Solitary fibrous tumors in the abdomen and pelvis: imaging spectrum with radiologic-pathologic correlation

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Learning objectives

To review and describe the spectrum of radiological findings found in fibrous solitary tumors of the abdomen and pelvis, emphasizing the ultrasound, tomography and magnetic resonance findings, as well as the clinical-pathological correlation.

Images for this section:

Fig. 1
Background

Introduction:

Solitary fibrous tumor (SFT) is an uncommon neoplasms of mesenchymal or submesothelial origin that represent less than 2% of all soft-tissue tumors. SFT was described by the first time in 1931; refer to pleural localization, being described later in other extrathoracic localizations: peritoneum, pericardium and other organs does not have a serosal layer like thymus, orbit, meninges, parotid gland and thyroid. Rarely affect liver parenchyma and pelvic structures. Recent advances in pathology have led to better understanding of the histogenesis, tumor distribution, and remarkable histologic heterogeneity of these tumors. As well, the clinical, radiological and immunohistochemical findings define the lesion, distinguishing of others tumors with similar appearance.

Natural history, clinical features and management:

Clinically, SFT manifests as an asymptomatic or slow-growing mass in middle-aged adults, without gender predilection; nevertheless, extrathoracic solitary fibrous tumors have been reported to occur slightly more frequently in women than men.

Common symptoms include pain, palpable mass, and neurologic or vascular symptoms. Also manifestations related to the size and location of the lesion like urinary obstruction or retention, bowel obstruction or constipation, and abdominal distention.

A small crew of these tumors (<5%) may manifest with hypoglycemia related to excessive production of insulinlike growth factor by the tumor. This is more commonly seen with tumors in the pelvis and retroperitoneum. Hypoglycemia secondary to malignant SFTs is mentioned in the literature as Doege-Potter syndrome.

Surgical excision is the treatment of choice in all cases of SFTs, with a 5-year survival rate of close to 100% with complete surgical excision. Rates of local recurrence are reported to be slightly higher with extrathoracic SFTs than intrathoracic, even in benign appearance tumors. On the other hand, antiangiogenic therapy has shown promising early results for the treatment of unresectable SFT. Neoadjuvant radiation therapy and systemic chemotherapy have also been tried, with variable success rates (Table 1 on page 5).

Pathologic Features:
At histopathologic analysis, SFTs are typically composed of juxtaposed hyper- and hypocellular spindle cell proliferation, dense collagenous stroma, and numerous thin-walled blood vessels with a staghorn configuration. Areas of hemorrhage and necrosis may be seen.

Immunohistochemical evaluation is essential to confirm the diagnosis. Typically the tumor specimens showed positivity for vimentin, CD34, and cytokeratin negativity. Several characteristic are predictive of aggressive clinical behavior: large tumors, cellular atypia, a high number of mitotic figures and more cellular pleomorphism (Fig. 2 on page 6).

Radiological Manifestations:

Ultrasonography (US): SFTs are seen as masses with variable echogenicity, being able to appear as hypoechoic or hyperechoic lesions with or without calcifications.

Computed Tomography (CT): is the initial exploratory modality of choice for detection of SFTs. SFTs appears as well-circumscribed, hypervascular masses that may displace or exert mass effects on adjacent organs such as the bowel, urinary bladder, vessels, and ureter. Central hypoenhancing or nonenhancing areas may be seen in the tumor, which represent necrosis or cystic change. Calcifications are rare and can be seen in large benign or malignant tumors. Besides, CT is important to detect local extent, including invasion into adjacent structures, and detection of regional and distant metastases.

Magnetic Resonance (MR): Typically STFs present intermediate signal intensity in T1-weighted images and heterogeneous low signal intensity with flow voids on T2-weighted images. Intense enhancement is seen after administration of gadolinium contrast material. Central nonenhancing areas may also be seen, which can represent areas of necrosis or cystic or myxoid degeneration.

Radiological features of solitary fibrous tumor of the liver:

Liver parenchyma involvement in SFT is rarely found, with fewer than 30 cases reported in the literature. The age of presentation varies with a range of 16-83 years and an average age at presentation of 55 years. There is a 2:1 female-to-male predominance. These tumors usually appear as large mass at presentation and do not have a lobar predilection. Likewise these tumors produce symptoms that can be directly attributed to the mass effect on neighboring organs and rarely can produce hypoglycemia (only 8 cases reported).
At imaging, STF typically manifest as large, well-defined, heterogeneously enhancing masses with or without areas of necrosis and a capsule. After administration of intravenous contrast material, early arterial enhancement is seen in the hypervascular component. The enhancement persists into the venous and delayed phases in the fibrous component. Calcification, cystic change, and a central scar have been reported. MR imaging include low to intermediate signal intensity on T1-weighted images and heterogeneous signal intensity with areas of low and high signal intensity on T2-weighted images. Areas of low T2 signal intensity correspond to the collagenous or fibrotic component.

The differential diagnosis includes hypervascular tumors like focal nodular hyperplasia, hepatocellular adenoma, hepatocellular carcinoma and fibrolamellar carcinoma (Fig. 3 on page 7).

**Radiological features of solitary fibrous tumor of peritoneum and pelvis:**

Pelvic SFT commonly arise from the pelvic peritoneum with clinical features similar to those of the other extrathoracic location. Generally seen in the 5th decade and do not exhibit any gender predilection. SFTs in the pelvis are often asymptomatic and large at presentation, with symptoms due to pressure effects on contiguous organs, presence of a palpable mass, or hypoglycemia. Malignant degeneration with recurrences has been reported in SFTs arising from the pelvic peritoneum, although the exact prevalence is unknown.

At imaging, pelvic SFTs appear as hypervascular masses with intratumoral cystic changes, necrosis or hemorrhage. MR imaging include heterogeneous low signal intensity with flow voids on T2-weighted images, representing fibrosis or collagen. Areas of central necrosis are generally seen in malignant tumors.

The differential diagnosis includes hypervascular pelvic masses like neurogenic tumors, GIST, sarcomas and inflammatory pseudotumor (Fig. 4 on page 8).

**Images for this section:**
Table 1: Summary table of solitary fibrous tumors in the abdomen and pelvis

<table>
<thead>
<tr>
<th><strong>Etiology</strong></th>
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<tr>
<td>• Mesenchymal cells</td>
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<tr>
<th><strong>Epidemiology - Incidence</strong></th>
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<tr>
<td>• Rare, less than 100 cases out of thoracic cavity</td>
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<table>
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<tr>
<th><strong>Age</strong></th>
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<tr>
<td>• Middle-aged adults (40-80 years)</td>
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<th><strong>Sex distribution</strong></th>
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<td>• Man – Women at certain locations such as the liver have been predominantly found in women</td>
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<table>
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<th><strong>Treatment</strong></th>
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<tr>
<td>• Surgical excision and in some cases have been tried neoadjuvant therapy: antiangiogenic, radiation and chemotherapy</td>
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<th><strong>Prognosis</strong></th>
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<td>• In general are benign tumors with an approximately 100% of survival rate at 5 years</td>
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</table>
Solitary fibrous tumors in the abdomen and pelvis: Pathologic Features

Juxtaposed hyper- and hypocellular spindle cell proliferation

Alternating fibrous areas and hyalinized thick-walled vessels, with strong CD34 reactivity, Bcl-2 and vimentin positivity and S100, actin, and keratin negativity

Fig. 2
Fig. 3: Differential diagnosis of solitary fibrous tumors of the liver
**Fig. 4: Differential diagnosis of solitary fibrous tumor of the pelvis**

<table>
<thead>
<tr>
<th>Solitary fibrous tumor</th>
<th>Inflammatory pseudotumor</th>
<th>Neurogenic tumor</th>
<th>GIST</th>
<th>Myxoid Liposarcoma</th>
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</thead>
<tbody>
<tr>
<td>Calcifications have been reported</td>
<td>Soft-tissue tumor that can show calcifications</td>
<td>Soft-tissue tumor. Usually follow the distribution of the sympathetic ganglia along paraspinal areas or arise from the adrenal medulla or the organ of Zuckerkandl</td>
<td>US: variable, generally hypoechoic with internal areas which can be homo or heterogeneous</td>
<td>US: Variable, Hyperechoic mass with heterogeneous structure</td>
</tr>
<tr>
<td>US: hypervascular mass with variable echogenicity</td>
<td>US: it has not been well-characterized, primarily it seems like a hypoechoic lesion</td>
<td>CT: Heterogeneous echogenicity</td>
<td>CT: Heterogeneous mass with internal cyst. Some myxoid liposarcomas may appear as complex cystic masses. Calcifications have been reported, however are rare</td>
<td>CT: Heterogeneous mass on T1 and T2-weighted images with internal low signal areas. Heterogeneously enhanced after administration of contrast material</td>
</tr>
<tr>
<td>CT: heterogeneous mass with or without areas of necrosis and capsule</td>
<td>CT: hypoattenuated or isoattenuated mass on unenhanced scans, and calcification has been observed. Variable enhancement patterns</td>
<td>MRI: variable. Usually hypointense relative to skeletal muscle on T1-weighted images, hyperintense on T2-weighted images, and heterogeneously enhanced after administration of contrast material</td>
<td>MRI: Shows a spectrum of appearances depending on the amount of fat and myxoid material, the degree of cellularity and vascularity, and the presence of necrosis. The appearance of the mass on contrast-enhanced T1 images could be: homogeneous, heterogeneous, or no enhancement, depending on the combination of the histology subtype</td>
<td>MRI: Low to intermediate signal intensity on T1-weighted images and heterogeneous signal intensity with areas of low and high signal intensity on T2-weighted images; these areas correspond to the collagenous or fibrotic component</td>
</tr>
<tr>
<td>The enhancement pattern have been defined like early in arterial phase on the hypervascular component and persists into the venous and delayed phases on the fibrous component</td>
<td></td>
<td>MRI: In general usually are hypointense on T1-weighted images and hyperintense on T2-weighted images</td>
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Imaging findings OR Procedure details

Case 1:

A 71-year-old man patient presented with a 3-days history of dizziness, confusion and hyperpnea. The examination revealed a palpable, non-painful, and tumescent mass in right hypochondrium.

Laboratory parameters, including liver function tests, revealed decrease of albumin serum, increase of Gamma-glutamyltransferase (GGT) and hypoglycemia of 35 mg/dl.

Ultrasound images showed a large hypervascular mass occupying the right lobe and segment 4 of the liver with displacement of surrounding organs (Fig. 5 on page 11).

Unenhanced and contrast-enhanced CT scan was obtained in arterial and venous phase. The scan confirmed the presence of a large, lobulated and well-defined liver mass of 27 x 23 x 20 cm of diameters. The lesion was localized in right lobe and segment 4 and shows exofitic growing thru peritoneal cavity producing displacement of gallbladder, pancreas, mesenteric vessels and bowel loops. Unenhanced CT revealed a hypoattenuating and slightly heterogeneous mass. The arterial phase showed heterogeneous enhancement which hyperdense areas and hypervascularization in the intrahepatic tumor with fewer enhancement on the peritoneal portion (Fig. 6 on page 12) The venous phase demonstrated homogenous enhancement primarily on intrahepatic portion (Fig. 7 on page 13).

MR imaging was performed showing heterogeneous signal intensity mass on T2-weighted images (Fig. 8 on page 14) and markedly low intensity signal on T1-weighted images with focal areas of high signal intensity. Gadolinium-enhanced T1-weighted in arterial phase shows early enhancement seen on the hypervascular component mainly in the peripheral intrahepatic portion and fewer or non-enhancement areas on the peritoneal portion of the tumor (Fig. 9 on page 15) these findings suggested areas of necrosis or cystic degeneration. On venous phase the intrahepatic portion as the same as unenhanced CT, showed homogeneous enhancement (Fig. 10 on page 23).

A percutaneous fine needle biopsy of the lesion was consistent with a solitary fibrous tumor based on immunohistochemical parameters, and posteriorly the intrasurgical biopsy confirms the results.
Case 2:

A 60-year-old man patient presented with non-painful abdominal mass. The examination revealed a palpable, non-painful and apparently lobulated mass in hypogastrium.

Laboratory parameters, including liver function tests, were unremarkable.

Ultrasound images revealed a large pelvic mass occupying mostly completely the hypogastric region. The mass was hypoechoic with internal hyperechoic areas that produced displacement of neighboring organs (Fig. 11 on page 21).

Unenhanced and contrast-enhanced CT scan was obtained in venous phase. The scan confirmed the presence of a large, lobulated and well-defined pelvic mass of 17 x 15 x 13 cm of diameters. The lesion was localized in Retzius space producing displacement of urinary bladder. Unenhanced CT revealed a hypoattenuated pelvic mass with coarse calcifications on it caudal portion (Fig. 12 on page 20). Contrast-enhanced CT demonstrated heterogeneous tumor with internal hypoattenuated areas (Fig. 13 on page 19).

MR imaging revealed heterogeneous signal intensity mass on T2-weighted images (Fig. 14 on page 18 and Fig. 15 on page 17) and markedly low intensity signal on T1-weighted images. Gadolinium-enhanced T1-weighted in arterial phase shows early enhancement with hypervascular regions and development of central vessels. In addition, is visualized a hypoenhancemented area in the central portion of the mass (Fig. 16 on page 16) Venous phase showed persistent enhancement correlates with hypercellular areas and fewer enhancement in others areas, these findings suggested areas of necrosis or fibrosis (Fig. 17 on page 22).

Surgery resection was performed and the pathological result was a solitary fibrous tumor based on immunohistochemical parameter.

Images for this section:
Solitary fibrous tumors of the liver: 
Ultrasound findings

Figure 5 a and b: Hypervascular liver mass occupying the right lobe and segment IV with displacement of the adjacent structures

**Fig. 5:** a and b: Hypervascular liver mass occupying the right lobe and segment IV with displacement of the adjacent structures
Fig. 6: Contrast-enhanced CT scan obtained in arterial phase. a and b) coronal and axial images shows a large liver mass with peritoneal extension. The mass shows heterogeneous enhancement and present internal hyperdense areas and hypervascularization in the intrahepatic portion (arrow) with fewer enhancement on the peritoneal portion (*)
**Fig. 7:** Contrast-enhanced CT scan obtained in venous phase. a and b) Coronal and sagittal images: well-defined liver lesion localized in the right lobe and segment IV shows exofitic growing to peritoneal cavity with displacement of gallbladder, pancreas, mesenteric vessels and bowel loops.
**Fig. 8:** a) Coronal T2-weighted image shows a large, lobulated and well-defined liver mass. The mass shows heterogeneous signal intensity b) Axial fat-suppressed T2-weighted MR image shows mixed signal intensity mass with peritoneal extension. The mass reveals areas of high signal intensity that might correspond with areas of necrosis, cystic or myxoid degeneration.
Fig. 9: Axial gadolinium-enhanced T1-weighted in arterial phase.

a) Early arterial enhancement is seen on the hypervascular component mainly on the peripheral intrahepatic portion

b) Less enhancement and non-enhancement areas on the peritoneal portion of the tumor
Fig. 16: Pelvic MR. Gadolinium enhanced T1-weighted. a) early arterial enhancement is seen on the hypervascular component, mainly on the periphery and in central vessels. b) heterogeneous enhancement in venous phase. c) The enhancement persists into delayed phases and show fewer central enhancement, indicating the presence of necrosis / fibrosis / degeneration
**Fig. 15:** Pelvic MR. a and b) Sagital and coronal T2-weighted images: lobulated and heterogeneous pelvic mass with absence of signal intensity in some parts of the tumor, indicating the presence of coarse calcifications inside the tumor.
Solitary fibrous tumors of the pelvis: Magnetic resonance findings

Figure 14. Pelvic MR. a) Axial STIR MR image and b) T2-weighted image shows a large, well-defined pelvic mass with markedly heterogeneous signal intensity

**Fig. 14:** Pelvic MR. a) Axial STIR MR image and b) T2-weighted image shows a large, well-defined pelvic mass with markedly heterogeneous signal intensity
Fig. 13: Contrast-enhanced CT scan obtained in venous phase. a and b) Coronal and sagittal images shows a large, lobulated, well-defined mass localized in Retzius Space displacing the bladder inferiorly.
Fig. 12: a and b) Unenhanced CT: Hypoattenuated pelvic mass in Retzius Space. The mass present coarse calcifications on it caudal portion (arrow)
**Fig. 11:** Transverse scan shows hypoechoic pelvic mass with internal hyperechoic areas. The mass displace contiguous structures.
**Fig. 17:** Pelvic MR. Gadolinium enhanced T1-weighted. a and b) Coronal and sagittal venous phase shows heterogeneous enhancement. The mass produce displacement of the bladder keeping the plane of cleavage.
Fig. 10: Axial gadolinium-enhanced T1-weighted in venous phase. a) homogeneous enhancement of the liver mass. b) heterogeneous enhancement which areas of low and intermediate signal mainly on the peritoneal portion corresponding with areas of necrosis or cystic or myxoid degeneration (*)
Conclusion

Abdominal fibrous solitary tumors are infrequent neoplasm, with a clinicobiologic variable behavior and recognition of the radiological findings might help to suggest the diagnosis as well as to collaborate with the decisions making as for treatment.

References


Personal Information