Primary Melanocytoma of the Lower Thoracic Spinal Cord: Case report and review of the literature

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Abstract

Background: Meningeal melanocytoma is a rare benign tumor, most frequently located in the posterior fossa and spinal canal. Our objective is to illustrate a case of this tumor that originated in the thoracolumbar area of the spine and had an uneventful clinical course after total resection.

Case description: We present the case of a 59 years old woman who presented with a medical history of ongoing neurological deterioration due to spastic paresis of the lower extremities. MRI of the thoracolumbar region identified a melanocytic melanoma as the underlying cause.

Conclusions: Melanocytic tumors of the central nervous system have a typical appearance on MRI scans, varying with the content and distribution of melanin. However, the differential diagnosis between malignant melanoma and melanocytoma still depends on pathological criteria. Spinal meningeal melanocytoma has a benign course, and it is amenable for gross total resection. The outcome is favorable following complete resection.

Abbreviations

WHO: World Health Organization; CNS: Central Nervous System; MRI: Magnetic Resonance Imaging; HPF: High-power field; MIB1: methylation-inhibited binding protein 1

Introduction

Primary melanocytic neoplasms of the meninges (meningeal melanocytomas) are uncommon neoplasms of the central nervous system [1]. Contrary to metastases from malignant skin melanoma, primary melanocytic lesions are very rare, with an incidence of 1 per 10 million for melanocytomas and 0.5 cases per 10 million for primary malignant melanomas [2]. This tumor entity was first described as meningeal melanocytoma by Limas and Tio in 1972 [3]. Melanocytomas are commonly solitary, low-grade neoplasms that do not invade surrounding structures. They are usually characterized by a benign clinical course, but local recurrence may occur [4].

Meningeal melanocytoma is an extremely rare pigmented tumor of the CNS deriving from leptomeningeal melanocytes, with the majority located in the posterior fossa and spinal canal [5,6]. Spinal meningeal melanocytoma is the histologically benign variant of a continuous melanocytic tumor spectrum in which primary malignant melanoma represents its malignant counterpart, their aggressiveness being entirely different [7]. It is generally considered as a slow-growing benign tumor, corresponding histologically to WHO grade 1 [8].

The differential diagnosis of melanocytomas includes primary or metastatic malignant melanoma [9], melanotic schwannoma [10], melanotic meningioma [11], and melanoblastosis [12].

Case Presentation

We report the case of a 59 years old woman, who presented with a mild to moderate degree, gradually progressive spastic weakness of the lower extremities. She also suffered from an ongoing incomplete cauda equine syndrome. The clinical course was of a few weeks duration.

A MRI of the thoracic and lumbar spine was performed, which revealed an intradural-extradural space – occupying lesion that was extending from the T11 to L1 spinal level.

MRI revealed a slightly hyperintense mass lesion, more readily apparent on sagittal images, which appear to enhance slightly after gadolinium contrast administration (Figure 1a, 1b).

Figure 1a: Preoperative sagittal MRI scan, without contrast, revealing the tumor mass relative to the normal spinal cord

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The patient was operated on, and a central laminectomy of T11 through L1 was executed. The dura matter was longitudinally opened in the midline and the mass was completely resected macroscopically (Figure 2a, 2b). The histopathologic examination recognized cancellous bone specimens, without remarkable histopathologic changes. The neoplasm consisted of spindle, epithelial polygonal cells, with round cystic nuclei and prominent nucleoli (Figure 3a, 3b). Melanin was recognized in the cytoplasm (Figure 3c) but nuclear pleomorphisms were not evident.

Figure 1b: Initial MRI scan, with contrast enhancement, which better delineates the margins of the tumor

Figure 2a: Intraoperative photograph, revealing the extent and color of the tumor mass, due to the presence of melanin

Figure 2b: Intraoperative photograph, showing macroscopically total tumor resection

Figure 3a: Hematoxylin- Eosin (H-E), 20 X (magnification) staining technique, verifying the aforementioned histopathologic features

Figure 3b: Similar histopathologic features, H-E staining technique, 40 X
Immunohistochemically, cellular staining for Vimentin (Figure 3d) and HMB 45-antibody were positive.

Figure 3c: Staining showing melanin pigmentation

Immediate postoperative MRI revealed total tumor excision (Figure 4a). The neurologic status of the patient appeared a slow but gradual improvement. Follow up MRI, performed at 12 months post-operatively, did not reveal any regression of the tumor (Figure 4b). In the meantime, the patient experienced mild improvement of her spastic paraparesis.

Figure 4a: Immediate postoperative MRI scan, documenting complete tumor resection

Figure 3d: Immunohistochemical staining, revealing immunopositivity for Vimentin

Differential diagnosis included metastatic malignant melanoma, in which case, angiogenesis and granulomatous territories are observed. Necrotic areas are frequent and diffuse in appearance, while tumor is recognized in the perivascular zone and at the periphery of tissue specimens. Cells share atypical features and nuclear pleomorphisms with atypical features were evident.

Melanotic schwannoma is also included in the differential diagnosis, but in this case there is immunopositivity for S100 protein.
In most cases, gross total resection is achievable [17,18]; however, in rare cases with dumbbell-shaped tumors involving the extraspinal region, a staging operation and subtotal resection should be attempted. During a mean follow-up period of 58.1 months, the symptoms were completely relieved in all the patients, and no tumor progression or recurrence was noted. Melanocytic tumors of the central nervous system have a typical appearance on MRI scans, varying with the content and distribution of melanin. However, the differential diagnosis between malignant melanoma and melanocytoma still depends on pathological criteria. Spinal meningeal melanocytoma has a benign course, and it is amenable for gross total resection. The outcome is favorable following complete resection.

Spinal melanocytomas are rare, especially when located in an intramedullary region, and are often not considered during differential diagnosis of intramedullary spinal cord tumors. The lack of distinctive imaging characteristics increases the challenge during preoperative diagnosis. The high recurrence rate of intermediate-grade intramedullary melanocytomas even after total tumor resection, and their histopathological features of aggressive behavior (for example, increased mitotic activity, infiltrative pattern) indicate close follow-up with serial MR imaging.

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**Conflicts of Interest**

The authors declare that there are no conflicts of interest.

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**References**


7. Jaiswal S, Vij M, Tungria A, Jaiswal AK, Srivastava AK, et al. (2011) Primary melanocytic tumors of the central nervous system: a neuroradiological and clinicopathological study of five cases and brief review of literature. Neurol India 59: 413-419. [Crossref]


