

Paraspinal Ancient Schwannoma of the Dorsal Ramus Nerve - A Case Report -

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Study Design: Case report.

Objectives: We report a case of paraspinal ancient schwannoma located at the upper thoracic level that mimicked an atypical lipoma or complicated epidermoid cyst.

Summary of Literature Review: Few case reports of paraspinal schwannoma have been reported and the incidence of ancient schwannoma in the paraspinal muscle layer is very rare.

Materials and Methods: A 39-year-old man complained of a growing palpable back mass for 5 years. He experienced aggravated chronic discomfort around the mass while lying down. Both T1- and T2- weighted magnetic resonance imaging (MRI) showed a well-capsuled and heterogeneous high-signal mass in the muscle layer at the level from the T1 to T4 vertebral bodies on the right side of the midline. The tumor was completely removed by en bloc resection.

Results: The pathologic examination revealed S-100 protein expression with degenerative changes. The lesion was diagnosed as an ancient schwannoma.

Conclusions: Schwannoma is one among the multiple possible causes of benign back masses. If a mass reveals a well-encapsulated heterogeneous mass on contrast MRI, a schwannoma should be suspected.

Key Words: Schwannoma, Back muscles

Schwannomas are mostly benign, slow-growing and solitary tumors arising from the Schwann cells in nerve sheath. It is assumed that 25–40% of schwannomas occur in the head and neck region.¹⁾ Although schwannomas arising from the dorsal ramus nerve are uncommon,^{2,3)} ancient schwannoma is extremely rare. We present an ancient schwannoma originating from the dorsal ramus nerve presenting as a large back mass in a 39-year-old man.

Case Report

This case report has been approved by the Institutional Review Board of Our Hospital. (IRB number: SCHUH 2019–14)

A 39-year-old male came to the outpatient clinic with palpable back mass on his right side of upper thoracic area. The mass, which had been growing slowly for five years, didn't induce severe pain, but caused him discomfort while lying down. There was no preceding illness and history of malignancy. On examination, the mass was firm, not

so movable and well-circumscribed. Also, there was no tenderness nor heating sense on palpation.

An MRI of the thoracic spine with contrast showed an isolated oval-shape mass measuring 5 x 4.5 x 9 cm. The mass was located in the paraspinal muscle and had well-circumscribed margin. T1 and T2 weighted images revealed heterogeneous high signal intensity from T1 to T4 vertebral bodies on right side of midline (Fig. 1). The preoperative diagnosis would be atypical lipoma or complicated cyst

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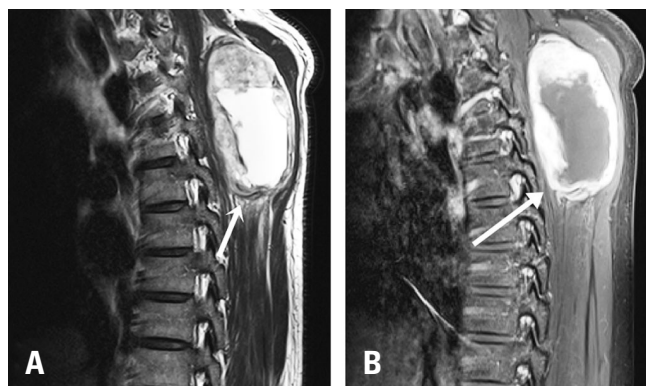


Fig. 1. Magnetic resonance imaging showing (A) a well-defined, irregular thick-walled cystic lesion and (B) central non-enhancement with peripheral irregular heterogeneous enhancement.

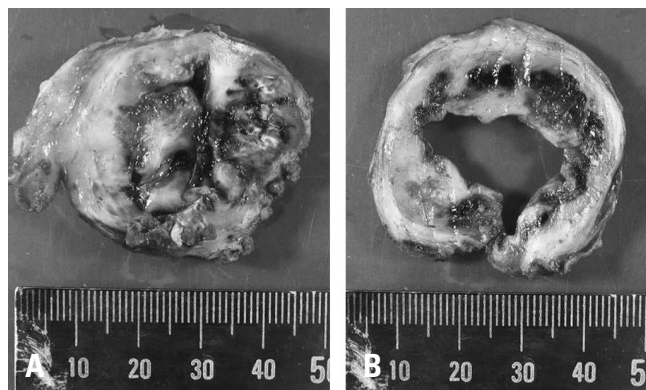


Fig. 2. The excised mass. (A) The mass was solitary, well circumscribed, and capsulated. It presented yellow-to-gray coloration, myxoid and soft stromal areas, and multiple tan/brown hemorrhagic foci. (B) On a cut section, cystic changes were more evident.

such as complicated epidermoid cyst or abscess.

The patient underwent open excision of the mass under general anesthesia. The mass was located on the right lamina of T2 and extended to the right transverse process.

The gross feature of the tumor was a solitary, well circumscribed and capsulated mass with a smooth surface. On cut section, it showed soft texture, yellow to gray colored, glistening stromal areas and multiple hemorrhagic foci with central cystic degeneration (Fig. 2). In the central portion, cellular component of the tumor was not identified and replaced with edematous, focally hyalinized and pinkish extracellular components at the low magnification (Fig. 3A). In addition, degenerative changes such as central cystic cavity, vascular hyaline change and intra- and extravascular thrombi were identified (Fig. 3B). In the peripheral portion of the tumor,

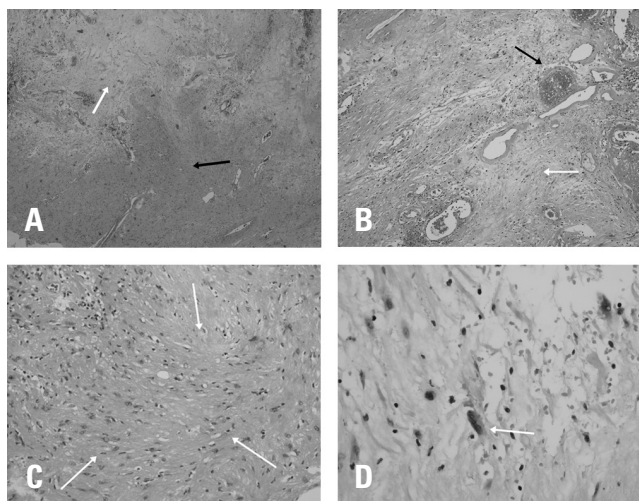


Fig. 3. Microscopic images showing (A) a hypocellular area in the central region (white arrow) and a more cellular area in the peripheral region (black arrow), but less cellular than a typical schwannoma (hematoxylin and eosin [H&E], $\times 40$). Both hyaline vascular changes (white arrow) and thrombi (black arrow) were present (H&E, $\times 100$). (C) The typical Antoni A area was not identified; instead, only a vague Antoni A-like area was identified in this case (H&E, $\times 200$). (D) Cellular features included bizarre nuclei (H&E, $\times 400$).

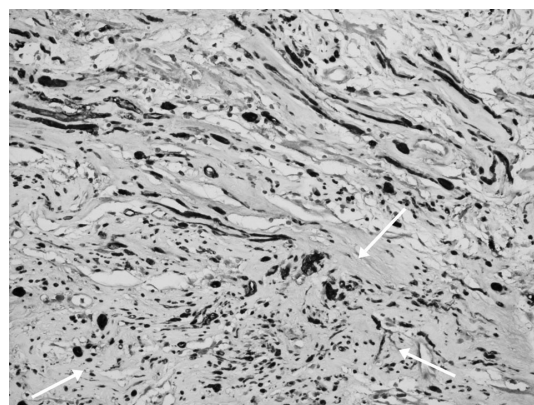


Fig. 4. Immunohistochemistry revealed diffuse, high S-100 protein expression.

a few cellular components were found and characterized by vague nuclear palisading with pinkish fibrillary cytoplasm, showing Antoni A-like arrangement (Fig. 3C). The cells with wavy-tapering nuclei and pinkish cytoplasm, which are typical features of the nerve sheath tumor, were identified in part. Hyperchromatic and bizarre nuclei were identified within the focal area of the tumor (Fig. 3D).⁴⁾ For the histologic confirmation of schwannoma, S-100 protein, a positive staining marker in the Schwann cell, was highly stained in the tumor cells (Fig. 4).

There was no recurrence within 1 year of follow-up.

Discussion

The most common types of benign back masses are lipoma, fibrous histiocytoma, nodular fasciitis, hemangioma, and schwannoma in regular sequence. Among these benign tumors, the chance of schwannoma is slim (about 5%).⁵⁾ Schwannomas are typically benign, slow-growing and solitary neoplasms of Schwann cell origin. They were first described by Verocay in 1908 and have since been given several names although schwannoma and neurilemmoma are the most common terms.⁶⁾

In spine area, schwannomas may occur at any level of the spinal axis but are mostly located in dura. However only a few cases of purely intramuscular paraspinal schwannoma have been reported previously^{2,3)} and only one ancient schwannoma was reported.⁷⁾ All of these extraspinal schwannoma in paraspinal region had no neurologic symptom. Because intramuscular Schwannomas originate from motor branch (dorsal ramus) nerves, neurological symptoms including pain, motor weakness, and paresthesia etc., are very rare.³⁾ Also these paraspinal schwannoma had a common feature which is well-capsulated heterogeneous mass on enhanced MRI.

These MRI findings could be confused with atypical lipoma or complicated epidermal cyst. Atypical lipoma was introduced to indicate the relatively benign nature of well-differentiated liposarcomas in the extremities. Thickened or nodular septa, associated nonadipose masses, prominent foci on high T2 signal, and prominent areas of enhancement are all findings reported to be suspicious of liposarcoma.⁸⁾ Epidermal cyst has traditional characteristics such as a fluid signal with variable low-signal components on T2-weight images and peripheral rim enhancement on enhanced images. Most complicated epidermal cysts show septation with thick and irregular rim enhancement in surrounding subcutaneous tissues.⁹⁾

Common histologic finding of schwannoma, although not specific, is the presence of Antoni A and Antoni B area. Antoni A area is characterized by centrifugally compacted arrangements of spindle cells and Verocay bodies while type B Antoni are characterized by the lack of tissue cellularity and myxoid with loosely arranged spindle cells.⁴⁾

An ancient schwannoma, introduced by Ackerman and Taylor, reveals partly loss of Antoni A areas with irregular nuclei and areas of hyalinization with hyperchromatism suggestive of degenerative changes.¹⁰⁾ In our case, significant degrees of degenerative change are identified such as cells with irregular shaped, bizarre and some hyperchromatic nuclei, cystic change, stromal edema, and hyaline change of vascular structure and formation of thrombi.

Schwannomas less than 1% become malignant, degenerating into a form of cancer known as neurofibrosarcoma.³⁾ Even though low chance of malignant transformation, surgical open excision is the treatment of choice for schwannoma.

Ancient schwannoma of the back is extremely rare. We present the first patient of a huge ancient schwannoma within the erector spinae muscles. Because of the rarity of intramuscular ancient schwannoma, the diagnosis can be easily overlooked. Therefore, we should check MRI with enhancement or other radiologic examination when palpable back mass is found. If the histologic findings suggested the presence of type A or B Antoni and the S-100 protein is expressed, clinicians should consider the possibility of schwannomas originating from dorsal ramus.

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후가지 신경으로부터 발생한 척추 주위 고대 신경초종 - 증례 보고 -

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연구 계획: 증례 보고

목적: 비전형적 지방종 또는 표피낭종과 유사한 상부 흉추부 주위 고대 신경초종 사례를 보고하고자 한다.

선행 연구문헌의 요약: 척추 주변부에 발생한 신경초종의 사례는 몇 예가 보고되기는 하였다. 그러나 척추 주변 근육 내에 발생한 고대 신경초종의 사례는 거의 없었으며, 매우 드물다.

대상 및 방법: 39세 남자환자는 5년 동안 점차 커지는 등의 종양을 호소하였다. 특히 누울 때 종양 주변부로 만성적인 불편감을 호소하였으며, 점차 심해지는 경향을 보였다. MRI상 T1 및 T2 증강 영상에서 흉추 1번에서 4번 부위 우측 척추 근육 내에 비균일한 고강도 신호를 보이는 종양이 관찰되었다. 그 종양은 일괄 절제술로 완전히 제거되었다.

결과: 병리조직검사 결과에서 세포의 퇴행성 변화와 함께 S-100 단백질 검출되면서 상기 병변은 고대 신경초종으로 확진되었다.

결론: 신경초종은 등에 발생하는 양성 종양의 많은 원인들 중 하나이다. 특히 조영 증강 MRI에서 종양이 주변 벽에 의해 잘 싸여있으면서 종양 실질의 비균일한 고강도 신호소견이 있을 때, 신경초종을 의심해야 한다.

색인 단어: 신경초종, 등 근육

약칭 제목: 척추 후가지 신경의 고대 신경초종

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