

Case Report

Paranglioma of the cauda equina region: A report of three cases

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Abstract

Background: Cauda equina paragangliomas (CEP) are rare tumors. Low back pain and sciatica are the main presenting symptoms. Magnetic resonance imaging (MRI) is the study of choice and treatment consists of total excision when feasible. Definitive diagnosis can only be made after immunohistochemical investigation. CEP is classified as grade I WHO and after total removal the prognosis is excellent. Nonetheless, after subtotal removal, tumor recurrence can occur.

Case Description: We present 3 cases of CEP, preoperatively diagnosed as an intradural mass on MRI and suspected to be ependymoma. All 3 patients presented with low back pain and variable sciatic pain. Total resection of the tumor was performed after which all patients fully recovered. There is no recurrence after 13, 11, and 5 years, respectively.

Conclusion: CEP is a rare tumor. We diagnosed 3 paragangliomas out of a series of 105 intradural extramedullary tumors in adults (1994–2005). No recurrence was seen after total resection. In retrospect, both the intraoperative appearance and the MR image were not completely typical for schwannoma or ependymoma, but final diagnosis can only be made histologically.

Key Words: Cauda equina, paraganglioma, spine

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INTRODUCTION

Extra-adrenal paragangliomas are rare neuroendocrine tumors, which can occur throughout the body. Parangliomas in the central nervous system occur in the carotid body, glomus jugulare, and the region of the cauda equina. Cauda equina paragangliomas (CEP) are rare tumors. The first case was, in retrospect, described in 1970.^[6] The origin of CEP is uncertain since the existence of paraganglia cells in the central nervous system remains

a point of debate.^[5] Histologically, CEP are benign tumors, which usually originate from the filum terminale.^[8] Low back pain is the main presenting symptom with often sciatica as well.^[1,3,5,8] Magnetic resonance imaging (MRI) is the study of choice and treatment consists of total excision when feasible. Definitive diagnosis can only be made after histological and immunohistochemical investigation. CEP are classified as grade I according to the World Health Organisation (WHO) and the prognosis is excellent. Nonetheless, tumor recurrence

after subtotal removal can occur. We diagnosed 3 CEP on a total of 105 intradural extramedullary tumors between 1994 and 2005 [Table 1], preoperatively diagnosed as an intradural mass on MRI.

CASE REPORTS

Case 1

A 37-year-old man presented with low back pain and bilateral sciatica without neurologic deficits. MRI showed a contrast enhancing intradural mass at L3–L4 with a diameter of 1.5 cm. Total resection of this vascular tumor was performed after which the patient made a full recovery. Histologic diagnosis confirmed the tumor to be a paraganglioma and there is no recurrence after 11 years.

Case 2

A 41-year-old man presented with low back and right-sided leg pain. Neurologic examination only revealed sensory loss in the right leg without motor deficits. MRI showed an intradural mass at L2–L3, roughly circular, with a diameter of 2 cm and with strong but inhomogeneous gadolinium enhancement [Figure 1]. At surgery, the highly vascular tumor was attached to the filum terminale and only minimal arachnoid adhesions with the nerve roots was seen. After total resection the patient made a full recovery. Histologic diagnosis was paraganglioma. Staging with MRI of the whole spinal cord, the brain and abdominal computed tomography (CT) scan was negative. There is no recurrence after 13 years.

Case 3

A 51-year-old man presented with low back pain with bilateral irradiation to the gluteal region. No neurologic deficits were present. MRI showed an intradural mass at L4, with a diameter of 1.5 cm, circular with somewhat irregular margins, moderate enhancement, and clearly located centrally in the dural sac [Figure 2]. A total resection of the tumor was performed [Figures 3 and 4]. Staging (MRI of the whole spinal cord, the brain, and abdominal CT scan) was negative. The patient fully recovered and there is no recurrence after 5 years.

Table 1: Intradural extramedullary tumors (1994–2005)

Tumor type	Number
Schwannoma/neurinoma	45
Meningioma	35
Ependymoma	13
Chordoma	3
Dermoid cyst	3
Enterogenous cyst	2
Cauda equina paragangliomas	3
Malignant neurinoma	1
Total	105

DISCUSSION

CEP is a rare tumor. The first case was described in 1970 as a “secretory ependymoma of the filum terminale.” In retrospect, this tumor proved to be a CEP.^[6] Since then some 220 cases have been reported. The origin of CEP is uncertain since the existence of paraganglia cells in the central nervous system remains a point of debate. In general, paragangliomas are neuroendocrine tumors that arise from neuroepithelial cell groups, called paraganglia. Histologically, these tumors are benign and characterized by the presence of abundant neurosecretory granules in their cytoplasm. The primary histological features of paragangliomas are the presence of “zellballen,” or a nesting of cell groups, and trabecular cords of cells within thin fibrovascular stroma.^[1,7,8] CEP are solid, highly vascularized, slow-growing, and well-encapsulated tumors, which usually originate from the proximal filum terminale, rather than the cauda equina nerve roots.^[1,2,8] Immunohistochemically, Glial fibrillary acidic protein (GFAP) staining is negative while Neuron Specific Enolase (NSE), synaptophysin, and chromogranin staining is positive. S-100 protein typically decorates the sustentacular cells.^[7,8] CEP represent 3%–4% of the lesions in this region and are commonly confused with ependymoma and schwannoma.

As was also the case in our small series, there is a male predominance. Low back pain is the main symptom in 50%–87% of patients, with sciatica in 20%–74%.^[1,3,8] The incidence of sensory and motor deficits, signs of intracranial hypertension, and sphincter dysfunction have been reported to be low.^[3] Despite their neuroendocrine origin, these tumors only rarely have functional hormonal activity.^[1,3-5,8] For diagnosis, MRI is the imaging study of choice and the tumor is usually hypo- or isointense to the conus medullaris on T1-weighted images, whereas it is hyperintense on T2-weighted images. CEP may



Figure 1: MRI (sagittal T1-weighted image with gadolinium) of patient #2 showing an intradural mass at L2–L3

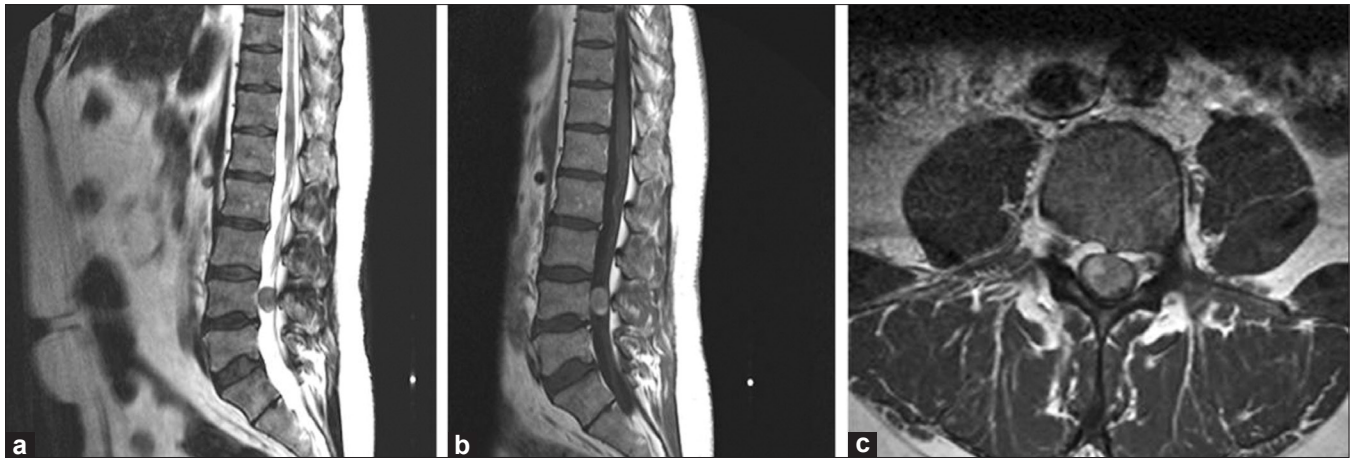


Figure 2: MRI of patient #3 showing an intradural mass at L4. (a) Sagittal T2-weighted image, (b) sagittal T1-weighted image with gadolinium, (c) axial T1-weighted image with gadolinium

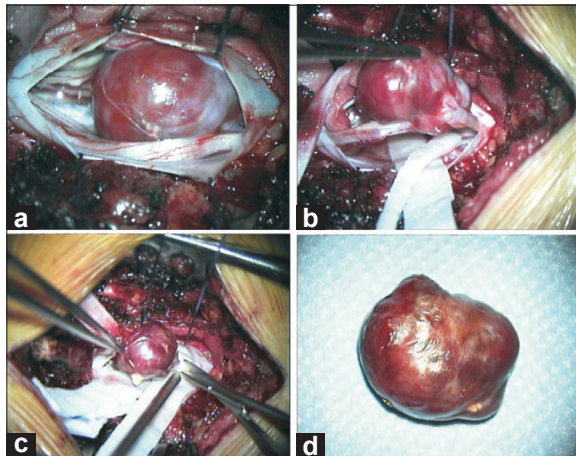


Figure 3: Intraoperative images of cauda equina paragangliomas (patient #3). (a) cauda equina paragangliomas after opening of dural sac; (b) cauda equina paragangliomas; (c) removal of cauda equina paragangliomas; and (d) cauda equina paragangliomas after resection

have cystic areas and after Gadolinium injection, there is a marked enhancement.^[8] On imaging studies, differential diagnosis with ependymoma, schwannoma, and even meningioma can be difficult. Compared to ependymomas, enhancement seems to be stronger due to the more vascular nature and in our patients, the tumors were rather circular instead of ovoid, and generally smaller at the time of diagnosis when most ependymomas extend over several vertebral levels. Compared to schwannomas, the central location and the absence of a dumbbell appearance may give a clue to the diagnosis. However, these characteristics are only relative and it is rare that diagnosis is made before surgery. Definitive diagnosis can only be made after immunohistochemical investigation.

Cauda equina paragangliomas present during operation as a soft, dark red, and well-circumscribed mass, attached

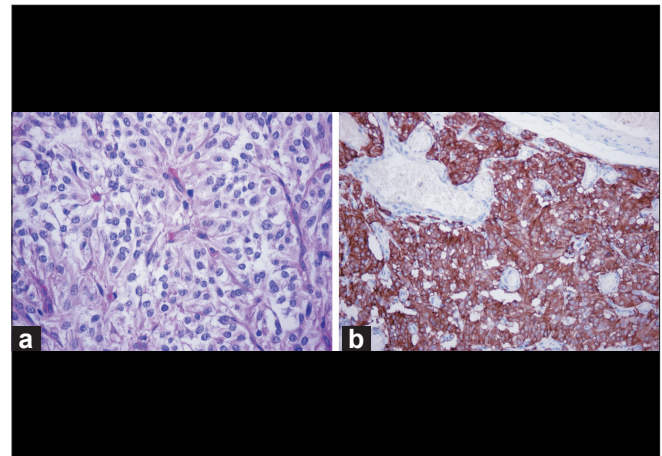


Figure 4: Histology. (a) HandE stain, showing the typical nesting (zellballen) pattern of the large polygonal cells, (b) strong synaptophysin expression in the tumor cells

to the filum terminale. Endocrine and vascular symptoms due to sudden catecholamine release during removal are rare compared to surgery for cranial paragangliomas.^[4] Total removal of the tumor should be the goal of surgery.^[1,3,8] However, cases have been described where local invasion or adhesion to nerve roots made a subtotal resection unavoidable. Complete surgical resection is considered curative and CEP is classified as grade I according to the WHO with an excellent prognosis. Nonetheless, recurrence can occur after subtotal removal. Although some authors suggest postoperative radiotherapy in case of subtotal resection, the effect of radiotherapy on recurrence prevention has not conclusively been demonstrated.

We diagnosed 3 CEP on a total of 105 intradural extramedullary tumors between 1994 and 2005. All the 3 cases presented with low back pain, as most often

reported in the literature. No recurrence was seen after total resection in these 3 patients. In retrospect, MRI was not completely typical for schwannoma or ependymoma, but final diagnosis can only be made histologically.

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