CK, EMA and Melan-A. MIB-1 for proliferative index was less than 1% (**Figure 2E**). Based on its mild morphology and low MIB-1 index, as well as immunophenotypic features, this case is the most consistent of a melanotic cellular Schwannoma.

## Melanotic schwannoma of thoracic spinal root



**Figure 1.** CT scanning for thoracic spinal. (A) dumb-bell shape mass across the intervertebral foramina, T7 vertebral body showed destructive change. (A) A well circumscribed dumb-bell shape mass, and the density is similar to the soft tissue (B).



**Figure 2.** Dense melanin pigments showed in the mass. (A) The tumor consisted of ovoid cells with mild atypical after depigmenting. Rare mitosis was seen. (B) Tumor cells were reactive to HMB-45 (C) and sporadic reactive for S-100. (D) MIB-1 revealed less than 1% cells were positive. (E) (A) H & E, 40 $\times$ , (B) H & E, 400 $\times$ , (C, E) IHC, 200 $\times$ , (D) IHC, 100 $\times$ .

### Prognosis and follow-up

The patient followed a quite benign course. 30 months follow-up did not show a proof of recurrent and metastasis.

### Discussion

Melanotic Schwannoma is a rare tumor, some of which is part of Carney Syndrome (companion with kollonema, the skin pigmentation and endocrine tumors or hormone secretion abnormality) and psammoma bodies are also figure out [1], some cases are sporadic. In this case there was no history of neurofibromatosis, derma nevus or melanoma, and without the evidence of other tumors or endocrine disturbance. However, differential diagnosis such as melanoma, or melanotic cellularity schwannoma or melanotic MPNST really needs account [2, 3].

Melanoma in vertebral canal is pretty rare, no matter primarily or metastatic. There is no special history of suspicious nevus or melanoma in this patient, and the morphology showed mild short spindle or ovoid cells hyperplasia without pleomorphism and bizarre giant cells, rare mitosis and no necrosis in all sections. Immunostaining showed S-100 sporadic positive, and HMB45 displayed the moderate positive, while melan-A was negative. Though the immunophenotype is consistent with the diagnosis of melanoma, the morphologic feature doesn't support this diagnosis, and the extremely low index of Ki-67 don't account for the melanoma yet.

The histological and immunostaining characteristics of melanotic schwannoma are different from the classical schwannoma, the former always be absent of Antoni A and Antoni B areas, GFAP often is negative too. But in this

case, the low grade melanotic MPNST needs to be excluded from melanotic schwannoma. Mention to a cellular nerve sheath tumor, sometimes, there doesn't have a clear threshold between two kinds of tumors. But since several documents proved melanotic schwannoma always followed an aggressive clinical course [6, 7], it's no need to clarify this is a melanotic schwannoma or a low grade MPNST.

The literature reported that melanotic schwannoma derived from the spinal nerve root had the dumb-bell appearance [4, 5], which was related with slow growth and low aggressiveness. Moreover, melanotic disorders have characteristics in MRI, which shows high density in T1 weight and low density in T2 weight. Hence, the differential diagnosis from the non-melanotic disorders is easy, which show the opposite MRI image. The image of this case has the appearance of dumb-bell, in accord with the neurogenic tumor. Even so, for the limited experience, we did not give the accurate diagnosis before the operation.

The prognosis of melanotic schwannoma is poor compared with the ordinary Schwannoma [6, 7]. In Zhang's opinion that the recurrence and metastasis rate of benign melanotic schwannoma was 18.2% and 9.1% respectively [8]. As a result, radical excision is the recommended therapy. And for this case, the patient showed a benign course, half year interval routine examination for 30 months did not show proof for recurrent or metastasis.

### Disclosure of conflict of interest

None.

**Address correspondence to:** Dr. Qin Chen, Department of Pathology, Women's Hospital, School of Medicine, Zhejiang University, Hangzhou 31006, Zhejiang, China. Tel: +86-57189713372; E-mail: 64945977@qq.com

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