Case report

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Melanotic schwannoma of the L5 root

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Abstract

Melanotic neoplasm of the central nervous system is rare and the majority of them are metastatic. Melanotic schwannoma (MS) is an unusual variant of nerve sheath neoplasm accounting for less than 1% of primary nerve sheath tumors. A case involving a 36-year-old man with MS at the L5 root is presented. Surgery, differential diagnosis, radiology, histology, and treatment of this rare entity are discussed.

Keywords

Intradural extramedullary spinal cord neoplasm, magnetic resonance imaging, melanotic schwannoma, neuroectodermal tumor

Introduction

Melanotic neoplasms of the central nervous system (CNS) are rare and the majority of them are metastatic.[1,2] Melanotic schwannoma (MS) is an unusual variant of nerve sheath neoplasm accounting for less than 1% of primary nerve sheath tumors.[3] It is composed of cells having the ultrastructure and immunophenotype of Schwann cells but containing melanosomes.[4] MS was described in 1932 by Miller as a distinct entity from a lesion of the thoracic sympathetic ganglion.[5] Since its description as an intraosseous lesion by Hodson in 1961,[5] approximately 100 MS cases have been reported in the relevant literature.[1,6] Psammomatous and sporadic forms of MS are described.[5] Psammomatous types are seen as a component in the Carney complex. This autosomal dominant multiple neoplasia syndrome is characterized by myxomas (cardiac, mammary, and cutaneous), mucocutaneous lentigines and blue nevi, and functional endocrine tumors (Cushing syndrome, precocious puberty, and acromegaly).[7] Psammomatous variants are usually seen in younger patients compared with sporadic ones, with a peak prevalence of the fourth decade.[1] The majority of these neoplasms behave benignly, but approximately 10% of MSs may undergo malignant degeneration,[8] and 15% of reported patients with MS have died of as a result of the tumor.[5]

Case report

A 36-year-old man presented with history of low back pain for approximately 3 months which was sudden in onset and severe in nature. The pain was radiating the right leg and affecting his daily life. His complaint worsened day by day, and after 2.5 months he developed right leg weakness and numbness. On neurological examination, there was right extensor hallucis longus paresis with 3/5 muscle strength. Right hemihypoesthesia at the right L5 and S1 dermatomes and slight hyperreflexia in Achilles were revealed. He had right paraspinal spasm. There were no neurocutaneous signs. Lumbar magnetic resonance imaging (MRI) revealed a round and well-defined, extraspinal $20 \times 20 \times 15\,mm^3$ mass lesion at the right L5-S1 neural foramen. The lesion was slightly hyperintense on T1 and T2 images. On sagittal spin echo sequences, remodeling was seen on the posterolateral area of the vertebral body. The lesion filled the right lateral recess on axial sequences (Figure 1). The mass lesion was at the neural foramen with these MRI characteristics, suggestive of a spinal nerve sheath tumor.

A right hemipartial laminectomy and flavectomy with inferior facetectomy exposed a $2 \times 2\,cm^2$ shining-black mass within a thin capsule arising from the right L5 radix at the right L3-S1 neural foramen with extraspinal extension. The bone around the lesion was destructed and the dura at its vicinity was very hard to

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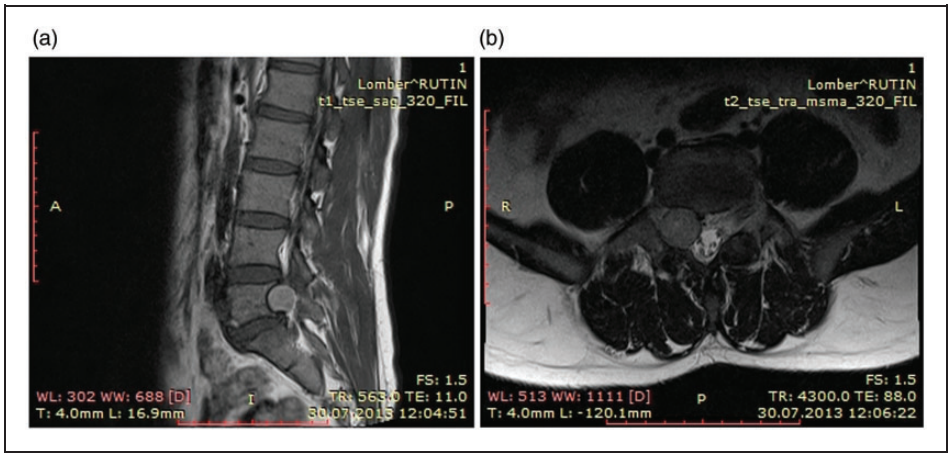

Figure 1. A hyperintense well-defined mass lesion and remodeling is seen at the L5 vertebral level (a); an axial sequence shows well-defined hyperintense mass lesion (b).

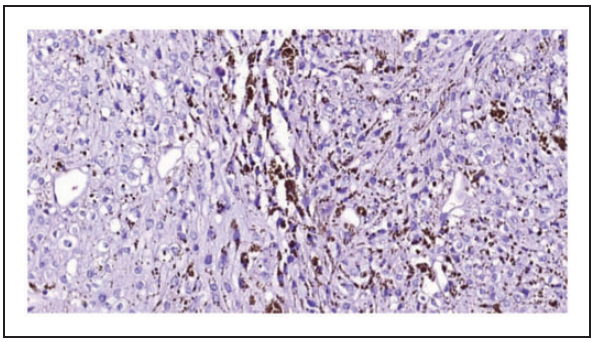

Figure 2. The histological examination revealed fusiform and epithelioid cells with highly pigmented cytoplasm.

separate from. The lesion was almost totally removed by a microneurosurgical technique under the operating microscope (Zeiss, OpMi Pentero 800, Germany). Histopathological examination revealed solid sheets of pigmented spindled and epithelioid cells composing the tumor. Tumor cells with dispersed cytoplasmic deposition of melanin associated with a psammomatous calcification and showing diffuse cytoplasmic and nuclear S100 immunopositivity were strongly suggestive of MS (Figure 2).

Marked improvement was observed postoperatively in pain radiating to the right leg. Paresis also improved to 4+/5 immediately after operation. After an uneventful postoperative period patient was discharged. A control MRI revealed no residual lesion in the tumor location after 6 months (Figure 3).

Discussion

Primary pigmented CNS lesions are rare. MS is a variant of schwannoma characterized by spindle and epithelioid cells which contain heavily pigmented granules.[9] MS arises from spinal nerve roots and sometimes from soft tissues. The majority of such lesions are benign. Only 10% of these tumors undergo malignant degeneration.[3] MS presents with about a 1/1 male to female ratio. Age at diagnosis is between 10 and 84 with the peak prevalence in the fourth decade in the literature.[1] MSs present in patients of younger ages than those with non-pigmented schwannomas.[10] Approximately half of these lesions are a component of Carney's syndrome.[6] It was demonstrated that MS recurred after resection in 18.2% of cases and metastasis was seen in 9.1%.[11] Radiotherapy (RT) and chemotherapy (ChT) are advocated after resection by some authors in the literature, particularly for those in whom incomplete resection of the tumor was performed.[8,12] But the presented case in this report showed regression in the residual mass after six months, and the residual lesion was also doubtful; the oncology department did not approve RT or ChT for this patient.

This rare entity with its features should be differentiated from classical schwannomas, pigmented neurofibromas, pigmented peripheral nerve sheath tumors, and primary or secondary malignant neoplasms. Correct diagnosis is crucial for planning a management. Microscopic total resection should be achieved to prevent local recurrences and probable metastasis.

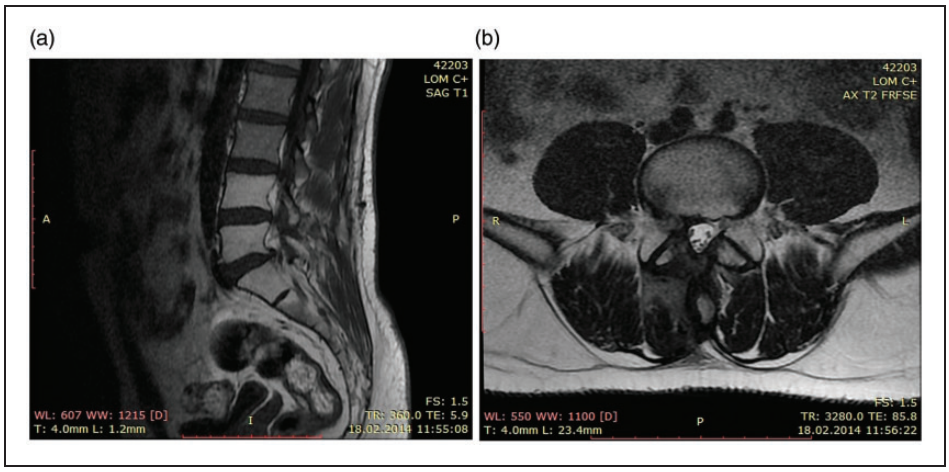

Figure 3. Sagittal (a) and axial (b) postoperative sequences show no residual mass.

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Conflict of interest

The authors declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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