

Case report

Ancient Schwannoma as an exceptional thoracic spinal tumor: A case report

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HIGHLIGHTS

- Schwannomas are the most common intradural extramedullary tumors of the spine.
- Ancient schwannoma is an uncommon variant of schwannomas.
- Ancient schwannomas infrequently occur in the spine.
- Ancient schwannoma of the thoracic spine is extremely rare.

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ABSTRACT

Schwannomas are mostly benign tumors arising from the nerve sheath. These tumors can be found anywhere in the body. Depending on their locations, they may cause compressive symptoms as well as cosmetic or functional defects. Ancient schwannomas, the rare variant of schwannomas, are the slow-growing tumors characterized with cystic necrotic degeneration areas in the neoplastic tissue. Ancient schwannomas rarely occur in the spinal canal, they are particularly unusual in the thoracic spine. Herein we present a 66-year-old woman with chronic back pain who is detected a cystic mass in her thoracic spine by magnetic resonance imaging and is diagnosed with ancient schwannoma by histological examination.

1. Introduction

Schwannomas, also called neurilemmoma are benign tumors which originate from embryonic neural crest cells of the nerve sheath. It has been reported that these tumors usually occur in the fourth to fifth decades with a slightly female preponderance [1]. Schwannomas can emerge anywhere along the neural network, however, the head, the flexor surfaces of extremities, and the trunk are mainly involved. Although the spine is not among the most frequent sites for schwannomas, spinal schwannomas are the most common intradural extramedullary (IDEM) tumors and they constitute approximately 30% of all intradural tumors [2]. These tumors are relatively common in the cervical and lumbar spine, but are extremely rare in the thoracic spine. The clinical picture is variable according to site of the tumor, however, back pain and radicular symptoms are usually prominent.

As distinct from common schwannomas, ancient schwannomas show multiple degenerative changes including calcification, cyst formation,

hemorrhage, and perivascular hyalinization. Magnetic resonance imaging (MRI) may demonstrate the degeneration areas within these tumors. Nevertheless, ancient schwannomas have neither clinical nor radiological specific features. Therefore, the definitive diagnosis is often required histological examination. We aimed to report this case with spinal mass that creates diagnostic difficulty, and is finally diagnosed with ancient schwannomas by surgical resection.

2. Case report

A 66-year-old female patient who has osteoporosis had recourse to the Endocrinology department with complaints of back pain. She stated that the pain was radiating to the left side and gradually worsening in recent months. Furthermore, the back pain had not improved although she had undergone left nephrectomy due to the disease of kidney stone about 1 year ago. The patient was taking alendronate 70 mg once a week for osteoporosis. Her family history was unremarkable. On physical

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examination, there was no pathological finding except the scar of incision on the left lumbar region and mild left costovertebral angle tenderness. Neurologic examination revealed negative babinski sign and normal deep tendon reflexes on both lower limbs. There were also neither sensory loss nor motor neurologic deficit. The comprehensive laboratory investigations also including the levels of serum creatinine, calcium, phosphate, parathyroid hormone, thyrotropin and thyroid hormones were normal. Lumbar spine X-ray only demonstrated degenerative changes of the vertebral bodies. On dual-energy X-ray absorptiometry, T-scores of both the lumbar spine and proximal femur were less than -3 as compatible with osteoporosis. Thereupon, MRI of the lumbar and thoracic spine was performed taking into account the possibility of nerve root compression or pathologic fracture of the vertebrae associated with osteoporosis because of she had a chronic intractable back pain.

MRI scans revealed a 35×9 mm sized mass occupying the spinal canal at T8-T9 level. The mass which is a well-circumscribed IDEM lesion was hypointense on T1-weighted images and heterogene hyperintense on T2-weighted images. The lesion that is compressing to the spinal cord from posterior showed peripheral rim enhancement around the central cystic area after contrast media injection (Figure 1). Thereupon, surgical excision was decided to provide the decompression of the spinal cord. Encapsulated cystic tumor was removed with en bloc resection following T8-T9 laminectomy. The operation was completed without any complications. The postoperative course was uneventful and she was discharged within a few days.

Histopathological examination revealed that the tumor consisted of elongated spindle cells with vesicular nucleus arrayed in a palisade pattern among of the large cystic cavities which are lined by single-layer squamous epithelium (Figure 2-A). Markedly nuclear pleomorphism was also exist in some of tumor cells. Immunohistochemical analysis

disclosed that the neoplastic cells were stained negatively for epithelial membrane antigen, pancytokeratin and desmin but diffuse positive for protein S-100 (Figure 2-B). Ki67 proliferation index was 1 %. As a result of these studies, ancient schwannoma was diagnosed.

3. Discussion

Schwannomas are nerve sheath tumors arising from Schwann cells. They are slow growing benign tumors and infrequently undergo malignant transformation. These tumors consist two types of the area of spindle cells as histopathologically: Antoni A and Antoni B. Antoni type A areas are highly cellular and composed with the compact bundles of spindle cells with the nuclei arranged in a palisade pattern. On the other hand, Antoni type B areas are characterised with hypocellular zones surrounded by a loose myxoid matrix. This difference also reflect to MRI viewing of schwannomas: Antoni type A areas usually reveal a strong gadolinium enhancement in contrast to Antoni type B areas which is lack of significant gadolinium enhancement.

Ancient schwannoma is an uncommon variant of schwannomas and accounts for less than 1% of all schwannomas [3]. As a probable consequence of their slow growth rate, ancient schwannomas display some degenerative changes histopathologically such as calcification, cystic necrosis, hemorrhage, intravascular thrombosis, perivascular hyalinization, and relative loss of Antoni type A areas in course of time. Because of these degenerative features and relatively high proportion of Antoni type B areas, ancient schwannomas show heterogeneous contrast enhancement in MRI. In our case, a cystic IDEM lesion that show peripheral rim-enhancement after gadolinium injection was also detected. Because of the atypical pattern of contrast enhancement in their MRI viewing, ancient schwannomas may be mistaken with various malignant

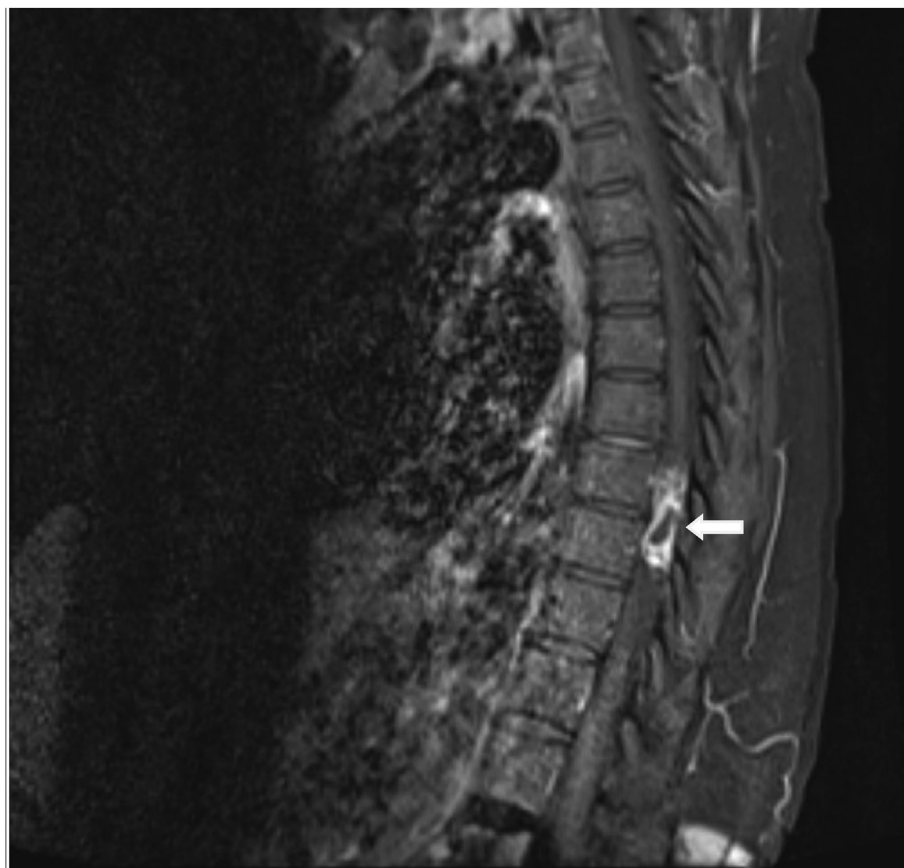
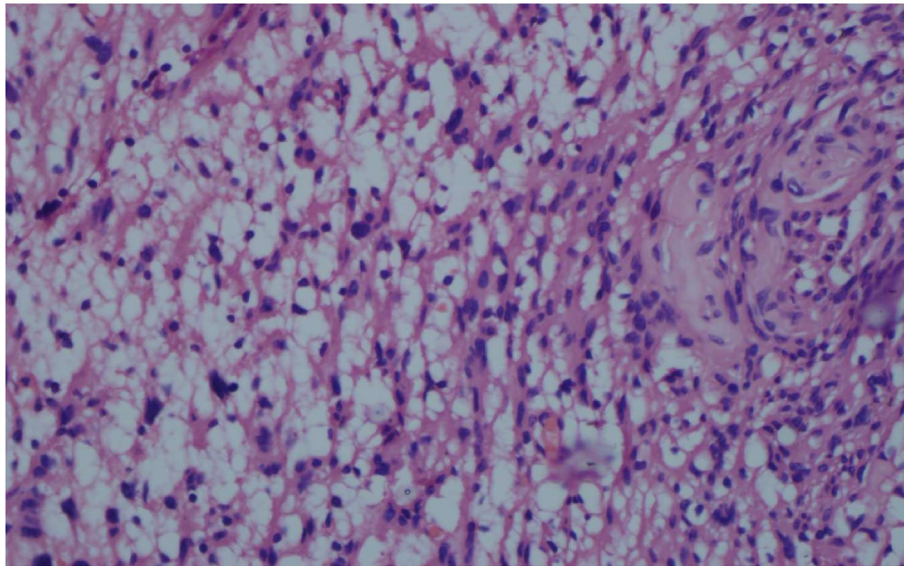


Figure 1. The cystic lesion that occupies the spinal canal at T8-T9 level and shows peripheral rim-enhancement after contrast media injection on T1-weighted sagittal image (white arrow).

a) H&E stained section of tumour showing elongated spindle cells with loose architecture (x40)



b) Diffuse and strongly positive immunohistochemical staining for S-100 protein (x20)

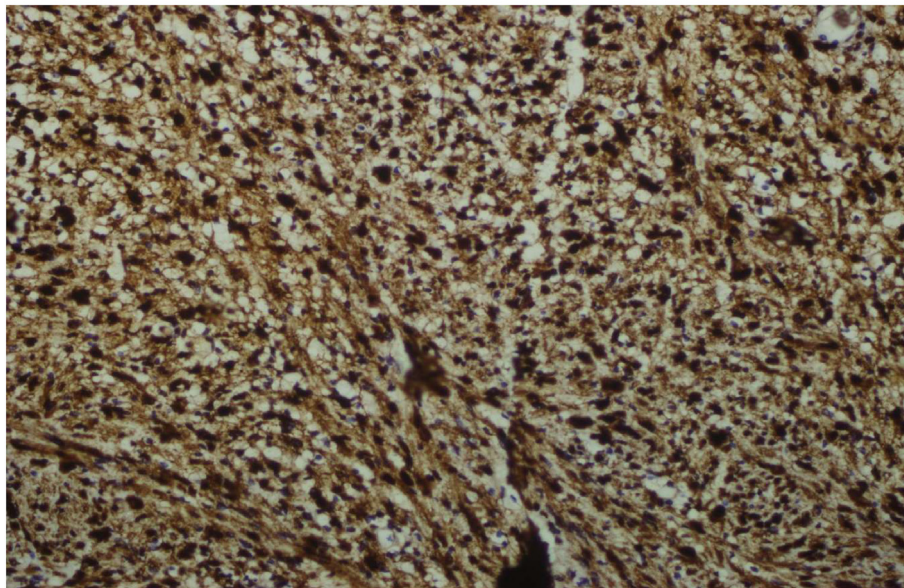


Figure 2. Histopathological view consistent with ancient schwannoma. a) H&E stained section of tumour showing elongated spindle cells with loose architecture (x40). b) Diffuse and strongly positive immunohistochemical staining for S-100 protein (x20).

mesenchymal neoplasia [4]. Other tumors setting in the spine such as ependymoma, cystic meningioma, cystic teratoma, and cystic lymphangioma should be also considered in differential diagnosis of spinal ancient schwannomas.

The clinical picture mostly varies depending on the location of the tumor. It can be occurred non-specific signs due to the compression of the surrounding tissues by the mass. The diagnosis may delay many years because these tumors usually have an indolent clinical course. Similar to other spinal tumors, ancient schwannomas setting in the spine may also imitate the findings of vertebral fracture or disc herniation via lead to localized or radicular pain. There were some misleading comorbidities such as osteoporosis and kidney stone in medical history of our case too.

Total excision of the mass is essential both to achieve the definitive diagnosis and to relieve the compressive symptoms if any. On the other hand, it must also be paid attention to safety of vital neurovascular structures in the spine during the surgery. Standard posterior midline approach with laminectomy is the preferable surgical technique in the most of case with spinal ancient schwannomas.

Unlike the common schwannomas, the characteristic histopathological feature of ancient schwannomas is the hypocellular areas which show markedly degenerative changes including cyst formation, hemorrhage, calcification, and necrosis. The spindle-shaped cells usually are surrounded with a loose extracellular stroma. Immunohistochemically, ancient schwannomas display the reactivity to protein S-100 likewise common schwannomas.

The long-term clinical outcome of spinal schwannomas is quite satisfying because of the benign biological behavior of these tumors if the mass can be totally removed [5]. The recurrence usually is not expected except schwannomas associated with neurofibromatosis syndromes. The mass was also completely excised in our case by posterior midline approach with T8-T9 laminectomy and there was no recurrence in the course of 12 months follow-up.

4. Conclusion

Ancient schwannoma is a rare clinical entity and it is exceptional in the thoracic spine. Due to the absence of any specific signs both clinically and radiologically for ancient schwannomas, many diseases may imitate these tumors. Despite their rarity, ancient schwannomas should also be taken into account in the differential diagnosis of spinal tumors.

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Author contribution statement

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Data will be made available on request.

Declaration of interests statement

The authors declare no conflict of interest.

Additional information

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