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Case report

Paraganglioma of the cauda equina: MR and angiographic findings

Jose Carlos Méndez, PhD, MD[a,*], *Rodrigo Carrasco, PhD*[b], *Maria Antonia Prieto, PhD*[c], *Eduardo Fandiño, PhD*[a], *Javier Blázquez, PhD*[a]

[a] *Department of Radiology , Hospital Universitario Ramón y Cajal, Madrid, Spain*
[b] *Department of Neurosurgery, Hospital Universitario Ramón y Cajal, Madrid, Spain*
[c] *Department of Anesthesiology, Hospital Universitario Ramón y Cajal, Madrid, Spain*

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ABSTRACT

Paragangliomas of the cauda equina are rare benign highly vascular tumors and occur almost exclusively in the cauda equina and filum terminale of the spinal cord. We present a case spinal paraganglioma of the cauda equina in a 75-year-old male with an emphasis on magnetic resonance imaging and conventional angiography findings.

Case report

A 75-year-old male was admitted with gradual onset of intermittant pain and numbness on his lower extremities with neurogenic claudication and urinary incontinence.

Magnetic resonance (MR) imaging demonstrated a large intradural extramedullary well-encapsulated tumor extending from L3 to L4 compressing the roots of the cauda equina, measured about 5 cm in craniocaudal diameter.

The tumor was isointense on T1-weighted sequences and T2-weighted sequences relative to the spinal cord (Figs. 1A and B). A hypointense rim was seen on T2-weighted images, attributed to the paramagnetic effect of hemosiderin or ferritin from previous hemorrhages; there were serpiginous structures of signal void around the mass on all sequences (Fig. 1B).

It presented marked contrast enhancement after gadolinium administration, with hypointense areas in both the upper and lower portions of the tumor (Fig. 1C). Perfusion-weighted MR images showed increased cerebral blood flow, consistent with hypervascular tumor (Fig. 1D).

A preoperative conventional angiography was planned to identify major arterial feeders, facilitate surgical removal, and reduce the operative risk.

Spinal angiography through the left T9 intercostal artery demonstrated a hypervascular lesion supplied by the anterior spinal artery of Adamkiewicz (Fig. 2). The signal void structures in MR were well corresponded with the feeding artery

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* Corresponding author.
E-mail address: josecarlos.mendez@salud.madrid.org (J.C. Méndez).

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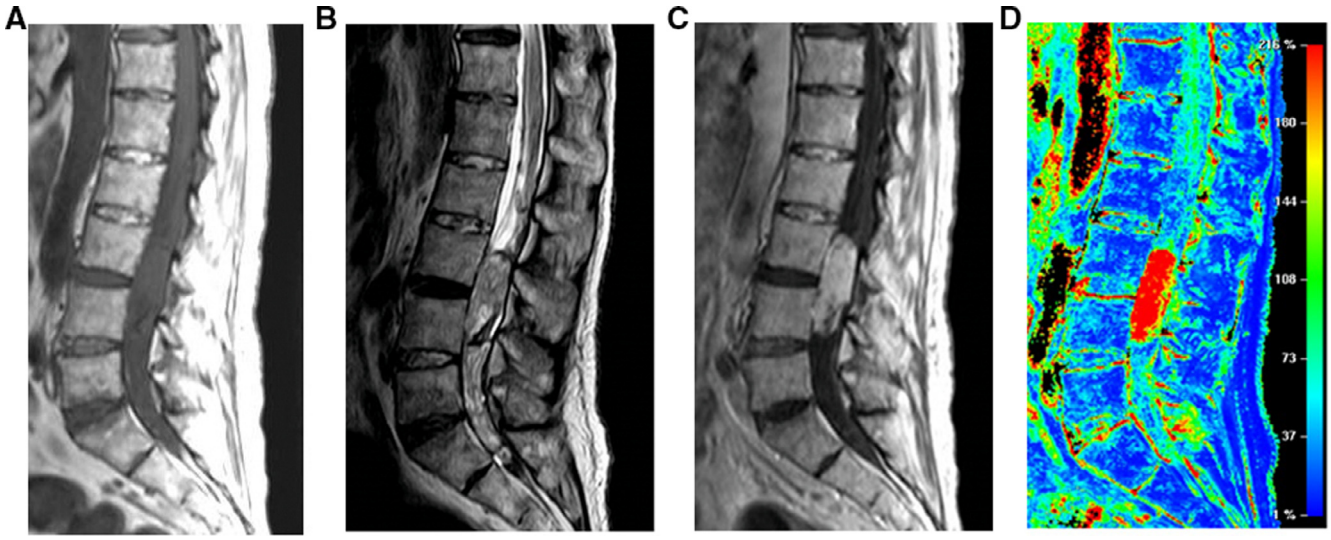

Fig. 1 – Paraganglioma of the cauda equina in a 75-year-old male. Sagittal T1-weighted (A), T2-weighted (B), gadolinium-enhanced T1-weighted (C) and, perfusion-weighted (D) magnetic resonance images revealing a large homogeneously isointense intradural lesion extending from L3 to L4, with heterogeneous enhancement after gadolinium. T2W imaging revealing a hypointense rim at the superior and inferior aspect of the lesión and flow voids cranial to the mass indicative of venous congestion or high vascularity of the tumor (B). Perfusion-weighted images showed increased rCBF in the lesión, suggesting high vascularization (D).

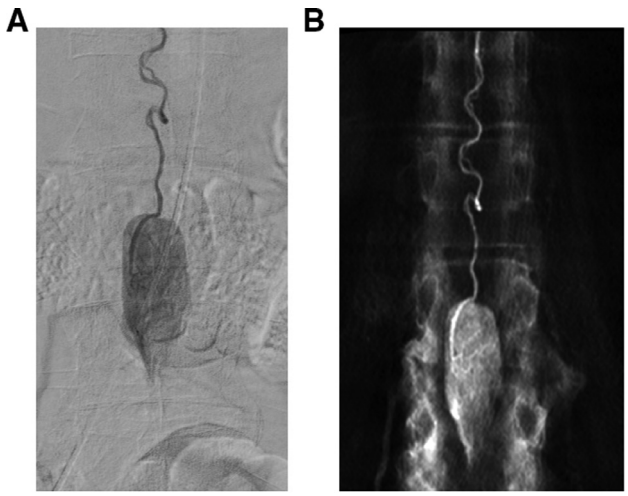

Fig. 2 – Selective spinal angiography via the left T12 intercostal artery. Conventional angiographic image (A), and CT reconstruction of 3D rotational angiography (B). Angiography revealed a highly vascular mass supplied by the anterior spinal artery (artery of Adamkiewicz). The lesión presented well defined margins and tumor stain in the late angiographic phase, like the appearance of a "silk cocoon."

on angiography. The lesion presented an intense early blush that persisted well into the arterial and venous phases, presenting a "silk cocoon" appearance in the late angiographic phase (Fig. 2A).

At surgery, a highly vascular well-circumscribed pinkish-gray tumor measuring 5.1 × 1.6 × 1.4 cm in size, was encountered. Several arterial feeders and arterialized draining veins, were also observed.

After coagulation and cutting of these vessels, the lesión was completely excised. Histopathologically the tumor had the typical appearance of a PG. The patient had an uncomplicated recovery and was symptom-free at 24 months.

Discussion

Spinal PG are extremely rare neuroendocrine tumors of the extra-adrenal paraganglionic system. They comprise 3%-4% of all spinal tumors, and about 200 cases of cauda equina and filum terminale PG have been reported in the literature. The first description of a PG of the filum terminale was published in 1970 by Miller and Torack [1]. Gelabert-Gonzalez published the largest review in the literature on cauda equina PG [2].

These lesions are most frequently located in the intradural extramedullary compartment and have a high affinity for the cauda equina or filum terminale.

These lesions are commonly encountered in the fifth and fourth decades of life with male predominance [3,4].

Patients typically present with clinical signs and symptoms referable to a lesion in the cauda equina, manifested by lower lumbar pain, sensory or motor loss of the lower extremities, and bowel and bladder dysfunction [3].

The MR imaging findings are generally nonspecific with the tumor relatively isointense with spinal cord on T1-weighted images and hyperintense on T2-weighted images, with intense heterogeneous gadolinium contrast enhancement [3–5].

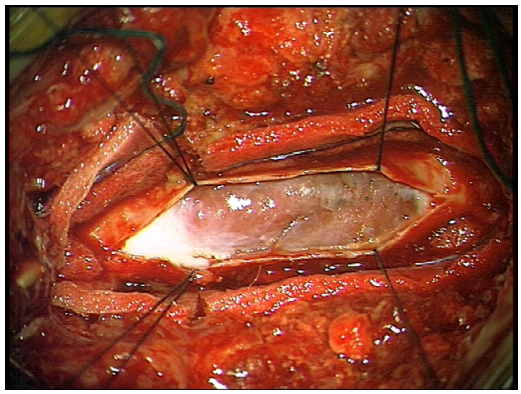

Fig. 3 – Intraoperative picture of the lesion, after durotomy (cranial to caudal from right to left). Hypervascular intradural extramedullary and well-marginated mass was observed.

The differential diagnosis of spinal PG includes nerve sheath tumor, meningioma, metastases, and myxopapillary ependymoma. Differentiation of PG from these tumors is frequently not possible on MR because of considerable overlap in their imaging findings. However, it should be appreciated that PG are vascular tumors and identification of features reflecting this quality is crucial [5,6]. On T2-weighted sequences intra- and peritumoral flow-voids and a salt-and-pepper appearance may indicate hypervascularity [7,8]. In addition, hypointense tumor margins on T2-weighted images, suggesting paramagnetic effects from hemosiderin, may also be seen.

Woo et al described "the polar sign" in T1 constrast-enchanced MR and T2-weighted MR images, representing subacute to chronic intratumoral hematomas within the lesion's superior and inferior poles [7] (Fig. 3).

Spinal digital subtraction angiography has been the gold standard in the evaluation of intraspinal tumor vascularity which is essential to the successful resection of potentially curable lesions and to perform preoperative embolization in selected cases [9]. Angiography reveals a well-defined hypervascular lesion with intense early blush that persists well into the arterial and venous phases ("silk cocoon appearance"), helping to presurgical planning and to differentiate PG from other tumors.

To the best of our knowledge, there are only four previous cases of primary spinal PG in the literature demonstrated by spinal angiography [10–13].

As seen in our case, spinal angiography demonstrates a good correlation with MR findings, and can be a very helpful procedure in the differentiation between primary spinal PG and other neoplasms of the *cauda equina region*.

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