Patterns of thoracic and extra-thoracic solitary fibrous tumors

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Learning objectives

The purpose of this exhibit is to illustrate and describe the spectrum of radiological findings found in solitary fibrous tumors (SFT), emphasizing the CT and MR characteristics. We aim to present a variety of SFT including the SFT of the pleura (the most common), thoracic extra-pleural, SFTs of peritoneum and pelvis including a rare case of vesical SFT, SFT of the neck/paranasal and SFT of the limbs.
Background

Introduction:

Solitary fibrous tumors (SFT) are uncommon tumors arising from mesenchymal cells representing less than 2% of all soft-tissue tumors.

SFT was described for the first time in 1931; refer to pleural localization, being described later in other extrathoracic localizations. The most commonly reported extrathoracic locations are the retroperitoneum, deep soft tissues of the proximal extremities, abdominal cavity, and head and neck. Although SFTs were thought to predominantly involve the pleura, it is now established that SFTs can originate in virtually any site of the body.

Epidemiologically rare reports of SFT outside the thorax have been made with less than 100 cases of extrapleural SFT have been reported worldwide.

The majority are benign, although up to 20% may be malignant.

Clinical features and management:

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

Most of the patients with SFT of the pleura (the most common type) are asymptomatic at the time of diagnosis, and the majority of these tumors are discovered on routine roentgenograms of the chest. In the remaining patients, the most common clinical symptoms are chest pain, cough and dyspnea. SFT may occur in benign and malignant forms, these latter showing locally invasive properties or relapsing after surgical resection.

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.

SFT in unusual locations (paraspinal, paramediastinal, intrafissural, abdominal), can pose interpretation problems or, indeed, point towards a different diagnosis.
The malignant potential of extrathoracic SFTs is similar to that of intrathoracic tumors, with malignant features detected at pathologic examination in up to 20% of cases in both categories. Most patients with extrapleural SFTs have a favorable outcome after surgical resection however rates of local recurrence are reported to be slightly higher with extrathoracic SFTs.

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. 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.

**Pathologic features:**

Pre-operative diagnosis can be obtained by a transthoracic cutting needle biopsy, but in most cases only pathological evaluation of the resected specimen supported by immunoreactivity of neoplastic cells for CD34 or CD99 allows confirmation of diagnosis. Concerning microscopic features, the most common architectural pattern is the so-called "patternless pattern", in which spindle cells with bland ovoidal vesicular nuclei, scarce cytoplasm, and connective tissue are arranged in a random pattern characterized by a combination of alternating hypocellular and hypercellular areas.
Findings and procedure details

General radiological Manifestations:

CT and MR findings are very dependent on tumor size.

**Computed tomography (CT):** CT scan is the key examination which more clearly shows the size and location of the tumor and aids in surgical planning. SFTs appear as well-circumscribed, hypervascular mass 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.

**SFT BY SITE:**

1. **THORACIC SFT - SFT OF THE PLEURA AND EXTRA-PLEURAL**

In case of small SFT of the pleura *(figure 1, 2, 3)*, CT more frequently typically demonstrates a homogeneous well-defined, non-invasive, lobular, soft-tissue mass, adjacent to the chest wall or within a fissure, showing an obtuse angle with the pleural surface.

Larger lesions *(figure 4, 5)* are typically heterogeneous and may not exhibit CT features suggestive of pleural tumors. Such lesions usually form acute angles with the adjacent pleural surface mimicking a subpleural pulmonary mass that could be misdiagnosed as peripheral lung cancer.

SFT of the pleura have been reported to exhibit intermediate to high attenuation on unenhanced CT scans. This attenuation has been attributed to the high physical density of collagen and the abundant capillary network within these lesions.
Intralesional calcifications are constantly associated with areas of necrosis and more easily seen in larger lesions.

In case of large masses, enhancement is typically intense and heterogeneous with central areas of low attenuation. Such intralesional geographic pattern has been shown to correlate with myxoid changes and areas of hemorrhage, necrosis, or cystic degeneration.

In the case of SFT of the pleura the mass effect of large lesions may produce atelectasis and displacement of brochi and vessels, but there should be no evidence of invasion into the lung or chest wall, nor multiple pleural seed. The lesions may grow to be very large, almost filling a hemithorax.

MR usually shows heterogeneous signal intensity on T1 and T2 weighted images with avid contrast enhancement following gadolinium administration. Areas of low T2 signal and lack of invasion of adjacent structures are some helpful features in suggesting this tumor.

Thoracic extra-pleural SFT generally present similar CT and MR characteristics as their pleural counterpart with no obvious connection with the pleural surfaces (figure 7, 8).

**Differential diagnosis of thoracic SFT:**

- The preoperative differential diagnosis that arises in a patient with a SFT of the pleura is essentially that of any mass lesion in the chest, ranging from carcinoma of the lung to various intrapleural sarcomas.
- The usual well-circumscribed appearance of the SFTP generally rules out malignant pleural mesothelioma since the latter invariably consists of multiple scattered pleural masses or is a more diffuse mass encasing the lung.
- The differential diagnosis becomes more difficult when SFT develops in particular sites, thus increasing the number of possible diagnoses.
- When located in the paraspinal area, SFT may appear indistinguishable from neurogenic tumours. In these cases it is important to evaluate the ribs: chest wall involvement by SFT is rare and usually manifests as sclerosis or cortical erosion at the costal level, a feature more typical of tumours of neurogenic origin.
- SFT of the pleura that have a mediastinic pleural origin can mimic a mediastinal neoplasm - in lesions of pleural origin the mediastinum is compressed and dislocated, contrary to what occurs in the presence of a mediastinal mass (which expands, compressing the pulmonary parenchyma without causing mediastinal shift)
- Tumors located within the fissural space may also be interpreted as pulmonary masses when they appear totally surrounded by pulmonary parenchyma.
2. 2. EXTRA-THORACIC SFT

Extrapleural SFTs typically manifest as large, slow-growing soft-tissue neoplasms.

SFT OF ABDOMINAL WALL, 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, 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 hypoglycaemia. Malignant degeneration with recurrences has been reported in SFTs arising from the pelvic peritoneum, although the exact prevalence is unknown. At imaging, pelvic SFTs (figure 9, 10) 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 large malignant tumors.

The urinary bladder is a very unusual location for solitary fibrous tumors. Patients may present with gross hematuria, abdominal pain, or urinary retention. On imaging, a heterogeneous vascular tumor that is either intravesicular or exophytic can be encountered (figure 11). The differential diagnosis includes inflammatory pseudotumor, leiomyosarcoma, sarcomatoid transitional cell carcinoma, and pheochromocytoma.

SFT OF THE NECK

SFT rarely originate in the intracranial or extracranial head and neck regions. Head and neck SFT have a good prognosis and are more likely to be benign than are SFT elsewhere.

Most solitary fibrous tumors of the head and neck arise in the nasal cavity or paranasal sinuses (figure 12, 13). They are also found in the nasopharynx, parapharyngeal space, buccal space, and larynx (figure 14, 15, 16).

Intracranial SFT are usually of dural origin and may have dural tails and cause hyperostosis of the overlying calvaria. The imaging differential diagnosis depends on the particular location of the tumor, but mainly includes meningioma, neurogenic tumor, and soft-tissue sarcoma.

Computed tomographic and magnetic resonance imaging generally show well-circumscribed tumors that enhanced strongly with contrast.

At MR imaging, these tumors appear as well-circumscribed, solid masses with low to intermediate signal intensity on T1-weighted images and heterogeneously high signal intensity on T2-weighted images. T2-weighted images may also show a low-
signal-intensity rim representing a pseudocapsule around the lesion. The tumors demonstrate variable enhancement, but enhancement is often intense because of their high vascularity.

**SFT OF SOFT TISSUE AND BONES**

Approximately 10% of SFTs arise from somatic soft tissues involving various sites, mainly the extremities and the head and neck, followed by the abdominal wall and other sites, including the bones and diaphragm (figure 17, 18, 19). Only a few reports have described SFT involving the extremities. Most occur in the proximal lower extremities.

Extra-thoracic SFT generally have a lobulated contour, are well defined, tending to displace adjacent structures. They also tend to be highly vascular with avid contrast enhancement.

Like SFT in other locations, intramuscular SFT have rich vascularity and are enhancing on both CT and MR images.

A wide range of benign and malignant tumors like desmoid tumor, fibrosarcoma, dermatofibrosarcoma protuberans, synovial sarcoma, neurogenic tumors, and malignant fibrous histiocytoma can mimic SFTs clinically and at imaging.
Fig. 1: Benign SFT of the pleura in an asymptomatic woman. Unenhanced (a) and enhanced (b) CT images demonstrating a small enhancing well-defined mass in the left hemithorax. Although the lesion doesn’t form a clearly obtuse angle with the adjacent pleural surface a smooth tapering margin is seen. The diagnosis of solitary fibrous tumour of the pleura was confirmed at surgery.

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Fig. 2: SFT of the mediastinal pleura. Axial unenhanced(a), arterial (b) and venous (c) phase CT images showing a slightly heterogenous enhancing pedunculated mass with smooth margins.

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**Fig. 3:** Malignant SFT of the pleura in woman, 82 years. Axial enhanced CT images (a) showing an inhomogeneous lobular enhancing mass in the left thoracic wall representing an exophytic component of the parietal pleura. One year later there are new pleural lesions (arrows), both with obtuse angles against adjacent pleural surfaces and one of them (in c) showing heterogenous enhancement.

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**Fig. 6:** Malignant SFT of the pleura. Contrast-enhanced CT scan demonstrates a homogeneous hypoenhancing spherical soft tissue mass abutting the parietal pleura. It also shows a heterogeneous large liver mass biopsy proven to be a SFT metastasis.

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**Fig. 4:** Large malignant SFT of the pleura. Axial (a) and coronal (b,c) enhanced CT images showing an extensive right pleural mass mimicking a mesothelioma. The diagnosis of solitary fibrous tumour of the pleura (SFT) was confirmed at surgery.

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**Fig. 5:** SFT of the pleura. Unenhanced (a) and enhanced (b) CT images showing a large inhomogeneous mass filling the entire left hemithorax with slight mediasinal shift to the right. There are spontaneous dense foci of hemorrhage and extensive areas of necrosis.

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**Fig. 7:** Dorsal thoracic SFT in man, 55 years. Axial unenhanced (a) and enhanced (b) CT shows a homogeneous lobular enhancing mass in the left posterior thoracic wall with smooth well-defined margins. This patient was submitted to resection with no evidence of recurrence to date.

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**Fig. 8:** Thoracic wall SFT. PA radiography (a) and enhanced axial CT images (b) showing a large well-defined exophytic mass with marked heterogeneous enhancement. The diagnosis of SFT was obtained by a transthoracic needle biopsy.

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**Fig. 9:** Typical imaging manifestations of SFT of the peritoneum. Axial (a) and coronal (b) contrast material-enhanced CT scan shows a well-defined intensely enhancing mass in the pelvis with central nonenhancing areas of necrosis (arrows).

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**Fig. 11:** SFT of the bladder. Axial (a) and coronal (b) CT images showing an intravesical heterogeneous vascularized mass. In the late phase axial images (c) the lobular contour is more conspicuous.

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**Fig. 10:** Atypical presentation of a SFT of the pelvis. Axial CT images showing a large heterogeneous mass with central necrosis and thick walls. It was biopsy proven to be a SFT of the pelvis.

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**Fig. 12:** SFT of the left nasal cavity. The left nasal cavity is filled with a solid, lobular, enhancing mass.

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**Fig. 13:** SFT of the right palatine tonsil. Enhanced CT images (a,b,c) showing a well-defined homogeneous enhancing mass at the right palatine tonsil. No adenopathy is present.

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**Fig. 15:** SFT of the parapharyngeal space. Axial CT after administration of intravenous contrast medium demonstrates a small homogeneous soft tissue mass with lobular borders in the right parapharyngeal space. Axial T1-weighted (b), T2-weighted (c) and gadolinium-enhanced T1-weighted MR images (d) showing a well-defined lesion hypointense on T1-weighted images, slightly hyperintense on T2-weighted images and avidly homogeneously enhancing.

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**Fig. 16:** SFT arising from the floor of the mouth. Axial T1-weighted (a), T2-weighted (b), and gadolinium-enhanced T1-weighted (c) MR images showing a large, well-defined mass. The mass has intermediate intensity signal on the T2-weighted image, isointense to muscle on the T1-weighted image showing moderate heterogeneous enhancement on the gadolinium-enhanced images.

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**Fig. 19:** Somatic SFT of the right supraclavicular fossa. MR images showing a large, well-defined enhancing mass at the right supraclavicular fossa. The tumor demonstrates signal isointensity relative to muscle on T1-weighted images (a), slight hiperintensity on T2-weighted images (b) and avid enhancement, with some non enhancing areas due to tissue necrosis on the contrast-enhanced MR images (c).

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**Fig. 17:** Axial T1-weighted (a) T2-weighted (b) and contrast-enhanced MR images show a well-defined, enhancing soft-tissue mass in the medial compartment of the right thigh. Feeding vessels have low signal intensity in all sequences due to flow void.

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**Fig. 18:** Axial T1-weighted (a) T2-weighted fat saturated (b) and contrast-enhanced MR images (c) show well-defined, heterogeneously enhancing soft-tissue mass in the lateral compartment of the thigh. TOF sequence MR images (d) show the hypervascularity of the lesion, biopsy proven to be a SFT.

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Fig. 14: SFT of the buccal space. Axial (a) and sagittal (b) CT images showing a spherical enhancing mass in the right buccal space, biopsy proven to be a SFT.

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Conclusion

Extrapleural SFTs are seen nowadays more commonly than their pleural counterparts. Findings on CT scans alone are generally nonspecific for SFT being dependent on tumor size. However, a frequent and unifying feature of SFT at MRI is the presence of low signal-intensity foci on T1- and T2-weighted images, corresponding to the collagen content. Smaller tumors tend to enhance homogeneously, whereas larger lesions may have central tubular or rounded low-attenuation areas due to cystic or necrotic change. When a central focus of heterogeneity and variable contrast enhancement is identified in CT or MRI images, malignant degeneration should be considered.
References

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