



Multiple Spinal Cord Melanoma: Case Report with Emphasis on the Difficult Preoperative Diagnosis

Multipl Omurilik Melanomu: Zor Preoperatif Tanının Vurgulanmasıyla Bir Olgu Sunumu

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ABSTRACT

Primary spinal melanoma (PSM) is rare and usually occurs in the cervical and thoracic regions. Here, we present a case of multiple spinal cord melanoma located at the level of cervical and upper thoracic spine in a 24-year-old male. The patient underwent a T6-T7 thoracic laminectomy and gross-total resection of the duty lesion. The histological findings of the tumor were consistent with an intermediate-grade melanocytoma. To the best of our knowledge, only two cases of multifocal primary melanocytic tumor have been reported in the literature. In this case study we review the pertinent literature and emphasize the importance and difficulties concerning the preoperative diagnosis of multiple PSM.

KEYWORDS: Primary spinal melanoma, Intermediate-grade, Multiple, Diagnosis

ÖZ

Primer spinal melanom (PSM) nadirdir ve genellikle servikal ve torasik bölgelerde oluşur. Burada 24 yaşında bir erkekte servikal ve üst torasik omurga seviyesinde olmuş bir multipl omurilik melanomu vakasını sunuyoruz. Hastaya T6-T7 torasik lamektomi ve gros total rezeksiyon yapıldı. Tümörün histolojik bulguları orta derece melanositom ile tutarlıydı. Bildiğimiz kadriyle literatürde sadece iki multifokal primer melanostik tümör vakası bildirilmiştir. Bu vaka çalışmasında ilgili literatürü gözden geçirip multipl PSM preoperatif tanısıyla ilgili zorlukları ve tanının önemini vurguluyoruz.

ANAHTAR SÖZCÜKLER: Primer spinal melanom, Orta derece, Multipl, Tanı

INTRODUCTION

Primary melanocytic tumors of the central nervous system (CNS) consist of a spectrum ranging from well-differentiated melanocytoma to malignant melanoma. PSM was initially reported by Hirschberg in 1906, and accounts for 38% of all CNS melanomas (9). So far, fewer than 60 cases have been reported in the English literature (6, 13). Intermediate-grade melanocytic tumors are not easily classified as melanocytoma or melanoma as they present some histological features associated with aggressive behavior, and moreover, only sporadic cases have been reported in the literature (2, 3). To the best of our knowledge, only two cases of multifocal primary melanocytic tumor have been reported in the literature (1, 9). In this study, we present a case of multiple spinal cord melanoma located at the level of the cervical and upper thoracic vertebrae, and conduct a review of the pertinent literature.

CASE REPORT

A 24-year-old male presented with a one-month history of progressive numbness and weakness of the left lower extremity. Physical examination revealed 3/5 muscle strength in the left leg accompanied by impaired sensibility below

the level of T-7. Magnetic resonance imaging (MRI) of the thoracic spine revealed intradural, extramedullary, and multifocal ovoid masses located at the level of upper thoracic vertebrae. The largest mass was approximately 1.2× 2.0 cm in size compressing the spinal cord at the T6-7 level. The masses were hyperintense on T1-weighted images, and isointense to hypointense on T2-weighted images. After administration of meglumine gadopentetate, homogeneous enhancement masses and strip enhancement leptomeninges were evident (Figure 1A-C). The cervical spinal MR imaging indicated the presence of multifocal snotty lesions at the level of lower cervical vertebra (Figure 2A-C).

The patient underwent a T6-T7 thoracic laminectomy and gross-total resection of the duty lesion. The surgeon observed a dura-based dark brown mass that compressed the edematous spinal cord (Figure 3A,B). Microscopically, highly pigmented spindle-cell lesions with moderate nuclear atypia and regions of necrosis were noted; additionally, the cells exhibited epithelioid and syncytial patterns. The nuclei were uniform and oval with small nucleoli. The lesional cells were immunopositive for S100 protein and HMB-45, whereas no expression of epithelial membrane antigen was detected (Figure 4A-D). The diagnosis based upon the pathological



Figure 1: Sagittal magnetic resonance images (MRI) of thoracic spine. (A) T1-weighted MRI shows multifocal ovoid masses located at the level of upper thoracic vertebrae, which has high signal intensity relative to that of the cord. (B) T2-weighted MRI shows homogenous signal hypointensity relative to that of the cord. (C) Contrast-enhanced T1-weighted MRI image shows homogeneous enhancement masses and strip enhancement leptomeninges.



Figure 2: Sagittal MRI of cervical spine shows multifocal snotty lesions located at the level of lower cervical spine, which has hyperintense signal on T1-weighted images (A), hypointensity on T2-weighted images (B) and noticeable enhancement after the injection of meglumine gadopentetate (C).

examination was melanoma, suggestive of an intermediate-grade melanocytoma.

Postoperatively, left lower-extremity strength and sensibility below the level of T-7 were markedly improved. The MR images of the thoracic spine obtained 1 week after surgery revealed the disappearance of the mass effect at the T6-7 level. A primary lesion was not observed upon subsequent abdominal color Doppler ultrasonography and skin examination, and no intracranial lesions were seen upon MRI of the brain. Therefore, a diagnosis of primary spinal intermediate-grade melanocytoma was made. A second surgery for the additional masses in the lower cervical vertebra and upper thoracic vertebrae was recommended, while the patient chose outpatient follow-up without further treatment.

DISCUSSION

According to the World Health Organization classification of primary CNS tumors, primary CNS melanoma includes diffuse leptomeningeal melanocytosis or melanomatosis, melanocytoma, and primary malignant melanoma. Primary CNS melanoma is rare, accounting for only 1% of all melanoma cases, and primary spinal melanoma is even more rare (7). Since first reported by Hirschberg in 1906, approximately 50 to 60 diagnosed cases have been reported in literature (4, 6, 13). These include melanocytoma, intermediate-grade melanocytoma and malignant melanoma. To the best of our knowledge, only two case of multifocal primary melanocytic tumor have been reported in the literature

(1, 9). The characteristics of these cases are summarized in Table I. Compared with previously reported cases, the patient presented in this study had a relatively longer lesion, extending from the level of lower cervical vertebra to upper thoracic vertebrae.

The clinical presentation of PSM is non-specific and predominantly consists of symptoms of progressive myelopathy. The slight predilection for the cervical and thoracic spine has been observed as intradural, extramedullary lesions. Currently, MRI is the best method for diagnosing spinal cord tumors. Melanoma characteristically exhibits hyperintensity on T1-weighted images, hypointensity on T2-weighted images, and homogeneous enhancement on T1 contrast-enhanced images. Melanoma does not always exhibit a homogeneous pattern on MR images and the degree of T1 and T2 shortening appears to be directly related to the melanin content (5). Our clinical and neuroimaging findings are similar to the available descriptions.

A preoperative diagnosis of PSM is difficult to achieve. According to the Hayward classification system, a diagnosis of primary melanoma is based on the absence of a melanoma outside the CNS, absence of a melanoma in other CNS sites, and histological confirmation of a melanoma (8). In addition, a number of other CNS lesions may be pigmented and produce melanin, including schwannoma, medulloblastoma, neurofibroma, meningioma, astrocytoma and pituitary tumours. Immunohistochemical and ultrastructural criteria can help differentiate between these entities, because primary

Table I: Summary of Reported Cases of Multifocal Primary Melanocytic Tumor of the Central Nervous System

Reference	Age (yrs), Sex	Location	Pathology	Treatment	Adjuvant Therapy	Outcome
Ali Y, et al., 2009 ¹	31, M	Bilateral cerebellopontine angles and the thoracic spine at the T5-T6 level	Meningeal melanocytoma	Gross-total resection of the thoracic lesion	None	Follow up at a few weeks after operation: a rapidly progressive coma, with signs of brainstem compression and hemodynamic instability, ultimately leading to his death.
Lee CH, et al., 2010 ⁹	39, M	Spreading from the level of C1 to the level of C6	Malignant melanoma	Gross-total removal	Local radiation therapy and adjuvant chemotherapy were given.	Follow up at 17 months after operation: no recurrence on MRI
Current report	24, M	Cervical and upper thoracic spine	Intermediate-grade melanocytoma	Gross-total removal of the duty lesion	Second surgery for the other masses but the patient did not accept further treatment.	Follow up at 2 weeks after operation: the left lower-extremity strength and the sensibility below the level of T-7 were markedly improved.

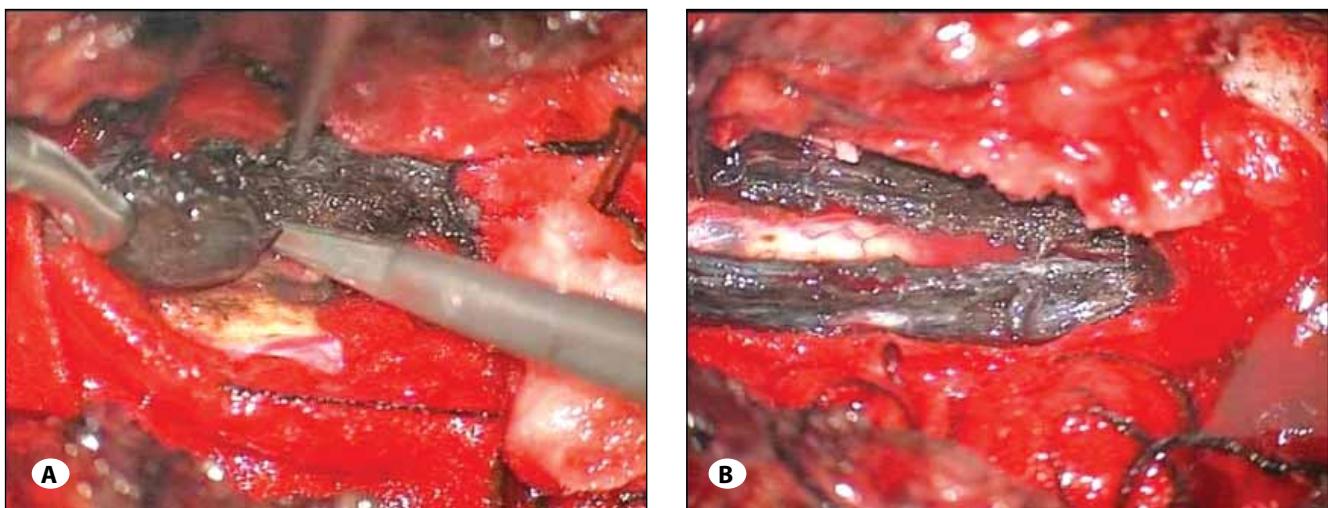


Figure 3: Intraoperative photographs showing the dura-based dark brown lesion compressing the edematous spinal cord (A), and the associated diffuse leptomeningeal hyperpigmentation (B).

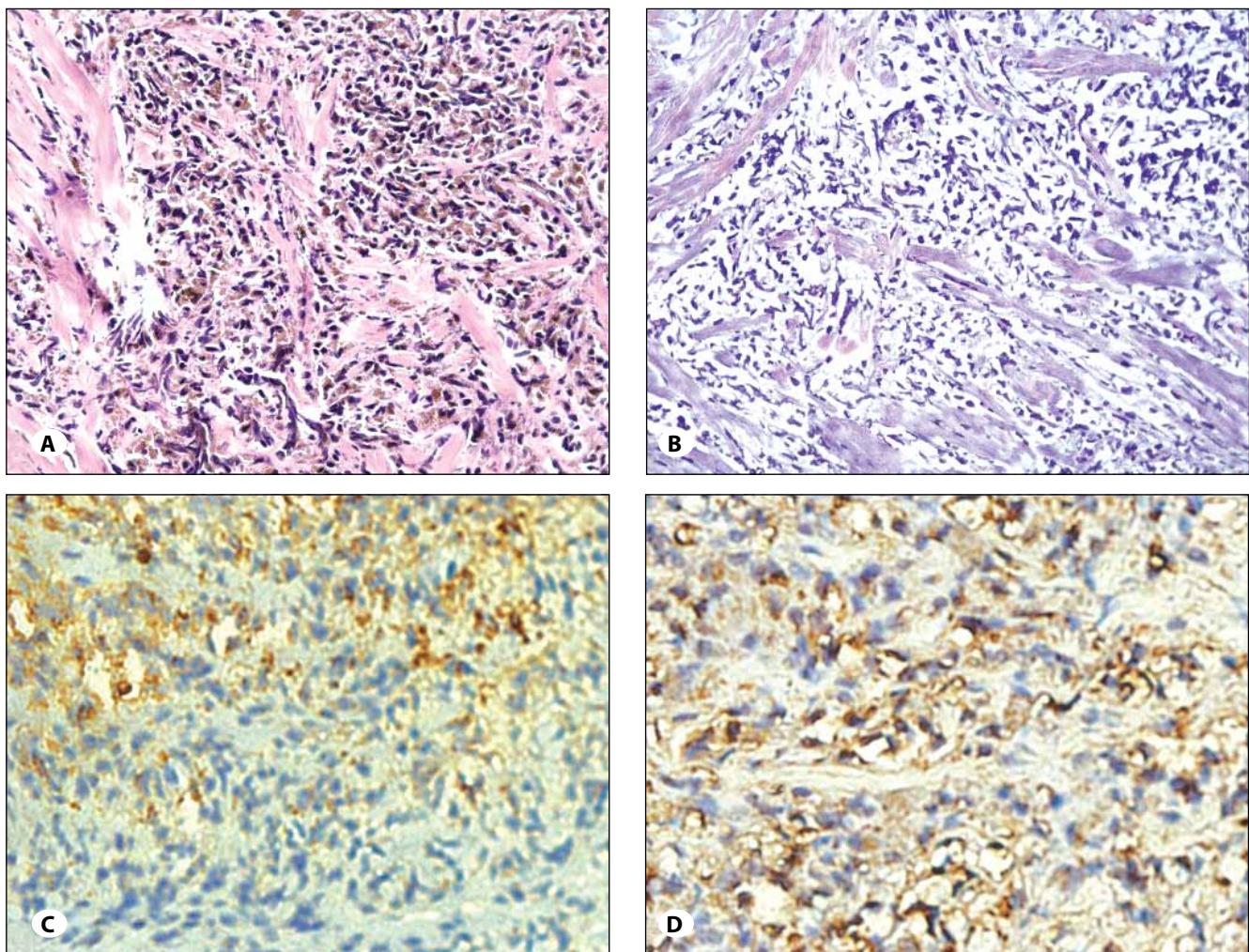


Figure 4: Histology of the PSM. (A) HE stain showing a moderately cell-dense collection of spindle cells with heavily pigmented cytoplasm, mainly arranged in an epithelioid and syncytial pattern. (B) Subjected to hydrogen peroxide bleaching showing the uniform and oval nuclei with small nucleoli. (C) HMB-45 immunopositivity of the tumor cells. (D) S100 immunopositivity of the tumor cells. A, B, C, D x400.

CNS melanoma demonstrates positivity for vimentin, S100 protein, and HMB- 45, whereas a negative immunoreaction to epithelial membrane antigen, cytokeratin, neuron-specific enolase, and Leu-7 is usually observed. It remains a diagnostic challenge to distinguish between a well-differentiated, intermediate-grade melanocytic lesion, and malignant melanoma. A detailed description of the histopathological criteria for distinguishing among the wide spectrum of melanocytic lesions was provided by Navas et al.(10) and modified by Brat and Quatresooz (2, 11).

Because the biological behavior of PSM is variable, complete resection is advised when possible. Adjuvant radiation therapy and Gamma Knife radiosurgery have been shown to control tumor growth and improve outcome following partial resection (1, 12). Ali et al. reported a case study of a 31-year-old patient presenting with a multifocal meningeal melanocytoma simultaneously affecting both cerebellopontine angles and the thoracic spinal cord, and exhibiting a remarkably aggressive clinical course and a poor prognosis (1). In this case study, we introduce a new entity, multiple intermediate-grade melanocytoma, which similarly appears to portend a poor prognosis. Therefore, careful monitoring of patients diagnosed with PSM is strongly advised, particularly for cases exhibiting multiple lesions.

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