

## Gangliocytic Paranglioma of Dorsolumbar Spine: A Rare Tumor at Rare Site

### Abstract

Spinal paragangliomas are rare benign tumors. The gangliocytic paragangliomas (GP) of spine are even rarer. The GPs are almost exclusively seen in duodenum. In spine, the usual site of affection is cauda equina region. The involvement of other spinal levels is far less often and includes thoracic and cervical region. We report this case of GP involving the conus medullaris region and reaching up to the upper part of cauda equina. The prognosis of spinal GP is excellent after total excision, as it is classified under WHO Grade I tumor. The patient made a full recovery after tumor removal. Clinical, radiological, and pathological characteristics of this rare tumor are discussed here along with.

**Keywords:** *Cauda equina, gangliocytic paragangliomas, spinal paragangliomas*

### Introduction

Gangliocytic paraganglioma (GP) was first reported in 1957 as ganglioneuroma by Dahl *et al.*<sup>[1]</sup> and named as GP by Kepes and Zacharias in 1971.<sup>[2]</sup> It is a rare, encapsulated, slow growing, benign tumor; having rare malignant potential with tendency to metastasize to regional lymph nodes. They are almost exclusively seen in the second part of duodenum.<sup>[3]</sup> GPs of the spine are extremely rare, where cauda equina is the most common site,<sup>[4]</sup> (conus-cauda region). In the present case, the tumor was localized at the level of conus medullaris extending up to the beginning of cauda equina, in dorsolumbar region (D12 to upper margin of L2). GP has generally been regarded as a neuroendocrine tumor and with histomorphological features of paraganglioma with mature ganglion cells.

### Case Report

A 42-year-old male patient presented with difficulty in walking since 15 days, pain and tingling sensation in the right leg since 6 months. The symptoms aggravated in night and were relieved after taking rest. No bowel or bladder complaints were reported. There was no previous history of hospitalization for any medical or surgical illness. The neurological evaluation showed

decreased motor power of right lower limb with no sensory deficits.

### Magnetic resonance imaging findings

An isodense, heterogeneous, intradural, extramedullary, enhancing mass lesion extending from lower margin of D12 to upper margin of L2 vertebra, at the level of conus medullaris extending up to the beginning of cauda equina was noted [Figure 1a and b]. No evidence of extraspinal invasion was seen. As the lesion was extramedullary and showed extension into the right neural foramina [Figure 1c], radiological impression was kept as a neurogenic tumor-Schwannoma/ependymoma/meningioma/neurofibroma. The clinical diagnosis was also kept the same.

### Intraoperative

Tumor was identified to be intradural, extramedullary with no extension in surrounding extraspinal, paravertebral soft tissue. Laminectomy and excision of tumor followed by duroplasty was done. Excised tumor was submitted for histopathology.

### Morphology

Multiple gray white tissue bits aggregating to 3 cm × 2.5 cm × 2 cm were received. Histopathological examination showed a capsulated tumor [Figure 2a] comprising of three distinct types of cell populations.

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Predominant cells were tumor cells (neuroendocrine cells) which were small, round to polygonal, with pale eosinophilic cytoplasm, round to oval nuclei, stippled chromatin, forming solid nests, and trabeculae [Figure 2b].

Ganglion cells having irregular contour, abundant cytoplasm, large vesicular nuclei with prominent nucleoli; were seen scattered and intermingled with neuroendocrine cells [Figure 2b].

Spindle (interstitial) cells, representing Schwann cells, were present along with branching vascular channels forming fine delicate network around the predominant cell type-giving rise to zellballen pattern [Figure 3a and b].

No evidence of pleomorphism, hyperchromasia, mitotic activity, and necrosis was noted.

Thus, the provisional histopathological diagnosis of “GP” was given.

### Immunohistochemistry

The tumor cells showed diffuse immunohistochemical expression of chromogranin, synaptophysin and neuron-specific enolase (NSE) supporting neuroendocrine differentiation. Ganglion cells expressed NSE, along with chromogranin and synaptophysin. Spindle cells showed

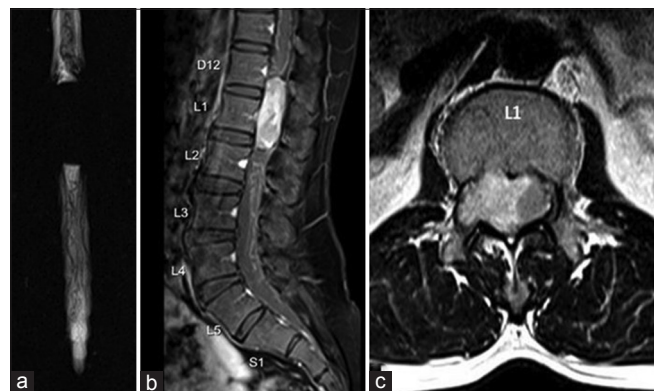


Figure 1: (a) Magnetic resonance myelogram showing displacement of conus to left by an intrathecal extramedullary mass. (b and c) Axial (b) and sagittal (c) postcontrast magnetic resonance imaging showing heterogeneous enhancing, lobulated, extramedullary, intradural lesion at the level of conus medullaris, up to upper part of cauda equina. Extension into right neural foramina with displacement of conus to the left is also seen (c)

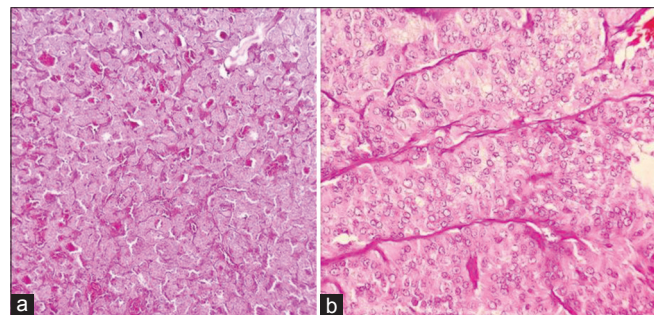


Figure 3: Zellballen pattern (a: H and E, ×100, and b: H and E, ×400)

S-100 positivity. The findings proved the microscopic diagnosis.

Figures 4 and 5 show immunohistochemical staining for chromogranin [Figure 4a], synaptophysin [Figure 4b], and neuron-specific enolase NSE [Figure 5a] highlighting neuroendocrine and ganglion cell components. Figure 5b: Immunohistochemical staining for S-100 is positive in spindle/interstitial cells 400×, IHC.

### Postoperative

The postoperative period was uneventful. Suture removal was done on 10<sup>th</sup> day. The patient was instructed to come every month for follow-up and third postoperative month for radiology (magnetic resonance imaging [MRI]). In spite of this, he came only once for check-up, i.e., 1 month postoperative, when he was clinically fine. After this, the patient was lost to follow-up.

### Discussion

Spectrum of intradural extramedullary tumors of the peripheral nervous system within the spinal canal in adults includes-Schwannomas, ependymoma, neurofibromas, or solitary metastasis and to a lesser extent paragangliomas. About 85%–90% of paragangliomas arise from the adrenal gland. The extra-adrenal paragangliomas can also occur in the central nervous system (CNS), where they are usually observed in the carotid body and glomus jugulare, also some other unusual sites being pineal region, sella turcica,

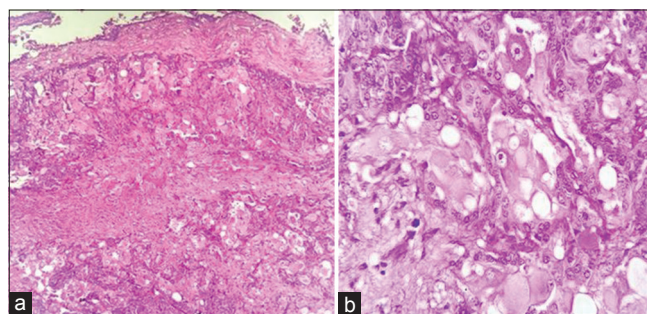


Figure 2: (a) Capsule with underlying tumor (H and E, ×40). (b) Ganglion cells (H and E, ×400)

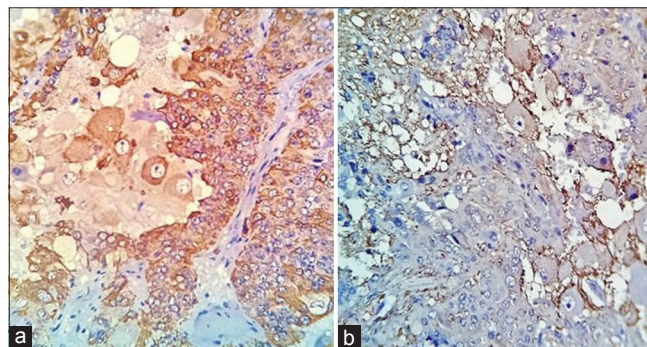
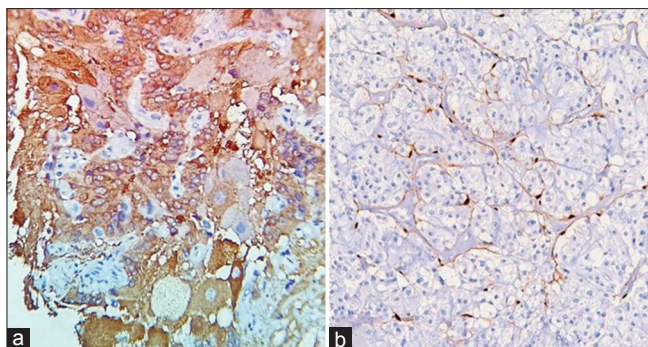


Figure 4: Immunohistochemical staining for chromogranin (a) and Synaptophysin (b) highlight neuroendocrine and ganglion cell components (IHC, ×400)





**Figure 5: (a) Immunohistochemical staining for neuron-specific enolase, neuron-specific enolase highlight neuroendocrine, and ganglion cell components. (b) Immunohistochemical staining for S-100 is positive in spindle/interstitial cells (IHC, x400)**

suprasellar region, or the orbit.<sup>[1]</sup> An uncommon localization is the spinal, where the incidence in the general population comes to be 0.07/100000 people.<sup>[5]</sup> Majority of spinal paragangliomas occur at cauda equina (conus-cauda region). Other spinal levels involved are thoracic and few in cervical region. Nearly mere 25% of the cauda equina paraganglioma show gangliocytic differentiation. The GP in the other spinal regions is even lesser.<sup>[4]</sup> The duodenum is the most common site of the GP, followed by the cauda equine of the spinal cord, respiratory system, jejunum, and esophagus. This case had localization at the level of conus medullaris extending up to the beginning of cauda equina, in dorsolumbar region (D12 – upper margin of L2). The histological differential diagnosis considered was an ependymoma, which has a poor prognosis. The distinctive triphasic appearance on histopathology essentially excluded the other possibilities.

### Histogenesis

The GP of the spine is considered to originate from neural crest cells which give rise to sympathoadrenal lineage, afterward undergoing divergent differentiation to form neuroendocrine and ganglion cell components.<sup>[6]</sup> Some authors have proposed that these lesions represent a hamartomatous proliferation when in duodenum, perhaps originating from different embryonal layers during the formation and migration of the pancreatic primordia.<sup>[7-9]</sup> It is based on the diversity of the elements composing GP. However, the fact that few of them have undergone malignant transformation with lymph node metastases argues in favor of a true neoplastic nature.<sup>[10]</sup>

The presenting complaints of spinal GPs are usually lumbar pain, motor or sensory loss in lower extremities, or bowel and bladder dysfunction. The MRI of intradural extramedullary lesions shows a similar image profile, thus making histologic examination the key to diagnosis. The usual management of this tumor is surgical resection. When the lesions are placed in the junction areas, such as present case, laminotomy is preferred over laminectomy,

considering the motion segments, and potential for the development of instability in the future. Adjuvant radiotherapy should be reserved for unencapsulated or incompletely excised lesions as it does not guarantee the prevention of tumor recurrence. However, follow-up in the form of clinical examination every month after the operative procedure till 6 months followed by every 6 months, and a radiological examination MRI at 3<sup>rd</sup> postoperative month followed by every 6 months is recommended.

### Conclusion

This case of GP, localized in dorsolumbar region, was seen at the level of conus medullaris extending up to the beginning of cauda equina. Their origin in CNS is still unclear. In view of rarity, a long-term follow-up and assessment of molecular pathogenesis are essential for therapeutic intervention, biological behavior, and monitoring the outcome.

### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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### Conflicts of interest

There are no conflicts of interest.

### References

- Dahl EV, Waugh JM, Dahlin DC. Gastrointestinal ganglioneuromas; brief review with report of a duodenal ganglioneuroma. *Am J Pathol* 1957;33:953-65.
- Kepes JJ, Zacharias DL. Gangliocytic paragangliomas of the duodenum. A report of two cases with light and electron microscopic examination. *Cancer* 1971;27:61-7.
- Kwon J, Lee SE, Kang MJ, Jang JY, Kim SW. A case of gangliocytic paraganglioma in the ampulla of vater. *World J Surg Oncol* 2010;8:42.
- Landi A, Mancarella C, Marotta N, Tarantino R, Lenzi J. Diagnosis and treatment of paragangliomas of the filum terminale, an extremely rare entity: Personal experience and literature review. *J Spine* 2013;S3:e001.
- Landi A, Tarantino R, Marotta N, Rocco P, Antonelli M, Salvati M, *et al.* Paraganglioma of the filum terminale: Case report. *World J Surg Oncol* 2009;7:95.
- Shankar GM, Chen L, Kim AH, Ross GL, Folkerth RD, Friedlander RM. Composite ganglioneuroma-paraganglioma of the filum terminale. *J Neurosurg Spine* 2010;12:709-13.
- Guarda LA, Ordonez NG, del Junco GW, Luna MA. Gangliocytic paraganglioma of the duodenum: An immunocytochemical study. *Am J Gastroenterol* 1983;78:794-8.

8. Perrone T, Sibley RK, Rosai J. Duodenal gangliocytic paraganglioma. An immunohistochemical and ultrastructural study and a hypothesis concerning its origin. *Am J Surg Pathol* 1985;9:31-41.
9. Burke AP, Helwig EB. Gangliocytic paraganglioma. *Am J Clin Pathol* 1989;92:1-9.
10. Okubo Y, Wakayama M, Nemoto T, Kitahara K, Nakayama H, Shibuya K, *et al.* Literature survey on epidemiology and pathology of gangliocytic paraganglioma. *BMC Cancer* 2011;11:187.