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## Spine

# A rare case of primary spinal cord melanoma

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### ARTICLE INFO

#### Article history:

Received 10 November 2017

Accepted 4 January 2018

Available online 20 February 2018

#### Keywords:

Neuroradiology

Melanoma

Spine

Malignancy

### ABSTRACT

Primary central nervous system melanoma is rare, accounting for approximately 1% of total melanoma cases (1-3). Primary central nervous system melanoma localized to the spinal cord is even rarer. The cervical and thoracic levels are most frequently involved. There are no pathognomonic imaging characteristics and diagnosis must be confirmed immunohistologically. The preferred treatment is gross total resection. Utilization of adjuvant radiotherapy and chemotherapy may improve disease-free survival. The prognosis for primary central nervous system melanoma is generally better than that of metastatic and cutaneous melanoma, although there are relatively few cases from which to draw conclusions. We report a case of a 64-year-old woman diagnosed with primary spinal melanoma of the thoracic spine treated with subtotal surgical resection followed by adjuvant radiation therapy.

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## Case report

A 64-year-old woman was seen for gradually worsening dysesthesia of the right lower extremity which subsequently progressed to involve the right upper extremity and left lower extremity. She had previously been treated for neck and mid back pain for several years. Magnetic resonance imaging (MRI) studies of the cervical and lumbar spine revealed no explanatory abnormalities. Initial nerve conduction studies were unremarkable. Her past medical and surgical history was noncontributory. Neurologic examination revealed normal strength in the extremities with hypesthesia to pin prick and light touch seeming to begin below the T9-T10 level. Deep tendon reflexes were mostly unremarkable, with one-fourth

right ankle jerk and equivocal Babinski on the left. The symptoms progressively worsened over several months, with decreasing strength resulting in falls, and hypesthesia to temperature, proprioception, and vibration of the distal lower extremities.

MRI of the thoracic spine showed an intramedullary mass at T8. The mass showed hyperintensity on T1 and T2 (Fig. 1A, sagittal; Fig. 1B, sagittal), moderate heterogeneous enhancement following gadolinium injection (Fig. 1C, sagittal), lack of flow voids, mild surrounding edema, and cord expansion. Given the intramedullary location and signal characteristics, the primary consideration was for an ependymoma or astrocytoma; less likely consideration included hemangioblastoma or metastatic disease. Because of edema and cord expansion, hemorrhage and demyelinating processes were less likely.

Competing Interests: The authors have declared that no competing interests exist.

The views expressed in this article are those of the author(s) and do not reflect the official policy or position of the Department of the Army, Department of Defense, or the U.S. Government.

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<https://doi.org/10.1016/j.radcr.2018.01.009>

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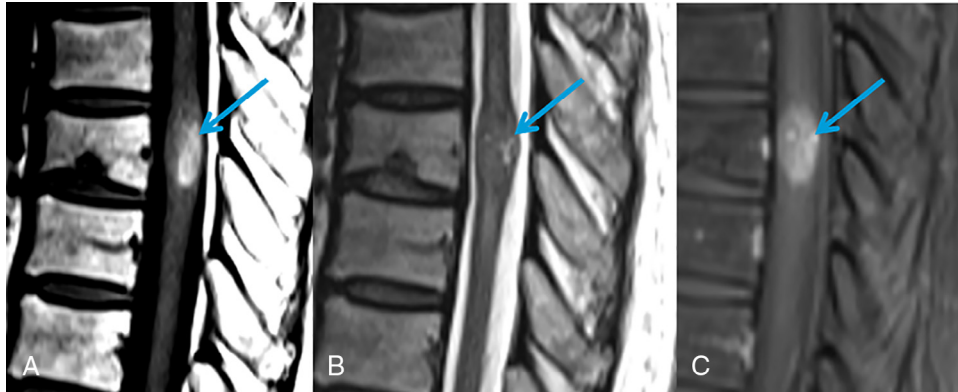


Fig. 1 – (A) Sagittal T1. (B) Sagittal T2. (C) Sagittal T1 with contrast.

The patient underwent T7-T9 laminectomy with near-total resection of a dark reddish-brown ventrally exophytic intramedullary tumor; subtotal resection was due to proximity of the tumor with the anterior spinal artery. The postoperative course was noneventful. Following the procedure, the patient underwent adjuvant radiotherapy to a dose of 50.4 Gy. Final pathologic evaluation returned a diagnosis of nonmelanocytic melanoma of the spinal meninges. The tissue was red-tan with pleomorphic melanocytic mitotic activity. The diagnosis was confirmed following expert consultation and specialized pathologic testing.

Following a thorough workup, including complete dermatologic examination and positron emission tomography, the absence of primary melanoma was confirmed.

The patient is alive (age 74), and at 8 years following the diagnosis, reports ongoing back pain and immobility requiring use of a wheelchair for most activities. Following over 7 years of stability, recent surveillance imaging shows enlargement of the residual enhancing mass at T7, but no clinical signs of metastasis.

## Discussion

Melanoma can occur anywhere there are melanocytes or cells with the potential to become melanocytes [1]. Primary central nervous system (CNS) melanomas are thought to arise from the neural crest [2,3]. The vast majority of CNS melanomas are metastatic. Metastatic melanoma is the third leading cause of all CNS metastases [4], behind breast and lung cancers. Primary CNS melanoma is very rare, accounting for approximately 1% of all melanomas [1,2,5,6], and 0.07% of all brain tumors [7]. The incidence has been estimated to be 0.005 cases per 100,000 population [8]. Primary spinal melanoma is even rarer. Spinal lesions most commonly are solitary, intradural extramedullary, and occur in the cervical and thoracic spine [6,8,9]. The case presented is even more rare as it presented intramedullary rather than intradural extramedullary. Primary CNS melanoma has a peak incidence in the fifth decade, having arisen in patients ranging from 15 to 80 years [6,8,10]. There is equal incidence between men and women [1]. The clinical presentation is nonspecific and dependent on the level of the

lesion. The most common sign is progressive motor weakness, but patients have presented with such symptoms as dysesthesias, weakness, abnormal reflexes, loss of bowel or bladder control, and pain [9,11,12].

MRI is the best imaging modality for evaluating spinal cord tumors. Distinction based solely on imaging characteristics remains difficult as there are no pathognomonic features. The differential includes other common lesions of the spinal cord, including ependymoma and astrocytoma, and other melanotic tumors such as benign melanocytoma or melanotic schwannoma. Characteristic imaging features on computed tomography include hyperdensity of the lesion, which may be suggestive of hemorrhage [2,4,13]. Characteristic imaging features on MRI differ from most intracranial tumors, and are characterized by hyperintensity on T1-weighted sequences, hypointensity on T2-weighted sequences, and homogeneous enhancement following infusion of gadolinium-based contrast agents [1,4,6,13,14]. These features overlap with other melanocytic tumors [1]. Furthermore, imaging characteristics may be variable, depending on the amount of hemorrhage and melanin present within the mass [1,6,15]. The imaging characteristics of the case presented varied somewhat from the classic description, in that the T7 intramedullary mass demonstrated hyperintensity on T2-weighted imaging. This is thought to be due to an increased amount of tissue edema or hemorrhage at the time of imaging.

The ultimate diagnosis must be made following histopathologic examination. Histopathologic confirmation is accomplished by demonstrating immunoreactivity for HMB-45 and S-100 protein. Microscopic features include the formation of tight nests surrounded by well-differentiated melanocytes with cytoplasm rich in melanin [16]. Differentiation of primary melanoma from metastatic disease must also be accomplished, and has important prognostic implications. A thorough search for additional foci should include complete skin examination, including mucosa, uvula, and genitalia [1]. Ophthalmologic and gastrointestinal examinations should also be performed [1,9]. The use of positron emission tomography with computed tomography following F-18-2-fluoro-2-deoxyglucose injection has been studied and found to be an accurate method of searching for additional foci of disease [2].

Hayward [17] published the most widely accepted criteria for diagnosis of primary CNS melanoma: (1) no malignant

melanoma identified outside the CNS; (2) involvement of the leptomeninges; (3) intramedullary spinal lesions; (4) hydrocephalus; (5) tumor in the pituitary or pineal gland; (6) a single intracerebral lesion. These criteria, along with other clinical features and histopathologic confirmation, usually allow for the diagnosis of primary CNS melanoma. The case presented meets these criteria.

Based on the literature, complete surgical resection offers the most promising outcome for primary spinal cord melanoma [1,6,8], with some reports documenting long-term survival following even subtotal resection [6]. The use of adjuvant radiotherapy is recommended in all cases [1,8,12,18]. The use of postoperative chemotherapy appears promising, especially after inadequate resection. However, there remain relatively few cases from which to draw conclusions [6,8,10]. Kim et al. found the mean total radiation dose to be 47 Gy (range from 30 to 60) for 11 of 26 cases reviewed [6]. In the case presented, the total dose was 50.4 Gy. Several authors have described treatment with dicarbazine, or alternatively vincristine, bleomycin, and cisplatin with mixed results [10,14,19,20].

Distinction between primary CNS melanoma and metastatic melanoma is crucial. The median survival for metastatic CNS melanoma has been reported to be between 3 and 6 months [6,21], in contradistinction to primary CNS melanoma which seems to offer a much better prognosis [6,8,11,20]. This may be related to the lack of CNS lymphatics, limiting the ability for metastasis [6]. Larsen et al. reported an average survival of approximately 7 years following surgery and radiation therapy [11], whereas Kim et al. reported a mean survival duration of approximately 30 months [6]. In the case presented, the patient is alive at 8 years following diagnosis and surgery, although she recently presented with worsening symptoms and enlargement of the remnant T7 mass.

The clinical course of primary spinal cord melanoma is unpredictable. Surveillance imaging is recommended at regular intervals [14,22]. Our patient has been followed up clinically and with serial MRI of the spine.

Primary spinal melanoma is extremely rare, with most cases occurring in the cervical and thoracic spine. The preferred treatment is surgical resection. Utilization of adjuvant radiotherapy and chemotherapy may improve disease-free survival. MRI imaging characteristics may vary, but generally include hyperintensity on T1-weighted imaging, hypointensity on T2-weighted imaging, and contrast enhancement. The prognosis for primary CNS melanoma is generally better than that of metastatic and cutaneous melanoma.

The views expressed in this article are those of the author(s) and do not reflect the official policy or position of the Department of the Army, Department of Defense, or the U.S. Government.

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