

Extradural Dorsal Spinal Paraganglioma: A Rare Case Report and Literature Review

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Abstract

Paragangliomas are neuroendocrine tumors originating from specialized cells of neural crest derivative. They are mostly found in the carotid and jugulotympanic region. Paragangliomas are extremely rare in the spine, and in literature, most cases are described as intradural extramedullary lesion in the cauda equine or filum terminale region. Extradural dorsal spine paragangliomas are even rarer, and till date, only 11 cases are reported in available literature. We describe a case of an adult woman who presented with mid dorsal region pain and sensory-motor spastic paraplegia. Imaging features were suggestive of D5 Pott's spine with epidural granulation tissue. Perioperatively, the tumor was moderately vascular and complete excision done. On HPE and IHC, diagnosis of paraganglioma was made. There was no evidence of clinical recurrence of the lesion at 6 months follow-up.

Keywords

- ▶ paraganglioma
- ▶ dorsal spine
- ▶ Zellballen's pattern
- ▶ immunohistochemistry

Introduction

Paragangliomas are neuroendocrine tumors of the extra-adrenal paraganglionic system. They are derived from chemoreceptor cells and usually seen in the carotid body and glomus jugulare, and their location in the spinal canal is rare. Most of them are found intradural, predominantly at the cauda equina.^{1,2} Epidural compression of the spinal cord by paraganglioma involving the dorsal spine occurs very rarely, and only a few number of cases have been reported in literature.³ These tumors are very rarely encountered in adolescents and adults, with the peak incidence in the fifth decade.⁴ We report a case of extradural dorsal spine paraganglioma in a 32-year-old woman.

Case History

A 32-year-old woman presented with a history of backache in the mid-dorsal region for 7 months and weakness of both the lower limbs for 1 month. General physical and systemic examinations were within normal limits. Neurologic

examination showed spastic paraplegia (0/5), hyperreflexia, and complete sensory loss in D5 and below. All the routine laboratory investigations including viral markers and chest X-ray were normal. Magnetic resonance imaging (MRI) of the dorsal spine revealed D5 vertebral body collapsed with lost intervening disc space and end plate irregularities with heterogeneous postcontrast enhancement along with D4–6 pre- and paravertebral soft tissue heterogeneous enhancement (▶ Fig. 1A–E). There was a preoperative impression of Pott's spine, and the patient prepared for surgery.

D5 laminectomy with complete excision of lesion, and D3–7 pedicle screw and rod fixation was done. The tumor was extradural, moderately vascular, bluish red, and soft to firm in consistency with complete collapse of D5 vertebral body. It was contiguous with the spinal roots but not adherent to the dura mater. The patient had an uneventful postoperative period and some sensory improvement in early postoperative period. The histopathologic examination (HPE) of the specimen showed neoplastic chief cells arranged in organized pattern, peripherally lined by

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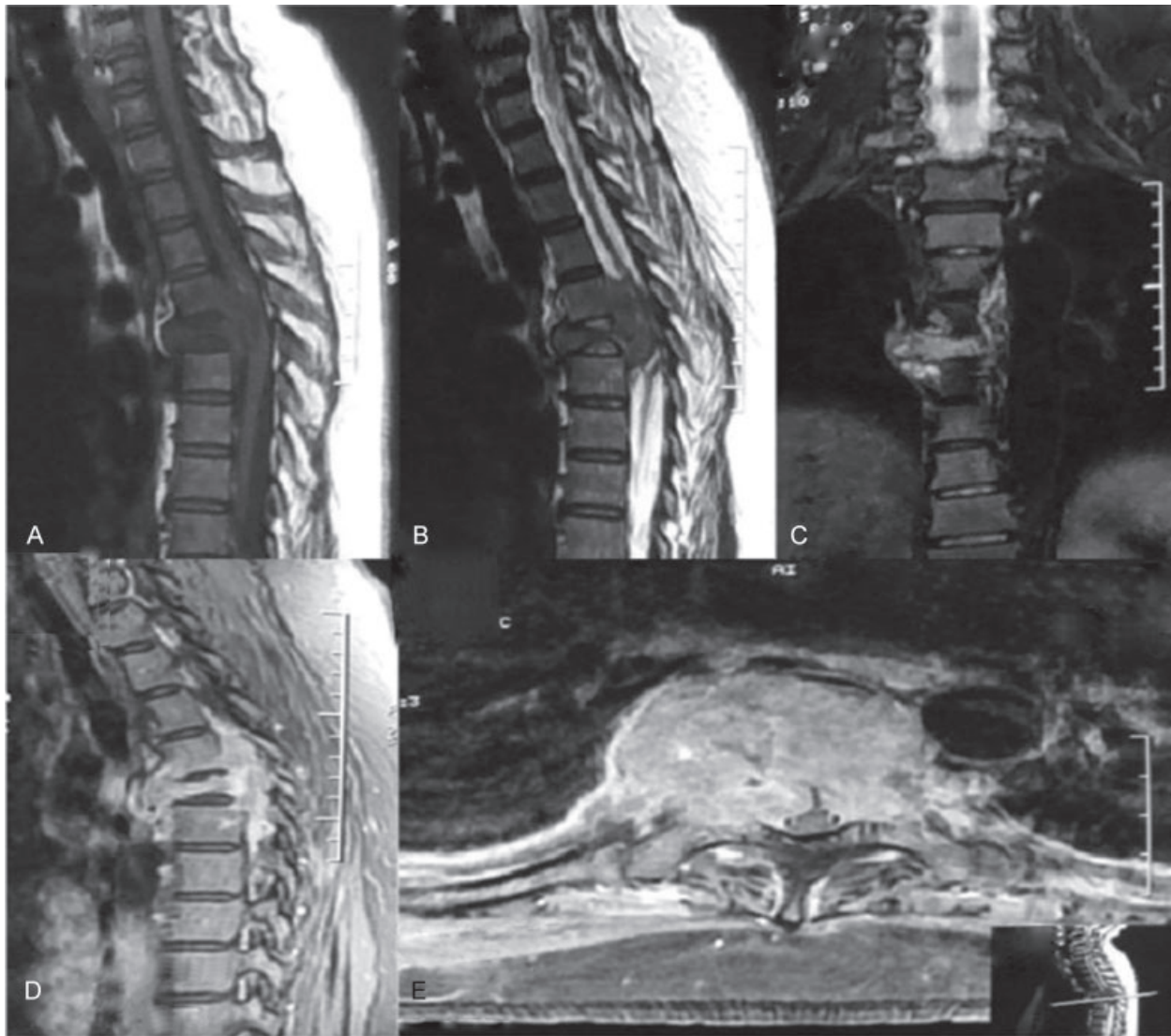


Fig. 1 Magnetic resonance images: (A) T1 sagittal image, (B, C) T2 sagittal and coronal image, and (D, E) contrast sagittal and axial image.

sustentacular cells separated by fibrous septa (Zellballen's pattern). Cells have nuclei with stippled chromatin, inconspicuous nucleoli, and eosinophilic granular to clear cytoplasm (►Fig. 2A). Immunohistochemistry (IHC) was performed for confirmation; positive reaction for chromogranin in the chief cells and S100 nuclear positivity in sustentacular cells (►Fig. 2B, C) were seen. Based on HPE and IHC, morphologic diagnosis of paraganglioma was established. There was no evidence of clinical recurrence of the lesion at 6 months follow-up, but later on the patient was lost to follow-up.

Discussion

Paraganglioma is a rare neuroendocrine neoplasm that may develop at various locations such as the head, neck, chest, and abdomen. The term "paraganglioma" was first given by Lerman and colleagues in 1972.⁵ Extra adrenal tumors are mainly found in the jugulotympanic region, and unusual intracranial locations include sellar, pineal, and petrous ridge

regions.^{1,6} The spinal location is rare. Most paragangliomas are found intradural, predominantly at the lumbosacral region.² Only 11 patients with extradural dorsal spine paraganglioma have been reported in the literature (►Table 1A–E).^{3,7–14}

Common clinical presentation of patients with spinal paragangliomas are back pain radiating down the leg, motor-sensory deficits, and bowel and bladder irregularity.^{7,15} The clinical presentation depends on the level of affected spinal cord and the degree of cord compression. Our patient presented with back pain in the mid-dorsal region and weakness of both the lower limbs. MRI finding of paragangliomas is common with other spinal tumors such as schwannomas, ependymomas, meningiomas, and dermoid, and paragangliomas appear isointense on T1-weighted images whereas hyperintense on T2-weighted images with contrast enhancement.¹⁶

Microscopically paragangliomas show typical nuclear monomorphism and "Zellballen's" pattern (large polyhedral chief cells arranged in nests), with granular argyrophilic cells. Perivascular pseudorosettes, areas of hemorrhage, and

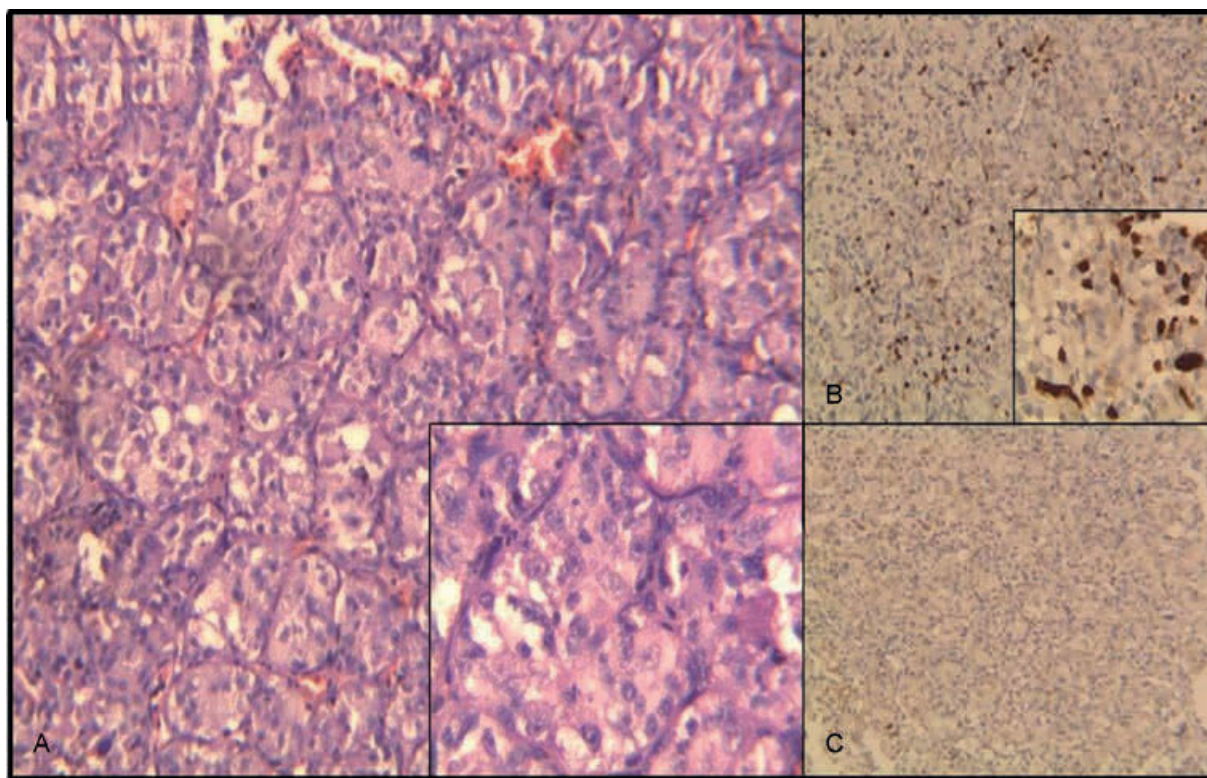


Fig. 2 Histopathology and immunohistochemistry images: (A) HPE $\times 100$ and $\times 200$, (B) S100 positive $\times 100$ and $\times 200$, and (C) chromogranin positive.

Table 1 Extradural dorsal paraganglioma: review of literature

No.	Year	Author	Sex	Age	Site	Clinical features	Surgical treatment
1	1983	Böker et al ⁷	F	35	T4–5	2 mo back pain, 4 wk paraparesis and sensory loss below T4	Total resection
2	1983	Böker et al ⁷	F	36	T11	10 mo back pain, sudden paraparesis and sensory loss below T11	Incomplete resection
3	1991	Cybulski et al ³	M	34	T8	4 mo midthoracic back pain, sudden paresthesia and stiffness at lower limbs	Stage 1: Incomplete resection Stage 2: Embolization Stage 3: Total resection
4	1996	Fitzgerald et al ⁸	M	30	T4	Back pain, numbness below T5, sustained ankle clonus in both legs	Total resection
5	1996	Noorda et al ⁹	F	52	T7–9	2y neuralgia in thoracic spine and lower extremities weakness, T7 local tenderness and decreased sensation	Stage 1: Embolization Stage 2: Total resection Stage 3: Anterior stabilization
6	2001	Shin et al ¹⁰	M	43	T6	1y recurrent back pain, sudden paresthesia of the lower extremities, urinary incontinence	Stage 1: Embolization Stage 2: Resection
7	2001	Shin et al ¹⁰	F	67	T11	Back pain for several years, lower abdominal pain for 1y	Incomplete resection
8	2002	Houten et al ¹¹	M	41	T9	2 wk gait disturbance and lower limbs, weakness, diminished sensation under T10, lower limbs hyperreflexia	Incomplete resection
9	2003	Jeffs et al ¹²	F	53	T12	10-mo history of headaches, facial flushing and palpitations associated with hypertension, mild hypesthesia in the right T12 dermatome	Grossly complete excision
10	2006	Conti et al ¹³	F	43	T1–4	Sudden spastic incomplete paraplegia and paresthesia at the lower limbs	Resected en bloc
11	2012	Mauricio et al ¹⁴	M	26	T10	Diffuse pain in thoracic region, with progressively diminished muscle strength and paresthesia in the left lower limb	Tumor resection with decompression and stabilization

Abbreviations: F, female; M, male; mo, month; wk, week.

necrosis may also be seen. Immunohistochemistry is positive for chromogranin, synaptophysin, neuron-specific (gamma) enolase, and S-100.¹⁷ Most paragangliomas are benign and cured by surgical removal. Only 3% have malignant potential with tendency to distant metastasis.¹⁸ Therefore, after gross total excision of tumor prognosis is good, and radiotherapy is required in case of locally invasive tumors or when complete excision of tumor is not achieved.

Although extremely uncommon, extradural dorsal spine paraganglioma should be kept as a differential diagnosis for epidural compressive lesions. Spinal paragangliomas are mostly benign neoplasms, and a complete surgical resection provides recurrence-free survival. Generous reporting of such type of cases helps us in better understanding of such a rare entity.

Note

The data required for the manuscript was provided by Department of Neurosurgery, King George's Medical University, Lucknow, Uttar Pradesh, India.

Conflict of Interest

None.

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