CT signs, patterns and differential diagnosis of solitary fibrous tumors of the pleura

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ABSTRACT First described by Klemperer and Rabin in 1931, solitary fibrous tumour of the pleura (SFTP) is a mesenchymal tumour that tends to involve the pleura, although it has also been described in other thoracic areas (mediastinum, pericardium and pulmonary parenchyma) and in extrathoracic sites (meninges, epiglottis, salivary glands, thyroid, kidneys and breast). SFTP usually presents as a peripheral mass abutting the pleural surface, to which it is attached by a broad base or, more frequently, by a pedicle that allows it to be mobile within the pleural cavity. A precise preoperative diagnosis can be arrived at with a cutting-needle biopsy, although most cases are diagnosed with postoperative histology and immunohistochemical analysis of the dissected sample. SFTP, owing to its large size or unusual locations (paraspinal, paramediastinal, intrafissural), can pose interpretation problems or, indeed, point towards a diagnosis of diseases of a totally different nature.

We present computed tomography (CT) features of SFTP in patients who had had surgical resection in order to discover any specific CT findings that might help in the diagnosis of these tumors.

KeyWords: CT; pleura; SFTP; fibrous tumour

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Clinical and pathologic signs

Solitary fibrous tumor of the pleura (SFTP), first described as a distinct clinical entity by Klemperer and Rabin in 1931 (1), is a mesenchymal neoplasm which usually involves the pleura, but it can occur in other thoracic areas (mediastinum, pericardium, and lung) as well as in extra-thoracic areas (meninx, epiglottis, salivary glands, thyroid, kidneys and breast) (2,3). SFTP occurs with equal frequency in both sexes and is more commonly found in the fourth, fifth, and sixth decades of life (4).

Most of the patients are asymptomatic at the time of diagnosis, and SFTP is discovered only on routine roentgenograms of the chest. In the remaining patients, the most common clinical symptoms are chest pain, cough and dyspnea (5,6). SFTP may occur in benign and malignant forms, these latter showing locally invasive properties or relapsing after surgical resection.

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 confirmatory diagnosis (5,7). 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. In the second most common pattern, tumor cells lie in close contiguity with irregular branching small vessels that result in a hemangiopericytoma-like appearance. Hyper-cellular areas may alternate with hypo-cellular fibrous areas, hemorrhagic, mixoid or necrotic areas (8). Tumor cells are immunoreactive for CD34 and CD99, also are variably positive for Bcl-2; usually cytokeratins and desmin are negative (9).

The purpose of this paper is to discuss computed tomography (CT) appearance of Solitary fibrous tumor of the pleura. We also discuss differential diagnostic considerations and pitfalls in diagnostic CT imaging.

Computed tomography (CT)

The chest computed tomography (CT) scan is the key examination, which more clearly shows the size and location of the tumor and aids in surgical planning.

Both the benign and malignant varieties of SFTP usually appear as well-delineated, often lobulated masses.

CT findings are strictly dependent on tumor size.

In case of small SFTP, CT more frequently typically demonstrates a homogeneous well-defined, non-invasive, lobular, soft-tis

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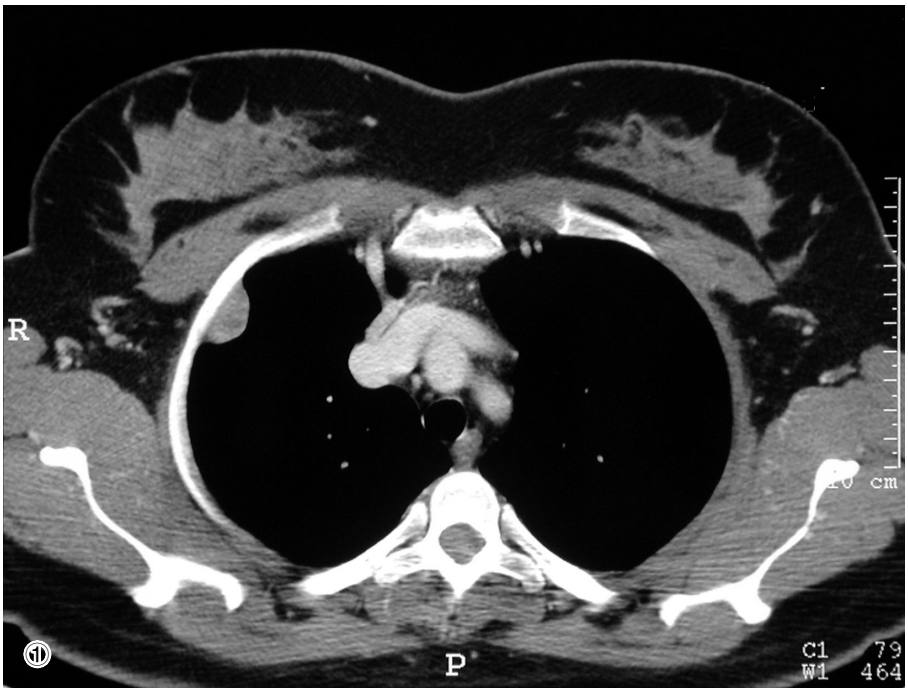

Fig.1 Benign SFTP in an asymptomatic woman. Enhanced chest CT scan demonstrates a small well-defined mass of the right superior hemithorax. The small mass forms an obtuse angle with the adjacent pleural surface.

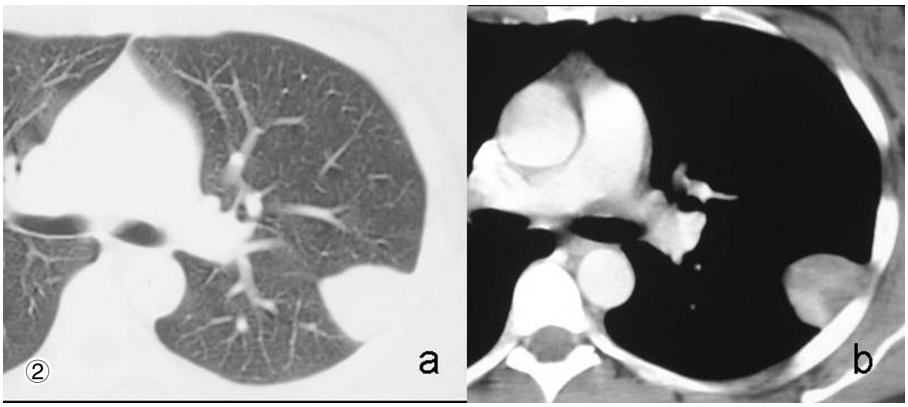

Fig.2 A 35-year-old man with incidental finding of intrafissural lung opacity on chest X-ray confirmed on axial computed tomography (CT) parenchymal (a) and mediastinal (b) windows. The diagnosis of solitary fibrous tumour of the pleura (SFTP) was confirmed at surgery.

sue mass, adjacent to the chest wall or within a fissure, showing an obtuse angle with the pleural surface (Fig 1,2) (10).

Larger lesions are typically heterogeneous and may not exhibit CT features suggestive of pleural tumors (Fig 3). Such lesions usually form acute angles with the adjacent pleural surface mimicking a subpleural pulmonary mass that could be misdiagnosed as peripheral lung cancer (5).

Dedrick et al. stated that a "smoothly tapering angle" of the tumor with the adjacent pleura (seen in 5 of their 6 cases) was a highly characteristic finding that could help in establishing the pleural location of the masses (Fig 4) (11).

SFTP 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 (5).

Intralesional calcifications (punctate, linear or coarse) are constantly associated with areas of necrosis and more easily seen in larger lesions (5,10).

In case of large masses, enhancement after contrast medium is typically intense and heterogeneous with central areas of low attenuation (Fig 5,6). Such intralesional geographic pattern has been shown to correlate with myxoid changes and areas of hemorrhage, necrosis, or cystic degeneration (3,10,12).

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 seeding. The lesions may grow to be very large, almost filling a hemithorax (Fig 7).

The preoperative differential diagnosis that arises in a patient with a SFTP is essentially that of any mass lesion in the chest, ranging from carcinoma of the lung to various intrapleural sarcomas (14). 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 difficoult when SFTP

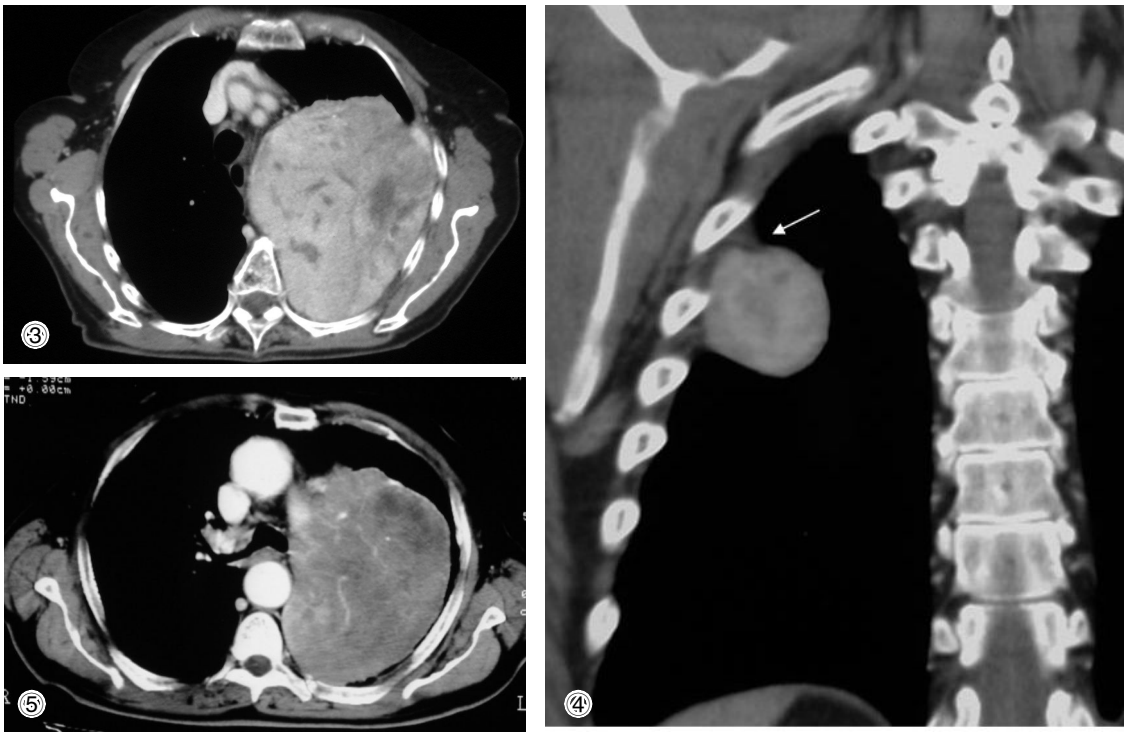

Fig.3 Malignant SFTP in a 65–year–old woman with dyspnea. Contrast–enhanced chest CT scan (mediastinal window) demonstrates a heterogeneously enhancing soft–tissue mass of the left superior hemithorax with internal focal and linear areas of low attenuation. Because of its large size, this lesion may raise questions of diagnostic interpretation, or even lead to a misdiagnosis.

Fig.4 Coronal CT images of a benign SFTP in an asymptomatic 63–year old man. Chest enhanced CT scan targeted to the visualization of the lesion (mediastinal window) demonstrates a heterogeneous spherical soft tissue mass abutting the parietal pleura. Although the lesion forms an acute angle with the pleura layers, a smooth tapering margin (arrow) is also seen.

Fig.5 An 81–year old man with cough, dyspnea and acute hypoglycemia. Contrast–enhanced chest CT scans shows heterogeneous enhancement pattern. Note the serpiginous branching linear areas of enhancement consistent with intralesional vessels and the geographical patterns of low attenuation within the lesion.

develops in particular sites, thus increasing the number of possible diagnoses.

First of all, when located in the paraspinal area, SFTP may appear indistinguishable from neurogenic tumours (Fig 8). In these cases, it is important to evaluate the ribs: chest wall involvement by SFTP is rare (5) and usually manifests as sclerosis or cortical erosion at the costal level, a feature more typical of tumours of neurogenic origin (5).

SFTP that have a mediastinic pleural origin can mimic a mediastinal neoplasm; differential diagnosis from a true mediastinal tumor is sometimes impossible (Fig 9,10).

Furthermore, multiplanar and volumetric reformatted CT images are crucial in the differential diagnosis of SFTP originating from the mediastinal pleura, which may mimic a thymic or a germ cell tumor.

In such cases, analysis of the mediastinum structures is also fundamental. In fact, 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) (12,13,14,15). Moreover three-dimensional CT angiography could be helpful in the accurate evaluation of the blood supply and in detecting the origin of SFTP.

Tumours located within the fissural space may also be interpreted as pulmonary masses when they appear totally surrounded by pulmonary parenchyma. Use of thin-slice multidetector CT with multiplanar reconstructions allows better visualisation of the fissure and its relationship with the tumour. Likewise, CT findings of fissural tails with obtuse tumour- fissure angles and a lentiform shape of tumor could correctly indicate a fissure originated SFTP (16).

Conclusions

SFTP are discovered incidentally on chest radiographs of asymptomatic patients.

The pre-operative CT differential diagnosis of any mass lesion of the chest ranges from the carcinoma of the lung to various intrapleural sarcomas and pleural mesothelioma, but SFTP should al-

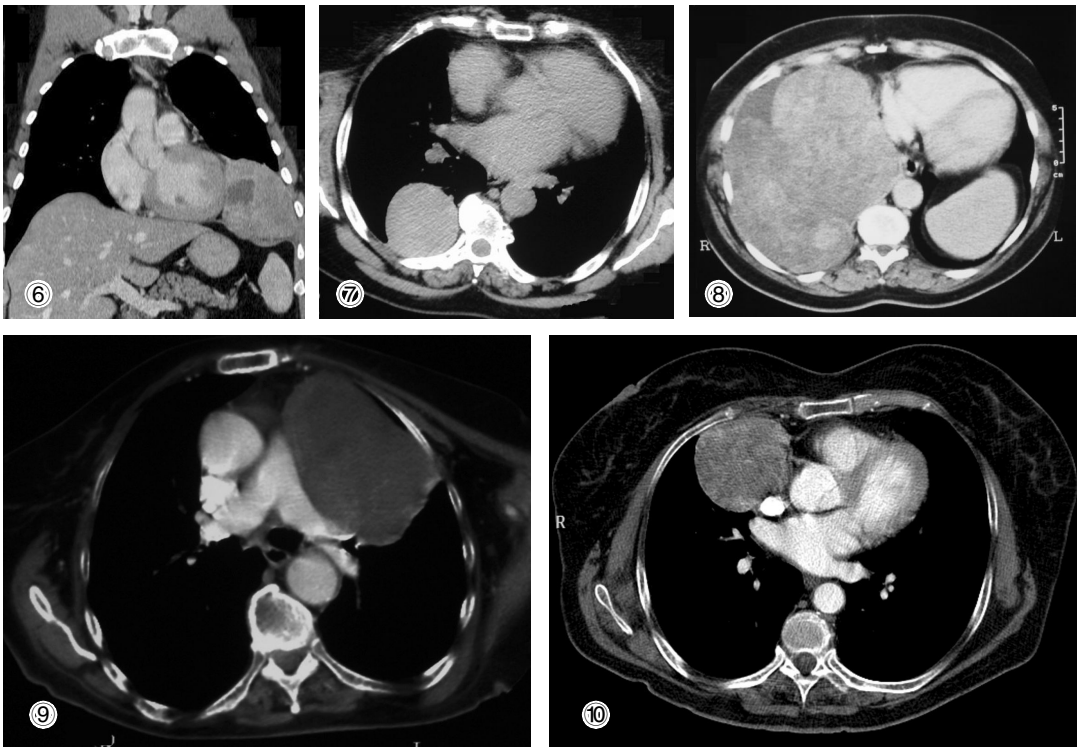

Fig.6 Coronal multi-detector CT after administration of intravenous contrast medium demonstrates a large heterogeneous soft-tissue mass with lobular borders. There is a marked enhancement, with visualization of a large oval area of focal low attenuation due to necrosis or cystic degeneration.

Fig.7 Unenhanced chest CT scan showing a right well-delineated spherical mass mimicking a peripheral neurogenic tumor.

Fig.8 Benign SFTP in a 64-year old woman with chest pain and dyspnea. Contrast-enhanced chest CT scan demonstrates a large inhomogeneous mass filling the entire right hemithorax with mediasinal shift to the left.

Fig.9 A benign localized fibrous tumor originating from the mediastinal pleura. CT shows the typical pattern of a mediastinal mass. At surgery, the mass was found to arise from the left mediastinal pleura.

Fig.10 Benign SFTP: Contrast-enhanced chest CT scan showed a huge heterogeneous soft-tissue mass abutting to the right atrium and ascending aorta.

so be considered.

The usual well-circumscribed appearance of the SFTP mass generally rules out malignant pleural mesothelioma since the latter invariably consists of multiple scattered pleural masses or a more diffuse mass encasing the lung.

A posterior paraspinal location might suggest a neurogenic tumor, while a more anterior and para-mediastinal location might raise the possibility of a thymic neoplasm, germ cell tumor, or teratoma.

When SFTP reaches a large size the diagnosis should be considered on respect to the absence of local invasion, lymphadenopathy, or metastatic spread in patients usually presenting in good health.

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