

Paraganglioma of cauda equina

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ABSTRACT

Spinal paragangliomas are rare benign tumors. Preoperative diagnosis is difficult, as they share common clinical and radiological features with other lesions at this site. We report an unusual case of paraganglioma in the cauda equina region in a 60-year-female. Ventricular ectopics were noted intraoperatively during surgical removal. Characteristic histology led to definitive diagnosis.

Keywords: Cauda equina, paraganglioma, ventricular ectopic

INTRODUCTION

Extra-adrenal paragangliomas are usually benign neoplasms, arising from specialized neural crest cells associated with paraganglia.^[1,2] Most common locations are carotid body and glomus jugulare tumors. Other locations that have been described include retroperitoneum, mediastinum, and gastrointestinal and respiratory tract.^[3] Spinal localization is uncommon with an incidence in general population calculated at 0.07 per 100,000 inhabitants.^[2,4] Clinical presentation is similar to other cauda equina lesions and shown to have no specific MRI findings.^[2,3] We report another case and discuss clinical, radiological and histological features in light of published data.

CASE REPORT

A 60-year-old woman presented with a 1-year history of low backache that radiated to right leg and worsened with movement. Evaluation revealed tenderness in lumbosacral region with the straight raise test of right leg producing pain. Sphincter dysfunction was absent. The neurologic examination was otherwise unremarkable. Lateral radiograph showed anterior marginal osteophytes in L1-L5.

MRI revealed a 2 cm X 1.5 cm well-circumscribed intradural tumor, isointense with spinal cord, below the conus at L3-L4 level on T1-weighted images [Figure 1]. A standard L3-L4 laminectomy was performed to gain access. The intradural, dark red, firm, capsulated tumor found attached to nerve roots was removed en bloc. Ventricular ectopics on lead III were observed intraoperatively on electrocardiogram. Histopathological study demonstrated the characteristic “zellballen” pattern of tumor cells associated with a delicate fibrovascular stroma [Figure 2], diagnostic of paraganglioma. The patient made an excellent postoperative recovery and has remained symptom free to date.

DISCUSSION

Intradural extramedullary lesions, by convention, include cauda equina masses. Possible pathologies at this site include nerve sheath tumors, meningiomas, primary CNS tumors like astrocytomas, ependymomas, medulloblastomas, germinomas, dural tumors like melanoma, solitary fibrous tumors, angioma, secondaries from lung, breast and melanoma, and tumor-like masses, which include epidermoid cyst, parasitic cyst and granulomatous lesions.^[5]

The first published case of spinal paraganglioma was in 1970 by Muller and Torack, although they denominated it as a secretory ependymoma.^[1,3,4] Clinically, cauda equina syndrome is unusual, despite the tumor taking up the whole diameter of the spinal canal. Sensory/ motor deficits and incontinence are relatively rare.^[4] No preoperative signs and symptoms of catecholamine hypersecretion have been reported as in the present case. Hence, blood tests to measure the levels of hormones were not deemed necessary.

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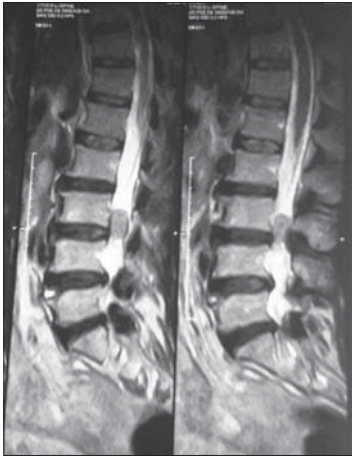


Figure 1: Sagittal T1-weighted image showing a well-delineated mass at L3-L4

Paragangliomas in spinal canal represent a distinct subtype of extra-adrenal paraganglioma. They are exceptional in being sympathetic rather than parasympathetic, in contrast to other extra-adrenal paragangliomas.^[2] Release of biogenic amines resulting from tumor manipulation explains the observation of ventricular ectopics intraoperatively. Intraoperative hypertensive crisis has also been described in previous reviews.^[1]

Paraganglionic tissue is not normally found in this region. The tumor being adherent to nerve roots as in our case conforms with the proposed theory that they arise from sympathetic neurons in thoracic and lumbar horns of the spinal cord, sending their axons to sympathetic trunk through communicating branches, or arise from heterotopic neurons lying along the branches.^[2,4,6]

MRI features, though shared by other intradural lesions, may be helpful, as serpentine dilatation of vessels between conus and lesion and hypointense peripheral rim are uncommon in other lesions.^[5] Functional imaging by scintigraphy with I-MIBG has drawback of relatively high levels of irradiation exposure.^[4]

Histologically, they are usually well encapsulated and show characteristic “zellballen” or nesting of large cells, circumscribed by vascular stroma, as observed in our case. Variants like gangliocytic, oncocytic and melanocytic have been described.^[1]

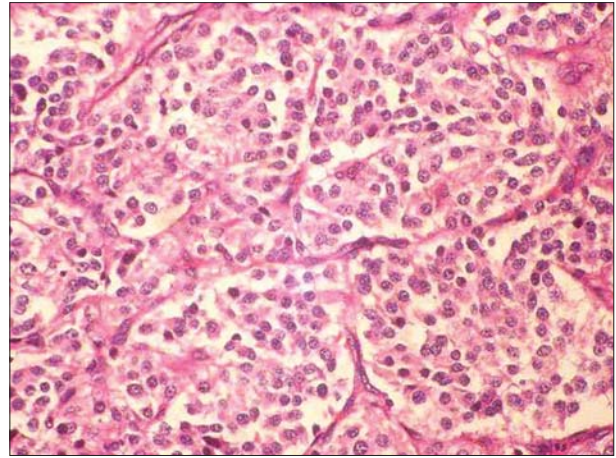


Figure 2: Histomorphology of paraganglioma showing characteristic “zellballen” pattern (H&E, 40x)

CONCLUSION

Sufficient reports of this lesion warrant its inclusion in more comprehensive differential diagnosis of intradural spinal lesion. Features of catecholamine secretion, even though rare, along with typical MRI features help distinguish spinal paraganglioma from other lesions. Histology provides the definitive diagnosis.

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