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Case Report

Two Case Reports and an Updated Review of Spinal Intraosseous Schwannoma

Fan Zhang, B.S., Feizhou Lu, Ph.D., Jianyuan 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[24]. Intraosseous lesions are rare, accounting for less than 0.2% of primary bone tumors[3], and the majority are located in the mandible and sacrum[6,8,9,16]. Other reported sites include the ulna, humerus, femur, tibia, ribs, patella, scapula, maxilla, the small bones of the hands, and vertebral bodies[2]. Spinal intraosseous schwannoma (SIS) was first reported in 1964[5]. 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 (Fig. 2C) and overexpression of S-100 protein (Fig. 2D), which confirmed a diagnosis of intraosseous schwannoma without originating nerve remnants. Intraoperative fluoroscopy revealed successful

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• Address for reprints : Feizhou Lu, Ph.D.
Department of Orthopedics, Huashan Hospital, Fudan University, Shanghai 200040, China
Tel : +86-13801707800, Fax : +86-21-62489191, E-mail : lufeizhou2012@126.com

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 reoccurence with relieved gait and sensation disturbance (Fig. 4G).

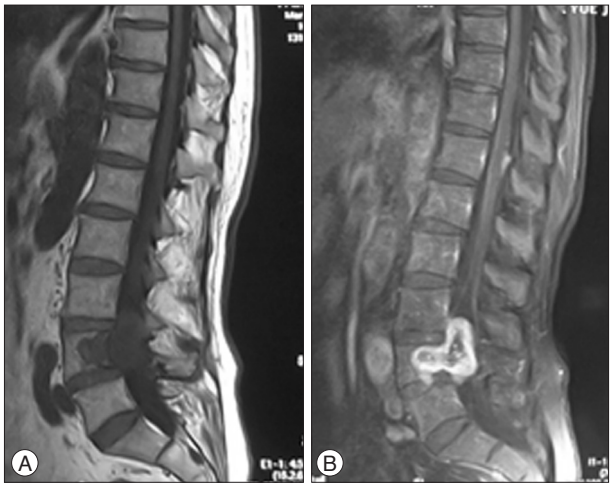

Fig. 1. Preoperative MR images from case 1 (71-year-old male), showing an abnormality in the L4 vertebra body. A : T1-weighted imaging. The tumor is isointense compared with the spinal cord. B : Gd-enhanced T1-weighted imaging showing irregular enhancement.

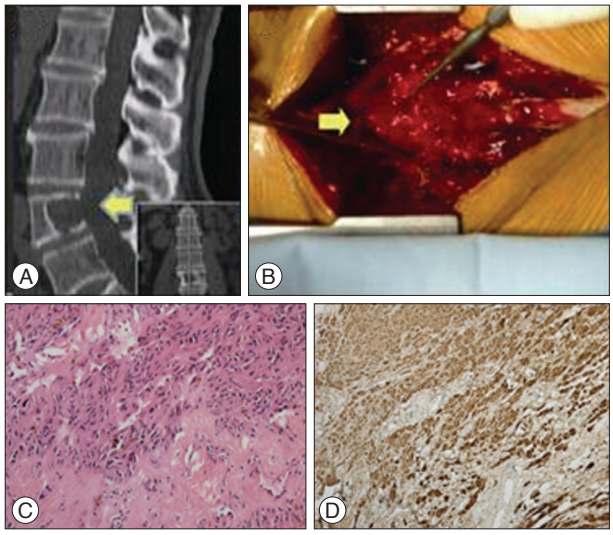

Fig. 2. Case 1, a 71-year-old male with L4 intraosseous schwannoma. A : High-signal intensity (arrow) on preoperative CT showing a tumor compressing the spinal cord. B : Intraoperative image showing the tumor (arrow) extending to the spinal canal. C : Histology revealed hypercellular (Antoni A) and hypocellular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

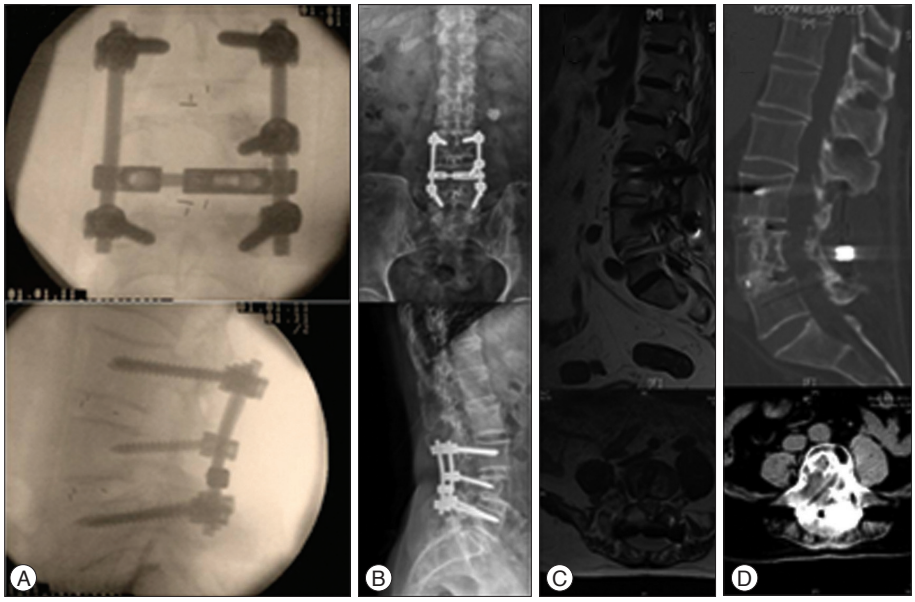

Fig. 3. Radiology images of case 1 from immediately after 2 years follow-up. A : Intraoperative fluoroscopy showed proper internal fixation (top : anteroposterior, bottom : lateral). B : Postoperative X-ray images showing that general fusion was realized without any internal fixation dislocation (top : anteroposterior, bottom : lateral). C : There was no reoccurrence 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).

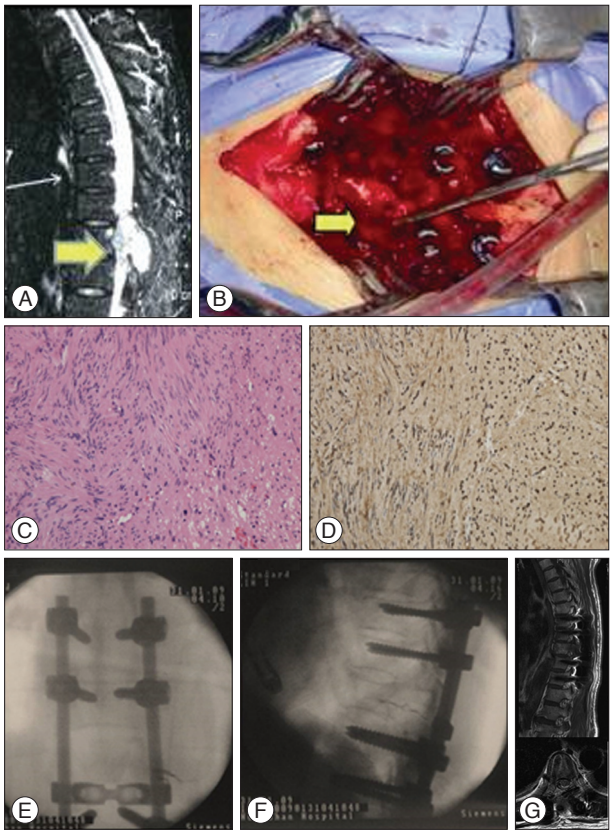

Fig. 4. Case 2, a 54-year-old female with T9 intraosseous schwannoma. A : T2-weighted MR imaging showing mixed-intensity tumor tissue (arrow). B : The tumor originated from posterior T9 without nerve root involvement (arrow), and we performed total piecemeal resection followed by posterior spinal reconstruction. C : Histology revealed hypercellular (Antoni A) and hypocellular (Antoni B) areas, indicating a typical schwannoma (H&E stain; ×200). D : Immunohistochemistry revealed S-100 protein overexpression (brown color, ×200). E and F : Intraoperative fluoroscopy demonstrated successful internal fixation (E : anteroposterior, F : lateral). G : There was no obvious sign of reoccurence at 4 year postoperatively according to the MRI images (top : lateral position, bottom : coronal).

DISCUSSION

Although SIS is quite rare, a number of cases have been published in the last 30 years[1-4,7,10-19,21,22,24,27]. 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[1] and Inaoka et al.[11] showed that Schwannoma originated from nerve root. It was defined as SIS by Park et al.[19]. 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[3,6,9,11,22-24]. 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[20,25]. In other words, the intraosseous origin of schwannoma must be nerves within bones that are free from adjacent neural tissue.[2,3] This view is supported by most reports in the SIS literature.[2,11]

Symptoms vary among SIS patients. Because most of these tumors enlarge slowly,[19] the patient's history may be considerably long, and most experience pain (14/25, 56%) depending on the tumor location.[24] 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.[22] 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.[19]

Radiological findings in SIS can also vary considerably, and differential diagnosis includes ruling out solitary myeloma, chordoma, chondrosarcoma, giant cell tumor, angioma, and aneurysmal bone cyst. SISs are sometimes found to primarily occupy the intraosseous 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.[3] 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[3,24]. 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[3]. SISs are not histologically different from schwannomas that develop elsewhere, even at the ultrastructural level[16], but the histological features of intraosseous neurilemmomas may be obscured in highly cellular lesions with subtle Antoni types A and B patterns[11]. Both types have long durations[16] 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 paravertebral tissue[16]. This is usually addressed with adequate cu-

Table 1. Previous reports of SIS

Ref.	Year	Author	Age (y)	Sex	Origin	Level/original location	Symptom	Treatment	Resection
7	1971	Dickson et al.	51	F	-	L3/vertebral body	Left thigh pain	Abdominal approach excision and fusion	Complete
10	1971	Gupta and Agarwal	37	M	-	-	Back pain, lower limb weakness	-	Complete
22	1975	Polkey	34	F	-	C6, C7/vertebral body	No neurological deficits, but neck pain after cervical injury	Posterior approach excision and fusion	Complete
16	1988	Naidu et al.	50	M	-	C3, C4/vertebral body, pedicle, and transverse process	Weakness in all limbs and burning sensation in both lower limbs	Posterior approach excision without fusion	Complete
1	1992	Barnowsky and Dalal	41	M	L4 root	L4	-	-	-
12	1994	Knapp et al.	65	F	-	L4, L5	-	-	Biopsy
18	1997	Nooraie et al.	46	M	-	T12, L1/vertebral body	Complaint of severe back pain after accident without abnormal neurological deficits	Posterior approach excision and fusion	Complete
13	1998	Ko et al.	-	-	-	T8	-	-	Operated
3	1998	Chang et al.	58	M	-	L4, L5/vertebral body	Severe pain and numbness of both lower extremities	1. Anterolateral retroperitoneal fusion and excision, 2. Posterior fusion	Complete
22	2000	Ramasamy et al.	37	M	-	T12/vertebral body	Back pain, weakness, and bilateral lower extremity numbness	Resection with anterior and posterior fusion	Complete
11	2001	Inaoka et al.	9	M	T10 root	T10/vertebral body and left transverse process	Nontender distention of the back without abnormal neurological deficits	Not mentioned	Complete
11	2001	Inaoka et al.	39	M	L5 root	L5/vertebral body and left transverse process	Moderate lumbar pain without abnormal neurological deficits	Not mentioned	Operated
24	2001	Schreuder et al.	39	F	-	C6/vertebral body	Neck pain and dysphagia	Anterior approach excision and fusion	Complete
15	2004	Mizutani et al.	73	F	-	C4	Discomfort in swallowing	-	Complete
17	2005	Nannapaneni and Sinar	42	M	-	C5/vertebral body	No neurological deficits	Anterior approach excision and fusion	Complete
26	2005	Singrakhia et al.	43	M	-	C3, C4/vertebral body	Increasing numbness around the right shoulder and deltoid weakness	Anterior approach excision and fusion	Operated
26	2005	Singrakhia et al.	45	M	-	C4/vertebral body and left transverse process	Progressive pain and weakness in the right upper limb	Anterior approach excision and fusion	Operated
10	2005	Gupta and Agarwal	30	F	-	L2/vertebral body	Complaints of backache and progressively increasing weakness in both lower limbs	Not mentioned	Complete

Table 1. Previous reports of SIS (continued)

Ref.	Year	Author	Age (y)	Sex	Origin	Level/original location	Symptom	Treatment	Resection
4	2007	Choudry et al.	30	M	-	T12/vertebral body	Back pain and deteriorating locomotor function, thinning of the leg	Thoracolumbar approach excision and fusion	Complete
19	2009	Park et al.	46	F	-	L4/vertebral body	Back pain and bilateral weakness in ankle dorsiflexion	Right transretroperitoneal approach excision and fusion	Complete
2	2009	Cetinkal et al.	55	F	-	T12/vertebral body, pedicle, lamina	Back pain and right leg pain and numbness	Posterior approach excision and fusion	Complete
15	2010	Mizutani et al.	44	F	-	C4/vertebral body	Bilateral upper limb paraesthesia	Right-sided anterior approach excision without fusion	Complete
14	2011	Kojima et al.	60	M	-	T9/lamina and spinous process	Back pain and progressive lower extremity weakness, bilateral lower extremity gait disturbance and numbness	Posterior approach excision and fusion	Complete
-	2012	Current study	71	M	-	L4/vertebral body and right transverse process	Right lumbocrural pain and gait disturbance, right leg paresthesia and weakness	Posterior approach excision and fusion	Complete
-	2012	Current study	54	F	-	T9/lamina and pedicle	Gait disturbance and bilateral leg paresthesia	Posterior approach excision and fusion	Complete

Table 2. Summary of SIS characteristics reported in Table 1

Cervical	7 (28%)
Thoracic	8 (32%)
Lumbar	9 (36%)
Originating from anterior elements	17 (68%)
Originating from posterior elements	2 (8%)
Excision with fusion	17 (68%)
Excision without fusion	2 (8%)

SIS : spinal intraosseous schwannoma

rettage beyond the original margins of the tumor without local adjuvant, which can result in long-term relief without a high recurrence rate[24]. Despite their tendency to be benign, SISs often result in spinal instability due to severe vertebral body invasion, and fusion with bone graft is required in approximately 68% of cases.

CONCLUSION

We report two cases of SIS, which is a rare differential diagnosis for intraosseous 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.

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