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Case Report

Spinal Intraosseous Schwannoma : A Case Report and Review

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A case of spinal intraosseous schwannoma (SIS) in the lumbar vertebra is reported. Clinical and radiologic characteristics of 16 reported cases of SIS were reviewed. SIS can be a rare differential diagnosis for intraosseous tumors.

KEY WORDS : Ancient · Intraosseous · Schwannoma · Spinal · Vertebral.

INTRODUCTION

Spinal schwannomas usually present as extramedullary, intradural tumors (70%)[14,25]. Intraosseous schwannomas are known to account for less than 0.2% of primary bone tumors[9]. Spinal schwannoma presenting as a vertebral intraosseous lesion is rare. We report a case of a spinal intraosseous schwannoma (SIS) of ancient type pathology associated with the expansile, osteolytic destruction of the entire vertebral body. Such presentations have rarely been reported in the 15 case reports[2,4-6,8,10,11,14,16,18,19,21-24,26,29,31-33].

An ancient schwannoma is a schwannoma with a long duration with degenerative change that is characterized by perivascular hyalinization, calcification, and cystic degeneration[15,28]. Ancient schwannomas are usually found in the head and neck, thorax, retroperitoneum, and lower extremities in elderly patients[15].

CASE REPORT

History and examination

A 48-year-old woman presented with back pain and bilateral grade III/IV weakness in ankle dorsiflexion. Magnetic resonance imaging and computed tomography (Fig. 1, 2) revealed a heterogeneously-enhancing, intraosseous tumor involving almost the entire L4 vertebral body. On T2-weighted imaging, the tumor was visualized as a well-encapsulated and lobulated mass, containing poorly-enhancing and high-intensity areas. The L4 vertebral body had developed a fracture because of the tumor. Since there was a sclerotic rim in the vertebral body surrounding the tumor, the radiologists suspected that the lesion was a slow-growing tumor such as an aneurysmal bone cyst and aggressive hemangioma, or a slow-growing metastatic lesion such as a thyroid tumor. Preoperative diagnosis of an ancient

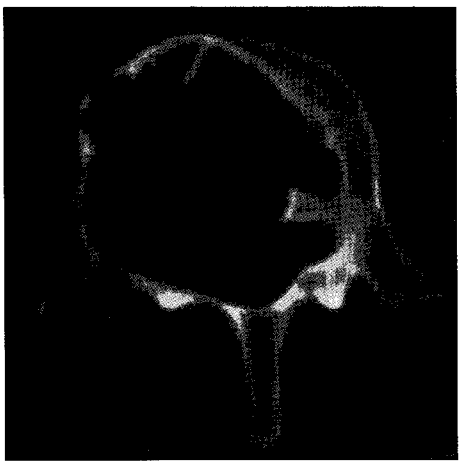

Fig. 1. Preoperative spine computed tomography reveals the completely intraosseous and expansile nature of the tumor.

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Fig. 2. T1-weighted, T2-weighted, and contrast-enhanced T1-weighted magnetic resonance imaging of the tumor. The poorly enhanced rim-like T2 high signal intensity areas represent the degenerated portion of the ancient schwannoma. Peripheral enhancement of the degenerative portion is found, and this is the cardinal radiologic characteristic of ancient schwannomas.

intraosseous schwannoma was difficult. The entirely intraosseous location hindered diagnosis of the schwannoma. The dural sac was severely compressed by the extradural tumor. Transfemoral spinal angiography revealed that the lesion was a hypovascular mass, and therefore, embolization was not done. No other hypermetabolic lesion was detected on whole-body and brain positron-emission tomography.

Operation

The tumor was successfully resected using a right transretroperitoneal approach in the left decubitus position. The tumor had caused thinning of the right-side wall of almost the entire L4 vertebral body. The tumor was exposed after the right vertebral wall was removed by a Kerrison punch. The tumor was fairly friable, soft, and grayish (Fig. 3). Frozen biopsy showed evidence of a low-grade spindle cell tumor. The tumor was well encapsulated and resected completely until the normal bone and disc space were exposed. A mesh cage with an allograft bone (45 cc) was inserted in place of the L4 vertebral body. Posterior lumbar interbody fixation in the L3 and L5 levels was performed in the prone position via a paramedian incision (Fig. 4).

Pathological findings

Gross pathological examination revealed that the tumor was composed of multiple gray-white glistening tissue. The tumor was pathologically classified as an ancient schwannoma. Histopathology revealed areas of nuclear palisading, with dense areas of tumor cells alternating with loosely textured myxoid tissue, which are consistent with Antoni type A and type B tissues, respectively (Fig. 5). Immunohistochemical testing for the S100 protein was diffusely positive, whereas those for Desmin and SMA were negative; these findings were suggestive of a schwannoma. Ki-67 was positive in less than 1% of the tumor cells, indicating that the lesion grows slowly. No malignant portion was noted in the specimen. Degenerative changes and thrombosed vessels were identified, thus confirming the diagnosis of ancient schwannoma.

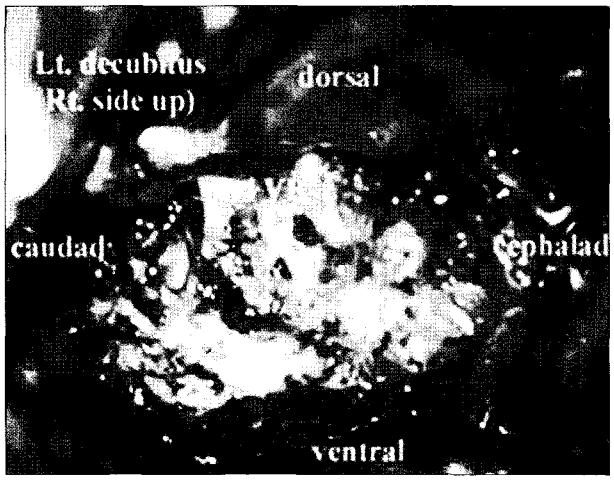

Fig. 3. Intraoperative photograph obtained after most of the film-like right L4 vertebral wall was removed by the punch. A friable, soft, and grayish mass with heterogeneity is found.

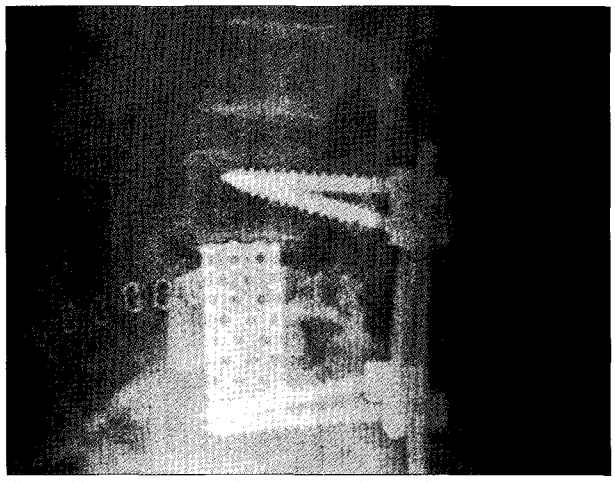

Fig. 4. After corpectomy and tumor removal, a mesh cage was inserted to the L4 body. Posterior lumbar interbody fusion was performed to the L3 and L5 body.

Postoperative course

Ten days after the operation, the patient's motor power for ankle dorsiflexion seemed to have improved to grade IV-/IV+. After three months, the patient's motor power improved, and she was ambulatory. Twenty-one months

after surgery, there was no evidence of recurrence clinically, and on a simple lumbar X-ray, the bony structure was not changed.

DISCUSSION

Method of review of spinal intraosseous schwannoma

An intraosseous schwannoma is a rare tumor. The first case of a SIS was reported from the Mayo Clinic by Cohen in 1964[6]. We retrieved 24 reported cases of SISs from the currently available English literature since 1960 with the search terms 'intraosseous', 'schwannoma', 'nerve sheath', 'vertebral invasion' and 'tumor' (Table 1)[2,4-6,8,10,11,14,16,18,19,21-24,26,29,31-33]. Tumors with pathologic diagnosis of schwannoma, nerve sheath tumor and malignant schwannoma were included[10,16,21,31]. Four cases of malignant peripheral nerve sheath tumors were excluded. Schwannomas with larger intraosseous portion than extraosseous portion were included[10,16,21,31]. However, 8 schwannoma cases with larger extraosseous portion than intraosseous portion were excluded, although intraosseous portions were present[3,30]. A case with insufficient clinical information was excluded[6]. The total number of cases included for the review was 16 including our case. Tumors were classified based on origin, level, border, location, size, completeness of resection and pathology.

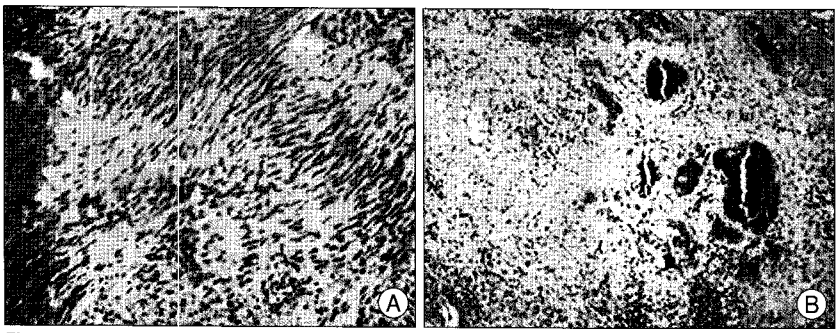

Fig. 5. Antoni A area (A) and Antoni B area with thrombosed vessels (B), a characteristic of ancient schwannomas (hematoxylin & eosin; ×200).

Demographics

Mean age of SIS patients was 41.1 years. There were 9 male patients and 6 female patients. In one report, age

Table 1. Spinal intraosseous schwannomas

No.	Year	Series	Age (y)	Sex	Origin	Level	Border	Size (cm)	Resection	Modified Sridhar classification	Pathology
1	1971	Dickson	51	F	-*	L3	Smooth	6×4.5×3	Complete	Type VII	Benign
2	1975	Polkey	34	F	-	C6, C7	Irregular	2.5×2.0	Complete	Type VII	Benign
3	1988	Naidu	50	M	-	C3, C4	Irregular	-	Complete	Type VII	Benign
4	1992	Barnowsky	41	M	L4 root	L4	Irregular	4.5	-	Type VII	Benign
5	1994	Knapp	65	F	-	L4, L5	Irregular	16×14×14.5	Biopsy	Type VII	Cellular schwannoma or sarcoma
6	1997	Ncoraie	46	M	-	T12, L1	Smooth	4×3	Complete	Type VII	Benign
7	1998	Ko	-	-	-	T8	Irregular	2.5	Operated	Type VII	Malignant
8	1998	Chang	58	M	-	L4, L5	Smooth	4×3	Complete	Type VI	Schwannoma with hemosiderin deposition, thrombosed vessels, and degenerative area (possibly ancient schwannoma)
9	2001	Inaoka	9	F	T10 root	T10	Irregular	4×3	Complete	Type VII	Benign neurinoma
10	2001	Inaoka	39	M	L5 root	L5	Smooth	5.5×5	Operated	Type VII	Benign neurinoma
11	2005	Nannapaneni	42	M	-	C5	Irregular	2×2×4.5	Complete	Type VI	Schwannoma with thrombosed vessels (possibly ancient schwannoma)
12	2005	Singrakhia	43	M	-	C3, C4	Irregular	3.5×2	Operated	Type VII	Benign
13	2005	Singrakhia	45	M	-	C4	Irregular	2.8×2.7	Operated	Type VII	Benign
14	2005	Gupta	30	F	-	L2	Irregular	5×3	Complete	Type VII	Benign
15	2006	Choudry	18	M	-	T12	Irregular	5.4×4×8	Complete	Type VII	Benign
16	2009	Park	46	F	-	L4	Smooth	3×3.5×2.5	Complete	Type VI	Ancient schwannoma

*Unknown or unspecified

and gender were not mentioned[19]. Slight male predominance and mean age was not different from frequency of male (61.2%) and mean age (44.3 years) of non-intraosseous schwannoma[7].

Origin of spinal intraosseous schwannoma

In the 3 cases cited in the literature, the sites of the tumor origin were the nerve roots or foramen[2,14,29]. In most cases exact origin of tumor could not be identified, however the fact that most SISs have some extraosseous portion suggests that origin of SIS is probably intraosseous invasion of extraosseous nerve sheath tumor.

Level of spinal intraosseous schwannoma

Most frequent level of SISs was lumbar region (44%). In general, thoracic level SISs were less frequent (25%) and cervical region SISs (31%) were more frequent despite the fact that thoracic vertebra is the longest segement. In previous non-intraosseous spinal schwannoma series, lumbar region was the most common location, and thoracic spinal schwannoma was two times more common than cervical schwannoma unlike SISs[7].

Border of spinal intraosseous schwannoma

Irregular border of spinal intraosseous schwannoma was once suggested as an evidence of higher invasive potential of SIS[13]. In our review, 11 SISs had irregular borders and incidence of irregular border was higher than schwannomas in other location suggesting more invasive characteristics of SISs[13].

Locations of SISs and suggestion of modification of benign spinal schwannoma classification to include SISs

In 2001, Sridhar suggested a classification system of benign spinal schwannoma including giant and invasive spinal schwannomas (Type I to V) (Table 2, Fig. 6) In the classification system of Sridhar, type V is nerve sheath tumor with erosion into vertebral body and lateral and posterior extension into myofascial planes[30]. Sridhar Type V is the only one type with characteristic of intraosseous schwannoma, invasion into vertebral body. However, this type also has large extraosseous portion and, mainly intraosseous schwannomas could not be properly classified based on Sridhar's classification. Thirteen of 16 SISs had vertebral body invasions and extraosseous portions in spinal canal and neural foramen, however lateral and posterior extensions into myofascial plane were not significant. Three SISs

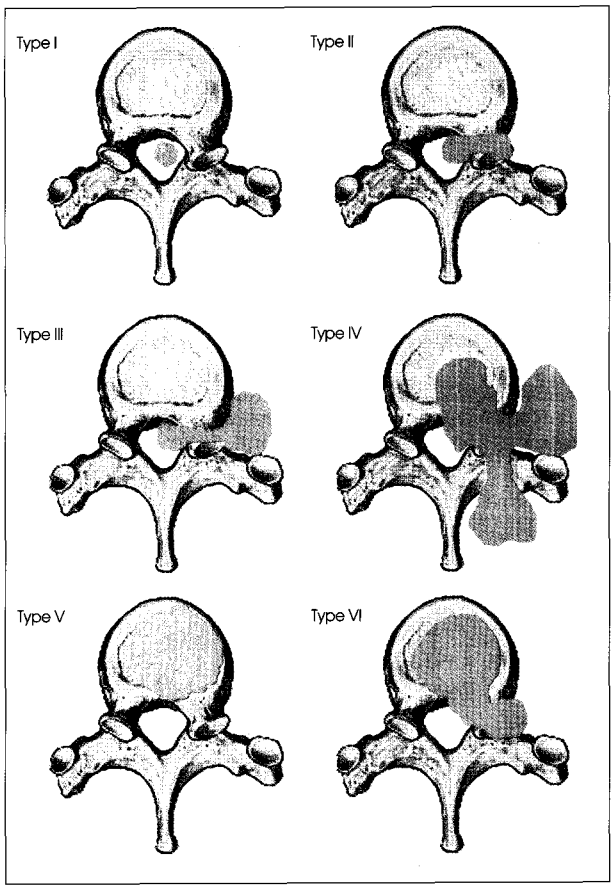

Fig. 6. Diagrammatic representation of the proposed modified classification of spinal schwannomas Types I to VII. Types I to V are identical to original classification of spinal schwannomas suggested by Sridhar. Types VI and VII are additional types in modified classification of spinal schwannomas (Table 2). Type II is not shown.

Table 2. Modified Sridhar classification of benign nerve sheath tumors

Original types	
Type I	Intraspinal tumor, < 2 vertebral segments in length; a : intradural; b : extradural
Type II	Intraspinal tumor > 2 vertebral segments in length (giant tumor)
Type III	Intraspinal tumor with extension into nerve root foramen
Type IV	Intraspinal tumor with extraspinal extension (dumbbell tumors); a : extraspinal component < 2.5 cm; b : extraspinal component > 2.5 cm (giant tumor)
Type V	Tumor with erosion into vertebral bodies (giant invasive tumor), lat & posterior extensions into myofascial planes
Additional types for spinal intraosseous schwannoma	
Type VI	Tumor in entirely intravertebral location without intraspinal portion
Type VII	Intraspinal tumor with erosion into vertebral bodies (invasive tumor) and extension into nerve root foramen

including our case were in almost entirely intraosseous intravertebral location without lateral or posterior extension. Thus, 16 SISs could not be classified based on Sridhar's spinal schwannoma classification system[4,23].

Therefore, we suggest two additional categories, type VI and type VII (Table 2, Fig. 6). Type VI is entirely intraosseous schwannoma without intraspinal portion. Type VII is intraspinal tumor with vertebral body invasion and extension into nerve root foramen.

The classification system including additional two categories was mentioned as 'modified Sridhar classification of benign nerve sheath tumor' and used for classification of reviewed SISs (Table 1). Our case was type VI, entirely intraosseous schwannoma.

Size of spinal intraosseous schwannoma

Sizes of intraosseous schwannomas were larger than non-intraosseous schwannomas. Five SISs spanned two vertebral bodies[4,18,22,24,26]. Criteria for giant schwannoma was suggested by Sridhar[30]. Intraspinal schwannomas spanning two vertebral bodies (Type II) and intraspinal tumor with extraspinal component > 2.5 cm are classified as giant schwannoma in Sridhar's classification. However, SISs are not included in Sridhar's classification and we suggest criteria of giant SIS would be SIS with more than two vertebral segments in length. Following this criteria, 5 among 16 cases are giant SIS.

Resectability

Ten of the 16 SISs were removed totally. SISs are not generally easy to remove because they have both intraosseous and extraosseous portions invading adjacent structures, including nerve roots, spinal cord and paravertebral tissue. This difficulty in resection was mentioned in similar tumors in location such as extradural schwannoma and giant invasive schwnnoma[3,30]. In addition, after removal of tumor, instability is frequently caused because of vertebral body invasion. Thus, as shown in our case, aggressive surgical approach and fusion were frequently required.

Pathology

All tumors except one case were benign schwannomas[19]. One ancient schwannoma, presenting case and one cellular schwannoma were found[18].

Our case was an ancient pathologic type SIS. In addition, lesions in the other 2 cases had ancient schwannoma features because the reports indicated thrombosed vessels, and 1 exhibited degenerative changes in the Antoni B areas[4,23]. An ancient schwannoma is a subtype of schwannoma characterized by degenerative change and is frequently large[1]. This subtype is believed to develop over a long period of time[12,17]. Consequently, In cases of intraosseous ancient schwannoma, long period of growth in intraosseous location might be related to ancient change and these three tumors with ancient schwannoma features were entirely intraosseous modified Sridhar type VI tumor. There were two case reports about malignant transformation of ancient schwannoma[20,27]. However, there was no malignant feature in our case. The ancient schwannoma in this report showed a rim-like degenerative portion and enhancement around it (Fig. 2). This was mentioned as the most accurate sign of an ancient schwannoma; however, this could not be a firm clue to exclude other diagnoses[15].

Formerly, the cellular type of schwannoma was known to have a higher rate of bone involvement compared to other types (19%)[18,34,35]. There was one possible cellular SIS[18]. There are pathologic similarities between a cellular schwannoma and an ancient schwannoma, including nuclear atypia and hypercellularity in both subtypes[12,17]. Common features of an ancient schwannoma and a cellular schwannoma might be the cardinal, characteristic pathological features of SISs because of the peculiar intraosseous environment.

CONCLUSION

We report a case of the ancient type of SIS. SIS can be a rare differential diagnosis for intraosseous tumor. Modification of classification of benign spinal schwannoma is suggested to include mainly intraosseous spinal schwannomas.

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