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An Unusual Case: Solitary Fibrous Tumor of the Lateral Abdominal Wall Muscles

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ABSTRACT

Background: Solitary fibrous tumor (SFT) is a mesenchymal tissue-based soft tissue tumor known for its potential aggressiveness. SFTs located in the abdominal wall are particularly rare. This report presents a unique case of SFT originating from the muscles of the abdominal wall.

Case Report: A 40-year-old male patient was referred to our hospital due to a mass in the left hypochondrium. Abdominal ultrasonography (US) revealed a heterogeneous, hypoechoic tumor with lobulated margins and limited blood supply. Magnetic resonance imaging (MRI) using Gadobutrol (Gadovist® 1.0, Schering AG, Berlin, Germany) demonstrated signal intensity heterogeneity and significant enhancement. The mass was excised en bloc. The specimen was immunopositive for Cluster of Differentiation 34 (CD34), Signal Transducer and Activator of Transcription 6 (STAT-6), desmin, and showed a 10.0% positivity for Ki-67. Follow-up examinations showed no evidence of tumor recurrence.

Conclusion: SFTs can display aggressive behavior as they often lack morphological indications of malignancy at the initial stages. For patients diagnosed with SFT, complete surgical excision is curative.

Keywords: Abdominal wall, magnetic resonance imaging, solitary fibrous tumors, ultrasonography.

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INTRODUCTION

SFTs are classified as fibroblastic/myofibroblastic tumors in the World Health Organization (WHO) 2013 classification and as fibroblastic neoplasms exhibiting moderate behavior (with rare instances of metastasizing) in the fifth edition of the classification of soft tissue and bone cancer. These tumors are equally prevalent among men and women, mostly presenting in the fifth and sixth decades of life.

The most frequent types of abdominal wall masses include desmoid tumors (30%), various forms of sarcomas (20%), metastases (18%), lipomas (6%), and endometriomas (4%). SFTs of the abdominal wall, however, are exceedingly rare.

Radiologically, distinguishing SFTs from hypervascular tumors or those with a predominantly fibrous composition can be challenging. Such tumors include leiomyosarcomas, neurogenic tumors, pheochromocytomas, lymphomas, desmoid tumors, malignant fibrous histocytomas,



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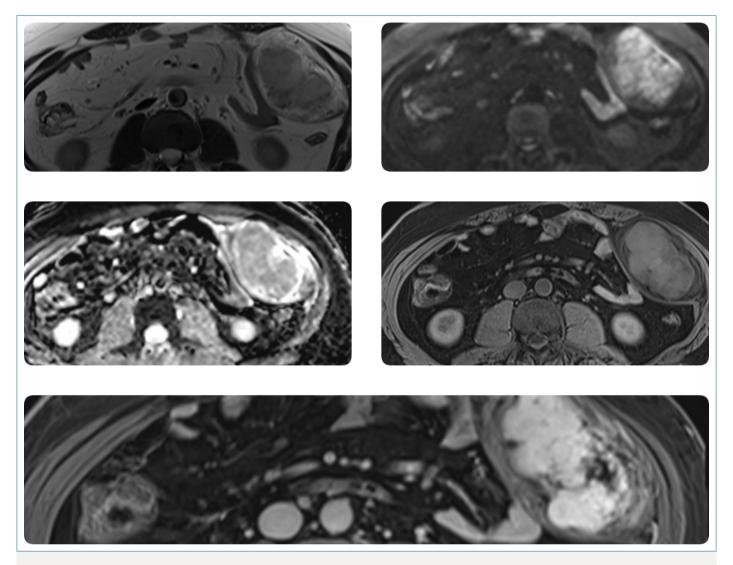


Figure 1. MRI displays a solitary fibrous tumor on a T2-weighted axial image, showing heterogeneously hyperintense areas alongside regions of low signal intensity, creating a white and black appearance within the rectus abdominis muscle on the left hypochondrium. The tumor exhibits hyperintensity on diffusion-weighted imaging and restriction on apparent diffusion coefficient (ADC) mapping, with progressive enhancement observed during the arterial, portal venous, and delayed phases.

mesotheliomas, and fibromas. Pathologically, SFTs can mimic other soft tissue neoplasms of various lineages, such as spindle cell lipoma, gastrointestinal stromal tumor, liposarcoma, and dermatofibrosarcoma protuberans, because the classic histomorphologic and immunohistochemical (IHC) profile is not always evident, making diagnosis difficult.

CASE REPORT

We report the case of a 40-year-old male patient who had noticed a mass in the left hypochondrium, right below the subcostal region, for several years. Physical examination revealed a firm mass within the muscles of the abdominal wall.

Initial assessment, including serum electrolytes, hematologic, and biochemical parameters, yielded nonspecific results. The patient was subsequently referred to the radiology department for Ultrasonography (US) and Magnetic Resonance Imaging (MRI). The US showed a heterogeneous hypoechoic solid lesion measuring 13.5 x 8 cm with a lobulated border and inadequate blood supply.

Dynamic MRI of the abdomen was performed using a 3.0-Tesla MRI system (Skyra; Siemens Medical Solutions, Erlangen, Germany), with an intravenous dose of 1 mmol/kg gadobutrol (Gadovist, Bayer Schering; 1 mmol/kg), followed by a 30-ml saline infusion.



Figure 2. Gross pathology of the specimen reveals a large, well-circumscribed tumor with a yellowish cut surface.

A vascular tumor was found in the left superficial aspect of the rectus abdominis muscle, exhibiting severe enhancement without evidence of invasion (Fig. 1). On T1-weighted sequences, it appeared hypointense with heterogeneous signal intensity, while on T2-weighted sequences, it was heterogeneously hyperintense with areas of low signal intensity. After the administration of contrast media, the mass showed intense enhancement from the arterial phase through to the late venous phase. The initial radiological diagnosis was suggestive of a desmoid tumor.

The mass was excised en bloc (Fig. 2). The specimen was immunopositive for Cluster of Differentiation 34 (CD34), Signal Transducer and Activator of Transcription 6 (STAT-6), and desmin, and also exhibited a 10.0% positivity for Ki-67. S100, actin, and Mucin 4 (MUC4) were all immunonegative. Three months post-surgery, there was no evidence of tumor recurrence.

DISCUSSION

SFTs can be located almost anywhere in the body, although they most commonly originate in the thoracic and abdominal cavities. Due to the absence of morphological indications of malignancy at the onset, SFTs can exhibit aggressive behavior. Solitary fibrous tumors, also known as benign mesothelioma or localized mesothelioma, were first identified in the pleura.

The term "solitary fibrous tumor" was introduced by Stout and Hamidi in 1951. Until 1990, SFTs were predominantly identified in the lungs and pleura. The first series of extrathoracic SFTs was published in 1991.¹

The current WHO classification categorizes SFTs as an intermediate (rarely metastasizing) fibroblastic/myofibroblastic neoplasm,¹ with a metastatic rate of 5–25%.²

Given that clinico-radiological characteristics are non-specific, diagnosis can only be confirmed through histological evidence.³

SFTs often present as well-defined, lobulated tumors that may displace nearby structures.⁴ On ultrasonography, SFTs might appear as homogeneous hypoechoic lesions, although variability can occur if cystic or myxoid degeneration is present within the lesion. Doppler flow may be minimal.⁵

The attenuation value of SFTs on unenhanced Computed Tomography (CT) images is determined by the lesion's collagen composition and cellularity.⁵ Lesions with higher cellularity appear isodense compared to adjacent muscle. Due to degeneration and core necrosis, larger SFTs are more likely to exhibit heterogeneous enhancement.⁴ In contrast, more aggressive SFTs tend to show heterogeneity following contrast injection compared to their more indolent counterparts.⁶

In lesions with myxoid or cystic degeneration, intralesional hypodensities are typical. While larger lesions display uneven attenuation, small SFTs show homogeneous attenuation.

Significant enhancement on contrast-enhanced CT scans indicates hypervascularity, whereas minor enhancement suggests hypocellularity. On T1-weighted images, SFTs often present with hypo- to isointense signals, and on T2-weighted images, they display variable signal intensity.⁴ Factors such as collagen content and low cellularity contribute to reduced-intensity foci on T1- and T2-weighted imaging.⁷

SFT most frequently occurs in the intraperitoneal, retroperitoneal, and pelvic regions of the abdomen. As observed in our case, abdominal wall SFTs can become symptomatic in the early stages, either due to their size or because they may protrude or appear asymmetrical from the exterior.

Immunohistochemical (IHC) markers expressed in SFT include CD34, vimentin, Cluster of Differentiation 99 (CD99), B-cell lymphoma 2 (BCL2), nuclear β -catenin, and epithelial membrane antigen (EMA).

Surgery is the primary treatment for SFT, with achieving adequate negative margins being crucial for reducing the risk of local recurrence. Adjuvant radiotherapy and chemotherapy are not commonly employed. Surgical planning and intervention vary significantly depending on the tumor's location and the involvement of adjacent structures.

CONCLUSION

Accurate diagnosis is critical since the treatment and prognosis for SFT differ significantly from those of their malignant counterparts. Therefore, clinicians and radiologists must consider SFTs in the differential diagnosis of abdominal wall tumors. For patients diagnosed with SFT, complete excision is curative.

Informed Consent: Written informed consent was obtained from patients who participated in this study.

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