**Solitary Fibrous Tumor of the Pleura: multimodal imaging findings with pathologic correlation**

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Learning objectives

• To exhibit key imaging findings of Solitary Fibrous Tumor of the Pleura (SFTP) with conventional x-ray, CT, US, MRI and PET-C
• To correlate imaging with pathologic findings
• To discuss the differential diagnosis of SFTP
Background

Summary:

SFTP is the second most common primary pleural neoplasm after mesothelioma. It is a slow growth tumor with mesenchymal origin and indeterminate malignant potential. It is usually an incidental finding with little or no symptoms, occasionally growing into large intrathoracic masses that may cause compressive symptoms. Knowledge of the key imaging findings in each imaging modality is mandatory to establish an accurate differential diagnosis with other solid pleural lesions.

History:

In 1870 Wagner provided the first report of Solitary Fibrous Tumor of Pleura (SFTP), but it was not recognized as a distinct entity until 1931, when Klemperer and Rabin published an accurate pathologic description (1). There has been wide variety of terms used to describe this neoplasm due to its controversial histogenesis.

The hypothesis of a neoplasm with mesothelial origin was suggested by Stout and Murray in 1942, where terms like "Localized mesothelioma" or "Benign mesothelioma" were incorrectly used (2). Others terms in the same line include: Localized fibrous mesothelioma, benign fibrous mesothelioma, solitary fibrous mesothelioma and fibrous mesothelioma.

However, other investigators showed that the mesothelial layer covering the tumor was not affected and they postulated that the epithelial cells seen could have been imbibed within growing fibrous mesenchymal tumors with a submesothelial origin (1).

The controversy on the origin of these tumors persisted for several decades and it is reflected by the variety of terms given to the neoplasm: submesothelial fibroma, subserosal fibroma, subpleural fibroma, submesothelioma, benign pleural fibroma, pleural fibromyxoma and pleural fibroma (3).

Currently, thanks to immunohistochemical techniques, there is a consensus that SFT is a distinctive mesenchymal neoplasm and it’s classified by the World Health Organization like a soft- tissue neoplasm of pluripotent fibroblastic or myofibroblastic origin that may arise anywhere throughout the body and may have a malignant potential (4).
Furthermore, in the revised classification hemangiopericytoma, which was previously considered a distinct vascular soft-tissue tumor, is now considered as a cellular variant of SFT and lipomatous hemangiopericytomas are actually regarded as fat-forming variant of solitary fibrous tumors(2)(4).

**Epidemiology:**

These tumors account for less than 5% of all tumors arising from the pleura (5), but it is the second most common primary pleural neoplasm. It also represents less than 2% of all soft-tissue tumors(6).

SFTP affect patients in the 6th and 7th decades of life(7), and majority of studies have showed equal frequency in both men and women (5). Although some authors have found a slight female predominance, this is still not significant (3).

It is difficult to know the true incidence and prevalence of solitary fibrous tumors because the majority of patients with these masses are asymptomatic. Approximately 800 cases of SFTP have been reported in the literature between 1931 and July 2002; and in 1978 Okike and colleagues estimated a prevalence of 2.8 per 100,000 individuals, in their institution Mayo Clinic (8).

Nevertheless, an increasing number of case series have been reported, with more than 1760 cases of SFTP until now (9)(10). This increase of incidence, could be attributed to widespread imaging use in clinical practice with resulting detection of a larger number of incidental tumors, and also could be due to the advancement of immunohistochemistry and electron-microscopy that has helped to identify extra-thoracic fibrous tumors (11).

Although solitary fibrous tumors most commonly occur in the pleura (5), numerous extra-thoracic sites of involvement have been reported (12). In this way, after its recognition in pleura and lung (12)(13), the solitary fibrous tumors(SFTs) were described in other serosal surfaces, like pericardium and peritoneum and was wrong called "localized fibrous tumor of the serosal cavities". Subsequently, SFTs has been discovered in almost every-where in the body, including abdomen (liver, kidney, retroperitoneum and urinary bladder), head (meninges and orbit), neck (thyroid and salivary gland) and soft tissues of breast and extremities. They have also been reported in the upper respiratory tract like the nose, paranasal sinuses, parapharyngeal tissues, nasopharynx and epiglottis. All of them share the characteristic of a mesenchymal origin, and are included under the term of Solitary Fibrous Tumor (SFT)(14).

**Pathogenesis:**
The most accredited hypothesis of etiopathogenesis is that these tumors originate from stromal cells (from submesothelial layer in case of pleura) with pluripotent fibroblastic or myofibroblastic phenotype cells (mesenchymal origin), whose growth is promoted by an aberrant reaction to inflammatory or hormonal stimuli (15).

Nonetheless, in 2013 a distinctive and specific gene fusion, NAB2-STAT6, has been identified in solitary fibrous tumors(16)(17). The same gene fusion was identified more recently in so called meningeal 'haemagiopericytoma', confirming that these tumors simply represent an example of solitary fibrous tumors, rather than a distinct entity (18)(2). These gene fusion results in notable over-expression of STAT6, which proves to be a specific immunohistochemical marker for solitary fibrous tumors. This is a remarkable finding but yet with undetermined significance in etiopathogenesis or genetic predisposition(4). Until now, there is only one report of SFTP occurring in family members (19).

There is no known association with tobacco, asbestos or any other environmental pollutants(20).

**Clinical Characteristics:**

Most of patients are entirely asymptomatic at the time of the diagnosis and SFTP is discovered as an incidental finding. Majority of symptoms are related with the size of tumor because they can grow to a large size, exerting mass effect in adjacent structures. In this way, large tumor are more likely to be symptomatic than small ones (21).

Most patients complain of nonspecific symptoms such as cough, thoracic pressure/pain, dyspnea and sensation of a mass moving within the chest (21). Signs of upper congestion might be find in case of superior vena cava compression. Also abdominal pain could occur in patients with supradiaphragmatic tumors. Less frequently, cases with hemoptysis and obstructive pneumonitis have also been reported. Systemic symptoms may occur including chills, sweats, weakness, and weight loss (3).

SFTP can be associated to paraneoplastic syndromes such as digital clubbing, Doege-Potter syndrome or Pierre-Marie-Bamberg syndrome, and they are most frequently found in malignant forms (14).

Digital clubbing and hypertrophic pulmonary osteoarthropathy (Pierre-Marie-Bamberg syndrome) are the most frequent paraneoplastic syndrome. It have been described in
10% to 20% of patients with benign or malignant SFTP and consist in pain, stiffness and swelling of joints, probably caused by an excessive release of hyaluronic acid (7)(11). These clinical features usually resolve within 2 to 5 months after excision of the tumor, but they can reappear in case of recurrence (22).

Doege-Potter syndrome consist in a refractory hypoglycemia, reported in less than 5-6% of patients with SFTP (22). Many theories have been proposed to explain this phenomenon, including increased glucose consumption by the tumor and proliferation of insulin receptors. Currently, the hypoglycemic mechanism demonstrated is a large form (high weight) of insulin-like growth factor 2 (IGF-2), which is probably an incompletely processed molecule of IGF-2, secreted from the tumor (23). A high serum level of insulin-like growth factor II, is typically associated with low levels of insulin, but these values returns to normality within 3 to 4 days after resection of the tumor (11).
Findings and procedure details

Imaging Appearance:

Radiography

SFTP usually appear as a peripheral opacity with well-defined borders and elongated shape (with the largest dimension in the longitudinal plane), that predominantly affects the middle or inferior hemithorax (Fig. 1 on page 18 A-B) (5). However, it is important to note that these tumors are widely distributed in the thorax.

They are typically in contact with the pleural surface, showing an "incomplete border sign", with an ill-defined margin, but may also be in fissure locations surrounded by lung with well-defined borders, mimicking a lung tumor (Fig. 2 on page 18 A-B) (24).

Despite they are pleural lesions and so they can show obtuse margins with thoracic wall (a typical sign of extrapulmonary location) (Fig. 5 on page 21 A-B), bigger lesions can present acute margins with thoracic wall, and can be confused with pulmonary masses (Fig. 5 on page 21 A-B), (3).

Change in shape and location during respiration or patient reposition, is also a typical sign in pedunculated lesions. SFTP also exhibits slow growth over time reaching enormous sizes (25).

Computed Tomography

Computed tomography (CT) is the best modality to evaluate tumor size and location, being also more sensitive to detect pedicle and encapsulation (5). SFTP typically appears in contact with the pleural surface, showing more displacement (mass effect) than invasion of the surrounding structures (Fig. 3 on page 19 C-D) (26). They have a variable size, being able to reach a large diameter that exerts mass effect over the mediastinum and adjacent lung, with passive atelectasis and displacement of bronchi and vessels.

CT usually demonstrates a solitary well-delineated, homogeneous and lobulated mass of soft tissue attenuation (Fig. 2 on page 18 C-E) (14). However, heterogeneous density may be observed because of hemorrhage, necrosis and cystic or myxoid degenerations (Fig. 5 on page 21 B-D). This heterogeneity is frequently manifested as areas of
low attenuation, with an incidence of 50-60% in benign lesions (BSFTP) and 100% in malignant solitary fibrous tumors (MSFTP), and can be attributed to the rapid growth of malignant variants that more often induces hemorrhage and necrosis (3).

Intermediate to high attenuation (respect to muscle) on unenhanced CT scans has also been described and it is attributed to the high density of collagen and abundant capillary network within these lesions (3). Calcifications are often seen (11-26%) regardless of their benign or malignant histologic features and they could be punctate, linear or coarse (Fig. 4 on page 20 D-F and Fig. 5 D) (1).

SFTP may form obtuse angles against the adjacent pleura (as described in other pleural lesions), but it is frequent to find acute angles, especially in large forms (Fig. 3 on page 19 C-D). Rosado found that 33% of the SFTP exhibited at least one obtuse or right angle and 3% exhibited only obtuse angles. Dedrick and colleagues described a smoothly tapering margin adjacent to the tumor helpful to establish the pleural location of these tumors, but unfortunately it is not present in all cases (26).

CT findings are highly dependent on tumor size. In this way, small SFTPs are homogeneous and show obtuse angle with the pleural surface, while large lesions are typically heterogeneous and may not exhibit CT features suggestive of focal pleural tumors.

Due to their rich vascularity these lesions have a significant enhancement on CT, which is typically heterogeneous (particularly in large lesions) with non-viable areas of low attenuation, that correspond to necrosis, myxoid or cystic degeneration (Fig. 5 on page 21 B-D) (3).

Lesions arising from the pleura, may show both mobility and deformability, with resultant change in location and shape on serial imaging (Fig. 1 on page 18 F-G). CT visualization of pedicle is infrequent, but it is possible to find changing patient position and may contain vascular supply of tumor (Fig. 2 on page 18E) (Fig. 3 on page 19) (14). It is also possible to find a sessile attachment (Fig. 4 on page 20 G).

A concurrent pleural effusion (serous or serosanguineous) is reported in a range between 0 and 12% of cases (10). In contrast, detection of hilar or mediastinal lymphadenopathies is not a feature of SFT and exclude its diagnosis. Sclerosis or pressure erosion on adjacent ribs have been reported in association with SFTP, but it is more characteristic of neurogenic neoplasms (26) (12).
Although, there are no pathognomonic features on CT that suggest malignancy, some finding are more characteristics in malignant forms (27). Malignant solitary fibrous tumor of pleura (mSFTP) typically appear as large masses exceeding 10 cm with heterogeneous attenuation (low-attenuation areas within the tumor) and patchy enhancement that could associate ipsilateral pleural effusion (32%) and irregularity or pleural enhancing (11)(26). Lesion multiplicity (Fig. 6 on page 22) and local invasion are also more likely for malignant forms (28).

Extrathoracic SFTs usually have similar characteristics on image that SFTP (Fig. 7 on page 25 and Fig. 8 on page 23).

**Magnetic Resonance**

MRI is superior to CT for the evaluation of tumor origin, extension and local invasion, particularly with respect to the chest wall and diaphragm, which is helpful for surgical planning (22). SFTP typically show low to intermediate signal intensity on T1-weighted and heterogeneous signal on T2-weighted, with low intensity images related to collagen content and low cellularity (reduced proton mobility), and areas with high signal intensity because of increased vascularity (edema), necrosis and cystic or myxoid degeneration (Fig. 1 on page 18 H-J) (5) (12). Prominent vascular structures may produce flow voids inside the tumor (3). Theses masses typically also have an intense enhancement after administration of gadolinium (Fig. 1 on page 18 H-J and Fig. 7 E-G)(14).

**Angiography**

Angiography has a limited role in the evaluation of SFTP, but is useful to determine the vascular supply of tumors, in case of preoperative embolization to reduce intraoperative blood loss (14)(5). Blood supply usually goes through the pedicle and may come from intercostal, internal mammary or inferior phrenic arteries, which is helpful to suspect extrapulmonary origin (3).

**Ultrasonography**

Ultrasonography can help to establish the extrapulmonary location of the thoracic mass and can be used to perform a US-guided a biopsy (3). At ultrasound these lesions are typically hypoechoic, and occasionally heterogeneous, correlating to the heterogeneity seen on images obtained with other modalities (Fig. 3 on page 19 E) (Fig. 4 on page 20 C) (Fig. 5 on page 21 E) (12) (14).
**F-FDG PET-CT**

Benign SFTP typically show a low level of FDG uptake at PET (SUV < 2.5), similar to mediastinal blood pool (Fig. 2 on page 18 F); while malignant SFTP tend to be hypermetabolic (27). Cardillo et al. reported that PET had 50% positive predictive value and 87.5% negative predictive value for malignant forms (28). Nevertheless, further evidence is needed with large-cohort multicenter studies to evaluate and validate the role of FDG PET/CT (12).

Large tumors may show heterogeneously uptake with hypometabolic areas of necrosis, cystic degeneration, myxoid changes, hemorrhage and calcifications (14). In this way, PET could be useful before needle biopsy, to target metabolically active areas within these masses and reduces the number of no diagnostic biopsies. In addition, preoperative FDG PET/CT can be performed in patients whose radiologic features suggest a malignant tumor, for identify distant metastasis (29) (27).

It is also important to take into account, that in case of intense FDG uptake, alternative diagnoses should be suspected, including mesothelioma and metastatic disease.

**Gross Pathologic Features:**

SFTP are usually well-circumscribed masses with round or ovoid shape and smoothly lobulated surface covered by a thin and translucent membrane with prominent blood vessels (Fig. 4 on page 20 H-I) (5)(30). They are typically solitary lesions, arising from visceral pleura in 70 to 80% of cases and from parietal one in the rest (3)(5)(14).

Nearly half of the tumors have a single pedicle of variable size (usually 1 cm long), which usually contain the vascular supply (Fig. 1 on page 18 K-L) (Fig. 3 F-G) (5). Other tumors are broad-based with sessile attachment (Fig. 4 on page 20 H-I), and adhesions to the adjacent pleural surfaces and pericardium, without signs of invasions are common (30).

On dissection, the surface appears greyish-white, pink-white, yellow-than, pink-red and less common brown, with whorled or nodular pattern including areas of necrosis, hemorrhage myxoid or cystic degeneration (Fig. 2 on page 18 G) (5) (30). These tumors have variable size (1 to 36cm) with mean of 7-9cm and also variable weight that can surpass the 5.2 kg (26).
Whereas most of the benign SFTP are small and pedunculated tumors attached from visceral pleura, the malignant variants are often sessile lesions, larger than 10 cm with necrotic and hemorrhagic degeneration, growing in atypical location (for example mediastinal pleura)(30).

**Histological Characteristics:**

SFTP originate from submesothelial connective tissues and have primitive pluripotential cells with mesenchymal differentiation. Intrapulmonary SFT, derive from invagination of the visceral pleura, interlobar septal connective tissue or pulmonary parenchymal fibroblast.

At histologic analysis, SFTP appear as low-grade neoplasms of variable cellularity (inversely related to collagen content) (3). Tumor cells are ovoid to spindle-shaped with round to oval nuclei which usually possess open chromatin (giving a vesicular appearance) and a discreet nucleolus. Faintly eosinophilic, vacuolated and scarce cytoplasm is also seen. Nuclear pleomorphism is variable and mitoses are usually rare (22).

Collagen ranges from wispy fibrils surrounding tumor cells in hypercellular areas, to thick and dense collagen forming sclerotic zones in hypocellular areas. These findings provide a wide spectrum of histological features, from predominantly fibrous lesions containing large collagenized areas to more cellular and less fibrous neoplasms (22).

Tumors are usually well vascularized with vessels of varying sizes, and degenerative features including myxoid changes, necrosis and hemorrhage may occur (14).

Based on histological characteristics, SFTP has been classified in different variants. The Fibrous form is the most frequent variant and have a heterogeneous microscopic appearance, alternating cellular and fibrous areas with spindle cells and thick-walled vessels. Cellular form of SFTs (previously called haemangiopericytoma) usually have a monotonous appearance, with moderate to high cellularity, little fibrosis, thin-walled branching vessels and cells with round to oval nuclei (2).

In addition to the fibrotic and cellular forms of SFT, several other unusual variants have been added to the list: Fat-forming variant of SFT (Before called Lipomatous haemangiopericytoma), Giant cell-rich variant of SFT (previously called Giant cell angiofibroma) and Fibrous Histiocytoma-like variant of SFT (Before called Deep fibrous histiocytoma) (2).
Ultrastructurally, SFTs usually show features of fibroblastic to myofibroblastic differentiation, although a recent studies demonstrated that these tumors are more heterogeneous in their cellular composition, containing fibroblasts, myofibroblasts, endothelial cells, pericytes and undifferentiated perivascular cells in varying proportions (2).

Commonly used criteria for malignancy are those described by England and colleagues, which include (a) high cellularity with crowding and overlapping of nuclei, (b) high mitotic activity of more than four mitotic figures per 10 high-power fields, and (c) pleomorphism based on nuclear size and nucleoli prominence (30); and may be associated with tumor necrosis or infiltrative margins. However, Lococo et al found that isolated pleomorphism can be detected without any other sign of malignancy, and that is generally considered as an intermediate or borderline form (28).

Because of the diversity of histological features, even large biopsy specimens may pose a diagnostic problem for experimented pathologist, being in many cases insufficient for diagnosis. (3)

**Immunohistochemistry:**

Immunohistochemically, SFTs, especially the fibrous form, express CD34 (a marker for normal endothelium and vascular tumors) in 80-90% of cases and CD99 in 70% (31). Bcl-2 marker of terminal differentiation (30%), epithelial membrane antigen (EMA) (30%) and smooth muscle actin (SMA) (20%) may occasionally be expressed. Vimentin are also positive in the majority of benign solitary fibrous tumors (32). They are usually negative for S-100 protein Desmin and Cytokeratins (2).

Expression of vimentin and CD34 and absence of expression of cytoplasmic keratins, provide evidence of mesenchymal origin. The anti-apoptotic proto-oncogene Bcl-2 is constitutively expressed by all benign and malignant SFTP, which suggests that these tumors may derive from a long-lived CD34-positive "fibroblastic" stem cell (22).

Cellular variant of SFT tend to be less frequently positive for CD34 and when positive, the staining is usually less strong than in fibrous SFT and often focal (2). Cytokeratin, S-100, and p53 proteins have shown increased expression in malignant solitary fibrous tumors (14).

Immunohistochemistry may be useful in differentiating the SFTP from mesotheliomas and sarcomas (10).
Diagnosis:

The diagnosis of SFTP is typically made after surgical excision (34). Nonetheless, surgical resection for diagnosis and treatment is acceptable only if the patient is operable, because operative morbidity and mortality are very small. In this way, some cases need preoperative diagnosis for chose the best disease management (21).

In these cases, a large-bore cutting needle biopsies (Tru-cut) should be performed. (3). It is recommendable to use CT contrast-enhanced or metabolic imaging as guide to target cellular and metabolically active areas, avoiding areas of fibrous tissue and necrosis (14). There is a low risk of pneumothorax, because the needle can usually be introduced into the mass avoiding aerated lung.

Percutaneous transthoracic fine-needle aspiration biopsy (FNAB) should not be performed, because rarely provides enough tissue to give a definitive diagnosis (9)(28); and this is explained by the heterogeneous and variable structure of SFTP.

The clinical value of biopsy before excision is still controversial. Rosado found that 32% of the benign and 20% of the malignant LFTP were accurately diagnosed with needle biopsy(3). Lahon found that CT-guided biopsy have a positive predictive value of only 39% (29). Currently, different authors recommend that biopsy should only be done if disease management will be substantially affected by the results or if surgical intervention is contraindicated and a diagnosis would alter treatment (9) (29) (27).

Bronchoscopy is only indicate to rule out other lesions (10).

Malignant Potential & Metastatic Pattern

The clinical presentation and histological features of SFTPs are usually benign but approximately 12-33% are locally aggressive or malignant (27) (11).

Malignant SFTP may occur de novo or degenerate from benign tumor. In fact, benign tumors may recur with histologic signs of malignancy several years after resection. Possibly malignant degeneration of SFTP is generated by chromosomal anomalies and genetic mutations of the pro-apoptotic gene p53 and the proto-oncogene BCL-2, leading an excessive proliferation rate (22).

A combination of imaging, pathologic and immunohistochemical features are used to predict malignant potential. Imaging features include tumor size > 10cm, interval increase
in size, extensive necrosis and hemorrhage into the tumor, pleural effusions, adjacent soft tissues infiltration and chest wall invasion (11). Pathologic findings include more than 4 mitoses per 10 high power field, increased cellularity, increased pleomorphism and extensive necrosis and hemorrhage (30)(22). Findings in immunohistochemical analysis include, loss of CD34 expression, cytokeratin expression, p53 expression and S-100 expression. Identification of these features needs additional diagnostic imaging to exclude metastatic disease and recurrence (14).

Solitary fibrous tumors may develop metastases in 35% of cases with a median period of 124 months, and 8% have metastases at moment of diagnosis (33). Although malignant forms are more likely to make metastases, metastatic disease have also been described in patients with benign SFT (7%) and on contrary, some cases of malignant SFT will not develop metastases (21%) (33).

Tumor size # 10 cm and mitotic count # 4 per 10 high-power fields are associated with a higher incidence of metastatic disease (30). Patients with extrathoracic SFT have also more possibilities to develop metastases (27%) compared with those with thoracic SFT (9%).

The most common sites of metastatic disease in patients with thoracic SFT are pleura, chest or abdominal wall, lung and peritoneum, but metastases have been observed in several organs includes liver, bones and brain. These metastases are usually manifested as solid and hypervascular lesions, and in case of bone metastases as lytic lesions (33).

**Differential Diagnosis:**

The preoperative differential diagnosis that arises in a patient with a SFTP is wide and needs to consider specific features to short up diagnosis list.

Radiographically, the presence of acute angles with the adjacent pleural surface may mimic a subpleural pulmonary mass that could be misdiagnosed as peripheral lung cancer. When the lesions are abutting in the diaphragm could be confused with relaxed diaphragmatic eventration. (24)

Once identified as an extra-pulmonary lesion, the differential diagnoses include neoplastic conditions as pleural metastases (Fig. 9 on page 24), malignant mesothelioma (Fig. 10 on page 26), Askin tumor, pulmonary Ewing's sarcoma (Fig. 11 on page 27), lymphoma, pleural lipoma (Fig. 12 on page 27) and synovial sarcomas (35); and also include tumorlike conditions (Fig. 12 on page 27, Fig. 13 on page 28) as loculated pleural effusion, thoracic splenosis and thoracic endometriosis (11)(36). The
usual well-circumscribed appearance of the SFTP generally rules out malignant pleural mesothelioma.

Tumors located within the fissure space may also be interpreted as pulmonary masses, because they appear totally surrounded by pulmonary parenchyma (37). That lead a wide imaging differential diagnosis including hamartomas, carcinoid tumors, bronchogenic carcinoma and necrotizing pneumonia (22)(14).

When SFTP is located in para-spinal area, may appear indistinguishable from neurogenic tumors (peripheral nerve sheath tumors) (Fig. 14 on page 28), which have a higher incidence. In these cases, it is important to evaluate the ribs, because neurogenic tumors are more likely to make sclerosis or cortical erosion at the costal level (37).

SFTP with mediastinal pleural origin can mimic a mediastinal neoplasm and sometimes make the differentiation is impossible. In these cases, is important to have into account that SFTP usually exerts mass effect and displaces adjacent structures, rather than expanding or infiltrating the mediastinum. Differential diagnosis of mediastinal masses includes thymic epithelial neoplasm, germ cell tumor, pericardial mesothelioma, sarcoma, lymph nodal mass and also neurogenic tumors (14).

**Treatment:**

Complete surgical excision is the treatment of choice for both benign and malignant SFTPs, because provides excellent long-term survival with low postoperative morbidity and mortality (29)(22).

The operative approach and extent of surgical excision is clearly dictated by size, location and spatial relations (21). The surgeon should save as much lung as possible, obtaining free-margin resection (R0) with a distance of 1 to 2 cm from the tumor (38). It is suggested an intraoperative checking of the surgical margins to confirm the radicalism of the excision (22).

Small (< 5 cm) pedunculated lesions without radiological malignant sings and located on the visceral pleura, are safely removed by video-assisted thoracoscopic surgery (VATS) (23). Extreme caution should be used to avoid contact between the tumor and the thoracoscopic sites, because of the potential metastasis and local recurrence at the port sites. Patients with VATS obtain less blood loss and operative time, as well as hospital stay because they have less postoperative pain and reduced respiratory impairment (9).
However, thoracotomy is mandatory in large tumors (> 5 cm) performing wedge resection, segmentectomy, lobectomy or pneumonectomy (38). Wedge resection is the most common surgical procedure used in pedunculated tumors arising from visceral pleura. Segmentectomy, lobectomy or even pneumonectomy may be required in large sessile lesions or 'inverted' tumors (intrapulmonary forms)(10). Due to the malignant variety of SFTPs, the spillage at the time of surgery should be avoided to reduce the possibility of local recurrence (22).

Pleurectomy may be needed for SFTPs adhered or originated from parietal pleura, and concomitant chest wall resection can be necessary in case of ribs and intercostal musculature invasion (22).

In large-sized tumors, preoperative percutaneous embolization is useful to reduce the perioperative blood loss. Postoperative mortality and morbidity are related with hemodynamic changes developed by decompress lung and mediastinal structures(10).

Recurrence, Follow-up and Prognosis:

Incomplete resection is known to be associated with decrease survival and increase recurrence rates (29). Relapse may occur in approximately 30% of cases, with a recurrence rate in benign SFTPs of 2.5-8% and in malignant ones 14-63% (29)(38). SFTPs with sessile attachment and tumors CD4-negative (reflect tumor dedifferentiation) have a significant higher risk of recurrence.

Recurrent disease typically affects the ipsilateral pleura and may also affect the lung (3), and most of them have been noted to occur within 2 years after surgical resection (11). Repeated resection is the best therapeutic option for local recurrences and have provided good outcomes (29).

Despite the well-known local recurrence rate after resection for malignant SFTP, the role for adjuvant therapy remains controversial. Some studies described that adjuvant therapy is probably not helpful because of the low cellular content and low mitotic rates characteristic in these tumors (3); but others reports describe long-term survivals with postoperative radiotherapy in patients with incomplete resection of the tumor and rewarding responses to ifosfamide and doxorubicin for recurrent inoperable SFTP (1).

A staging system has been proposed to serve as a guideline for surgical management and adjuvant chemotherapy by Perrot et al. which divides these tumors principally into four classes: benign pedunculated SFTP; benign sessile SFTP; malignant pedunculated SFTP and malignant sessile SFTP(1). All of them require surgical resection. Benign
pedunculated SFTPs have a risk of recurrence of 2%; no adjuvant chemotherapy is required but annual imaging is recommended. Benign sessile SFTPs have a risk of recurrence of 8%; no adjuvant chemotherapy is required but semi-annual imaging for the first 2 years and subsequent annual imaging is recommended. Similarly, malignant pedunculated SFTPs have a risk of recurrence of 14%; no adjuvant chemotherapy is required but semi-annual imaging for the first 2 years and subsequent annual imaging is recommended. However, malignant sessile SFTPs have a risk of recurrence of 63%; adjuvant chemotherapy may be considered and semi-annual imaging for the first 2 years and subsequent annual imaging is recommended (22).

Otherwise, Lahon et al has suggested that in mSFTP with sessile morphologic type or CD34-negative immunostaining results, adjuvant therapy to avoid recurrence should be considered (29).

In conclusion, until now there is no adequate data to support the usage of radiotherapy and chemotherapy in the treatment of SFTPs.

Complete surgical resection represents the most important prognostic factor for SFTP (9). Benign SFTP have an excellent long-term survival, with 96% of patients alive after 5 years. In malignant SFTP the long-term survival is lower, mainly because of local recurrences, with a 5 year survival rate of 68% (29).

Although no formal follow-up guidelines exist at present, it is known that solitary fibrous tumors of the thorax tend to recur within the first 2 years after resection; consequently, a long-term follow-up with CT every 6 months for 2 years and yearly thereafter is mandatory after complete resection to exclude tumor recurrence or metastatic disease (29).
Fig. 1: Benign Solitary Fibrous Tumor of Pleura

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Figure 2: 49 year-old male with an incidental finding of a chest mass. Chest X-ray (A,B) depicted a nodular lesion over left upper lobe with obtuse angles and a sharply marginated medial border and an ill-defined lateral border, consistent with incomplete border sign (blue arrow) which supports extrapulmonary location. A CT (C-G) was later performed depicting a 2 cm extrapleural non-infiltrating mass with single pedicle (red arrow). No pleural effusion was found. Fusion images of PET-CT (F) revealed a low glucose uptake (SUVmax 1.4), similar to mediastinal blood pool. All these findings strongly suggested a benign solitary fibrous tumor of the pleura. The lesion was removed by VATS. Pathology specimen showed a well defined subpleural whitish nodular mass. Histologic analysis confirmed the diagnosis.

Fig. 2: Benign Solitary Fibrous Tumor of Pleura

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Fig. 3. 59 year-old woman with previous history of smoking and COPD. Chest X-ray (A-B) showed a left subpulmonary mass (red arrows) that mimic an elevated left hemidiaphragm. Chest CT (C-D) showed a 10 cm extrapulmonar mass above the left hemidiaphragm (Green arrows). The lesion was heterogeneous and showed a pedicle on its superior aspect (red arrowhead). Ultrasound (E) also depicted its heterogeneity and clearly showed its relation with the diaphragmatic pleura. It was suspected to be a SFTP and was surgically removed. Pathologic specimen (F-G) showed a smooth surface with a single pedicle (blue arrowhead). The inner structure (F) showed hypervascularized regions, hemorrhagic foci and necrosis. Histologic analysis confirmed the diagnosis of benign SFTP.

Fig. 3: Benign Solitary Fibrous Tumor of Pleura

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Fig. 4. 63 year-old woman with a huge chest mass incidentally discovered on a routine preoperative chest X-ray (A,B) that revealed an enormous right subpulmonary mass. Ultrasound (C) depicted an heterogeneous lesion, predominantly hypoechoic with calcifications and hyperechogenic areas, as well as anechoic lines due to vessels and cystic degeneration. Chest CT (D-E) showed well defined, extrapulmonary and heterogeneous mass of 17 cm lenght. Calcifications (arrows) and enhancing regions (arrowhead) were seen inside the mass. A sessile attachment (blue arrow) is seen in the inferior aspect of the mass on CT image (G). Pathologic specimen (H-I) showed a smooth surface with sessile atleastment (blue arrow). On dissection, the surface appears yellow-than with reddish hypervascularized zones, hemorrhagic foci and necrotic zones. Histologic analysis confirmed the diagnosis of **malignant solitary fibrous tumor of the pleura**.

**Fig. 4: Malignant Solitary Fibrous Tumor of Pleura**

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**Figure 5.** 92 years old man with recurrent and symptomatic hypoglycemias that underwent an abdominal CT with suspicion of an insulnoma. Chest X-ray (A) showed a huge subpulmonary mass mimicking right diaphragmatic elevation (red arrow). Contrast enhanced CT (B-E) depicted a well-defined and heterogeneous mass in close contact with right diaphragm. There are huge cystic/necrotic areas (green arrow), gross calcifications (yellow arrow) and prominent vessels (blue arrows) that arise from right phrenic artery. Ultrasound (F) was performed to guide a percutaneous biopsy and depicted a predominantly hypo-echoic and heterogeneous mass with Doppler flux inside the tumor. Histological analysis confirmed a benign SFTP. Hypoglicemia was probably a paraneoplastic phenomenon (Doege-Potter syndrome). The patient did not underwent surgery due to advanced age.

**Fig. 5: Benign Solitary Fibrous Tumor of Pleura**

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Fig. 6: Local recurrence of a Malignant Fibrous Tumor of Pleura. 81 years-old man that had had a partial resection of a mSFTP two years before (previous CT non-available). Contrast-enhanced CT (A-D) showed multiple, rounded and well defined masses (red arrows) with pleural attachment. Lesions have soft tissue density with no significant contrast enhancement, suggesting necrosis / cystic degeneration. Ipsilateral pleural effusion is also observed (arrowhead), being also a typical feature of mSFTP.

Fig. 6: Malignant Solitary Fibrous Tumor of Pleura

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**Fig. 8:** Solitary Fibrous Tumor of Peritoneum

A 52 years-old woman with non-specific abdominal pain. An abdominal ultrasound was performed showing an heterogeneous pelvis mass with hypoechoic foci and Doppler flow inside (A). Contrast enhanced CT showed a lobulated heterogeneous mass with cystic parts and intense contrast enhancement (blue arrow) (B - E). On serial CT imaging, the mass showed change in location and shape (red arrows on C, D, E), which is typical of pedunculated lesions. An ultrasound guided biopsy (Tru-cut) was performed and histology confirmed a SFT of peritoneum.
Figure 9: Pleural Metastases

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Figure 7. Benign SFT of peritoneum. 46 years old woman with an incidental finding in abdominal US performed because of abdominal discomfort after hysterectomy due to uterine myomatosis. Abdominal US (A and B) showed a round and well circumscribed isoechoic mass (36mm) with presence of Doppler flow. Non contrast (C) and contrast enhanced CT (D) demonstrated a solitary well-delineated, homogeneous and round mass of soft-tissue attenuation (blue arrows) (E) with significant enhancement (F). On MRI it had an intermediate signal intensity on T2FS (G) and T1FS (H) and an intense enhancement after administration of gadolinium (I), showing a discrete anterior pedicle attached to parietal peritoneum (red arrows) (confirmed by surgery and pathology). Gross pathology found a well-circumscribed mass (51 x 45 x 25 mm) covered by a thin and translucent membrane with prominent blood vessels with single pedicle (J). Histology confirmed benign SFT peritoneal.

Fig. 7: Benign Solitary Fibrous Tumor of Peritoneum

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**Fig. 10:** Pleural Mesothelioma

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**Fig. 11:** Ewing’s Sarcoma

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Fig. 12: Chest Wall Lipoma and Loculated Interlobar Effusion

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Fig. 14: Chest Wall Neurogenic Tumor

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**Fig. 13:** Lung Abscess

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Conclusion

SFTP is the second most common primary pleural neoplasm. It is usually an incidental finding with little or no symptoms with indeterminate malignancy risk. Knowledge of the key imaging findings in each imaging modality and awareness of their differential diagnosis is mandatory for a correct diagnosis.
Personal information


References


