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Solitary Fibrous Tumor of the Pleura: Histology, CT Scan Images and Review of Literature over the Last Twenty Years

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Introduction

Solitary fibrous tumor of the pleura is a rare neoplasm. In Literature up to 800 cases [1-3] have been reported, and these numbers show its rarity, despite of mesotheliomas, the most pleural tumors represented. Males and females are equal distributed and the same is true for age. No correlation with exposure to asbestos, tobacco or others environmental agents, were found for its development. Solitary fibrous tumor of the pleura occurs as localized neoplasms of the pleura and was initially classified as "localized mesothelioma". Recently, with the aid of the electronic microscope and immunohistochemistry, has been possible emphasize that their origin is mesenchymal and not mesothelial, so the term "localized mesothelioma" has been replaced with "solitary fibrous pleural tumor" STFP [4]. In the past STFP were described only in the pleura but recently it has been found located also in other sites [5,6] such as abdomen, liver, peritoneum, retroperitoneal spaces, meninges, orbit, thyroid, salivary glands and soft tissues including the breast [7-10]. The STFP can be associated to other synchronous or metacron neoplasms like prostate, lung, breast, endometrial carcinoma and thyroid. Although most of STFP are benign neoplasms, a part of these could have a malignant behavior. The clinical behavior is unpredictable and probably is due to their histological and morphological features. STFP may remain silent for many years before turning into malignant form [11].

Clinical Features

Often SFPTs are accidentally discovered by radiological investigation like X-Ray [12,13]. Most patients with SFPTs become symptomatic when these tumors reach large size [2,10,14]. About 54-67% of patients with benign SFPTs are symptomatic, while 75% of cases of malignant SFPTs, show symptoms [15]. These symptoms are cough, chest pain and dyspnoea. If there is obstruction of the airway, also hemoptysis and pneumonia [2,10] could be seen. Paraneoplastic syndromes, represented by digital hypocratism and Pierre-Marie-Bamberg syndrome, are observed in 10-20% of cases and specially with large size SFPTs. Symptoms usually vanish after the neoplasm has been removed, but may reappear in case of recurrence [10,16,17]. In less than 5% of patients with SFPTs an increase of insulin-like factor II type occur and this causes refractory to therapy hypoglycaemia (Doege-Potter syndrome) [10,18,19]. The incidence of Doege-Potter syndrome in SFPT is similar in both sexes and there no differences in both benign and malignant forms.

Some patients may also present gynecomastia or galactorrhoea [1]. Sometimes large size SFPTs may appear with an unusual clinical presentation, as the two cases reported by Santambrogio et al. [20] and Shaker et al. [21]. The first described a patient with large size SFPTs manifested with syncope episodes of coughing. The second author reported the case of a woman with lower limbs edema and dyspnea caused by a bulky SFPTs compression to the right atrium and the inferior vena cava.

Histological Characteristics

England [10] suggested SFPTs originate from sub-mesothelial connective tissue. Histologically SFPTs occur such as low-grade neoplasms with variable cellularity. The cancer cells present oval or fusiform shape with oval nuclei and chromatin is well distributed. Electronic microscopy has identified at least 3 pathological patterns: "subverted order" is the most common. In this type the cells and the surrounding collagen do not exhibit any architecture. The second most common pattern is "hemangiopericytoma-like", in which dense anastomoses are highlighted among blood vessels. The third one is the least frequent and refers to angiofibroma-like, fibrosarcoma-like patterns and synovialsarcoma-like.

Histologic differential diagnosis is difficult and includes spindle-cell melanoma, sarcomatoid mesothelioma and soft tissue sarcomas. Recently immunohistochemistry has revealed to be extremely useful for differential diagnosis. Perrot et al. [2] summarized the most important immunohistochemical characteristics: in SFPTs vimentin is positive and keratin negative. CD34 is + in most benign SFPTs as well in malignant one, while it reremains negative in most of other tumors of the lung. Lately, a cytogenetic procedure

has also contributed: SFPTs is often associated with trisomy 8 and trisomy 21 and this is helpful for diagnose and differentiate fibrous pleural tumor from mesothelioma and other sarcomas. Genomic hybridization has shown that these chromosomal abnormalities are more frequent in SFPTs larger than 10 cm and this correlation may suggest that gene mutations can promote the growth of neoplasia. Also SFPTs maliciousness prediction is not easy. England et al. [10] defined malignancy criteria (Table 1).

Table 1: Malignancy criteria by England et al. [10].

A	Abundant cellularity with dense and overlapping nuclei
B	High mitotic activity with more than 4 mitotic figures per fields
C	Pleomorphism with cytonuclear atypia
D	Large necrotic or haemorragic areas
E	Pleural effusion
F	Atypical localization

Perrot et al [11] according to the revision of the literature divided SFPT in four stages, correlating histologic features with clinic presentation:

Stage 0: SFPT pedunculated without signs of malignity

Stage 1: SFPT sessile without signs of malignancy

Stage 2: SFPT pedunculated with histologic signs of malignancy

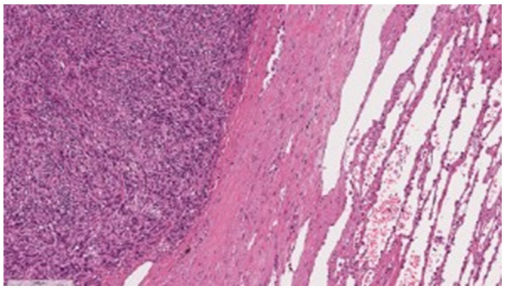
Stage 3: SFPT sessile with signs of malignancy

Stage 4: Multiple and metastatic SFPT

The malignancy criteria used in this classification include the presence of hypercellularity, atypia, cellular pleomorphism, increased number of mitosis, high number of mitosis per field, necrosis and invasion

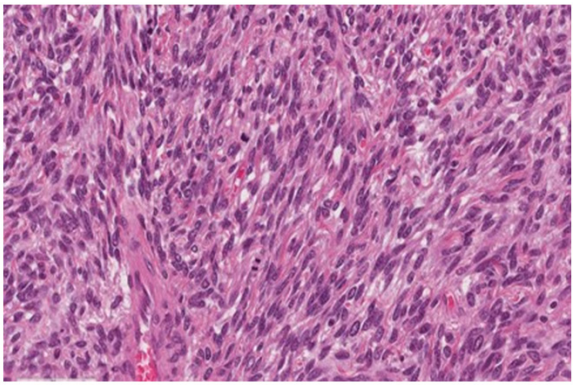
stromal / vascular.

Macroscopic Description Of Histological Slides (Figures 1-3)



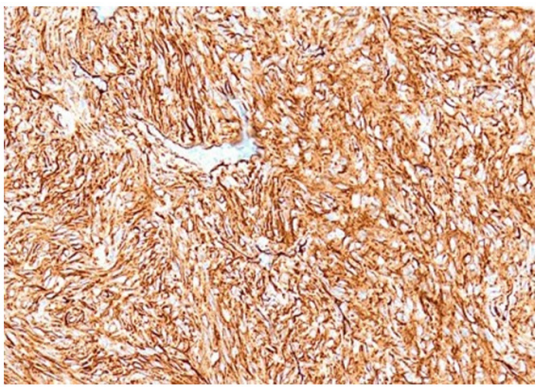
Macroscopic description: 5 gray-whitish rounded suture shrubs: the first cm 12x7.2x7 size, the second cm 13x9x8 size, the third cm 10x9x7 size, the fourth cm 5x5x1.5 size and the fifth one cm 2.5x1.8x1 size. All fragments include irregular surface, with fibrous pseudocapsula, in appearance lobulated with mucoid areas on the surface of the section; multiple nodules are surrounded by unhindered lung parenchyma rhymes. The nodes macroscopically described are constituted by proliferation of fused cells, circumscribed by fibrous pseudocapsules.

Figure 1: Hematoxylin-eosin stain 100x .



Macroscopic Description: these neoplastic cells have bulky, ovoid, monomorphic nuclei; mitotic 14/10 HPFs (two evident mitosis in photo 2); Focal areas of necrosis are present. There are also areas of sclerosis and stromal and perivascular healing.

Figure 2: Hematoxylin eosin stain 400x.



Macroscopic Description: Proliferation cells were found to be intensely positive to immunohistochemical reaction for CD34 (see photo 3, 200x) and for CD99 (+).

Figure 3: Hematoxylin-Eosin stain 200x.

By gentle courtesy of Luisa Delsedime, Pathology, University of Turin

Diagnosis

Chest X-Ray

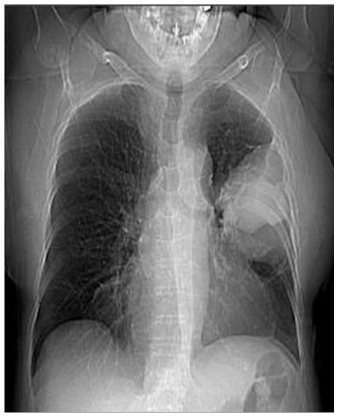


Figure 4: Rx of the Chest.

Chest X-ray shows a well-defined mass that is localized typically in the surface of the lung. SFPT is often located in the middle and lower lobes [22] (Figure 4).

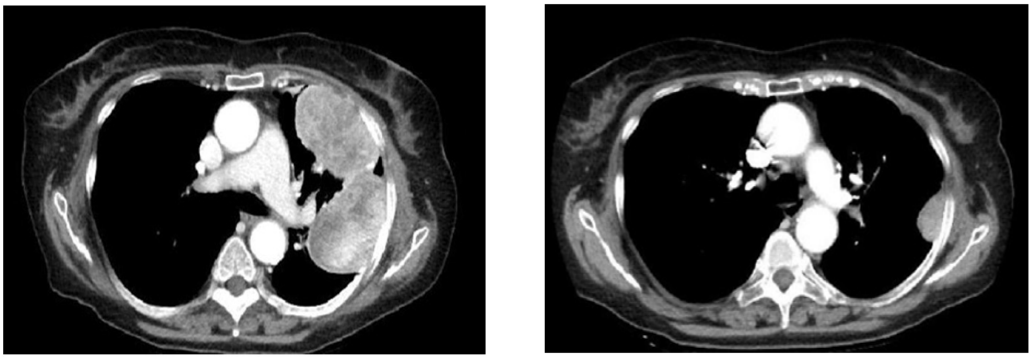


Figure 5: CT-Scan. Pleural nodulations on the left side.

Chest-CT-Scan

SFPT CT scan typically demonstrates homogeneous nodulations, well defined on the pleural surface. In some cases it results difficult to distinguish SFPT from interlobar masses [23]. In some SFPT calcifications can also be observed, regardless of the benignity or malignancy of the lesion [10,11] and it can be difficult to differentiated them from a bronchial carcinoid [24]. With CT Scan is not possible differentiate a benign lesion from malignant one and even the dimensions do not correlate with the behaviour of the neoplasm [25,26] (Figure 5).

Magnetic resonance

Magnetic resonance imaging (RM) has a limited use in pleural diseases. RM investigation is better to TC for delineate morphology and the relationship large-size SFPTs with adjoining mediastinal structures, like great vessels and with the diaphragm [11,27-29].

Angiography

Angiography is an important diagnostic tool. It allows localizing the SFPTs vascular peduncle [30]. The demonstration of arterial supply from the frenic artery, intercostal artery or from the internal mammary vessels, can be very helpful to determinate the extrapulmonary origin of large-size SFPTs.

Chest-ultrasound

The indication to perform an ultrasound chest scan for diagnoses SFPTs or its evaluation isn't shared by many authors, except for a biopsy trans-thoracic for typing [31].

PetWith18Fluoroosoxyglucose

Recently has been documented that FDG-PET can be used to diagnose and for follow up of treated patients. Cardillo et al. [32]. In their study confirmed the high negative predictive value of PET for assessment of the malignancy behaviour of the lesions. In the case of multiple SFPTs and a high metabolic rate highlighted by FDG-PET, a malignant nature must be suspected [33].

Differential Diagnosis

The main differential diagnosis of malignant SFPT includes mesothelioma, neurogenic sarcoma, synovial sarcoma, haemangiopericitoma, fibrosarcoma and malignant fibrous hystyocytoma [34-36]. A great help for diagnosis comes from immunohystochemical analysis.

Therapy

Surgical treatment

The best treatment for SFPT is radical and complete surgical excision of the neoplasm because of the high rate of recurrences [37]. Pedunculated tumors can be resected with a wedge resection, but sometimes even a lobectomy is necessary, partial pleurectomy or even a resection in block with part of chest wall. If the neoplasm is adherent to the parietal pleura, dissection must be used by extrapleuric via [16,38].

Regarding surgery access, it can be used postero-lateral toracotomy, anterior thoracotomy as well as the approach by Video-Assisted Thoracoscopic approach, according to the size of the tumor and its location.

Adjuvant chemotherapy

Currently, adjuvant chemotherapy is used after surgery resection and it's recommended in malignant SFPT, especially in recurrent forms.

Neo-Adjuvant chemotherapy

In SFPT neo-adjuvant chemotherapy is limited due to difficulty of obtaining a pre-operative histologic diagnosis [39].

Prognosis

Prognosis of SFPT is generally good (88%), but approximately 12% of the cases evolves in spread of

intrathoracic or non-resectable recurrence. The relapse occurs up to 17 years after resection and is usually located in the same emithorax [35]. Intrathoracic recurrence can be lethal for mediastinum compression and for the inferior vena cava obstruction [18]. Metastases spread occur by hematogen via and generally they are localized to liver, central nervous system, spleen, peritoneum, adrenal gland, gastrointestinal tract, kidney and bone.

After resection of malignant sessile SFTP the risk of relapse is higher. Most SFPT relapses of malignant sessile occur within two

years from resection and approximately about 50% of the relapses is the cause of death. Therefore is recommended check with Chest X-Ray or CT- Scan every six months during the first 2 years and then once a year. In case of recurrence is indicated surgical treatment [39]. The most important predictors are morphologic and histological indicators [4].

De Perrot [11] proposed a SFPT's classification based on their characteristics and prognosis:

A. A recurrence of 2% in pedunculate benign tumors

B. B recurrence of 8% in sessile benign tumors

C. C recurrence of 14% pedunculated malignant tumors

D. D recurrence of 63% and mortality of 30% within the first 2 years in malignant sessile FSTP

E. E pleural effusion

F. F atypical localization

However more than dimension histological features are important [11] and the most important indicator of clinic outcome is the complete recability of the tumor at the first presentation (Table 2).

Table 2: SFPT clinical manifestations related with frequency of symptoms.

	Benign	Malign
Clinical manifestation		
Symptomatic	+	+++
Casual or Accidental Discovery	++	+/-
Pain	+/-	+++
Dyspnea	+/-	+++
Macroscopic characteristics		
Atypical Locations	Infrequent	Frequent
Size <10 cm	Infrequent	Frequent
Sessile	Infrequent	Frequent
Peduncolate	Frequent	Infrequent
Necrosis	Rare	Frequent
Bleedind	Rare	Frequent
Calcification	Frequent	Rare
Microscopic characteristics		
Ipercellularity	Rare	Frequent
Cellular Pleomorphysm	Rare	Frequent
High mitotic index	-	Frequent
Necrosis	Rare	Frequent
Invasion of the surrounding tissue	-	Frequent

Conclusion

SFPT is a rare disease with uncertain behaviour, curable in most cases. From a biological point of view, we need more knowledge to optimize the clinical outcome. Immunohistochemistry is very useful. Indeed, in addition to the reaction with CD34, the recent introduction of protein STAT36 seems to allow a better and more accurate diagnosis. The best treatment remains the surgical resection, especially earlier and radical at the presentation. Because of the uncertain behavior after surgery, a long follow-up is mandatory and in case of recurrences it is necessary to consider the option of a re-surgical approach.

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