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Exophytic Intramedullary Hemangioblastoma Presenting as an Extramedullary Mass on Myelography

Peter Corr, Thomas Dicker, and Michael Wright

Summary: Exophytic intramedullary tumors can look like extramedullary tumors on myelography, as in this case. Contrastenhanced sagittal and axial MR shows the true origin of these tumors.

Index terms: Hemangioblastoma; Medulla oblongata; Spinal cord, neoplasms

Common causes of well-defined benign intradural masses include nerve sheath tumors and meningiomas (1). We present a case of an exophytic intramedullary hemangioblastoma that mimicked an extramedullary mass on myelography.

Case Report

A 56-year-old man had a 12-month history of progressive lower limb pain and weakness. There was myelopathy with upper motor neuron signs in both lower limbs and a T-7 sensory level with sacral sparing and a lower thoracic scoliosis. Cerebrospinal fluid examination was normal except for a slightly elevated protein.

Myelography (Fig 1) showed a well-defined smooth mass within the posterior thoracic intradural space contiguous with a normal-caliber spinal cord. Sagittal and axial T1-weighted spin-echo magnetic resonance (MR) sequences before and after administration of gadopentetate dimeglumine and sagittal gradient-echo sequences were performed. A posterior intradural mass that was isointense with the spinal cord was noted on the unenhanced sequence (Fig 2A). A focal high-intensity cord signal contiguous with the intradural lesion was noted on the gradient-echo sequence (Fig 2B). The lesion as well as the adjacent cord enhanced intensely after injection of contrast material (Fig 2C and D). Postcontrast T1-weighted sequences of the brain and the rest of the cord were also performed and showed normal findings. The preoperative diagnosis was a meningioma or neurofibroma.

At laminectomy, a vascular tumor was noted in the intradural space arising from the spinal cord with a large encapsulated intramedullary component. The tumor in-

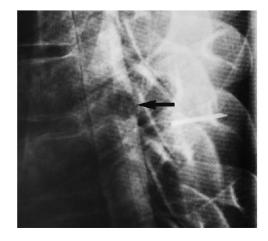


Fig 1. Myelogram shows a smooth posterior extramedullary mass with anterior displacement of the cord (*arrow*).

cluding the intramedullary component was successfully removed. Histology confirmed a hemangioblastoma.

Discussion

This case illustrates the importance of axial MR imaging in the radiologic evaluation of intradural tumors. We failed to appreciate that the enhancing focal cord signal was caused by the intramedullary component of the tumor and not by compressive myelomalacia from an intradural mass, as we initially thought. Exophytic intramedullary tumors rarely cause intradural masses (1).

Spinal hemangioblastoma is an uncommon tumor with a prevalence varying from 1.6% to 5.8% of all spinal cord tumors (2, 3). These tumors predominate in young males and are usually intramedullary in location. A review of 85 proved spinal hemangioblastomas found only eight tumors (9.4%) that were both in-

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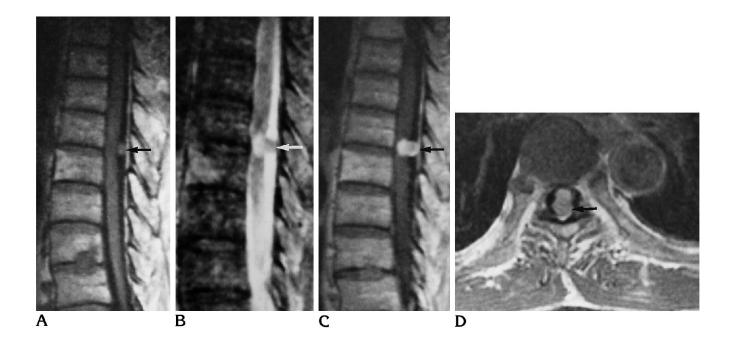


Fig 2. *A*, Sagittal T1-weighted MR image (500/30 [repetition time/echo time]) shows small dorsal extramedullary mass (*arrow*). *B*, Sagittal gradient-echo image (300/20, 20° flip angle) shows high-intensity cord signal (*arrow*). *C*, Sagittal and *D*, axial T1-weighted images after contrast administration show true exophytic nature of the intramedullary tumor (*arrow*).

tramedullary and extramedullary, as in our case (2).

Hemangioblastomas consist of thin-walled, closely packed blood vessels with interspersed large stromal cells (4). This accounts for their intense contrast enhancement after injection of gadopentetate dimeglumine, as seen in our case. Although not seen in our patient, the presence of enlarged dorsal draining pial veins, seen as serpinginous flow voids on MR, and associated intramedullary cysts can be a helpful pointer to the diagnosis (5).

Most spinal hemangioblastomas are solitary; however, in 20% of patients they are multiple and usually associated with von Hippel–Lindau disease (6). MR imaging of the brain, especially the posterior fossa and remainder of the cord, is recommended to exclude further lesions. Rarely, exophytic intramedullary tumors present as extramedullary masses on myelography; however, MR will help define their origin and extent.

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