Spinal Meningeal Melanocytoma in the S-1 Nerve Root Sheath with Paraspinal Extension Mimicking Schwannoma

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BACKGROUND: **Spinal melanocytomas are variants that can be extradural or intradural and are most often located in the intradural extramedullary compartment of the main thecal sac as in schwannomas. However, origin of this variant from the pure peripheral compartment of rootlets is exceedingly rare.**

CASE DESCRIPTION: **The authors present a case of spinal melanocytoma with confusing pathologic and radiologic features. This patient presented clinically with severe radiating pain on the right lower extremity. Before surgery, clinical and radiologic findings were consistent with a schwannoma. However, on operation, a coal-black pigmented lesion mimicking old blood clot was found inside the right S-1 root sheath, attached to the dura. The mass was completely removed and consequently the patient's symptoms improved.**

CONCLUSIONS: **The spinal melanocytoma should be included as a differential diagnosis before giving a presumptive diagnosis of schwannoma or hemorrhage for the patient with a space-occupying lesion at a peripheral rootlet. Intraoperative gross morphology and histopathologic findings facilitate differential diagnosis.**

INTRODUCTION

Spinal melanocytomas are usually slowly growing tumors that usually present with symptoms of mass effect, leading to progressive neurologic deficits (13). The clinical manifestation occurs mostly in the third to sixth decades of life, and there is no gender predominance (5).

Melanocytomas arise from leptomeningeal melanocytes in the central neuraxis (1). Approximately 50% of the cases are found intracranially and the other half in the spinal canal. Spinal melanocytomas are variants that can be extradural or intradural and are most often located in the intradural extramedullary compartment of the main thecal sac as in a schwannoma. However, origin of this variant from the pure peripheral compartment of rootlets is exceedingly rare.

We present a case of a single S-1 root melanocytoma and review the literature, with special attention to differential diagnosis.

Key words
- Meningeal melanocytoma
- Sacral spine
- Schwannoma
- Spine tumor

Abbreviations and Acronyms
CT: Computed tomography
MRI: Magnetic resonance imaging

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CASE REPORT

A 57-year-old man presented with right leg sciatica along the L-5 dermatome for 2 years, and the pain had exacerbated in the previous month. Neurologic examination on admission revealed sensory disturbance at the L-5 dermatome and grade 4 weakness of the ankle and toe dorsiflexion. Magnetic resonance imaging (MRI) of the spine showed an intradural tumor around the right S-1 root that was high signal intensity on T1-weighted imaging and low signal on T2-weighted imaging. The tumor displayed homogeneous well enhancement on contrast-enhanced MRI (**Figure 1**) and showed iso-density on computed tomographic (CT) scans (**Figure 2**). At first, we suspected nerve root hematoma due to nerve root block from other hospitals. But patient denied any previous procedure.

The patient underwent hemilaminectomy of S-1 and the upper S-2 through a posterior approach. Coal-black pigmented lesions were found next to the right S-1 root, attached to the nerve sheath. Linear incision over the nerve sheath and a sudden gushing of darkish hematoma was noted (**Figure 3**). Scattered pigmented particles were removed using a tumor forceps. Using a nerve stimulator, motor function of the nerve rootlet was checked and one rootlet running upper and lateral to the mass was dissected. With further careful resection, decompression of the nerve rootlet was completed.

Histologic evaluation showed spindle or epithelioid cells with prominent nucleoli, heavy melanin pigmentation without nuclear pleomorphism, mitosis, and necrosis (**Figure 4**). Immunohistologic evaluation of the tumor cells yielded positive results for HMB-45 (melanocytic marker), S-100 protein, and Vimentin, and were negative for glial fibrillary acidic protein (**Figure 5**).

The radiating pain resolved dramatically after the operation. No symptoms or recurrence have been seen during a 1-year follow-up.

DISCUSSION

Intradural spinal melanocytoma involving a single nerve root (S-1) is an extremely rare case. Within the spinal canal, the usual location of a meningeal melanocytoma is the cervical region. It can be also found in the

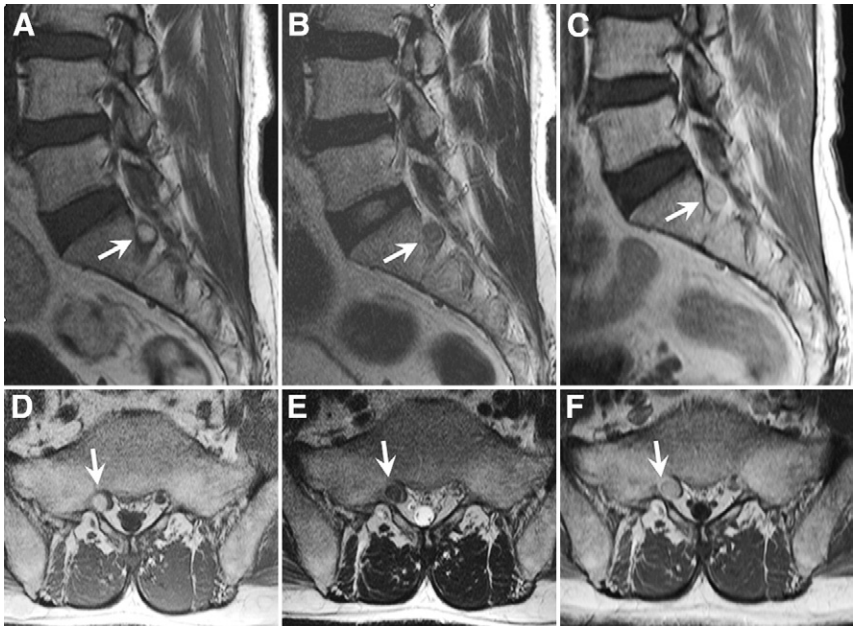

Figure 1. T1 (**A, D**), T2 (**B, E**), and contrast-enhanced T1 (**C, F**) sagittal and axial MRI show 1-cm-size mass (white arrow) around the right S-1 root.

lumbar region, either extradurally or intradurally. This tumor originates from the spinal roots. Orukaptan et al. (10) in 2000 reviews found 66 cases of meningeal melanocytoma, and only 36 were spinal meningeal melanocytoma. We found only one case report of spinal meningeal melanocytoma involving L-3 nerve root (4).

Gross meningeal melanocytoma appears as a localized, solitary, nodular lesion, with black or brown pigmentation, firmly attached to the dura (6). There could be small hemorrhages and superficial siderosis (3).

On CT scan, these tumors are iso- to high-density lesions that are enhanced by the addition of contrast media (9). MRI shows iso- to high intensity on T1-weighted images and low intensity on T2-weighted images, but this varies with the degree of melanization and hemorrhage (9). The tumor enhances uniformly with Gd injection and may show blooming of low signal related to melanin susceptibility effect.

In our case, a single nerve root was involved, which could confuse the diagnosis with that of intradural nerve root hematoma or melanotic schwannoma. Other differential diagnoses for melanocytic neoplasms of the central nervous system include benign melanocytomas, highly aggressive metastatic malignant melanomas, melanotic meningiomas, and melanotic ependymomas (2).

Spinal meningeal melanocytoma can be confused with intradural nerve root hematoma, which is also a rare condition. MRI of intradural hematoma show hyperintense on T1- and T2-weighted images, with variable contrast enhancement (12).

Schwannomas tend to have signal intensity equal to or less than that of the spinal cord on T1-weighted images and mild to marked hyperintensity on T2-weighted images (8). Focal areas of even greater hyperintensity on T2-weighted images often correspond to cystic portions, whereas hypointensity may represent hemorrhage, dense cellularity, or collagen deposition. Schwannomas enhanced very strongly, most of the time with an irregular appear-

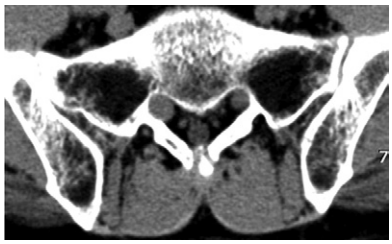

Figure 2. Axial CT reveals isodensity mass around the right S-1 root.

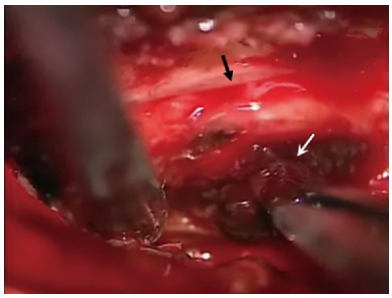

Figure 3. Intraoperative images show a coal-black pigmented mass (white arrow) next to right S-1 root (black arrow), which was attached to the dura.

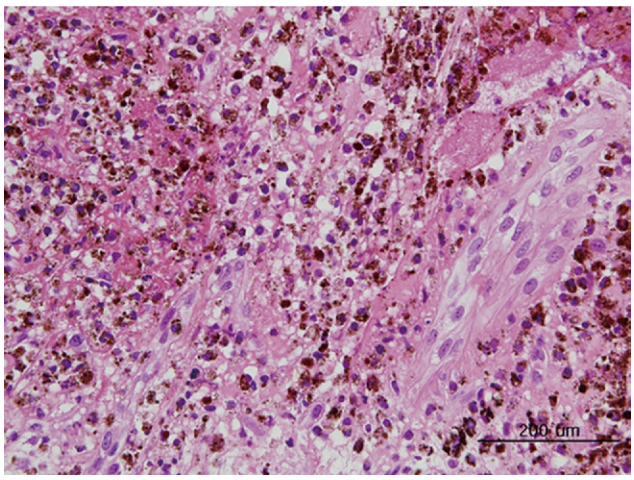

Figure 4. The microscopic examination (×40) shows spindle or epithelioid tumor cells containing variable amounts of melanin pigment on H&E (hematoxylin & eosin) stain.