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Case Report

Recurrent solitary fibrous tumor of lumbar spine with vertebral body involvement: imaging features and differential diagnosis with report of a case

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ABSTRACT

Solitary fibrous tumors (SFTs) of the spine are exceedingly rare tumors of mesenchymal origin. Most spinal SFTs arise from the thoracic spine, followed by cervical spine, and last lumbar spine with only 6 cases reported in literature. SFTs represent a wide range of neoplasms, ranging from benign to malignant. These tumors can develop a late recurrence, even after a decade or more of initial presentation, requiring long-term follow-up. We present a case of recurrent SFT of the lumbar spine with vertebral body involvement, presenting more than a decade after initial resection. It was initially misdiagnosed as a paraganglioma. To the best of our knowledge, there have been only 3 previous cases reporting SFT with vertebral body involvement.

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Introduction

Solitary fibrous tumors (SFTs), previously grouped together with hemangiopericytomas, are rare soft-tissue tumors mostly occurring in the thoracic region but can arise anywhere in the body [1–3]. Spinal SFTs are a rare entity with the most common location being thoracic spine followed by cervical and lastly lumbosacral spine [4–10]. SFTs are mesenchymal in origin as proved by immunohistochemistry; however, histologically, these appear as haphazardly arranged spindle cells with interspersed hypocellular collagenous areas. A histopathologic differential diagnosis for SFT

includes sarcomatoid carcinoma that displays additional nuclear atypia and a malignant glandular component. Synovial sarcoma is another differential that demonstrates more primitive and hyperchromatic chromatin with increased mitotic activity. CD34 is usually negative in synovial sarcoma [11]. Mesenchymal chondrosarcoma may show a similar vascular pattern, however, would demonstrate cartilaginous growth [11]. Nuclear labeling for STAT6 is highly specific for SFT and is related to the NAB2-STAT6 gene fusion [11].

Most of these lesions have a benign course, but local recurrence and metastasis have been reported in a number of cases [12]. In terms of malignant potential, pleural SFTs are

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more likely to be malignant when compared with extrapleural tumors [13]. We present a case of spinal SFT involving lumbar vertebral body. It was previously misdiagnosed as a paraganglioma and only appropriately characterized after recurrence.

Case report

A 73-year-old man with history of treated prostate cancer, presented originally with complaint of left lower-extremity weakness and numbness that was progressively getting worse over the last few years. Additional complaints included chronic low back pain and constipation.

Physical examination revealed diminished sensation along the left groin, anterior thigh and calf. The strength in right lower extremity was reported as 4 of 5, and left lower extremity was reported as 3 of 5. The patient was originally diagnosed in 1992, with a lumbar spinal tumor known to be a paraganglioma, for which he had undergone surgical resection at the time, followed by combination intensity-modulated radiation therapy and proton therapy in 2002. The patient was followed by a physician from an outside facility since 2012, and in 2015, he was referred to our facility for a second opinion, as the lumbar lesion was thought to be increasing in the interim (Figs 1A and B). He presented to our facility with the above mentioned chief complaint, where computed tomography (CT) imaging displayed expansion of canal by isodense soft-tissue mass with accompanying osseous destruction (Figs 2A-D). Further evaluation with a magnetic resonance imaging (MRI) scan demonstrated the mass to be hypointense on T1-weighted and T2-weighted imaging, with T1 postcontrast imaging showing homogeneous enhancement of the lesion (Figs 3A-C).

The patient then underwent anterior L1 corpectomy, durotomy repair, anterior T11-L2 fusion, posterior T11-L3 fusion for removal of lesion at the L1 vertebral level. The mass was approximately 6.9 × 6.5 × 2.5 cm in size. Pathology specimens

were obtained from the L1 vertebral body and the adjacent dura. These specimens demonstrated spindle cell lesion with interspersed collagen and small vascular spaces (Fig. 4). A synovial sarcoma was ruled out, as a positive CD34 stain was observed (Fig. 5). Nonetheless, a confirmatory histopathologic diagnosis of SFT was further displayed by a positive STAT6 stain (Fig. 6).

Discussion

SFT is a soft-tissue neoplasm of mesenchymal origin. These rare lesions usually arise from an intrathoracic location; however, literature has reported extrathoracic locations as well, such as spinal cord, head and neck, extremities, abdominal and pelvic organs, and retroperitoneum [14-16]. To the best of our knowledge, there have been only 3 reported cases of SFT resulting in vertebral body involvement [5,17,18].

SFTs have been previously grouped together with heman-giopericytomas. As pleura was the most common site of involvement, the neoplasm was initially thought to be mesothelial in origin; however, further characterization with immunohistochemical stains as well as its ubiquitous presence resolved the matter and determined the mesenchymal origin of the tumor [19]. SFT can present in a wide range of age groups with peak incidence noted in fifth to seventh decade of life, with no gender predilection [20,21]. The histology is usually comprised of collagenous matrix with arrays of haphazardly arranged, uniform elongated spindle cells [21]. Immunohistochemistry displays positive stain for CD34 in SFTs [21]. With the identification of CD34 and STAT6 in SFTs, the diagnosis has become somewhat less elusive than it used to be.

Furthermore, radiographic features on plain films demonstrate erosive soft-tissue mass in spinal column or appendage. A contrast-enhanced CT will show a smooth, well circumscribed homogeneously enhancing soft-tissue mass with possible erosion of adjacent osseous structures. No



Fig. 1 – (A) T1 sagittal precontrast image from a study in 2014 demonstrates hypointense lesion with irregular margins and osseous destruction at L1 vertebral level. **(B)** T1 sagittal postcontrast image from a study in 2014 demonstrates mild heterogeneous enhancement of the lesion at L1 vertebral level.

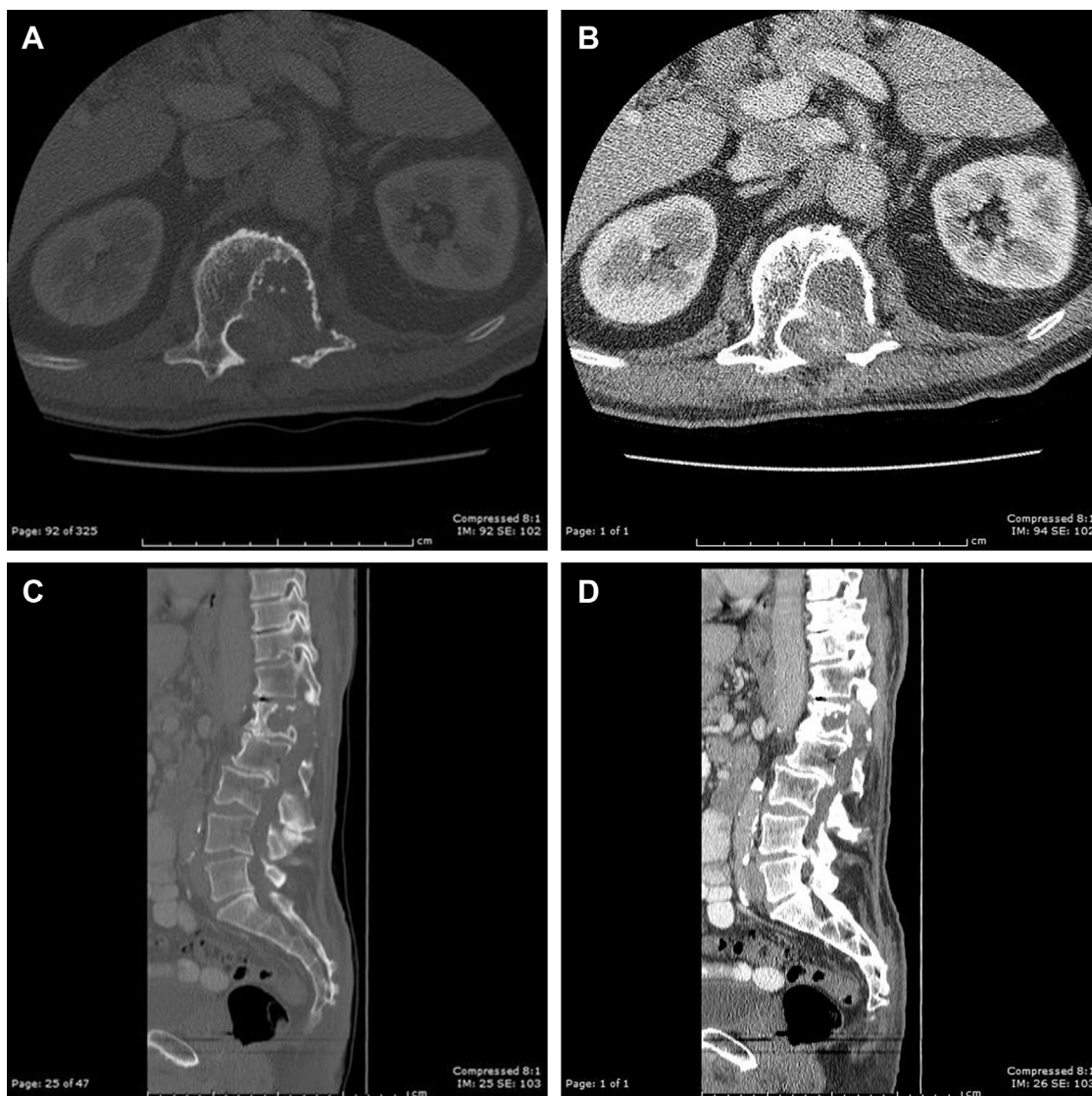


Fig. 2 – (A) Axial CT image demonstrates isodense soft-tissue mass with adjacent osseous destruction involving the L1 vertebral body. (B) Axial CT image in soft tissue window again demonstrates the soft-tissue mass with extension in to the spinal canal and adjacent osseous destruction. (C) Sagittal CT image demonstrates soft-tissue mass with accompanying osseous destruction with extension of the mass into the spinal canal at L1 vertebral level. (D) Sagittal CT image in soft-tissue window again demonstrates soft-tissue mass with accompanying osseous destruction with extension of the mass into the spinal canal at L1 vertebral level.

phleboliths or calcific components are usually identified on CT. However, larger tumors have areas of necrosis represented by hypoattenuated areas on CT [22]. MRI findings of SFT demonstrate a hypointense multilobular mass expanding into or eroding adjacent bone on T1 and a hypointense mass on T2-weighted imaging. T1 postcontrast images show vivid, homogenous enhancement of the mass [23,24]. Short T1 inversion recovery imaging would demonstrate osseous invasion as a hyperintense component with the normal marrow being suppressed. However, variable characteristics on MRI have been reported for areas of necrosis and hemorrhage within the mass; with increased signal intensity on T1-weighted imaging suggestive of subacute hemorrhage, whereas heterogeneous pattern of enhancement on

postcontrast images with increased signal intensity on T2-weighted imaging is more suggestive of necrotic foci [24]. In addition, positron emission tomography scan may be helpful in differentiating benign from malignant SFT and identifying any metastasis present at the time of presentation [24]. Differential diagnosis for an SFT based on radiographic presentation includes a meningioma, schwannoma, chordoma, aggressive hemangioma, vascular metastases, and rarely an angiosarcoma. A meningioma would usually be well circumscribed, nonerosive, with calcifications and would be more common in women [25]. As for a schwannoma, one would visualize an avidly enhancing mass in the spinal canal with neural foramen involvement leading to bony remodeling and foraminal enlargement. In addition, a schwannoma

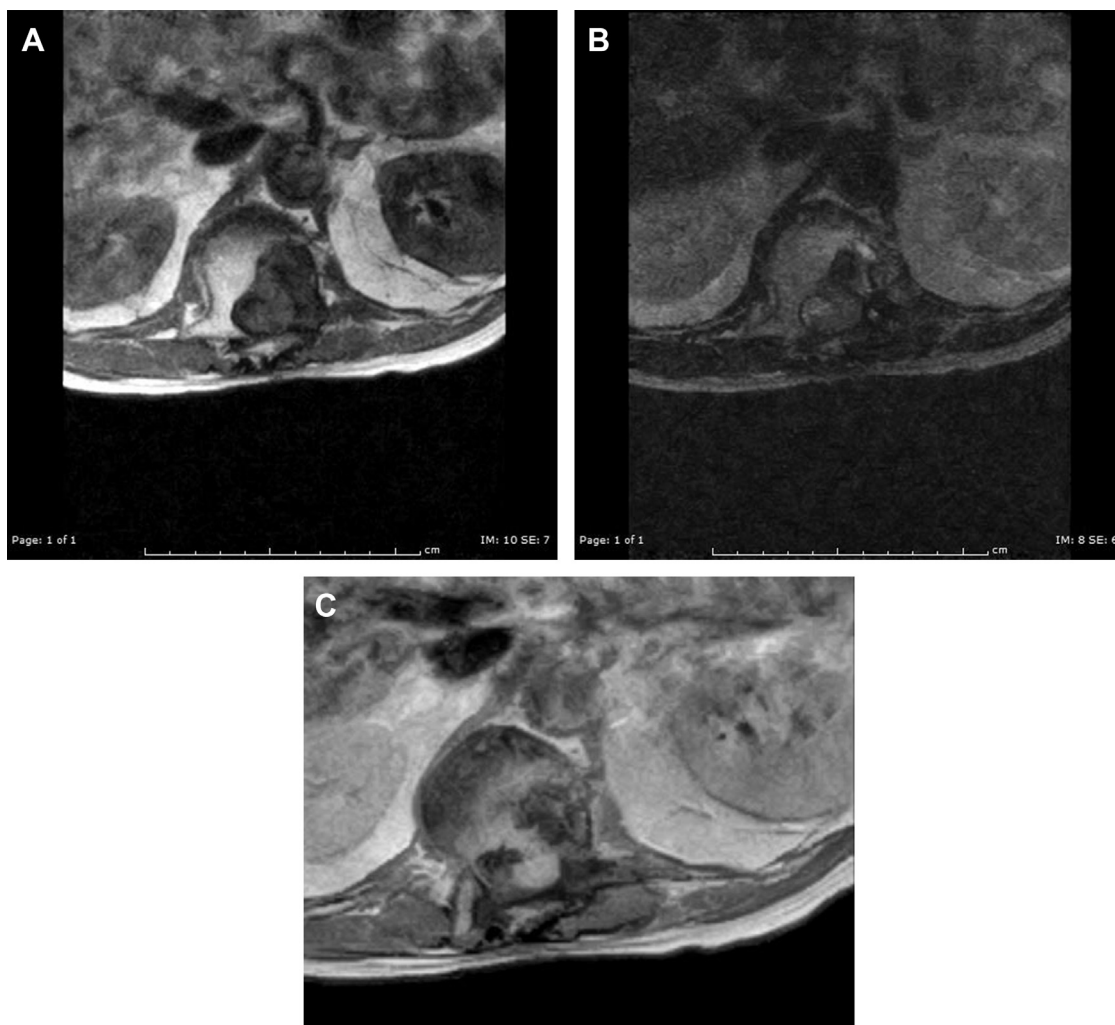


Fig. 3 – (A) Axial precontrast T1-weighted sequences demonstrate L1 epidural soft-tissue mass eroding the adjacent bone with the mass appearing hypointense on T1-weighted sequences. (B) Axial T2-weighted sequences show the soft-tissue mass appearing profoundly hypointense with redemonstration of osseous destruction at the L1 vertebral level. (C) Axial postcontrast T1 images show homogeneous vivid enhancement of the soft-tissue mass involving the L1 vertebral level.

would be hyperintense on T2 [26]. Furthermore, a chordoma is most commonly seen in the sacrum with its presentation as an extraosseous and/or epidural soft-tissue mass with the

destructive process centered in the bone [27]. An aggressive hemangioma shows thickened vertical trabeculae within area of circumscribed osteolysis, with the lesion demonstrating

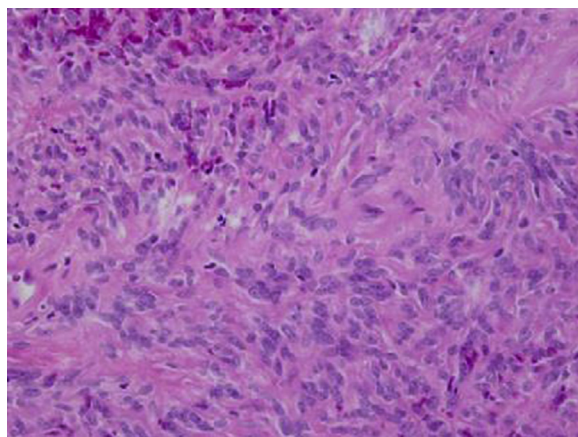


Fig. 4 – Spindle cell lesion with interspersed collagen and small vascular spaces.

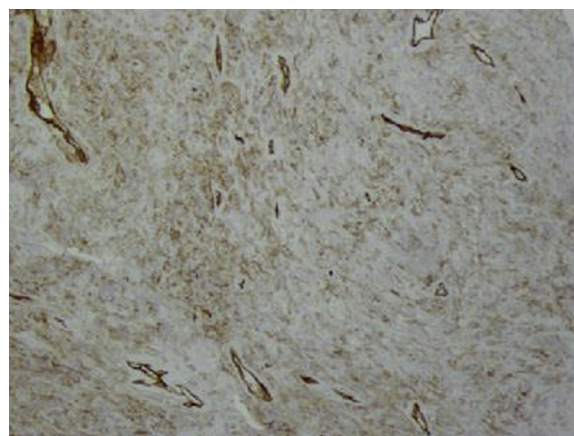


Fig. 5 – Positive CD34 stain.



Fig. 6 – Positive STAT6 staining.

avid enhancement with an associated epidural or paravertebral soft-tissue mass [28]. Renal cell and thyroid carcinomas are known for causing vascular metastases; however, these are usually centered in the cancellous bone with involvement of the posterior elements of the vertebral body [29].

SFT represents a diverse group of neoplasms in terms of its malignant potential; ranging from completely benign tumor with complete cure on resection to malignant neoplasm with recurrence and even distant metastasis [30]. A number of indicators have been defined to characterize SFT into typical (with benign outlook) or malignant neoplasm. High mitotic index, nuclear pleomorphism, necrosis, and large tumor size have all been associated with a more malignant course; however, no clear correlation between histologic characteristics and clinical outcome has been established [31]. A study focusing on delayed recurrent SFT demonstrated that 5 of 12 cases of recurrence were initially diagnosed with a typical SFT; thus suggesting a need for prolonged follow-up even with histologically benign disease initially [26]. SFTs are known for a late recurrence. A study aiming to determine outcomes for late recurrence reported a median time of 12 years to the first recurrence [26]. Our case corroborates this finding as well, as our patient presented with symptoms after almost a decade of initial resection. The most common pattern of recurrence is local, but a substantial percentage of patients can present with distant metastasis, so a history of SFT in a patient presenting with a mass in an unrelated location should still prompt consideration for recurrence even after a decade or more of initial diagnosis.

This case also emphasizes the importance of considering diagnosis other than metastatic prostate cancer in patients with vertebral body involvement and back pain. With prostate cancer being one of the more common causes of symptoms in this scenario, additional diagnostic considerations can be overlooked.

Conclusions

SFTs of the spine, especially lumbar spine, are exceedingly rare with only a few published cases. Spinal SFTs can present with a

late recurrence causing local invasion and destruction of vertebral body even after a decade or more of initial resection. So in a patient with history of SFT, possibility of recurrence should always be kept in mind. Furthermore, prolonged follow-up is recommended once a diagnosis of SFT is established.

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