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AJNR Am J Neuroradiol 1997, 18 (1) 180-182

<http://www.ajnr.org/content/18/1/180>

This information is current as
of April 17, 2024.

MR of Spinal Meningeal Melanocytoma

Edward J. Czarnecki, Richard Silbergleit, and Jorge A. Gutierrez

Summary: We report an unusual case of meningeal melanocytoma, a rare pigmented lesion of the leptomeninges, that occurred within the leptomeninges of the thoracic spinal canal. The lesion showed increased signal intensity on T1-weighted MR images relative to gray matter, was isointense with gray matter on T2-weighted images, and enhanced mildly but homogeneously after administration of contrast material.

Index terms: Meninges, neoplasms; Spine, neoplasms

Meningeal melanocytoma is a rare primary pigmented lesion of the leptomeninges (1). Although similar to melanotic meningioma in its benign nature, melanocytoma has its own unique imaging, histologic, and ultrastructural makeup. These characteristics allow for differentiation between meningeal melanocytoma, melanotic meningioma, and melanoma (primary or metastatic). We present a case of meningeal melanocytoma that occurred within the leptomeninges of the thoracic spinal canal.

Case Report

A 69-year-old woman had a 9-day history of unsteadiness accompanied by a subjective feeling of leg weakness, which had progressed during the preceding week. She denied any pain or bowel/bladder dysfunction, and there was no significant medical history. Results of physical examination and laboratory studies were unremarkable. Findings on contrast-enhanced computed tomographic (CT) scans of the brain were normal. Myelography and postmyelography CT showed an intradural extramedullary lesion at T8–9 (Fig 1A). Further examination by magnetic resonance (MR) imaging showed the lesion to have increased signal intensity relative to the cord on T1-weighted images and isointense signal intensity on fast spin-echo T2-weighted images. The lesion showed diffuse mild homogeneous enhancement in addition to the inherent noncontrast T1 high signal intensity (Fig 1B–E). Our preoperative diagnosis was a melanotic meningioma. The patient underwent thoracic laminectomy with total resection of the intradural extramedullary lesion. The gross

specimen was jet black in appearance (Fig 1F). The histologic and electron microscopic evaluation revealed a meningeal melanocytoma (Fig 1G). After surgery, both the unsteadiness and the subjective feeling of leg weakness resolved. The patient underwent postoperative dermatologic and ophthalmologic consultation, which revealed no other melanotic lesions.

Discussion

Melanocytes are neural crest in origin. They are found throughout the leptomeninges, but are most concentrated on the surface of the brain stem and upper cervical spinal cord. These leptomeningeal melanocytes give rise to the meningeal melanocytoma.

Melanotic tumors of the central nervous system are rare and are most commonly metastatic in origin. Primary tumors of the leptomeningeal melanocytes do occur and are usually malignant, with about 25% associated with neurocutaneous melanosis syndrome (2). A primary benign meninges-based melanocytic tumor was described by Limas and Tio in 1972 (1). They used the term *meningeal melanocytoma* to describe a benign tumor of melanocytic origin arising from the leptomeninges.

We found fewer than 30 of these cases in the literature (3, 4). The lack of strict diagnostic criteria before 1972 has resulted in some confusion about the total number of legitimate cases of melanocytoma. Of those reported, only a few have been studied by MR imaging. Melanocytomas have been shown to occur anywhere along the neural axis, but they are most commonly found in the region of the foramen magnum, the posterior fossa, or Meckel's cave, or adjacent to cranial nerve nuclei.

Of the reported melanocytomas that have been studied with MR imaging, one was not adequately described, two were isointense with

Received December 18, 1995; accepted after revision March 8, 1996.

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AJNR 18:180–182, Jan 1997 0195-6108/97/1801-0180 © American Society of Neuroradiology

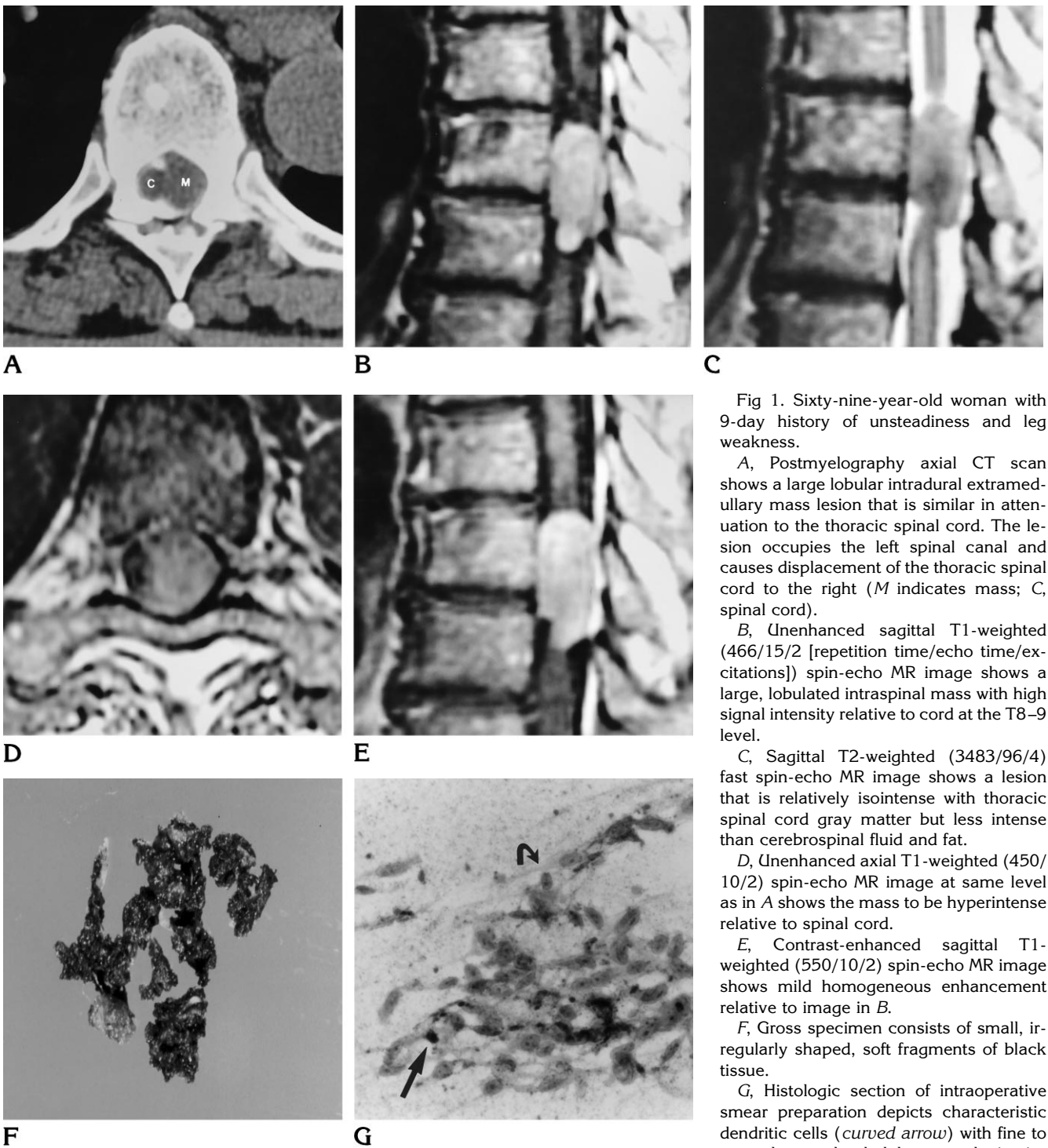


Fig 1. Sixty-nine-year-old woman with 9-day history of unsteadiness and leg weakness.

A, Postmyelography axial CT scan shows a large lobular intradural extramedullary mass lesion that is similar in attenuation to the thoracic spinal cord. The lesion occupies the left spinal canal and causes displacement of the thoracic spinal cord to the right (*M* indicates mass; *C*, spinal cord).

B, Unenhanced sagittal T1-weighted (466/15/2 [repetition time/echo time/excitations]) spin-echo MR image shows a large, lobulated intraspinal mass with high signal intensity relative to cord at the T8-9 level.

C, Sagittal T2-weighted (3483/96/4) fast spin-echo MR image shows a lesion that is relatively isointense with thoracic spinal cord gray matter but less intense than cerebrospinal fluid and fat.

D, Unenhanced axial T1-weighted (450/10/2) spin-echo MR image at same level as in A shows the mass to be hyperintense relative to spinal cord.

E, Contrast-enhanced sagittal T1-weighted (550/10/2) spin-echo MR image shows mild homogeneous enhancement relative to image in B.

F, Gross specimen consists of small, irregularly shaped, soft fragments of black tissue.

G, Histologic section of intraoperative smear preparation depicts characteristic dendritic cells (*curved arrow*) with fine to coarsely granular dark brown melanin pigment (*straight arrow*) (hematoxylin-eosin, magnification $\times 200$).

gray matter on T1-weighted images with homogeneous enhancement on postcontrast images, one had high signal intensity on T1-weighted images with low signal intensity on T2-weighted images, and one was isointense with gray matter on T1-weighted images and hyperintense on T2-weighted images (2–6). The differences in MR signal intensities between the reported lesions most likely relate to the degree of melanin content. Paramagnetic free radicals known to exist in melanin are thought to be responsible for the associated T1 and T2 shortening by the proton-electron dipole-dipole proton relaxation enhancement mechanism. The degree of T1 and T2 shortening appears to be directly related to the melanin content. Intracellular methemoglobin and fat also have high signal intensity relative to gray matter on T1-weighted images and isointense to low signal intensity on T2-weighted images. In our case, the lack of blood or fat attenuation on the postmyelography CT scan, the difference between fat and lesion on fast spin-echo T2-weighted images, and the enhancement on postcontrast MR images helped to discriminate melanocytoma from these other entities. CT of melanocytoma has been shown to be somewhat variable, with most having the same attenuation as gray matter before contrast administration and showing variable enhancement after intravenous administration of contrast material.

On gross examination, most meningeal melanocytomas appear black. Other reported colorations include red, brown, white, tan, and blue. Melanocytomas have unique light microscopic, electron microscopic, and immunoperoxidase staining characteristics. Electron microscopic and immunoperoxidase staining allow for differentiation between melanocytoma and melanotic meningioma. Melanocytomas lack the desmosomes and interdigitating cellular processes found in meningiomas. Reports of immunoperoxidase staining show that epithelial markers are absent in melanocytomas but present in meningiomas (7). Differentiating metastatic or primary melanoma of the central

nervous system from melanocytoma can be difficult on frozen section analysis. Histologically, melanocytomas lack necrosis and significant mitotic activity. They closely resemble uveal tract melanocytomas.

Clinically, patients with meningeal melanocytoma do much better than patients with primary or metastatic melanoma of the leptomeninges (5). Complete surgical resection can be curative. Because there can be some overlap histologically between malignant melanoma and melanocytoma, a thorough physical examination to search for a primary cutaneous, mucosal, or ocular melanoma is recommended.

In summary, meningeal melanocytoma must be considered when one encounters a hyperintense meningeal-based lesion on T1-weighted images that is isointense to hypointense relative to gray matter on T2-weighted images and that shows enhancement after administration of contrast material. This differential diagnosis will allow the surgeon to make decisions regarding aggressive surgical management and alert the pathologist to the possibility that this may be a benign melanocytic tumor and not a primary or metastatic melanoma.

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