An Unusual Case of Conus Nonfunctioning Paraganglioma

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ABSTRACT

Paragangliomas are neuroendocrine tumors with differentiation of chromaffin cells originated from neural crests. Paragangliomas most commonly occur in the head and neck regions (more than 90% of cases). Spinal paragangliomas are uncommon and difficult to diagnose preoperatively due to the absence of any specific radiological and clinical findings.

A 39-year male patient presented with severe back pain with inability to lie down for 2 days. Neurological examination revealed paraparesis with brisk lower limb reflexes. His magnetic resonance imaging (MRI) lumbosacral spine suggestive of homogeneously contrast enhancing intradural mass confined to L1 to L2 level. The patient underwent laminectomy with excision of the highly vascular intradural lesion. Histopathological examination confirmed as paraganglioma.

Lumbar spinal paragangliomas are diagnostic dilemma not only due to the rarity of these lesions, but also due to radiological features mimicking other common lesions at this location like ependymoma and schwannomas. Characteristic features on T2-weighted MRI and gradient echo imaging can help in preoperative diagnosis, though these features are not specific of these lesions. Preoperative consideration of these lesions helps in taking precautions during excision as these tumors can be functioning paragangliomas.

Keywords: Chromaffin cells, Neuroendocrine tumors, Paraganglioma.

INTRODUCTION

Paragangliomas are neuroendocrine tumors with differentiation of chromaffin cells originated from neural crests. Carotid and glomus jugulare tumors constitute >90% of extra-adrenal paragangliomas. Spinal paragangliomas are uncommon. Usually confined to the cauda equina and filum terminale. They are commonly encountered in adolescents and adults, with the peak incidence in the fifth decade. These tumors typically present with mononeuropathy or sometimes paraparesis.

CASE REPORT

A 39-year male patient was admitted to the hospital with low back pain for two and a half year. The patient has developed neurogenic claudication since 4 weeks, which was progressive. The patient had severe back pain for 2 days, which was aggravated after lying down. He was more comfortable in sitting position. There was no sensory abnormality on neurological examination. Tone in upper limb was normal while there was hypertonia in lower limbs. Power was medical research council grade 5 in upper limb and grade 4/5 in both lower limbs. Knee and ankle reflexes were brisk in both lower limbs and plantar responses were extensor. Perianal sensation and anal tone was normal. The straight leg elevation test was negative on both sides.

Magnetic resonance imaging scan of the lumbosacral region revealed 3.5 × 2 cm sized lesion at L1 to L2 level, which was homogeneous isointense on T1-weighted images and heterogeneous hyperintense on T2-weighted images. Serpiginous flow voids, like arteriovenous malformation, were noted in the upper margin of the tumor and it was very much prominent on T2-weighted images (Fig. 1). The lesion was intradural in location with homogeneous enhancement on intravenous gadolinium injection (Fig. 2).

The patient underwent L1 and L2 laminectomy. On dural opening, well-circumscribed, pinkish brown and

**Fig. 1:** Sagittal MRI revealed 3.5 × 2 cm sized lesion at L1 to L2 level serpiginous flow voids, in the upper margin of the tumor prominent on T2-weighted images.
firm lesion was noted. It was easily separated from the surrounding roots. Major arterial feeder supplying the tumor was disconnected and the lesion was separated from its originating nerve root. Gross total resection of the tumor achieved, which was about 3.5 × 2 cm in size (Fig. 3).

Histopathological examination of the specimen showed dilated vessels on its surface, Gamma-Gandy bodies and inflammation. Hemosiderin pigmentation was detected in the pseudocapsule (Fig. 4). No intratumoral cyst was seen despite the large size. Immunohistochemistry study of the tumor tissue revealed positivity for synaptophysin, chromogranin, S100 was focally positive and Ki 67 was low.

The patient made a good postoperative recovery. There have been no signs of recurrence after 2 years follow-up.

DISCUSSION

Paragangliomas are neuroectodermal tumors of the autonomic paraganglia, derived from the chromaffin cells of neural crest origin. The first authors to describe this pathological entity were Miller and Torack in 1970, denominating it a secretory ependymoma, whereas Lerman was the first to coin the term paraganglioma in 1972.2 Their location within the central nervous system is unusual.

Spinal localization of these tumors is unusual; the estimated annual incidence is reportedly 0.07 per 100,000 in the general population.3 Spinal paragangliomas are believed to originate from sympathetic neurons placed in the thoracic and lumbar spinal cord, for which the path to the efferent sympathetic chain is represented by spinal communicating branches.4-6 Another theory is that tumors may arise from heterotopic neurons that lie along these proximal branches. In the lumbar region, they may arise from paraganglia that are located in the cauda equina. Because ependymal cell participation in their development cannot be ruled out, some authors speculate that some diffuse neuroendocrine system constituents can be the result of a local differentiation from the tissue that was not derived from the neural crest.7 Systemic manifestations, which are frequently associated with pheochromocytomas because of catecholamine release, are rarely described in cauda equina paraganglioma because of a supposed inability of these tumors to secrete hormones into the bloodstream or the inefficacy of the released substances to provoke symptoms.8 These tumors are often described as highly vascular lesions and hemorrhagic manifestations, namely subarachnoid hemorrhage, are often reported.1,8,9 As paragangliomas of cauda equina are rare, these tumors are often mistaken preoperatively for ependymoma or schwannomas.10 The gold standard for diagnosis is MRI, although a correct diagnosis may pose some problems because the neuroradiological features of the lesion are generally nonspecific. The lesion on T1-weighted sequences is usually hypo-
isointense to the conus medullaris and hyperintense on T2-weighted sequences; sometimes heterogeneous signal intensity has been observed.11 Hypointense tumor margins on T2-weighted sequences and gradient echo imaging may indicate paramagnetic effects from hemosiderin or ferritin because of previous hemorrhages.7,10-12 Paragangliomas are hypervascular lesions that produce punctate areas of flow void dispersed in a matrix of increased signal intensity. Intralesional punctate and linear low signal and multiple signal voids capping the tumor were likely to be findings of dilated vessels. Contrast-enhanced T1-weighted sequences demonstrated marked enhancement,10 and serpiginous fill voids around the tumor was a frequent feature, suggesting that dilated vessels or congested veins attributed to the hypervascularity. High signal intensity was observed in the cerebrospinal fluid (CSF) beneath the tumor, probably secondary to CSF circulation blockage, CSF protein elevation, or CSF pulsation reduction.13

When performed, selective spinal angiography may help the diagnosis because it demonstrates a highly vascular mass in the early arterial phase. The vascular blush becomes homogeneus in the late arterial phase. Preoperative embolization of the vessels feeding the tumor may be performed to minimize intraoperative bleeding.14

Currently, scintigraphy with I-131 metaiodobenzylguanidine is a functional imaging method for adrenal and extra-adrenal PGL localization. Although this method provides a high level of sensitivity and specificity, it is also inconvenient because of elevated radiation exposure, limited spatial resolution, and failure to identify nonsecreting PGLs, which are the most of those situated in the lumbar region.4 Preoperative cauda equina paraganglioma diagnosis is almost impossible, except in the presence of systemic manifestation with the possibility to detect high urinary biogenic amines or their metabolites.6

The classical morphological features of paraganglioma are tumor cells having granular cytoplasm with rich vascular networks. S-100 positive sustentacular cells could often be demonstrated. Unusual morphological features like co-existing cortical hyperplasia, vacuolar degeneration of tumor cells, presence of pheochromoblasts (small cells) and calciospherites, melanin pigmentation, cystic and oncocytic changes may be seen.15

Gross total excision of the lesion is the treatment of choice as there is no role of chemotherapy. However, incompletely resected and aggressive lesion may recur or distant metastasize. Role of radiotherapy in these cases is doubtful. The median free interval between surgical removal and local recurrence is estimated from 6 years to 20 years, so the long-term follow-up is mandatory whenever complete removal has not been achieved.

CONCLUSION

Lumbar spinal paragangliomas are diagnostic dilemma not only due to the rarity of these lesions, but also due to radiological features mimicking other common lesions at this location like ependymoma and schwannomas. Characteristic features on T2-weighted MRI and gradient echo imaging can help in preoperative diagnosis, though these features are not specific of these lesions. Preoperative consideration of these lesions helps in taking precautions during excision as these tumors can be functioning paragangliomas.

REFERENCES