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 equina 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. 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. These tumors are very rarely encountered in adolescents and adults, with the peak incidence in the fifth decade. 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 heterogenous 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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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. Extra adrenal tumors are mainly found in the jugulotympanic region, and unusual intracranial locations include sellar, pineal, and petrous ridge regions. The spinal location is rare. Most paragangliomas are found intradural, predominantly at the lumbosacral region. Only 11 patients with extradural dorsal spine paraganglioma have been reported in the literature (►Table 1A–E).

Common clinical presentation of patients with spinal paragangliomas are back pain radiating down the leg, motor-sensory deficits, and bowel and bladder irregularity. 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.

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

Table 1 Extradural dorsal paraganglioma: review of literature

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

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-100.16 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**
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**Conflict of Interest**
None.

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**References**