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Primary Solitary Fibrous Extradural Tumor of Thoracic Spine: A Case Report

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ABSTRACT

Background Data: Solitary fibrous tumor/hemangiopericytoma (SFT/HPC) is a rare central nervous system (CNS) tumor. Its incidence in the spine is even rarer. It is diagnosed histopathologically as preoperative imaging reveals no pathognomonic features. It is most common in the thoracic spine and may have a poor prognosis despite surgery and adjuvant therapy. We present a case of thoracic extradural spinal primary SFT to be added to the list of the few reported cases.

Purpose: To report a rare tumor of extradural SFT in the thoracic spine.

Study Design: A case report.

Patient and Methods: We report one of a middle-aged female who presented with sensory and motor weakness for 2 months. With relevant work-up and an MRI imaging, an extradural lesion was noted in the spine. The patient underwent an en bloc resection of the lesion with a smooth postoperative course and the specimen was analyzed for histopathology.

Result: The histopathological analysis of the specimen showed features suggestive of a SFT; immunohistochemistry (IHC) was correlated with the histopathological diagnosis. At the 3-month follow-up, the patient had improvement in the power of the lower limbs and spasticity from grade 7/11 to grade 10/11 according to thoracic myelopathy JOA score.

Conclusion: SFTs are rare tumors whose clinical course is unpredictable. They commonly occur in intradural extramedullary and rarely extradural sites. The goal of treatment is complete excision. (2020ESJ212)

Keywords: Solitary fibrous tumor, Hemangiopericytoma, Spine, Rare, Extradural.

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INTRODUCTION

Solitary fibrous tumor/hemangiopericytoma (SFT/HPC) is a rare CNS tumor described by Stout and Murray in 1942.[1,16,17] They are thought to arise from pericytes surrounding the blood vessels (pericapillary cells or Zimmerman's pericytes), with a monomorphic population of compact polygonal or fusiform cells and branching stromal vascular pattern (dilated slit-like "staghorn" blood vessels surrounded by network of reticulin fibers).[5] Although commonly seen in the chest and abdomen, it can occur anywhere in the body.[14,18] It accounts for less than 1% CNS tumors and 2%–4% of primary meningeal tumors. [3,5,8] To the best of our knowledge, 185 cases of SFT/HPC in the spine have been reported in English literature, most of which are intradural extramedullary.[5] They can occur as either primary tumors of the spine or metastatic lesions, which are localized to the vertebral body, or more commonly can be adherent to the meninges.[5] Due to its rarity, limited information exists with regard to its natural history and management. This is supported by most of the reported cases mostly published as case reports. We report a single case of primary solitary fibrous extradural tumor of the thoracic spine.

CASE REPORT

A 44-year-old female presented with backache associated with progressive weakness in both lower limbs and hypoesthesia and paresthesia below the 5th rib for 2 months. On clinical examination, the patient had spastic paraparesis with a 7/11 score according to the thoracic myelopathy Japanese Orthopedic Association Score (JOA) with superficial sensory loss below the 5th rib without any bowel and bladder involvement. A contrast-enhanced Magnetic Resonance Imaging (MRI) showed a well-defined extradural lesion measuring 61, 24, and 10 millimeters in dimensions in the posterior epidural region extending from D2 to D5. It also extended into left D3-D4 and left D4-D5 neural foramina. It was hypo- to isointense on T1 and hyperintense on T2 weighted images MRI with a near homogenous enhancement of contrast. The lesion caused the anterior displacement of the cord with severe thinning and altered intensity within the cord (Figure 1). Positron Emission Tomography (PET) scan was conducted which ruled out metastasis.

The patient underwent a D2–D4 laminectomy in a supine position under general anesthesia and fluoroscopic guidance. A long spindle-shaped globular mass with a smooth surface in the extradural space, which was moderately vascular and adherent to the dural surface with a definitive plane of cleavage between dura and tumor, was observed. En bloc resection of the tumor was achieved (Figure 2A). The patient showed no neurological deterioration in the immediate postoperative period. Postoperative MRI showed complete excision of the lesion (Figure 2B). Histopathology revealed sheets of fascicles of spindle cells with mixed cellular architecture. Hyalinized collagenous stroma was seen. The cells were described as oval- to spindle-shaped with vesicular nuclei and scanty cytoplasm (Figure 3). The bony tissue in the proximity of the lesion on analysis was free of tumor tissue. IHC was conducted, showing a positive correlation with vimentin, CD34, and MIB-1. This suggested a diagnosis of SFT grade 1. At a 3-month follow-up, the patient had improvement in the power of the lower limbs and spasticity with a 10/11 score according to the JOA score at the last follow-up.

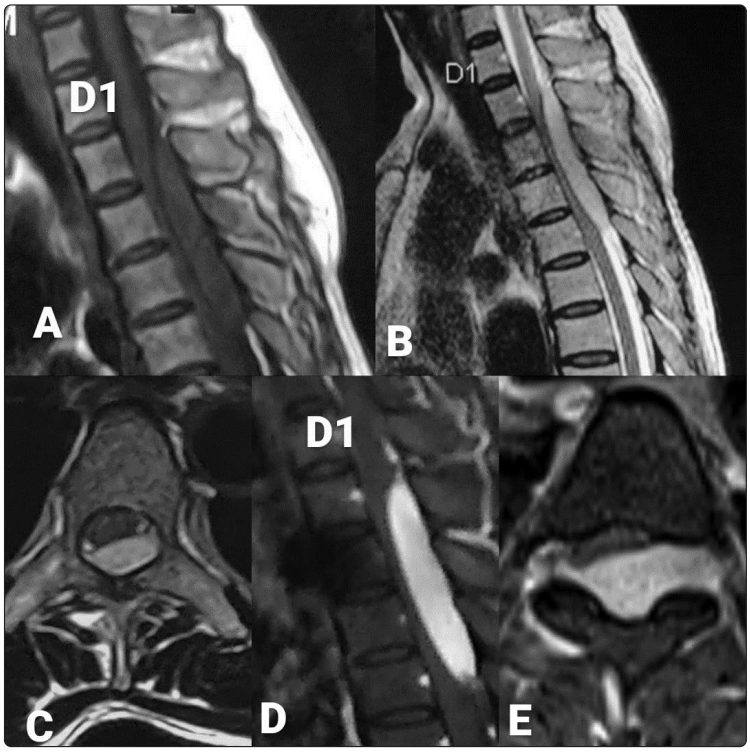

Figure 1. MRI of the thoracic spine: (A) Sagittal section T1 hypo- to isointense extradural lesion from D2 to D5. (B) Sagittal section T2 hyperintense lesion with anterior displacement of the cord with severe thinning and altered intensity within the cord. (C) Axial T2 showing compression with anterior displacement of cord at level. (D) Sagittal T1 with a near homogenous enhancement of contrast. (E) Axial T1 contrast image with extension into left D3-D4 and left D4- D5 neural foramina.

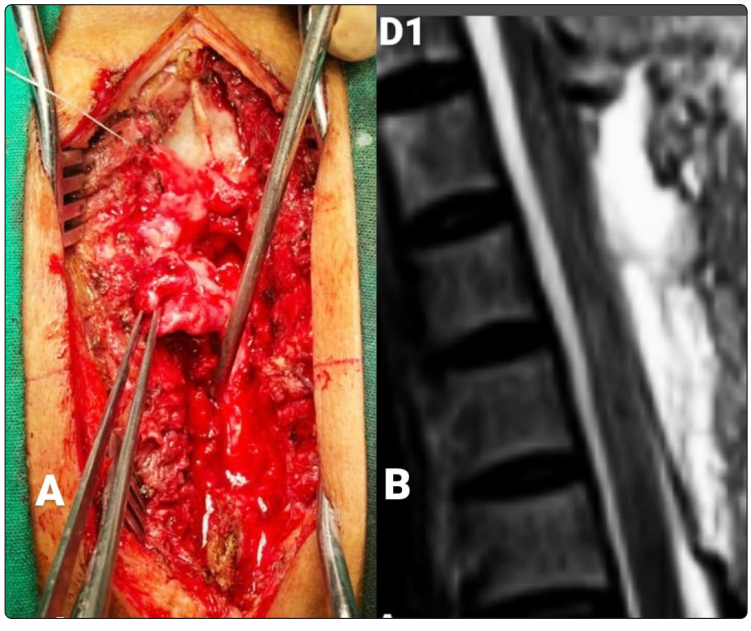

Figure 2. Intraoperative and postoperative MRI. (A) Globular mass with a smooth surface in the extradural space adherent to the dural surface with a definitive plane of cleavage between dura and tumor. (B) Complete excision of lesion with postoperative changes.

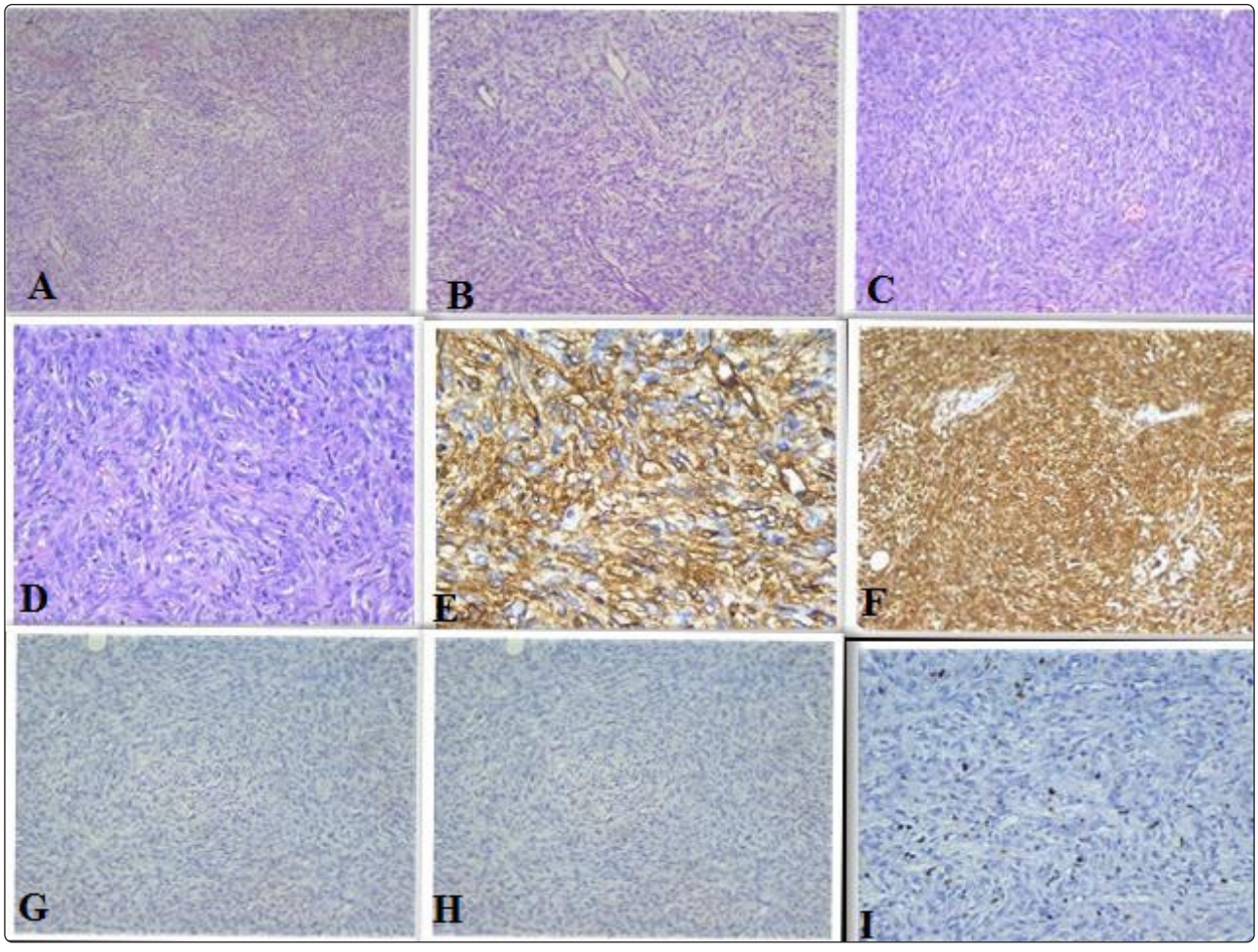

Figure 3. Histopathological slides. (A–D) Tumor composed of sheets and fascicles of spindle cells with hypercellular and hypocellular areas and collagenous stroma. The cells are oval to spindle with vesicular nuclei and scanty cytoplasm. Stroma shows staghorn blood vessels. Mitoses and necrosis were not seen. (E) Tumor cells are positive for CD34. (F) Tumor cells are positive for vimentin. (G & H) S100 and EMA are negative. (I) Mib1 is 3%-4% positive in tumor.

DISCUSSION

SFTs are rare tumors of dendritic mesenchymal origin.[13,17] Early literature on SFT/HPC showed that they were termed angioblastic meningiomas before they were recognized as separate entities.[11,16] For years they have been grouped with HPC. Neuropathologists maintained the distinction as their clinical and pathological manifestations were quite different.[4,5]

In 1931, Klemperer and Rabin described them as localized forms of primary neoplasm of pleura.[10,14] In 2007, they were thought to be of mesenchymal origin of CNS tumors.[9,16] In 2016, in its 4th edition, WHO merged the two as histologically both lesions have the same cellular features and they provided a grading system for them.[12,16] These lesions show NAB_2-STAT6 DNA fusion (molecular genetic profile) similar to hemangiopericytoma generated by genomic inversion of 12q13 locus that causes STAT6 nuclear expression detected by IHC.[14,15] Carneiro and colleagues reported the first 2 cases of spinal SFT treated by surgery.[6] Most SFTs are benign exhibiting benign histology and biological behavior. Malignant lesions though seen have very low incidence with obvious nuclear abnormalities, many mitotic figures, and surrounding infiltration.

Malignant transformation of benign lesions has been reported.[6]

Spinal hemangiopericytomas are classified into 3 types and 5 subtypes based on the location as follows: type 1, extradural (1A, intracanal; 1B, extracanal); type 2, intradural (2A, extramedullary; 2B, intramedullary); type 3, intra- to extradural and paravertebral type.[16] They are classified based on 3D morphology for dumbbell tumors based on their anatomical relationship to surrounding structures as follows. Type I tumors are located only in the spinal canal with intradural and extradural growth patterns. Type II consists of epidural tumors and includes three subtypes: A, foraminal; B, paravertebral; C, foraminal and paravertebral, distinguished according to the degree of extraforaminal spread. Type III includes intra-/extradural tumors with foraminal (IIIa) and paravertebral spread (IIIb). Type IV tumors are extradural and intravertebral, invading only the vertebral body. Type V lesions are extradural with laminar invasion and extralaminar spread. Type VI tumors show multidirectional erosion of the bone. Additional classification indicates the degree of craniocaudal tumor invasion related to the number of intervertebral foramina (i.e., IF 2: tumors involve two intervertebral foramina).[5]

The most recent 2016 WHO classification of SFT/HPC was based on protein and nuclear structure into 3 grades. Grade 1 is a highly collagenous spindle cell lesion with relatively low cellularity, which encompassed SFT. Grade 2 is more cellular, less collagenous tumors with plump cells and "staghorn" vasculature. Grade 3 can be linked to anaplastic HPC variety diagnosed based on 5 or more mitoses per 10 high power fields.[5]

SFTs affect the adult age group with slight male predominance. Most of them are frequently localized in the thoracic region followed by the cervical and then lumbar regions. There have also been reports of involvement of sacrum and junctional levels or multiple levels. They are mostly localized in the intradural extramedullary region. Clinical manifestations of SFTs are usually nonspecific and are mostly related to the site and size of the lesion. The main signs are local pain associated with weakness and paresthesia of the limbs and other pressure symptoms in the nerve distribution areas.[14]

Spinal SFTs need to be differentiated from schwannoma, meningioma, and hemangioblastoma. Imaging does not have any pathognomonic features to determine its origin and diagnosis can be confirmed only by histopathology.[2]

MRI is nonspecific and lesions are iso- to hypointense on T1 and T2 with the presence of vascular flow voids and heterogeneous contrast enhancement. Tumor degeneration can produce cystic necrosis causing hyperintensity on T2 signals.[14] The absence of calcification, hyperostosis, and relatively narrow base and nodularity help in differentiating it from meningioma as they share distinct similarities. Angiography is rarely needed and only plays a role in large tumors for preoperative embolization.[16]

The biology of SFTs remains unclear; 10%–20% show invasive or malignant features and are seen to recur or metastasize.[14] Treatment of choice in the case of SFT is surgery.[2] En bloc resection or surgical gross total resection when possible is shown to be acceptable for grade 1 lesions and GTR followed by postoperative radiotherapy for grade 2-3 lesions. Grade 3 is a predictive factor for recurrence. Recurrence has been documented with subtotal resections. For grade 1 tumors with GTR, there appears to be no role of adjuvant postoperative therapy.[7]

While debating the prognosis of the tumor, factors that are of prognostic significance are the proliferation index, mitotic picture, the histopathological grade of the tumor, the extent of resection, and the malignant nature of the lesion. The tumors' response to radiotherapy is poor irrespective of the tumor being benign or malignant. The extent of resection and delayed recurrence play a role in the prognosis. Long-term follow-up is recommended.[6]

CONCLUSION

SFTs are rare tumors whose clinical course is unpredictable. They commonly occur in the intradural extramedullary sites and rarely in extradural sites. The goal of treatment is complete excision.

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الملخص العربي

الورم الليفي الأولي خارج الام الجافية في العمود الفقري الصدري: تقرير حالة

البيانات الخلفية: الورم الليفي الانفرادي / ورم الخلايا الوعائية (SFT / HPC) هو ورم نادر في الجهاز العصبي المركزي (CNS). بل إن حدوثه في العمود الفقري اكثر ندره. يتم تشخيصه من الناحية النسيجية حيث أن التصوير قبل الجراحة لا يكشف عن أي سمات مرضية. هو الأكثر شيوعًا في العمود الفقري الصدري وقد يكون له تأثير سيئ على الرغم من الجراحة والعلاج المساعد. نقدم حالة من حالات SFT الأولية في العمود الفقري الصدري خارج الام الجافية ليتم إضافتها إلى قائمة الحالات القليلة المبلغ عنها.

الغرض: الإبلاغ عن ورم نادر من SFT خارج الجافية في العمود الفقري الصدري.

تصميم الدراسة: تقرير حالة.

المرضي والطرق: أبلغنا عن امرأة في منتصف العمر أصيبت بضعف حسي وحركي لمدة شهرين. مع المتابعة ذات الصلة والتصوير بالرنين المغناطيسي، لوحظ وجود آفة خارج الام الجافية في العمود الفقري. خضع المريض لاستئصال كتلة من الآفة مع مسار سلس بعد الجراحة وتم تحليل العينة لتشريح الأنسجة.

النتائج: أظهر التحليل التشريحي المرضي للعينة سمات توحي بـ SFT ؛ ارتبطت الكيمياء النسيجية المناعية (IHC) بالتشخيص المرضي للنسيج. في متابعة لمدة 3 أشهر ، كان لدى المريض تحسن في قوة الأطراف السفلية والتشنج من الصف 11/7 إلى الصف 11/10 وفقًا لدرجة اعتلال النخاع الصدري JOA.

الخلاصة: إن SFTs هي أورام نادرة لا يمكن التنبؤ بمسارها السريري. تحدث بشكل شائع في مواقع خارج النخاع داخل الام الجافية ونادرًا ما تكون خارج الجافية. الهدف من العلاج هو الاستئصال الكامل.