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Case report

Fat-forming solitary fibrous tumor of the sacrum: A case report and literature review ☆,☆☆

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ARTICLE INFO

Article history:

Received 1 December 2020

Revised 19 April 2021

Accepted 21 April 2021

Keywords:

Solitary fibrous tumor

Fat-forming

Lipomatous

Hemangiopericytoma

Spine

ABSTRACT

Fat-forming variant of solitary fibrous tumor (SFT) is a rare mesenchymal neoplasm. Here we report the case of a 33-year-old woman who developed pain and muscle weakness from the posterior aspect of the right hip to lower extremity. Imaging examinations revealed a mass with fatty components and hypervascular solid components filling the sacral spinal canal and sacral foramen. The sacral mass was resected and histological examination of the specimens revealed patternless proliferation of short spindle-shaped cells with staghorn blood vessels. A number of mature adipocyte-like cells were also observed. The tumor cells were positive for STAT6 and the nuclei of the adipocytes were also positive, which was diagnostic for fat-forming SFT.

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Introduction

Solitary fibrous tumor (SFT) is a neoplasm of mesenchymal origin. As SFT was originally described in the pleura, those occurring at other sites are sometimes referred to as extrapleu-

ral SFT. Fat-forming SFT is a rare type [1] that is more common in middle-aged adults, with no gender predilection. Retroperitoneal areas and the deep soft tissues of the lower extremity, especially the thigh, are predominantly affected [2]. Only one case of fat-forming SFT in the spine region has previously been reported [3]. Here we present the case of a 33-year-old patient

☆ Acknowledgements: We thank Forte Science Communications for English proofreading this paper.

☆☆ Competing Interests: None

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<https://doi.org/10.1016/j.radcr.2021.04.052>

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who underwent surgery for a sacral mass that was confirmed histopathologically and immunohistochemically as this tumoral subtype.

Case report

A 33-year-old woman with a five-year history of occasional right lower extremity pain presented with pain, numbness, and muscle weakness from the posterior aspect of her right hip to the lower extremity after lifting a heavy object one month prior to presentation. Other than a Cesarean section, her past medical history was unremarkable and she did not drink alcohol, smoke, or use illicit drugs. Medications included pregabalin and brotizolam.

On physical examination, she had a right-sided limp. She was afebrile, had normal vital signs, and the remainder of the examination was unremarkable. Laboratory data, including tumor markers, were within normal limits.

Radiographs of the sacrum showed an indistinct first sacral hiatus and osteosclerosis from the upper edge of the right S1 vertebral body to the lower edge of the S2 sacral foramen (Fig. 1). Magnetic resonance (MR) images revealed a $7 \times 2.5 \times 3$ cm lobular intraspinal mass within the sacral

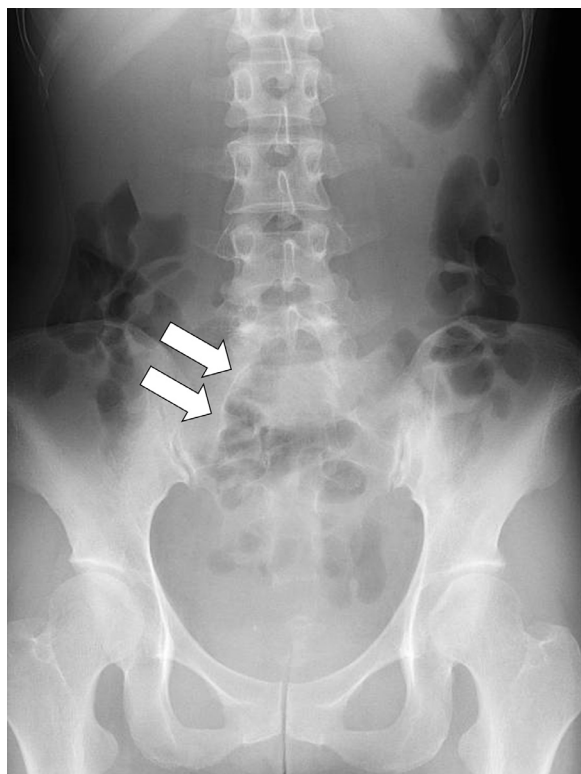


Fig. 1 – Radiograph of the sacrum. The first sacral hiatus is asymmetrical, and is obscured on the right side. Enlargement of the first and second sacral hiatus is suspected. Osteosclerotic changes are seen at the margins (arrows). The lumbar spine and the sacroiliac joints are intact.

canal with scalloping of the S1 and S2 vertebral bodies. The mass showed inhomogeneous hyperintensity on T1-weighted imaging (T1WI) (Fig. 2a). Regions that exhibited hyperintensity on T1WI showed hypointensity on fat-suppressed T1WI, compatible with fat components (Fig. 2b). The mass showed avid heterogeneous contrast enhancement (Fig. 2c). T2WI showed areas of intermediate signal with flow voids as well as areas of high signal (Fig. 2d). The apparent diffusion coefficient (ADC) value of the mass was 0.875×10^{-3} mm²/s. Dynamic contrast-enhanced computed tomography (CT) showed arterial hypervascularity within the lesion (Fig. 2e), and areas of hypodensity suggesting fat components. The leading imaging differential consideration was schwannoma.

CT-guided biopsy of the mass revealed spindle-cell-shaped proliferation expressing CD34 antigen. Immunohistochemistry was positive for STAT6 and Bcl-2 favoring a diagnosis of SFT. Two-stage surgery was planned and the tumor vessels were embolized before resection. Intraoperatively, the findings were of an elastic and soft epidural mass that was pulsatile and bled easily. Only the S1 nerve was adherent to the tumor. Specimens showed patternless proliferation of short spindle-shaped cells, with conspicuous small vessel growth, some of which were staghorn-like (Fig. 3a). Many mature adipocyte-like cells were also observed (Fig. 3b). Cell atypia was mild and the mitotic figure was unremarkable. On immunohistochemistry, the tumor cells were positive for STAT6. The nuclei of the adipocytes also showed positive images, suggesting adipocytic differentiation of the tumor. Bcl-2 was diffusely positive, CD34 was partially positive, S-100 was partially positive, and CDK4 was negative. The diagnosis of fat-forming SFT was based on the histological findings. No other lesions were noted and no additional treatment was given.

Discussion

SFT is a rare soft tissue tumor of mesenchymal origin. It was first recognized on serous surfaces such as the lungs, pericardium, pleura, and peritoneum, and later found to occur at various other sites [4]. The types of SFT include conventional and fibrous variants, which show a variety of morphological characteristics rich in collagen fibers; and cellular variants, which are enriched in dendritic vessels and have fewer collagen fibers. In some cases, calcification of collagen fibers or myxomatous changes may be present in areas of low cell density. The cellular variant corresponds to what has traditionally been considered hemangiopericytoma but is now grouped under the same concept because cellular variant SFT and hemangiopericytoma can occur simultaneously in the same tumor, migrate to each other at the time of recurrence, or show indeterminate histological features.

Conventional or cellular variant SFTs can have mature adipocytes without atypia, which was previously termed lipomatous hemangiopericytoma but is now regarded as a fat-forming variant of SFT [2,5]. Fat-forming SFT occurs with a male-to-female ratio of 3:2 and occurs predominantly in the retroperitoneum and deep soft tissues of the lower extremities and trunk. SFTs and fat-forming SFTs share similar clinical features. Malignant fat-forming SFT has also been reported [6].

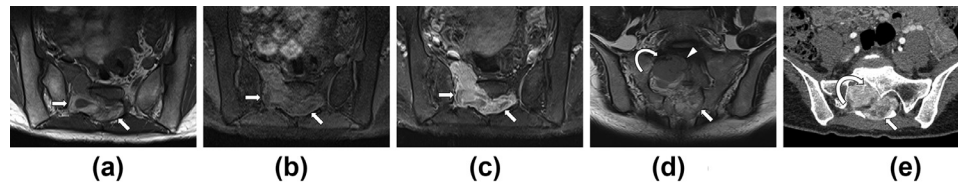


Fig. 2 – Axial T1-weighted fast spin echo (FSE) (TR 548; TE 9.2) MR image (a) shows a lobular mass expanding the sacral spinal canal with areas of high signal intensity (arrows) identical to that of subcutaneous adipose tissue. Axial fat-suppressed T1-weighted FSE (TR 560; TE 8.7) MR image (b) shows decreased signal intensity (arrows) confirming the fatty component. Axial fat-suppressed contrast-enhanced T1-weighted FSE (TR 560; TE 8.7) MR image (c) demonstrates strong contrast enhancement of the mass, except for focal areas of fat-suppressed low signal intensity (arrows). Coronal T2-weighted FSE (TR 3200; TE 87) MR image (d) shows areas of intermediate signal intensity (arrowhead) and high signal intensity (arrow) in the mass. The former is accompanied by signal voids (curved arrow), suggesting hypervascular cellular-rich components. The latter suggests fatty components. Axial contrast-enhanced CT image obtained during the arterial phase (e) shows strong contrast enhancement of the mass and a vessel (curved arrow) corresponding to the flow voids seen in (d). The mass contains areas of hypodensity suggestive of fat (arrow).

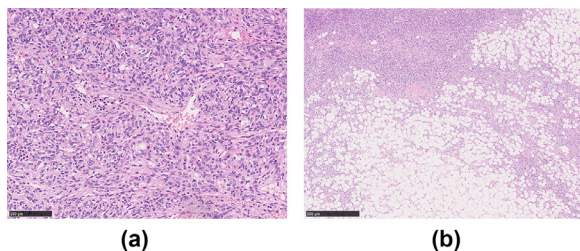


Fig. 3 – Representative hematoxylin and eosin-stained specimens of the tumor. (a) Short spindle-shaped tumor cells show patternless proliferation with various degrees of collagen fibrillation. There is an increase in the number of small blood vessels, some of which have a staghorn-like appearance. (b) Many mature adipocyte-like cells are also seen.

Fat-forming SFT in the spinal canal is extremely rare, with only one previously reported case, a 38-year-old man with a lobular intraspinal tumor displaying the signal intensity characteristics of fat, located within the lumbosacral canal [3]. The lesion extended from L4 to S2, expanding the canal and causing posterior scalloping of the L5 and S1 vertebrae. Initially considered to be a liposarcoma, the tumor was confirmed histopathologically as fat-forming SFT.

Radiologically, our case showed similar findings to the previously reported case, with fatty and hypervascular components. The differential diagnoses for an intraspinal tumor involving the sacrum include angioliipoma, lipoma variants, angiomyoliipoma, perivascular epithelioid cell tumor (PEComa), dedifferentiated liposarcoma, and schwannoma with fatty degeneration. Angioliipoma is a benign neoplasm comprising both mature fatty tissue and abnormal vascular elements, which has a high ADC value and a predilection for the dorsal thoracic epidural space but rarely the sacrum. Lipoma variants, angiomyoliipoma, and PEComa can have a low ADC value; however, the sacrum is not the predilection site of these tumors.

Dedifferentiated liposarcoma was unlikely because the bony scalloping observed in the present case suggested a slow-growing benign lesion. Finally, schwannoma with fatty degeneration was unlikely because of the marked arterial enhancement.

It is noteworthy that in the present case the immunohistology of mature adipose-like cells was positive for STAT6, which indicates adipogenesis due to tumor differentiation rather than fat deposition due to degeneration. It is well known that STAT6 immunohistochemistry is useful in the pathological examination of SFTs when the histology is modified by unusual features, including such as fat forming [7,8].

In conclusion, we present a case of fat-forming SFT affecting the sacrum. This rare tumor can be included in the differential diagnosis when hypervascular and cellular-rich components are observed in addition to fatty components. In the pathological diagnosis, immunostaining for STAT6 is useful.

Patient consent

This case report was approved by the institutional review board, and written informed consent was waived because of the retrospective design.

REFERENCES

- [1] Nielsen GP, Dickersin GR, Provenzal JM, Rosenberg AE. Lipomatous hemangiopericytoma: A histologic, ultrastructural and immunohistochemical study of a unique variant of hemangiopericytoma. *Am J Surg Pathol* 1995 Published online. doi:10.1097/0000478-199507000-00002.
- [2] Guillou L, Gebhard S, Coindre JM. Lipomatous hemangiopericytoma: A fat-containing variant of solitary fibrous tumor? Clinicopathologic, immunohistochemical, and ultrastructural analysis of a series in favor of a unifying concept. *Hum Pathol* 2000 Published online. doi:10.1053/hupa.2000.9777.

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- [3] Aftab S, Casey A, Tirabosco R, Kabir SR, Saifuddin A. Fat-forming solitary fibrous tumour (lipomatous haemangiopericytoma) of the spine: Case report and literature review. *Skeletal Radiol* 2010 Published online. doi:[10.1007/s00256-010-0991-4](https://doi.org/10.1007/s00256-010-0991-4).
- [4] Ronchi A, Cozzolino I, Zito Marino F, et al. Extrapleural solitary fibrous tumor: A distinct entity from pleural solitary fibrous tumor. An update on clinical, molecular and diagnostic features. *Ann Diagn Pathol* 2018 Published online. doi:[10.1016/j.anndiagpath.2018.01.004](https://doi.org/10.1016/j.anndiagpath.2018.01.004).
- [5] Doyle LA. Sarcoma classification: an update based on the 2013 World Health Organization Classification of Tumors of Soft Tissue and Bone. *Cancer*. 2014 Published online. doi:[10.1002/cncr.28657](https://doi.org/10.1002/cncr.28657).
- [6] Zhang LP, Zhang L, Wang G, Adhikari BK, Liu Q, Zhang W. Pericardial malignant solitary fibrous tumour with right atrial invasion – a case report and literature review. *J Int Med Res* 2019 Published online. doi:[10.1177/0300060519843734](https://doi.org/10.1177/0300060519843734).
- [7] Yoshida A, Tsuta K, Ohno M, et al. STAT6 immunohistochemistry is helpful in the diagnosis of solitary fibrous tumors. *Am J Surg Pathol* 2014 Published online. doi:[10.1097/PAS.000000000000137](https://doi.org/10.1097/PAS.000000000000137).
- [8] Creytens D, Ferdinande L. Diagnostic utility of STAT6 immunohistochemistry in the diagnosis of fat-forming solitary fibrous tumors: To the editor. *Appl Immunohistochem Mol Morphol* 2016 Published online. doi:[10.1097/PAI.000000000000166](https://doi.org/10.1097/PAI.000000000000166).