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Primary leptomeningeal melanoma in an adolescent: Case report and review of the literature

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A previously healthy 16-year-old male patient came to the emergency department with a six-week history of left posterior hip pain following a head-on ATV collision. After failing conservative therapy, he was found to have weakness and atrophy of the left lower extremity on exam. MRI demonstrated an epidural mass that was hyperintense on T1 and hypointense on T2. Biopsy of the mass revealed a melanocytic lesion; further lesions were identified in the lungs, which biopsy proved to be malignant melanoma. Dermoscopy of the entire body, anoscopy, and retinal exam were negative for melanoma; a whole-body PET scan did not reveal any further lesions or evidence of a primary lesion. Given the lack of a primary lesion, this was thought to be a primary leptomeningeal melanoma. Primary melanomatous tumors of the spine are rare entities, with fewer than 40 cases described in the literature. This case was unique due to the patient's very young age and the presence of metastases on presentation.

Introduction

Primary CNS melanoma is a rare entity, first described by Virchow in 1859. In 1976, Hayward established a guide for classification of primary CNS melanoma (1), which described how primary CNS melanoma is considered a diagnosis of exclusion; that is, it requires the presence of melanoma in the CNS with no primary melanoma located in mucosal, retinal, or cutaneous tissues. CNS melanomatous tumors range from benign melanocytomas to malignant melanomas. Location in the spine is especially rare, with fewer than 40 cases described in the literature (2-3). Although the tumors can be located anywhere along the leptomeningeal axis, they are typically found in the cervical or thoracic spine (4, 5).

It is thought that primary melanoma of the spinal cord arises from either leptomeningeal melanoblasts accompanying vascular sheaths or from neuroectodermal rest cells during embryogenesis (2, 6). In several large series, patient age ranges were between 15-71 years, with a peak in the 5th decade of life (7-9). Therefore, this case was unusual because of the age at diagnosis and the metastatic nature on presentation. A search of the literature revealed only one case presenting with brain metastases (7) and none with lung and lymph-node involvement.

Case report

A 16-year-old male Caucasian patient with no significant past medical history presented to the emergency department with a six-week history of posterior, sharp left hip pain. He had recently been involved in a head-on ATV-versus-trail-bike collision. At the time of the incident, he was not wearing a helmet, remained mounted on the ATV, and was able to ambulate without obvious difficulty immediately after. He was prescribed oral NSAIDs, with only minimal relief of symptoms over the course of the next two weeks.

On followup, his PCP suspected sciatica and subsequently prescribed oral corticosteroids. Five days later, the

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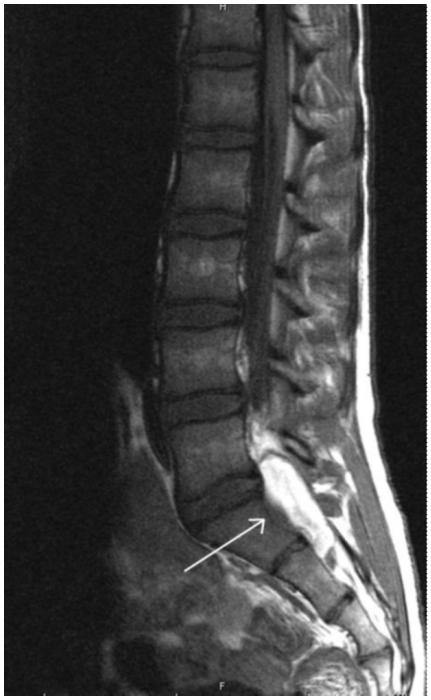

Figure 1. 16-year-old male with primary leptomeningeal melanoma. A T1-weighted image of the lumbar spine shows a hyperintense epidural mass.

patient returned to the emergency department with severe worsening of the hip pain. During that visit, plain films of the lumbar spine and left hip were obtained but were unremarkable. The patient was seen in PM&R for further evaluation. Atrophy of the left gastrocnemius and soleus muscles was identified on physical exam. Additional imaging (obtained with noncontract MRI of the lumbar spine) showed a 4.5x1.8cm tubular structure in the spinal canal that was hyperintense on T1-weighted images (Fig. 1) and hypointense on T2-weighted images (Fig. 2). The lesion had

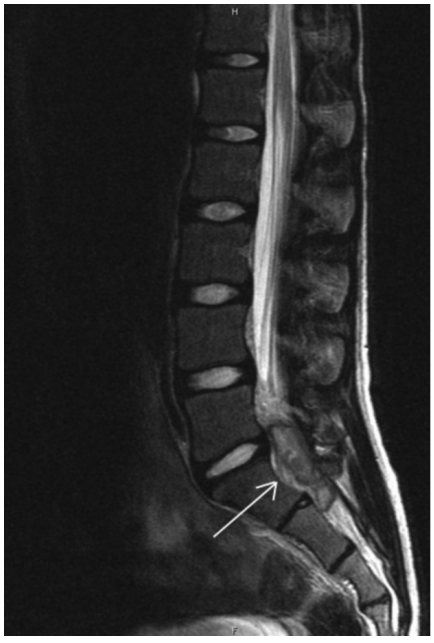

Figure 2. 16-year-old male with primary leptomeningeal melanoma. A T2-weighted image of the lumbar spine shows a hypo- to isointense epidural mass.

significant mass effect on the left sacral nerve roots, with hemorrhage extending into the S1-S2 and S2-S3 neuroforamina and accompanying severe central spinal stenosis. Given the history of recent trauma and patient age, the mass was suspected to most likely represent an epidural hematoma.

The patient was brought to the OR for emergent evacuation of the suspected hematoma and complete L1 hemilaminectomy. A grossly bluish, sticky, and well-organized clotted mass was identified, removed, and sent to pathology. The specimen was read as consistent with a melanocytic neoplasm; due to the low proliferation rate, the tumor was thought to be more likely a benign melanocytoma. However, given the possibility of metastasis, CT images of chest/abdomen/pelvis and MRI of the brain and total spine were obtained. MRI of the brain and total spine found no significant abnormalities. CT revealed multiple small pulmonary nodules. In order to rule out metastatic disease, two pulmonary nodule biopsies were obtained and interpreted as malignant melanoma; the lesions appeared black when removed (Figs. 3-4). A thorough full-body dermoscopy, retinal exam, and anoscopy did not identify any melanocytic lesions or suspicious moles. Therefore, the primary lesion was thought to be of spinal origin, most likely lumbosacral leptomeningeal or an epidural melanocyte stem-cell cluster. After the initial lesion was resected,

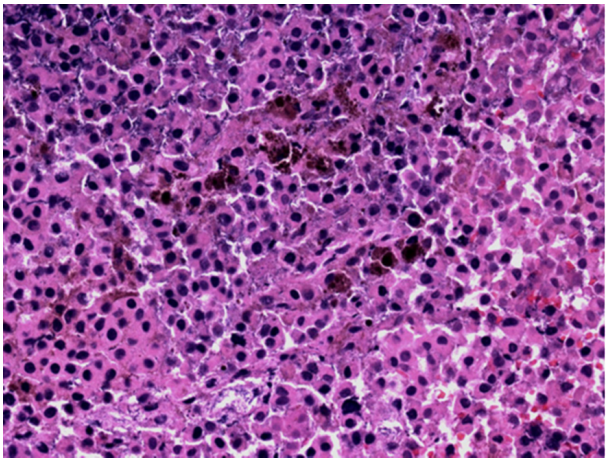

Figure 3. 16-year-old male with primary leptomeningeal melanoma. A slide from the resection of the epidural lesion at the base of the spine shows a largely necrotic tumor. The better preserved cells are dyscohesive, with round pyknotic nuclei and moderately abundant eosinophilic cytoplasm. Mitoses are not seen, and the MIB-1 rate (a measure of the cellular reproductive rate) was very low. Abundant dark brown patches suggest a melanocytic neoplasm.

molecular testing showed that the patient's melanoma was c-kit positive; the patient was subsequently treated with imatinib, and he clinically improved over the next several months. At the time of this report, the patient was in remission from his melanoma.

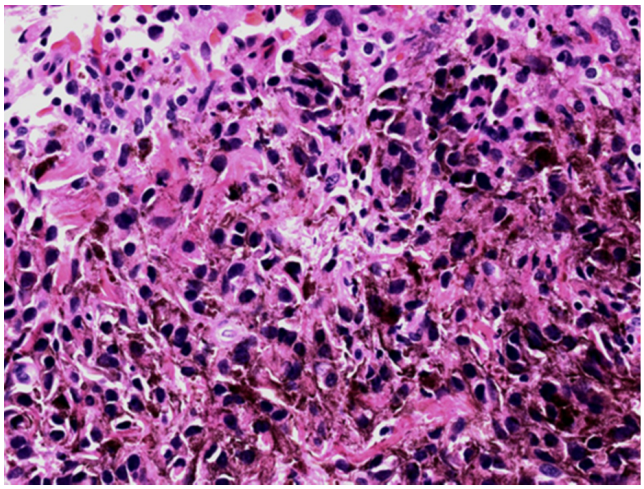

Figure 4.16-year-old male with primary leptomeningeal melanoma. Immunohistochemistry for the melanocyte-specific marker Melan-A shows the characteristic cytoplasmic staining, confirming the diagnosis of a melanocytic neoplasm.

Discussion

MRI findings of spinal-cord lesions can help suggest the presence of melanomas, melanocytomas, and other pigmented lesions (6, 10). However, differentiating between pigmented tumors and other entities can be confusing. Tumors with high T1 signal and low to isointense T2 signal and iso- to hyperintensity on CT should be suspected as melanomas. This imaging pattern occurs in malignant melanomas due to the paramagnetic effect of stable free radicals and the metal-scavenging effects of melanin (11). However, this pattern can be seen in other lesions as well, and melanomas in general can be quite heterogeneous. Differential diagnosis would also include subacute epidural hematoma. This was initially suspected in this case due to the history of trauma; it presents with high signal intensity on T1 due to the presence of intracellular and extracellular methemoglobin, which has a short T1 due to dipole-dipole interactions (11). Hyperintensity on T1-weighted imaging is seen in fat or hemorrhage as well as in melanoma, whereas hypointensity on T2-weighted imaging could conceivably be seen in subacute hemorrhage. This case demonstrates that melanoma should always be on the differential diagnosis of a mass lesion showing characteristic imaging findings, regardless of the patient's age, other demographics, or risk factors.

References

1. Hayward R. Malignant melanoma and the central nervous system. A guide for classification based on the clinical findings. *J Neurol Neurosurg Psychiatry* 1976 39:526–530 [PubMed]
2. Kim M, Yoon D, and Shin D. Primary spinal cord melanoma. *J Korean Neurosurg Soc*. 2010 Aug;48(2):157-161. [PubMed]
3. Kounin G, Romansky K, Traykov L, et al. Primary spinal melanoma with bilateral papilledema. *Clinical Neurology and Neurosurgery* 2005 107(6) 525-527 [PubMed]
4. Wadasadawalaa T, Trivedia S, Guptab T, et al. The diagnostic dilemma of primary central nervous system melanoma. *Journal of Clinical Neuroscience* 2010; 17(8)1014–1017 [PubMed]
5. Larson T, Houser O, Onofrio B, et al. Primary spinal melanoma. *JNS* 1987 66(1) 47-49 [PubMed]
6. Farrokh D, Fransen P, and Faverly D. MR findings of a primary intramedullary malignant melanoma: Case report and literature review. *AJNR* 2001 22: 1864-1866 [PubMed]
7. Brat DJ, Giannini C, Scheithauer BW, Burger PC. Primary melanocytic neoplasms of the central nervous systems. *Am J Surg Pathol* 1999;23:745-54. [PubMed]
8. Liubinas S, Maartens N, and Drummond J. Primary melanocytic neoplasms of the central nervous system. *Journal of Clinical Neuroscience* 2010 17(10) 1227–1232 [PubMed]
9. Yu J, Zhao, D, Chen S, et al. Primary melanoma of the cervical spine with cerebral metastases: Case report and review of the literature. *Journal of International Medical Research* 2002 40(3) 1207-1215 [PubMed]
10. Demirci A, Kawamura Y, Sze G. et al. MR of parenchymal neurocutaneous melanosis. *AJNR* 1995 16:603–606 [PubMed]
11. Ginat D, and Meyers S. Intracranial lesions with high signal intensity on T1-weighted MR images: Differential diagnosis. *Radiographics* 2012 32(2) 499-516 [PubMed]