Solitary Fibrous Tumour of the Spine: Case Report and Histopathological Review

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Abstract: Solitary fibrous tumour (SFT) is an uncommon type of spindle cell tumour that affects soft tissues. Due to the rarity of spinal SFTs, they are often overlooked by healthcare providers, leading to frequent misdiagnosis. The clinical signs of spinal SFT are not specific and can vary based on tumour size and location. Typically, the main symptom is localized pain, which can be associated with limb numbness and other symptoms caused by pressure. Computed tomography scan was used to assess the extent of tumour involvement in the spinal canal and to identify any affected tissues. Magnetic resonance imaging is the most sensitive imaging method, and it is usually similar to disc extrusion or sequestered disc fragments. Surgical removal is the primary treatment for spinal SFT, and additional therapies, such as chemotherapy and radiotherapy, are considered for cases in which the tumour is not fully resected or inoperable.

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Introduction

Solitary fibrous tumour (SFT) is a rare spindle cell soft tissue tumour. To date, fewer than 100 cases have been reported, predominantly in adults (range of 10–83 years; median of 51.5 years). The incidence was slightly higher in men than in women (56.4% vs. 43.6%). The rarity of spinal SFTs often leads clinicians to overlook their existence, resulting in misdiagnosis (Lang et al., 2018). SFTs can occur in various parts of the body, with 24.5% of them located in the central nervous system.

Among spinal locations, the thoracic spine is the most prevalent (56%), followed by the cervical spine (31%), and lumbar spine (13%) (Zhang et al., 2019; Verla et al., 2020). These tumours are mesenchymal in nature and may originate from dendritic mesenchymal cells, sharing a similar NAB2-STAT6 DNA fusion pattern with haemangiopericytoma (Lang et al., 2018).

Herein, we report the case of a 30-year-old male patient reporting of numbness in his legs, particularly on the left side, over the past year. Informed consent was obtained from the patient.

Case report

A 30-year-old male presented with a year-long history of progressive numbness in his legs, which appeared to be more pronounced on the left side. The symptoms had gradually worsened over time, prompting him to seek medical attention. Despite this, he did not report any significant motor deficits. Upon physical examination, there were no signs of weakness, and he was able to perform movements such as dorsiflexion on both sides with full strength. The sensory examination, however, revealed reduced perception of vibration sensation in the left leg when compared to the right leg, suggesting a sensory

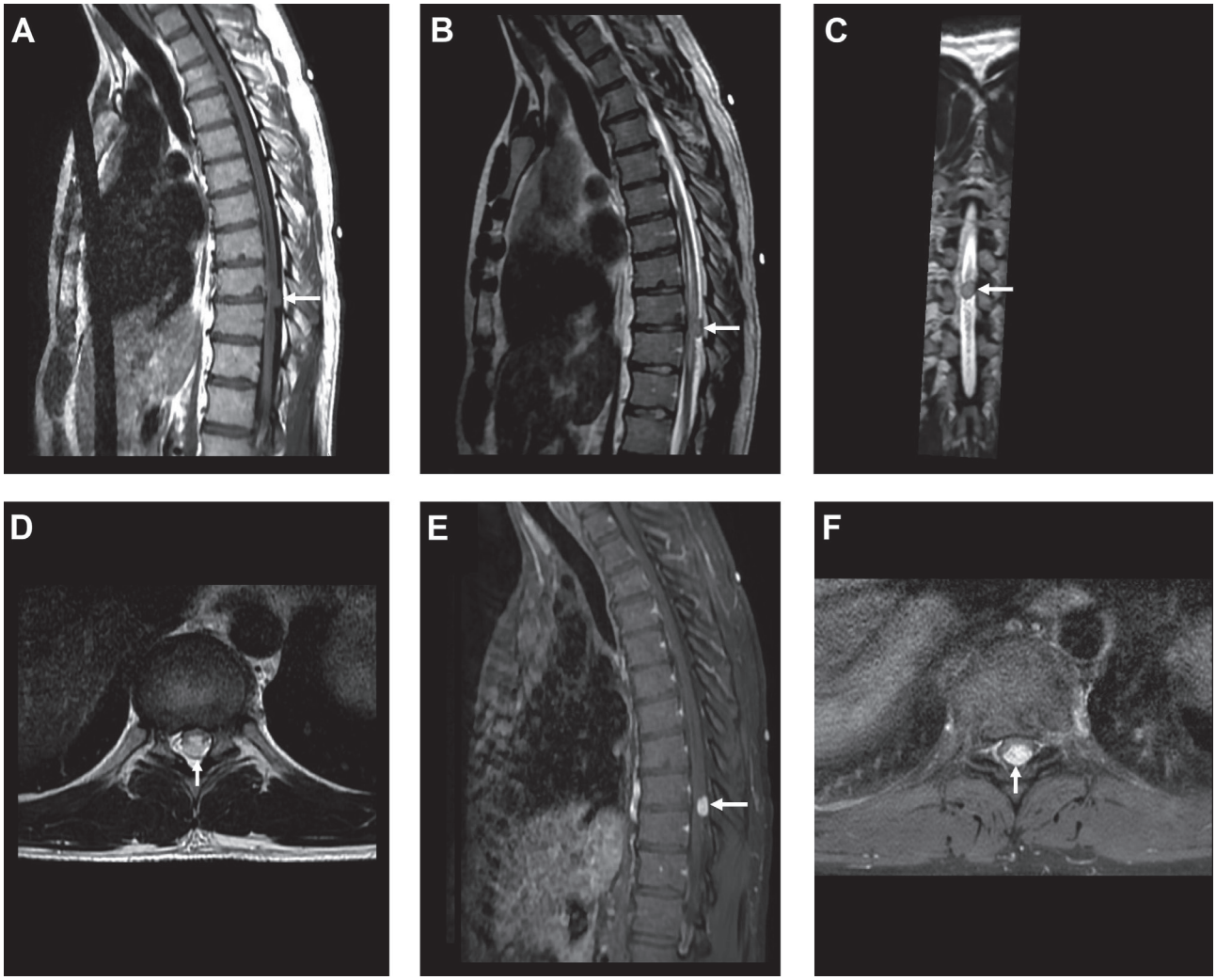

Figure 1: Thoracic spine magnetic resonance imaging. Sagittal T1 weighted-image (A); T2 weighted-image (B); coronal T2 weighted-image (C); axial T2 weighted-image (D); sagittal (E) and axial (F) T1 FAT SAT with contrast. Intradural extramedullary enhancing tumour located dorsally to the spinal cord at the T9-10 level (white arrows), with spinal cord infiltration and spinal cord edema/pre syrinx.

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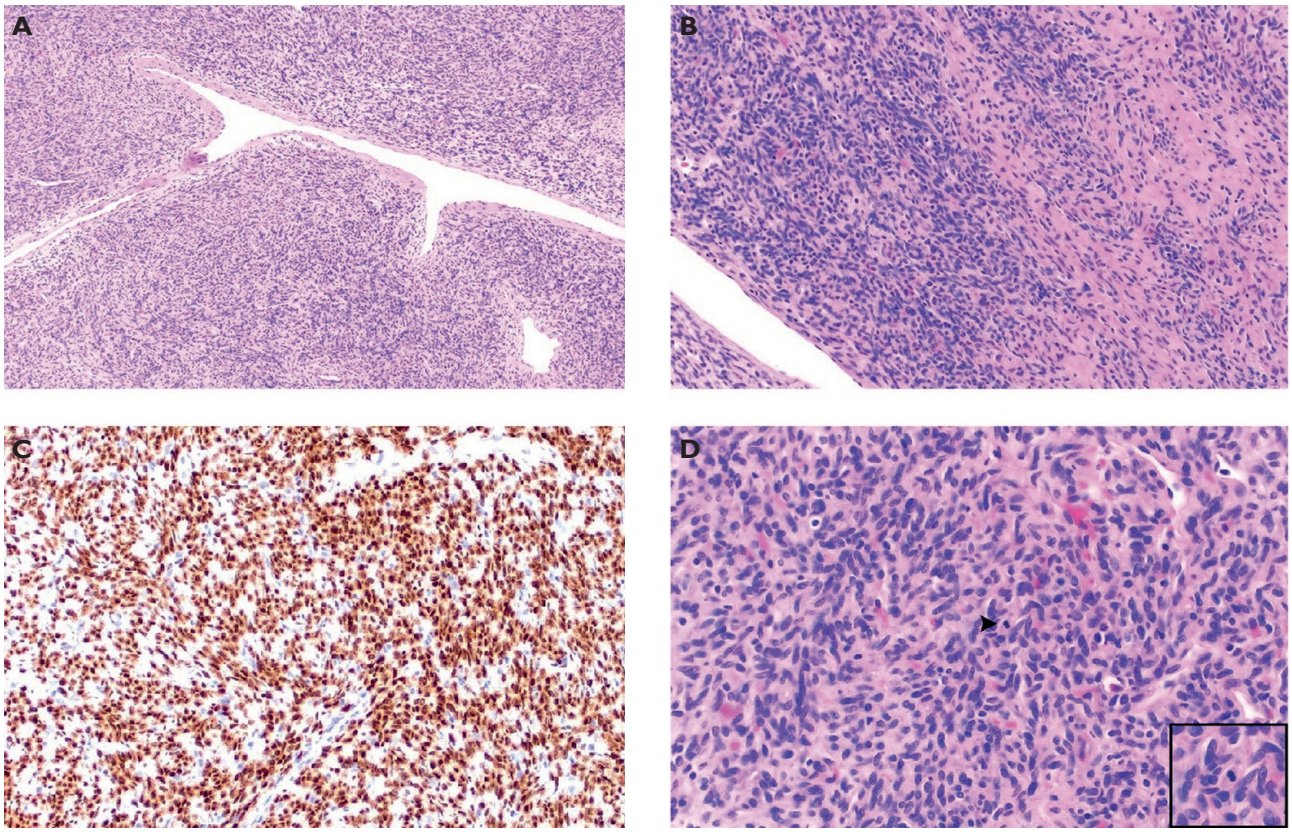

Figure 2: Histologic sections of solitary fibrous tumour. A) The tumour is composed of ovoid spindled cells and has scattered, thin-walled, dilated vessels with a "staghorn" appearance (haematoxylin [H] and eosin [E] stain, 200× magnification). B) The tumour shows regions of variable cellularity. Within regions of higher cellularity, the tumour cells are haphazardly arranged (left). Whereas in regions of lower cellularity, there is dense collagen deposition (right) (H and E, 200× magnification). C) Nuclear expression of STAT6 within the tumour cells (STAT6 immunohistochemical stain, 200× magnification). D) Haphazardly arranged tumour cells with a mitotic figure (arrowhead). The inset image on the right highlights the mitotic figure with greater magnification (H and E, 400× magnification).

disturbance. Despite this sensory loss, the patient was still able to walk independently without any assistance, and there were no abnormalities noted in his motor coordination.

Further neurological evaluation revealed hyperactivity of the deep tendon reflexes, particularly in the lower extremities, which suggested an upper motor neuron involvement. The absence of motor weakness and the patient's ability to ambulate independently were reassuring, but the sensory findings warranted further investigation into the cause of his symptoms.

To determine the underlying pathology, the patient underwent magnetic resonance imaging (MRI) of the spine. The imaging revealed an intradural extramedullary enhancing tumour located at the dorsal aspect of the spinal cord between the T9 and T10 vertebral levels (Figure 1). This location and the characteristics of the tumour raised concerns about a mass effect on the spinal cord or nerve roots, which could account for the sensory changes observed.

The tumour was subsequently surgically excised through a posterior approach. The operation proceeded without complications, and a biopsy was taken for further histopathological analysis. The results from the biopsy confirmed the diagnosis of a solitary fibrous tumour (SFT), classified as central nervous system (CNS) World Health Organization (WHO) grade 2. Molecular analysis revealed the presence of an NAB2-STAT6 gene fusion (NAB2-exon4:STAT6-exon2), a characteristic genetic alteration frequently seen in solitary fibrous tumours (Figure 2). This fusion is often associated with the pathogenesis of SFTs and provides valuable information for both diagnosis and potential therapeutic strategies.

Postoperatively, the patient experienced significant improvement in his sensory symptoms, with gradual restoration of vibration sensation in the left leg. Follow-up care, including regular MRI surveillance, will be necessary to monitor for any recurrence of the tumour. The patient's motor function remained intact, and he continued to walk independently, with no further deficits observed during his recovery.

Discussion

The clinical manifestations of spinal SFT are nonspecific and may vary depending on the location and size of the lesion. The primary symptom is often localized pain, which may be accompanied by limb numbness and other pressure-related symptoms. Although spinal SFTs are typically found in the intradural extramedullary area, they can also occur extradurally or intramedullary and are often accompanied by exophytic growth (Lang et al., 2018).

Computed tomography (CT) can be used to assess the extent of SFT involvement within the spinal canal and to determine any involvement of surrounding tissues (Zhang et al., 2019). However, MRI remains the most sensitive imaging modality, often demonstrating radiographic similarities to disc extrusion or sequestered disc fragments on T1- and T2-weighted imaging (displaying low signal intensity). Nevertheless, avid contrast enhancement is typically observed, indicating a well-circumscribed homogeneous lesion (Kim et al., 2020; Verla et al., 2020). Given its nonspecific clinical and radiographic features, spinal SFTs may resemble other pathologies, such as schwannoma, meningioma, and osteosarcoma (Verla et al., 2020). Biopsy or surgical resection is essential for accurate diagnosis (Lang et al., 2018).

Histopathologically, typical SFTs exhibit clear tumour cell boundaries with an uneven distribution of cell-rich and cell-sparse areas. Immunohistochemically, SFTs tend to express mesenchymal and vascular endothelium-related antigens, including CD34, CD99, vimentin, and bcl-2, and typically show negative staining for EMA, SMA, GFAP, and S-100. Moreover, recent studies have identified STAT6 as a characteristic tumour marker for SFT diagnosis, and the combination of STAT6 with CD34, CD99, and bcl-2 aids in the differential diagnosis of related lesions (Zhang et al., 2019).

The primary treatment for spinal SFT is complete surgical excision, with adjuvant therapies like chemotherapy or radiotherapy, considered for incomplete resections or inoperable cases (Kim et al., 2020). Approximately 10–20% of SFTs show invasive or malignant features, leading to recurrence or metastasis (Lang et al., 2018; WHO Classification of Tumours Editorial Board, 2021). Consequently, long-term follow-up is recommended for all SFTs (WHO Classification of Tumours Editorial Board, 2021). Although benign SFTs have a favourable prognosis, with a 5-year survival rate of approximately 100%, studies indicate that 63% of patients with malignant SFTs experience recurrence following surgery and succumb within two years (Lang et al., 2018).

Conclusion

SFT of the spine is a rare entity with fewer than 100 cases reported, making early diagnosis challenging. Its clinical presentation is often nonspecific, with symptoms such as localized pain or limb numbness, which can mimic other spinal conditions. Diagnosis relies heavily on imaging modalities like MRI, but histopathological analysis and immunohistochemistry, particularly markers such as CD34 and STAT6 are essential for confirmation. Complete surgical resection remains the primary treatment approach, with a generally favourable prognosis for benign cases. However, long-term follow-up is crucial due to the risk of recurrence, even in low-grade tumours, and the potential for distant metastasis in higher-grade cases. This report underscores the importance of considering SFT in the differential diagnosis of spinal tumours and highlights the need for awareness among clinicians to prevent misdiagnosis.

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