Two Case Reports and an Updated Review of Spinal Intraosseous Schwannoma

Fan Zhang, B.S., Feizhou Lu, Ph.D., Jinyuan Jiang, B.S., Hongli Wang, M.S.

Department of Orthopedics, Huashan Hospital, Fudan University, Shanghai, China

We report two rare cases of spinal intraosseous schwannoma (SIS) with sustained myelopathy symptoms and provide an updated review regarding SIS in the literature. A 71-year-old man experienced right lumbocrural pain and gait disturbance accompanied with paresthesia and right leg weakness. Imaging examinations revealed a mass with lesions in L4 vertebral body causing bone destruction and spinal cord compression. Complete resection of the well-demarcated tumor and posterior fusion were performed. A 54-year-old female reported bilateral gait disturbance, paresthesia, and numbness without weakness, and imaging revealed a posterior mass from T9 causing spinal cord compression and bone erosion. The tumor was completely separated from the spinal nerve root. The tumors from both patients were confirmed as schwannomas. Tumor recurrence was not observed at the 2–4 year follow-up. Although rare, SIS should be considered during differential diagnosis and can affect treatment planning. SIS symptoms vary depending on tumor location, and fusion is frequently necessary for spinal reconstruction after complete tumor resection.

Key Words : Spinal intraosseous schwannoma · Myelopathy · Differential diagnosis.

INTRODUCTION

Schwannoma (neurilemmoma) is a benign tumor that arises mainly in sensory nerve sheaths. Intraosseous lesions are rare, accounting for less than 0.2% of primary bone tumors, and the majority are located in the mandible and sacrum. Other reported sites include the ulna, humerus, femur, tibia, ribs, patella, scapula, maxilla, the small bones of the hands, and vertebral bodies. Spinal intraosseous schwannoma (SIS) was first reported in 1964. SIS is an extremely rare lesion, and the diagnosis remains unclear, mostly because of its origin. This is the focus of controversy between some authors, whereas the pathological diagnosis and surgical treatment are quite similar. Correctly diagnosing SIS without resection is difficult given the broad range of symptoms. However, appropriate diagnosis is important to properly plan surgical intervention. Magnetic resonance imaging (MRI) with contrast can be useful in preoperatively diagnosing SIS, and histopathology is very informative for resected specimens. Here, we report two cases of intraosseous schwannoma involving the body of L4 and the posterior structures of T9 that were diagnosed following myelopathic symptoms.

CASE REPORT

Case 1

A 71-year-old man experienced right lumbocrural pain and gait disturbance accompanied by paresthesia and right leg weakness for 6 months. Neurologic examination revealed impaired right leg motor function (grade 3/5) with diminished feeling on the right side caudally from the lumbar L4 sensory dermatome; however, his nerve reflexes were normal. Enhanced magnetic resonance imaging (MRI) (Fig. 1) showed a mass with lesions in the vertebral body (L4) and spinal canal compressing the lumbar spinal cord. Computed tomography (CT) scan revealed a slowly growing tumor with severe vertebrae destruction (Fig. 2A, arrow). Piecemeal resection and decompression were performed. After total laminectomy and facetectomy of L3–5, a well-demarcated tumor was exposed extending into the spinal canal (Fig. 2B, arrow) without nerve involvement or dural adhesions. The spine was stabilized with pedicle screws and rods after the tumor was completely resected. Histological characteristic of the tumor revealed Antoni A and B tissue and overexpression of S-100 protein, which confirmed a diagnosis of intraosseous schwannoma without originating nerve remnants. Intraoperative fluoroscopy revealed successful fusion.
internal fixation (Fig. 3A). Postoperative images taken at the 2-year follow-up showed no obvious evidence of recurrence and general bony fusion (Fig. 3B, C, D), and the patient's gait and sensation in the right lower extremity showed good recovery.

Case 2
A 54-year-old female reported a 4-month history of gait disturbance and paresthesia of both lower extremities. Physical examination revealed numbness in both legs without obvious weakness (grade 5/5); nerve reflexes were normal. MRI (Fig. 4A) showed a mass (arrow) that appeared to originate from the posterior elements of T9 and extended into the spinal canal and paravertebral areas, extruding the spinal cord. T9 vertebrae bone erosion was observed on CT. The imaging results led to a differential diagnosis of primary benign/malignant bone tumor or metastatic tumor. The tumor was completely separated and surgically resected from the spinal nerve root with a clear border, and a posterior fusion with allograft bone was performed to stabilize the spine. Pathological characteristics of the tumor confirmed a benign schwannoma (Fig. 4C, D). Intraoperative fluoroscopy demonstrated successful internal fixation in the proper position for posterior interbody fusion (Fig. 4E, F). According to the MRI images at the 4-year follow-up, there was no obvious sign of recurrence with relieved gait and sensation disturbance (Fig. 4G).

Fig. 1. Preoperative MR images from case 1 (71-year-old male), showing an abnormality in the L4 vertebra body. A : T1-weighted imaging. The tumor is isointense compared with the spinal cord. B : Gd-enhanced T1-weighted imaging showing irregular enhancement.

Fig. 2. Case 1, a 71-year-old male with L4 intraosseous schwannoma. A : High-signal intensity (arrow) on preoperative CT showing a tumor compressing the spinal cord. B : Intraoperative image showing the tumor (arrow) extending to the spinal canal. C : Histology revealed hypercellular (Antoni A) and hypocellular (Antoni B) areas, indicating a typical schwannoma (H&E staining, ×200). D : Immunohistochemistry showing S-100 protein over-expression (brown color, ×200).

Fig. 3. Radiology images of case 1 from immediately after 2 years follow-up. A : Intraoperative fluoroscopy showed proper internal fixation (top : anteroposterior, bottom : lateral). B : Postoperative X-ray images showing that general fusion was realized without any internal fixation dislocation (top : anteroposterior, bottom : lateral). C : There was no recurrence observed on the MRI at the 2-year follow-up (top : lateral, bottom : coronal). D : Follow-up CT images taken 2 years later revealed that general fusion was achieved without any internal fixation dislocation (top : lateral, bottom : coronal).
DISCUSSION

Although SIS is quite rare, a number of cases have been published in the last 30 years. We looked at images in the cited articles and found classic pre- and intra-operative images of SIS to ensure that there was no connection between the tumor and nerve tissue, but the photos were not always convincing. Our report aims to provide a clear definition of SIS and a review of this rare disease. Because the symptoms associated with SIS can vary depending on their location in the spinal cord and because they can overlap with those manifested in other conditions, incorrect diagnosis remains a problem. A relatively complete summary of SIS cases described from 1971–2012 is shown in Tables 1, 2. Though the cases reported by Barnowsky and Dalal and Inaoka et al. showed that Schwannoma originated from nerve root. It was defined as SIS by Park et al. The majority of reports do not hold this view, and we determined those cases were probably intraosseous invasions of extraosseous nerve sheath tumors. It is known that neurilemmomas can involve bone by three possible mechanisms: 1) an extraosseous tumor causing secondary bone erosion; 2) a tumor arising centrally within the bone; and 3) tumor origination in the nutrient canal followed by growth into a dumbbell-shape that enlarges the spinal canal. Of these, only the second mechanism could occur with intraosseous neurilemmoma; the small nerves that give rise to these tumors have been described in the human vertebrae. In other words, the intraosseous origin of schwannoma must be nerves within bones that are free from adjacent neural tissue. This view is supported by most reports in the SIS literature.

Symptoms vary among SIS patients. Because most of these tumors enlarge slowly, the patient’s history may be considerably long, and most experience pain (14/25, 56%) depending on the tumor location. Neurological compression symptoms develop when the tumor perforates the bone cortex and causes spinal cord protrusion, but specific symptoms can differ depending on the level of the lesion. SISs are most commonly found in the lumbar region (38%), followed by thoracic (32%) and cervical (28%), which is different from the results reported by Park et al.

Radiological findings in SIS can also vary considerably, and differential diagnosis includes ruling out solitary myeloma, chondroma, chondrosarcoma, giant cell tumor, angiomia, and aneurysmal bone cyst. SISs are sometimes found to primarily occupy the intraosseus region with or without extravertebral and spinal canal involvement, and a hollowed out vertebral body with a single, thin, bulging cortex perforation is suggestive of intraosseous origin. Generally, intraosseous schwannoma appears on radiological images as a lytic defect with bone erosion lacking new periosteal bone formation and calcification/ossification, although a narrow sclerotic zone may be present between the tumor and bone. Vertebral intraosseous schwannomas gradually increase in size, resulting in pedicle and vertebral body erosion that widens the foramen and vertebral scalloping. Histological conformation is mandatory for a diagnosis of SIS. Proliferation of slender spindle cells with oval nuclei and focal palisading nuclei (Antoni A) and degenerated hypocellular areas (Antoni B) with hemosiderin deposition and thrombosed blood vessels are suggestive of schwannoma. SISs are not histologically different from schwannomas that develop elsewhere, even at the ultrastructural level, but the histological features of intraosseous neurilemmomas may be obscured in highly cellular lesions with subtle Antoni types A and B patterns. Both types have long durations and similar degenerative characteristics, including perivascular hyalinization, calcification, and cystic degeneration.

The gold standard for benign bone tumors is marginal resection. Unfortunately, this is often difficult to achieve in SISs, which have both intra- and extraosseous components that invade adjacent structures, including nerve roots, spinal cord, and para-vertebral tissue. This is usually addressed with adequate cu-
<table>
<thead>
<tr>
<th>Ref</th>
<th>Year</th>
<th>Author</th>
<th>Age (y)</th>
<th>Sex</th>
<th>Origin</th>
<th>Level/original location</th>
<th>Symptom</th>
<th>Treatment</th>
<th>Resection</th>
</tr>
</thead>
<tbody>
<tr>
<td>7</td>
<td>1971</td>
<td>Dickson et al.</td>
<td>51</td>
<td>F</td>
<td>-</td>
<td>L3/vertebral body</td>
<td>Left thigh pain</td>
<td>Abdominal approach excision and fusion</td>
<td>Complete</td>
</tr>
<tr>
<td>10</td>
<td>1971</td>
<td>Gupta and Agarwal</td>
<td>37</td>
<td>M</td>
<td>-</td>
<td>-</td>
<td>Back pain, lower limb weakness</td>
<td>-</td>
<td>Complete</td>
</tr>
<tr>
<td>22</td>
<td>1975</td>
<td>Polley</td>
<td>34</td>
<td>F</td>
<td>-</td>
<td>C6, C7/vertebral body</td>
<td>No neurological deficits, but neck pain after cervical injury</td>
<td>Posterior approach excision and fusion</td>
<td>Complete</td>
</tr>
<tr>
<td>16</td>
<td>1988</td>
<td>Naidu et al.</td>
<td>50</td>
<td>M</td>
<td>-</td>
<td>C3, C4/vertebral body, pedicle, and transverse process</td>
<td>Weakness in all limbs and burning sensation in both lower limbs</td>
<td>Posterior approach excision without fusion</td>
<td>Complete</td>
</tr>
<tr>
<td>1</td>
<td>1992</td>
<td>Barnowsky and Dalal</td>
<td>41</td>
<td>M</td>
<td>L4 root</td>
<td>L4</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>12</td>
<td>1994</td>
<td>Knapp et al.</td>
<td>65</td>
<td>F</td>
<td>-</td>
<td>L4, L5</td>
<td>-</td>
<td>-</td>
<td>Biopsy</td>
</tr>
<tr>
<td>18</td>
<td>1997</td>
<td>Nooraie et al.</td>
<td>46</td>
<td>M</td>
<td>-</td>
<td>T12, L1/vertebral body</td>
<td>Complaint of severe back pain after accident without abnormal neurological deficits</td>
<td>Posterior approach excision and fusion</td>
<td>Complete</td>
</tr>
<tr>
<td>13</td>
<td>1998</td>
<td>Ko et al.</td>
<td>-</td>
<td>-</td>
<td>T8</td>
<td>-</td>
<td>-</td>
<td>Operated</td>
<td>-</td>
</tr>
<tr>
<td>3</td>
<td>1998</td>
<td>Chang et al.</td>
<td>58</td>
<td>M</td>
<td>-</td>
<td>L4, L5/vertebral body</td>
<td>Severe pain and numbness of both lower extremities</td>
<td>1. Anterolateral retroperitoneal fusion and excision, 2. Posterior fusion</td>
<td>Complete</td>
</tr>
<tr>
<td>22</td>
<td>2000</td>
<td>Ramasamy et al.</td>
<td>37</td>
<td>M</td>
<td>-</td>
<td>T12/vertebral body</td>
<td>Back pain, weakness, and bilateral lower extremity numbness</td>
<td>Resection with anterior and posterior fusion</td>
<td>Complete</td>
</tr>
<tr>
<td>11</td>
<td>2001</td>
<td>Inaoka et al.</td>
<td>9</td>
<td>M</td>
<td>T10 root</td>
<td>T10/vertebral body and left transverse process</td>
<td>Nontender distention of the back without abnormal neurological deficits</td>
<td>Not mentioned</td>
<td>Complete</td>
</tr>
<tr>
<td>11</td>
<td>2001</td>
<td>Inaoka et al.</td>
<td>39</td>
<td>M</td>
<td>L5 root</td>
<td>L5/vertebral body and left transverse process</td>
<td>Moderate lumbar pain without abnormal neurological deficits</td>
<td>Not mentioned</td>
<td>Operated</td>
</tr>
<tr>
<td>24</td>
<td>2001</td>
<td>Schreuder et al.</td>
<td>39</td>
<td>F</td>
<td>-</td>
<td>C6/vertebral body</td>
<td>Neck pain and dysphagia</td>
<td>Anterior approach excision and fusion</td>
<td>Complete</td>
</tr>
<tr>
<td>15</td>
<td>2004</td>
<td>Mizutani et al.</td>
<td>73</td>
<td>F</td>
<td>-</td>
<td>C4</td>
<td>Discomfort in swallowing</td>
<td>-</td>
<td>Complete</td>
</tr>
<tr>
<td>17</td>
<td>2005</td>
<td>Nannapaneni and Sinar</td>
<td>42</td>
<td>M</td>
<td>-</td>
<td>C5/vertebral body</td>
<td>No neurological deficits</td>
<td>Anterior approach excision and fusion</td>
<td>Complete</td>
</tr>
<tr>
<td>26</td>
<td>2005</td>
<td>Singrakhia et al.</td>
<td>43</td>
<td>M</td>
<td>-</td>
<td>C3, C4/vertebral body</td>
<td>Increasing numbness around the right shoulder and deltoid weakness</td>
<td>Anterior approach excision and fusion</td>
<td>Operated</td>
</tr>
<tr>
<td>26</td>
<td>2005</td>
<td>Singrakhia et al.</td>
<td>45</td>
<td>M</td>
<td>-</td>
<td>C4/vertebral body and left transverse process</td>
<td>Progressive pain and weakness in the right upper limb</td>
<td>Anterior approach excision and fusion</td>
<td>Operated</td>
</tr>
<tr>
<td>10</td>
<td>2005</td>
<td>Gupta and Agarwal</td>
<td>30</td>
<td>F</td>
<td>-</td>
<td>L2/vertebral body</td>
<td>Complaints of backache and progressively increasing weakness in both lower limbs</td>
<td>Not mentioned</td>
<td>Complete</td>
</tr>
</tbody>
</table>
We report two cases of SIS, which is a rare differential diagnosis for intrasosseous tumor. Proper diagnosis requires radiological tests, gross intraoperative findings, and postoperative histological results. Symptoms vary depending on tumor location, and fusion is necessary to stabilize the spine after the tumor is completely excised.

### References

12. Knapp TR, Struk DW, Munk PL, Bainbridge TC, Bhimji SD, Poyn PY:...
Spinal Cord Compression due to Intraosseous Schwannoma | F Zhang, et al.