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## Case Report

# Intradural-extramedullary solitary fibrous tumor of the thoracic spine: A case report ☆,☆☆

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## ABSTRACT

Spinal solitary fibrous tumors are extremely rare neoplasms and of those, intradural extramedullary location is even rarer. A 64-year-old male presented to the emergency department with worsening right leg pain over 1 month. Whole spine magnetic resonance imaging revealed a well-circumscribed mass with low T1 and markedly low T2 signal intensity at the level of T1-2. Spine computed tomography showed no evidence of calcification or acute hemorrhage. Surgical removal was performed and the final diagnosis was intradural extramedullary solitary fibrous tumor.

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## Introduction

Solitary fibrous tumors (SFT), also known as hemangiopericytomas are rare mesenchymal tumors that arise from pericapillary cells that were first described in 1931 by Klemperer and Rabin [1–3]. Most common location is the visceral pleura but various unusual extrapleural sites of SFTs have been reported. Of those, spinal SFT remains a rare entity, having only 90 cases reported in literature [4]. Most spinal SFTs are intramedullary. Intradural extramedullary location is even rarer [5,6]. We herein describe radiological findings of a rare case of intradural extramedullary SFT in the spinal cord.

## Case report

A 64-year-old male presented to our emergency department with worsening right leg pain over 1 month. Neurological examination revealed right-sided paresthesia and hypoesthesia to touch sensation below T5 dermatome level. Other physical examinations were within the normal limits. Right knee and ankle jerk were hypoactive. Babinski sign was present bilaterally. There was no motor weakness.

Lower extremities computed tomography (CT) angiography showed nonspecific lower extremity arteries. Whole spine magnetic resonance imaging (MRI) was obtained and showed

☆ Declaration of competing interest: None.

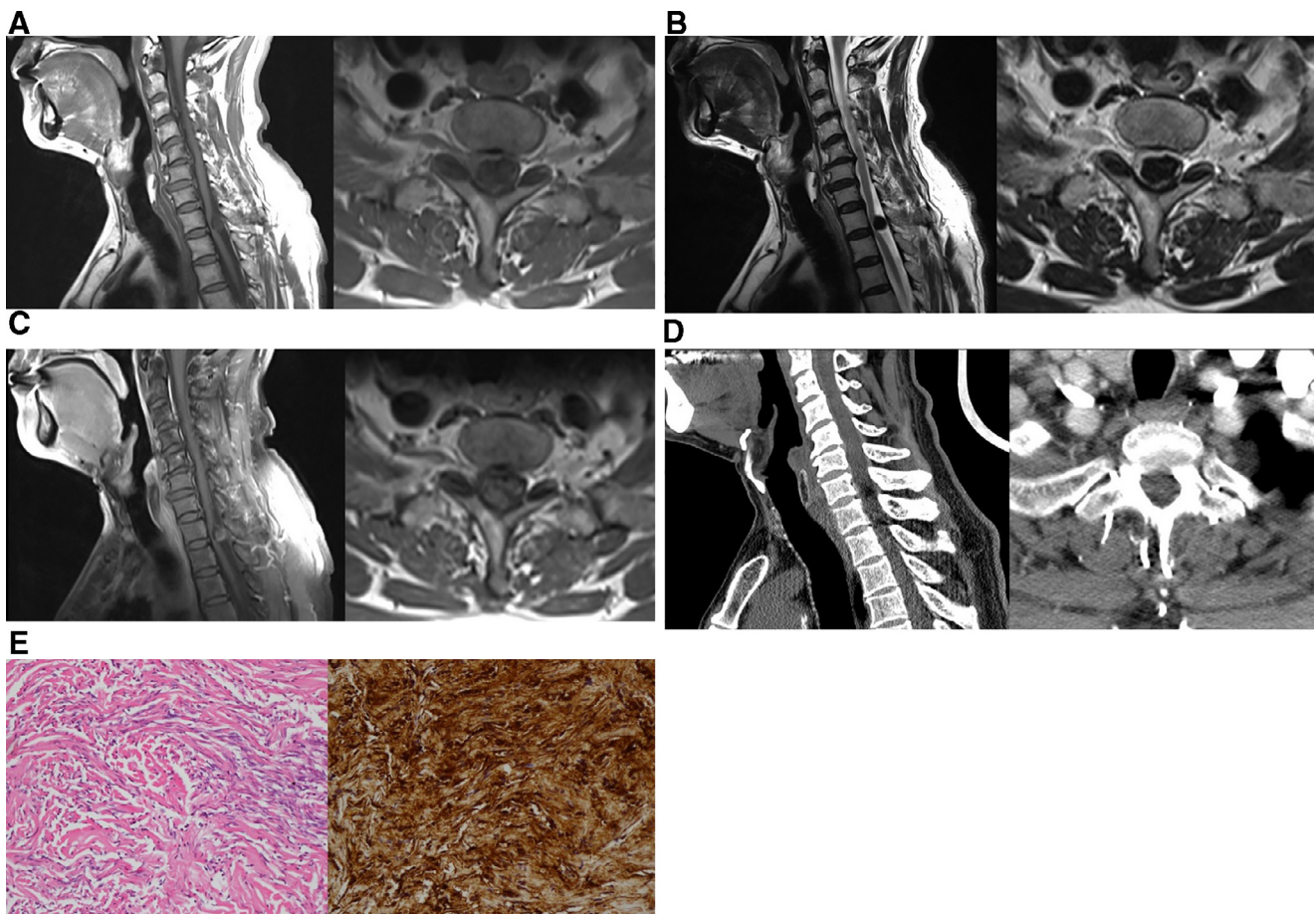
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**Fig. 1** – Intradural extramedullary SFT at thoracic spine in a 64-year-old man. (A) (Left) Sagittal and (right) axial MRI T1 sequence of the spine shows well circumscribed mass (1.4 × 0.7 cm) with low T1 signal intensity in posterior epidural space, lying dorsal and left lateral to the spinal cord at the level of T1-T2. (B) (Left) Sagittal and (right) axial MRI T2 sequence of the spine shows markedly dark T2 signal mass located intradural extramedullary at the level of T1-T2. Dorsal mass is compressing the spinal cord to the right side anterolaterally. (C) (Left) Sagittal and (right) axial MRI Gadolinium-enhanced sequence of the spine shows mild heterogeneous enhancement of the intradural extramedullary mass at the level of T1-T2. (D) (Left) Sagittal and (right) axial CT of the spine shows no evidence of calcification or acute hemorrhage. Mass is isodense and is difficult to delineate with adjacent structures. (E) (Left) Hematoxylin and eosin (H&E) stain, original magnification ×200, shows spindle cells embedded in a thick collagenous stroma. (Right) Immunohistochemical stain for CD34, original magnification ×200, shows strong and diffuse immunopositivity with CD34.

a well-circumscribed mass like lesion (1.4 × 0.9 × 1cm) in the epidural space, lying dorsal, and left lateral to the spinal cord at the level of T1-T2 (Fig. 1A, 1B, 1C). Mass showed low T1 signal intensity, low T2 signal intensity, and heterogeneous mild enhancement with gadolinium administration. The mass was pushing the spinal cord to the right side and anterolaterally. Since the margin between the spinal cord and tumor was clear, we could confirm that the location of the mass was intradural extramedullary.

Cervical spine CT showed no evidence of calcification or acute hemorrhage. The isodense mass was difficult to delineate with adjacent structures (Fig. 1D). The mass was surgically removed and grossly it appeared as a grayish solid hard mass. Histologically it showed spindle cells in an abundant collagenic stroma without necrosis and positive CD34 immunostain with negative S100 and Epithelial membrane antigen (EMA) (Fig. 1E). Final diagnosis was intradural extramedullary SFT.

Follow-up MRI image was done at 2 weeks after surgery and showed a post-op fluid collection but no remnant tumor. MRI scan at 6 months follow-up showed no recurrence. The patient had good recovery after the operation and had an uneventful course without any sign and symptoms suggesting recurrence of tumor. Follow-up neurological examination of the patient performed at 12 months postoperatively showed normal sensation and strength.

## Discussion

SFT occurring in the spinal cord is extremely rare. The most common location being thoracic spine followed by cervical spine and lastly lumbosacral spine [6,7]. In contrast to intracranial SFTs, which arise most frequently from the dura,

the spinal SFT of the spinal cord are most commonly intramedullary [4,5].

Histologically, SFTs are composed of spindle cells within a background of collagen stroma, often in a whorled pattern or patternless [2,4,8]. The diagnosis is confirmed by characteristic positive immunohistochemical staining for CD34 and negative staining for other differentials such as EMA and S-100 which when positive indicates meningioma and schwannoma, respectively [2,5,9].

From literature review, only 20 cases of thoracic intradural extramedullary SFT have been reported [6]. Incidence seems to be almost double in men than women. Mean age was 39 years old. These tumors usually have a stationary growth rate but it can compress the spinal cord and induce neurological symptoms even in early stages. Most produced symptoms consisted of pain, myelopathy, radiculopathy, and motor or sensory deficit [3,5].

Masses that can be found in intradural extramedullary space of the spine are commonly meningioma or schwannoma. Schwannoma is more common in the lumbar region and usually presents as heterogeneously high T2 signal dumbbell-shaped mass with obvious cystic degeneration and marked enhancement [9]. Meningioma usually presents with isointense T1 and T2 signal mass with characteristic dural tail sign and thickened dura [3,6,9]. Thoracic spine followed by cervical spine is the most common location for spinal meningioma. Presence of calcification, hemorrhage, or severe fibrosis was contemplated due to prominent low T2 signal intensity of the mass. From the initial MRI scan, calcified meningioma was the first differential diagnosis considering the thoracic location of the mass and low T2 signal. However, cervical spine CT showed no evidence of calcification or acute hemorrhage.

Final diagnosis was SFT in the spinal cord which is known to present as a well-circumscribed encapsulated mass with characteristic markedly dark T2 signal and isointense to hypointense T1 signal on MRI [1,3,5,9]. Prominent low T2 signal is thought to be due to rich collagen content of SFT [1]. It is also known to show strong homogeneous enhancement due to its rich vascularity and may accompany peritumoral edema, which was not present in our case. Mild enhancement pattern in this case may be due to high collagenous content with relatively low cellularity and vascularity.

In this case, the patient presented with right leg pain and sensory deficits. These symptoms are probably due to the space occupying effects of the mass. Compression of the spinal cord from the dorsal direction can compromise various sensory pathways such as dorsal column–medial lemniscus pathway and anterolateral tracts or adjacent nerve roots, which could induce sensory symptoms. Sparing of motor function in this case may be due to rather lateral location of the corticospinal tract.

Primary management option for spinal SFT is complete surgical excision. If resection is incomplete or if the tumor is inoperable, adjuvant therapies, such as chemotherapy or radiotherapy can be considered. SFTs are usually slow growing benign tumor but rarely can undergo malignant transformation [8]. Just like other malignancies, malignant SFTs will tend

to show more irregular margins, more heterogeneous signals, heterogeneous enhancement pattern, invasion into adjacent structures such as vertebra, metastasize to other organs such as lung and locally recur even after complete resection [2,7,10]. Malignant transformation, recurrence, or metastasis can occur many years after initial diagnosis and after total resection. Thus, long term follow up with MRI is necessary to detect any signs malignant transformation or recurrence after surgery [7,10].

In conclusion, spinal intradural extramedullary SFT is an extremely rare neoplasm among spinal cord lesions. Distinctive dark T2 signal on MRI allows SFT to be differentiated from other spinal cord tumors. MRI also provides information about its location, extent, and compression of the cord or nerve roots. Radiological long-term follow-up is also required to detect malignant transformation or recurrence after surgery. Understanding radiological features of spinal SFT, and its differential diagnoses will aid in its diagnosis provide useful information for preoperative surgical planning.

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