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Case Report

Melanotic Schwannoma of Spine: Illustration of Two Cases with Diverse Clinical Presentation and Outcome

Abstract

Melanotic schwannomas (MS) are rare variants of schwannomas the occurrence of which is described in case reports only. They usually arise from posterior spinal nerve roots and less commonly from other cells of neural crest origin. Although they are relatively benign tumors in young, aggressive behavior is reported. They occur as isolated tumors or as part of a syndrome named Carney complex. We try to describe the pathology, diagnosis, management, and prognosis of MSs in two different cases: one cervical intramedullary with no recurrence on 5-year follow-up and the other one extramedullary in lumbar region with early recurrence and aggressive course. A brief review of literature is done.

Keywords: *HMB-45, melanotic, schwannoma, spine*

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Introduction

Schwannomas are benign neoplasms of schwann cells. Melanotic schwannomas (MS) are rare variants and their occurrence is described in case reports only. Although commonly arise from posterior spinal nerve roots, they can occur in other locations, such as the sympathetic chain, acoustic nerve, cerebellum, orbit, choroid, soft tissues, heart, oral cavity, esophageal wall, stomach, bronchus, retroperitoneum, uterine cervix, and parotid gland. They arise sporadically or as part of Carney complex in association with other lesions. Usually, presents with features of compression of spinal cord or exiting nerve roots. Magnetic resonance imaging (MRI) is the diagnostic modality of choice. Although these are benign tumors usually, aggressive behavior and malignancy have been reported. We report two cases - one in lumbar region; extramedullary lesion with recurrence and aggressive course and another one cervical intramedullary lesion with no recurrence following excision in 5-year follow-up.

Case Reports

Case one

A 35-year-old male was referred to us with the diagnosis of recurrent spinal tumor. He was diagnosed to have spinal intradural extramedullary tumor about 10 months back and underwent L2–L3 laminectomy and excision of the tumor at another hospital after which he improved neurologically but again presented with severe back ache and foot drop. MRI was repeated which showed recurrence of the lesion with extension to extradural tissues and paraspinal muscles. Initial histopathology report was MS.

We did exploration and during surgery, there was blackish deposit in intramuscular plane and extradural tissues. Tumor was extending intradurally. All the nerve roots were adherent to the tumor and there was blackish tumor deposits on the roots and arachnoid. Near total excision of tumor and arachnoid leaving the small deposits on the nerves was done.

Histopathology showed ovoid to spindle cells containing brownish pigment positive for HMB-45, S-100 and vimentin, with prominent nucleoli suggestive of MS [Figures 1-5].

Case two

A 25-year-old male was evaluated in another hospital for nonspecific neck pain, MRI scan was showing fusiform enlargement of the cervical cord by an intramedullary lesion at C2 vertebral body level, which was hyperintense on T1 and intense contrast enhancement. He underwent C2 laminectomy and excision of the lesion. There was a blackish

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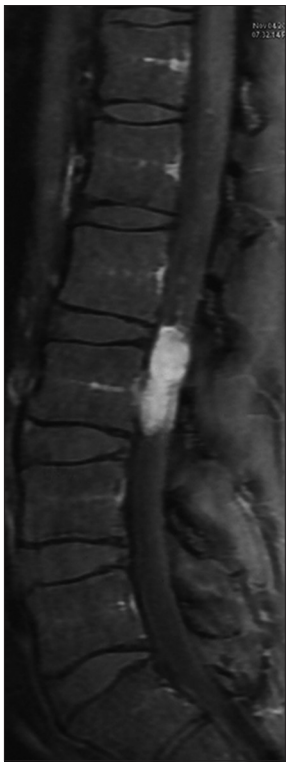

Figure 1: Ti sagittal with intense contrast enhancement

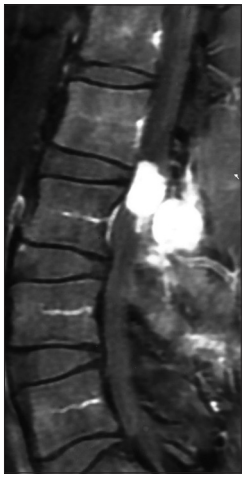

Figure 3: T1 sagittal contrast showing recurrence

pigmented lesion on the surface of the cord going deep intramedullary. Histopathology showed cells with cytoplasm rich in melanin positive for S100 and HMB-45, nuclear pleomorphism and prominent nucleoli suggestive of melanotic schwannoma. He is on follow-up for the last 5 years, and there is no evidence of recurrence on follow-up imaging [Figures 6-9].

Discussion

MS is a rare circumscribed nerve sheath tumor of melanin producing schwann cells[1] the tumor was originally described by Millar in 1932 as a "malignant melanotic

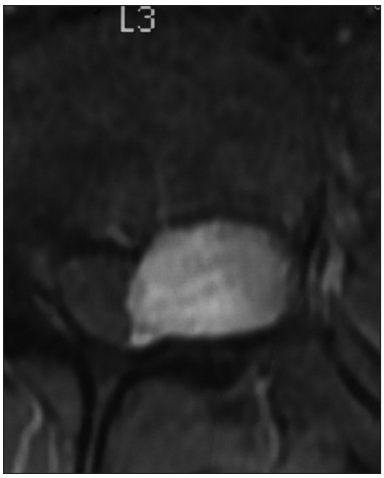

Figure 2: T_1 Axial with contrast enhancement

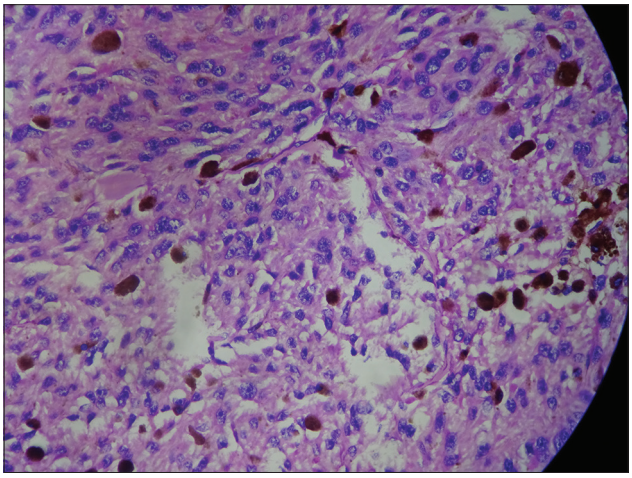

Figure 4: Intracellular melanin deposits (H and E)

tumor of sympathetic ganglion cells."[1,2] Hodson in 1961 suggested that it was a form of schwannoma.[3] Because of its tendency to involve somatic and autonomic nerves as well as sympathetic ganglia, uniform composition of schwann cells and frequent association with neurological symptoms, the lesion is classifiable as peripheral nerve sheath tumor.[1] Histogenesis of melanin in schwann cell rests in the fact that the neural crest cells migrate and differentiate into divergent tissues such as melanocytes, schwann cells, neurons of peripheral nervous system, adrenal medulla, and calcitonin-producing c-cells of thyroid.

MS is a rare variant of schwannoma arising most commonly in posterior spinal nerve roots.[4-6] They can occur in other locations such as the sympathetic chain, acoustic nerve, cerebellum, orbit, choroid, soft tissues, heart, oral cavity, wall of esophagus and stomach, bronchus, uterine cervix, retroperitoneum, and parotid.[5-8]

MS occurs in patients 10–84 years but peak incidence is seen in the fourth decade with slight female predominance −1.4–1.0.[1]

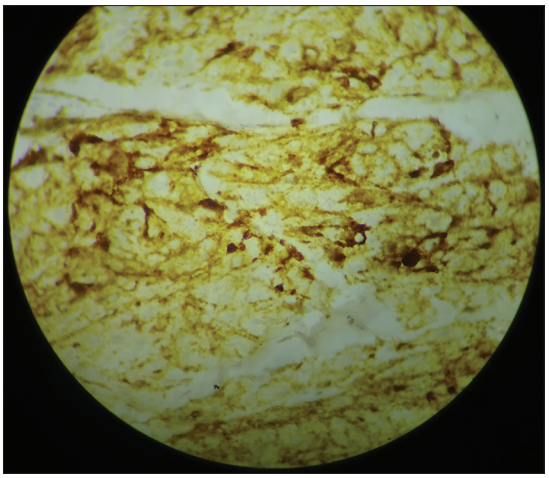

Figure 5: Immunohistochemistry positive for HMB-45

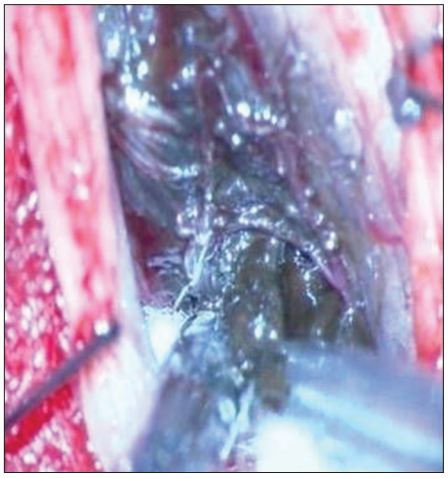

Figure 7: Intraoperative image showing blackish tumor inside the cord

Dorsal location represents 30.5% of the spinal MS. Clinical presentation is related to the involvement of a nerve (pain or sensory abnormality) or due to mass effect as in extramedullary spinal tumors.[5,9] Bone erosion may be seen particularly in spinal nerve root tumors.[1]

Psammomatous MS (PMS) is a distinct clinical variant associated with Carney complex. About 55% of patients with PMS have Carney complex.[10] It is an autosomal dominant multiple endocrine and lentiginosis syndrome featuring tumors such as PMS, pigmentation of skin, lips, and external genitalia (65%), myxoma of heart (65%), skin and breast, endocrine hyperactivity including ACTH-dependent Cushing's syndrome caused by primary pigmented nodular adrenocortical disease, sexual precocity, and congenital osteo-chondromyxoma.[10]

As in all spinal cord pathologies, MRI is the investigation of choice. Typically, the lesions are hyperintense on T1 and hypointense on T2 due to paramagnetic free radicals in melanin. Non-MSs are hypointense on T1 and hyperintense on T2.[11] Both enhance on contrast, and it helps to differentiate tumor from spinal cord edema. Chronic

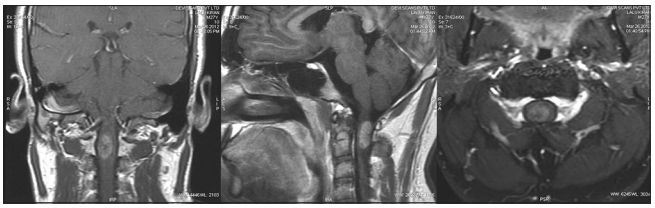

Figure 6: Contrast enhancing intramedullary lesion

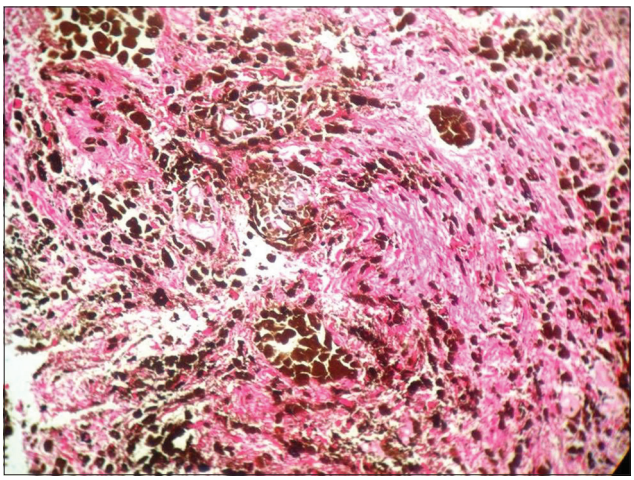

Figure 8: Intracellular melanin (H and E)

hematomas are hyperintense on T1 and T2 while subbacute hematomas are difficult to distinguish.[8,11] Computed tomography scan may show bone scalloping or destruction and foraminal widening.

The possible etiologies of intramedullary melanotic schwannomas include: (1) Occasionally intramedullary spinal nerve fibers can occur, displacing Schwann cells centrally during embryogenesis. (2) Perivascular bundles of peripheral nerves are a normal occurrence and have the potential to displace Schwann cells intramedullary. (3) Neoplastic extension of Schwann cells through the insertion site of the dorsal root. (4) Neoplastic differentiation of neuroectodermal cells in the pia to form intramedullary MS. (5) Disordered migration of neural crest cells during neural tube closure.[11]

Grossly, these tumors are circumscribed and are enveloped by a thin fibrous membrane which may in part be infiltrated by tumor infiltration. Often lobulated, these are soft, firm or rubbery with black, brown, or gray cut section; sometimes with areas of hemorrhage or necrosis.[10] Bone destruction if present is associated with malignancy.[1]

Microscopy shows cellular lesion composed of spindle-shaped and epithelioid cells arranged in lobules or fascicles, cells containing melanin pigments. Clusters of spindle-shaped cells with long interdigitating cytoplasmic process. Melanosomes in all stages of maturation, most often Stage 2–4 are found within the cytoplasm of these tumor cells.[1]

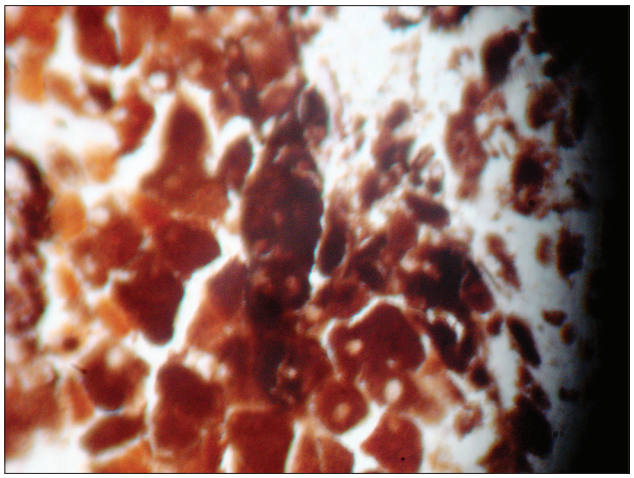

Figure 9: Positive for HMB-45

Immunohistochemistry shows that both benign and malignant melanotic schwannomas are immunoreactive for vimentin, S-100, and HMB-45. Melan-A, melanoma cell adhesion molecule and microphthalmic transcription factor reactivity is also seen.[1,12] Negative for glial fibrillary acidic protein (GFAP) but one case of +ve GFAP is reported.[13]

Differential diagnosis includes conventional schwannoma, pigmented lesions such as pigmented neurofibroma, meningeal melanocytoma, metastatic melanoma, and clear cell sarcoma. Conventional schwannoma lacks melanin pigment and psammoma bodies. The distinction of MSs from pigmented neurofibroma is difficult.[14] They show microscopic pigmentation only, lack psammoma bodies and fat, scanty cytoplasm and nonuniform immune staining for S-100.[1] MSs and melanocytomas may be found to represent a lesion continuum.[12] Melanocytomas arise in the cranial and spinal leptomeninges. Of great clinical importance is differentiating MS from metastatic melanoma. Paraspinal site is rare for metastatic melanoma, rarely totally black, are obviously cytologically malignant and lack psammoma bodies and fat.[1] Carney describes that the dendritic appearance of cells in MS is seldom seen in metastatic melanoma.[10]

All available literatures suggest complete surgical excision and careful follow-up whenever possible.[15] Periodic follow-up is needed because of high risk of recurrence, malignant transformation, and metastasis. Local recurrence and seeding of the subarachnoid space may be seen.[6,15] Zhang *et al*. demonstrate that the chance of recurrence following resection is 18.2% and metastasis in 9.1% of cases.[8] Hence, radiotherapy is recommended following subtotal excision.[11]

More studies are necessary to understand the natural history, prognosis, and best management strategies.

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Conflicts of interest

There are no conflicts of interest.

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