

Acute Paraplegia Revealing a Hemorrhagic Cauda Equina Paraganglioma

Abstract

Cauda equina paragangliomas are rare neuroendocrine benign and slow-growing tumors. Acute paraplegia, occurring because of sudden intratumoral hemorrhage, represents an extremely rare clinical picture of this disease. We report the case of a 64-year-old male presenting with a 5-day acute lower back pain, sciatica, and leg weakness. Spinal imaging showed an intradural mass of the cauda equina region and an emergent surgical treatment was achieved. The lesion was removed “*en bloc*,” and subarachnoid blood was noticed during surgery. The postoperative course was uneventful, with complete regression of pain and progressive motor recovery. The histological study revealed typical microscopic and immunohistochemical features of paragangliomas.

Keywords: Acute, cauda equina, paraganglioma, paraplegia

Introduction

Paraganglioma (PGL) is a rare neuroendocrine benign tumor. It derives from neuroepithelial cells' group called paraganglia, which migrate from the neural crest.^[1] Cauda equina PGLs (CEP) are rare, accounting for 3.5%–4% of all tumors in this region.^[2] Usually presenting as a slow-growing tumor, fast neurological impairments are uncommon and only five cases have been reported to our knowledge. We report an unusual case of hemorrhagic PGL of the cauda equina revealed by acute paraplegia.

Case Report

A 64-year-old male presented with intense lower back pain, bilateral S1 sciatica, and lower limbs' weakness evolving acutely for 5 days. He has no medical history. At examination, the patient was suffering from unbearable lumbar pain requiring morphine, he was confined to a wheelchair, and he had a flaccid paraplegia with abolished patellar and Achilles reflexes bilaterally. There was neither sensory impairment nor sphincter disorder.

A lumbar computed tomography (CT) scan was performed first, because of its availability and showed a spontaneous hyperdensity inside the spinal canal at the L2 and L3 levels, with calcifications. The magnetic resonance imaging (MRI) was performed as soon as possible, revealing

an intradural extramedullary lesion at the L2 and upper of L3 levels [Figure 1]. The quality of images was altered because the patient could not lie in prone position for a long while. The T2-weighted images (T2WI) demonstrated a hypointense oval-shaped mass facing the body of L2 and the L2–L3 disc. On the T1-weighted images (T1WI), the tumor was isointense with heterogeneous contrast enhancement. Evoked diagnoses were ependymoma, neurinoma, and hemangioblastoma.

Urgent surgery has been performed in this patient, consisting of an L2 and L3 laminectomy then straight midline dural opening. The tumor was located deep in the cauda equina roots [Figure 2]. It was a well-encapsulated dark red and glossy lesion covered by nerve roots, with superficial calcifications. There was no adherence to the dura. Xanthochromic cerebrospinal fluid was encountered. After a circumferential sharp dissection from the nerve roots, the tumor was found firmly attached to the filum terminale. This attachment was coagulated and cut, and then the tumor was totally removed “*en bloc*.”

After surgery, the pain subsided completely and the patient gradually recovered a normal mobility. The pathological studies concluded to a PGL [Figure 3].

Discussion

The first case of cauda equina PGL was described in 1972 by Lerman *et al.*, while

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an earlier case was misdiagnosed as a “non-secretory ependymoma of the filum terminale.”^{3,4]}

PGLs of the central nervous system are uncommon. Most of them are spinal PGLs, especially in the cauda equina. CEP represents 3.5%–4% of all neoplasms in this region. The tumor may dwell either on a nerve root or on the filum terminale.^{1,2]} CEPs are predominantly seen in men in the fourth to fifth decade. Symptoms are dominated by lower back pain and sciatica, while motor and sensory deficits are rare and sphincter and erectile dysfunction very exceptional. Commonly, diagnosis is delayed by more than 1 year after presentation (up to 15 years), which reflects the nonspecific symptoms and the benign nature of the tumor. Cauda equina syndrome is uncommon although the tumor may occupy the entire spinal canal diameter.^{5]} Systemic manifestations due to the excess of catecholamine releasing, as in pheochromocytoma, are exceptional in CEP.^{2]}

PGLs are known to be highly vascularized tumors.^{2,5]} To our knowledge, acute paraplegia as a result of hemorrhage from a CEP has been reported in only five cases, as resumed

in Table 1.^{5-9]} However, it is useful to notice that bleeding from a CEP can be provoked by iatrogenic maneuver such as epidural steroid injection^{7]} or in the postoperative course after PGL removal,^{10]} thus causing sudden paraplegia. Moreover, repeated subarachnoid hemorrhage has been reported,^{11]} even with consequent superficial siderosis of the CNS, due to chronic deposition of hemosiderin. It has to be noted, otherwise, that calcification is an extremely rare finding in PGL,^{12]} and this feature was observed in our patient.

CT scan of the lumbar region may show the intracanal spinal lesion, but MRI is the gold standard for diagnosis. Typically, the CEP presents as an oval-shaped intradural mass, well circumscribed, in isosignal on T1WI, hypersignal in T2WI and showing a more-or-less homogenous marked enhancement after gadolinium injection.^{2]} A hypointense tumor rim on T2WI, called the “cap sign,” indicates hemosiderin deposition from previous hemorrhage.^{12]} Furthermore, the high vascularity of PGLs is supported by the presence of serpiginous flow void structure overhanging the upper pole of the tumor or all around it, consisting



Figure 1: Sagittal magnetic resonance imaging of the lumbar spine showing an intradural tumor, isointense on T1-weighted images with peripheral contrast enhancement (a) and hypo-intense on T2-weighted images (b)

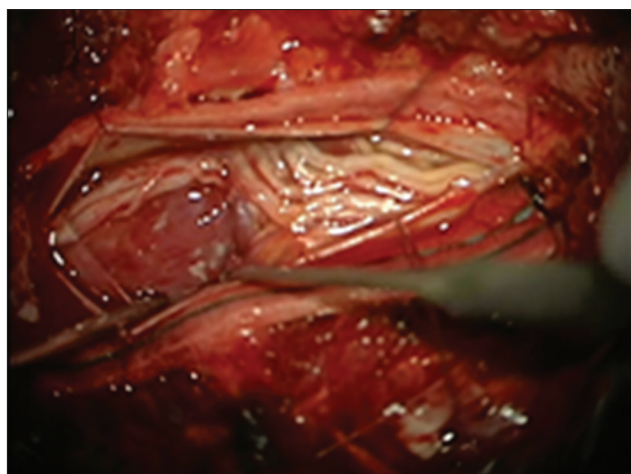


Figure 2: Intraoperative picture: The tumor is intradural, reddish and covered by the cauda equina roots

Table 1: Cases review of hemorrhagic paragangliomas of the cauda equina revealed by acute paraplegia

Case	Age (years) sex	Symptoms	Duration	Evidence of hemorrhage	Treatment	Evolution
Ma <i>et al.</i> , 2012 ^{6]}	51 male	Lumbosciatica and acute paraplegia	6 months	MRI, CT scan, intraoperative	Laminectomy and GTR	Partial recovery
Pikis <i>et al.</i> , 2013 ^{7]}	37 male	Lower back pain and urine retention	1 year after steroid injection	MRI	Laminectomy and GTR	Full recovery
Woo <i>et al.</i> , 2014 ^{8]}	60 male	Lower back pain, acute lower limb weakness and urine retention	3 months	MRI, intraoperative	Laminectomy and GTR	Full recovery
Nagarjun <i>et al.</i> , 2016 ^{9]}	36 female	Acute fl accid paraparesis, urine retention and sensory loss	4 days	MRI, intraoperative	Laminectomy and GTR	Partial recovery
Walsh <i>et al.</i> , 2016 ^{5]}	47 male	Acute left lower limb weakness and urinary retention	3 days	MRI	Laminectomy and GTR	Full recovery
Our case	64 male	Lumbosciatica and acute paraplegia	5 days	MRI, CT scan, intraoperative	Laminectomy and GTR	Full recovery

MRI – Magnetic resonance imaging; CT – Computed tomography; GTR – Gross total resection

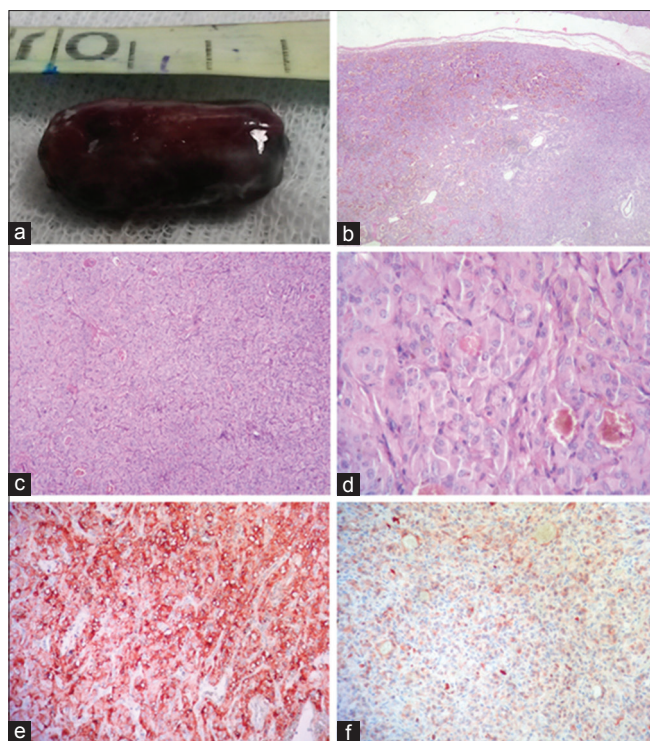


Figure 3: Encapsulated oval-shaped tumor (a). Tumor proliferation arranged in anastomosed trabecular structures, surrounded by a thin capsule (b: H and E, ×4, c: H and E ×10). polygonal cells with round nuclei and granular cytoplasm, gathered in clusters: The Zellballen pattern (d: H and E, ×40). Mitotic figures are rare. The tumoral stroma's type is endocrine with a capillary network. Some hemorrhage foci are noticed. Immunohistochemical staining showed the expression of synaptophysin by tumor cells (e) and positivity of S100 protein in sustentacular cells (f)

of dilated vessels.^[12] These radiological features are nonspecific of the PGL and differential diagnoses are still possible: ependymoma (myxopapillary type), neurinoma, meningioma, and hemangioblastoma.

Intraoperatively, CEPs are usually soft, reddish, well-encapsulated, and well-circumscribed masses and have a vascular pedicle. Total removal should be the goal of surgery. Whenever possible, the tumor should be resected in one piece when manipulating the tumor because it can trigger a hypertensive crisis.^[4]

Typically, the histopathological findings are the so-called “Zellballen” (nests of small polyhedral cells) and trabecular cords of cells, closely circumscribed by a richly vascular stromal network. The predominant cell type is the chief cell, which has an abundant eosinophilic and granular cytoplasm, containing biogenic amines; the second cell type is the sustentacular or supporting cell, which is spindle shaped.^[13] The loss of normal paraganglionic architecture and the paucity or absence of sustentacular cell have been associated with more aggressive or malignant behavior.^[13] Immunohistochemical staining is the key to diagnosis. The chief cells are positive for neuron-specific enolase, chromogranin, and synaptophysin. Tumor cells are

negative for epithelial antigen membrane and glial fibrillary acidic protein (GFAP). The characteristic finding is that sustentacular cells are sensitively and reliably expressed by S100 protein.^[10] The main differential diagnosis consists of ependymoma, showing positivity for GFAP and mucin with a different pattern for S100 protein (positive in tumor cells).^[7,10]

Gross total resection represents the curative treatment for these tumors, and no adjuvant treatment is needed. When a subtotal resection is performed for local invasion, local recurrence and even distant metastatic spread may be observed.^[1] Tumor recurrence may occur lately, many years after surgery, this justifies a long-term follow-up. Postoperative radiotherapy can be considered for incompletely resected tumors, but resistance is possible.

Conclusion

PGLs of the cauda equina region are very rare. They are benign neuroendocrine slow-growing tumors, and they present mostly with chronic lower back pain and sciatica. Acute paraplegia is an extremely rare clinical manifestation of cauda equina PGL and reflects the hypervascular nature of this tumor. Surgical total removal is the only treatment of such lesions, and the histological examination makes the diagnosis. The prognosis is excellent if total resection is achieved.

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The patient is willing to have his case enrolled and his data published. Also, our institute approves this study and publication.

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Conflicts of interest

There are no conflicts of interest.

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