Solitary fibrous tumors of the pleura mimicking mediastinal tumours: a case series

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Learning objectives

The aim of this work is to present 5 cases of Solitary Fibrous Tumor of the Pleura (SFTP), arising from mediastinal pleura, mimicking an anterior/middle mediastinal tumor in Patients who had undergone surgical exeresis, in order to demonstrate the appearance of these lesions and their findings at chest X-ray and Computed Tomography (CT).
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.

Literature indicates also that sometimes such expansive lesions can occur in other thoracic (mediastinum, pericardium and pulmonary parenchyma) and extra-thoracic sites (such as 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 chest X-ray performed for other reasons. 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.

Sometimes a precise pre-operative diagnosis can be obtained by biopsy (by histologic sampling) performed by cutting needle-biopsy, but in most cases diagnosis is provided by post-operative histology and immunohistochemistry on the surgical specimen [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 and 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 [8,9].
Imaging findings OR Procedure details

Imaging findings

The most common findings described in Literature are divided by imaging technique.

**Chest X-ray**

Chest radiographs of Patients with small SFTP typically demonstrate a well defined solitary mass, which may be visualized in the lung periphery and typically abuts a pleural surface or can be located within a fissure [5]. The shape is often elongated or roughly lenticular, with the largest dimension in the longitudinal plane. When large, they cause opacity of the corresponding hemithorax, which may be subtotal or massive.

**Computed Tomography**

CT findings are strictly dependent on tumor size. The smaller SFTP appear as homogeneous well-defined masses, only occasionally lobulated, in contact with the pleural surface and forming obtuse angles with it. When larger, they appear heterogeneous and connected by acute angles [10].

Very large lesions are typically heterogeneous and may not exhibit CT features suggestive of focal 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 [5,11,12].

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 to 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 with calcification, enhancement after contrast medium is typically intense and heterogeneous with central areas of low attenuation.

**Magnetic Resonance**

In Magnetic Resonance imaging SFTP show variable signal intensity: it has been described as iso-ipointense to muscle on T1 and ipointense on T2-weighted images [5,13,14], but Ferretti et al. reported 4 cases of SFTP with heterogeneous
signal intensity on T2-weighted images [13]. Signal intensity increased with intravenous gadolinium. It has been suggested that this variable signal intensity mainly depends on the relative amount of collagen and fibroblasts, and presence of areas of hemorrhage, necrosis or cystic degeneration in the tumor. Intense enhancement of a SFTP is generally due to high vascularity [4].

**Ultrasonography**

In ultrasonography SFTP appear as well-circumscribed homogeneous low echogenicity or heterogeneous tissue masses; sometimes they exhibit hypoechoic and hyperechoic areas in the absence of cystic degeneration and of intralesional calcifications [15].

Case reports

**CASE 1**

A 74-year-old woman without any evidence of known asbestos exposure, presented with a history of haemoptysis. In her past medical history there was ischemic heart disease and a 26 pack-year history of cigarette smoking.

Chest radiographs showed a well-defined, homogeneous density projecting before and upwards the left hilum (Figures 1A and 1B).
CT scans demonstrated a 5.5 x 4.2 x 4 cm left upper lobe mass with well-delineated lobular contour that abutted the mediastinal surface; no signs of invasion of surrounding lung parenchyma and mediastinal structures, lymph nodes, calcifications or pleural effusion were apparent (Figure 1C). After the administration of intravenous contrast material, scans showed significant homogeneous enhancement of the mass (Figure 1D).

This mass was visible on a previous CT scan of the chest performed 5 years before, in which it was a 2.5 cm lesion.

Physical examination, routine hematochemical analysis, spirometry and arterial gas evaluation were normal. Bronchoscopy disclosed no abnormality.

No fine-needle aspiration (FNA) was performed. A presumptive diagnosis of mediastinal slow growing tumor (thymoma) was made.

At surgery, a segmental resection of the left upper lobe revealed a 48 g pedunculated mass.

Pathologic examination demonstrated a SFTP.
The Patient had an uncomplicated postoperative recovery, and remains well 3 years after surgery.

**CASE 2**

A 56-year-old woman, asymptomatic, underwent an occasional chest X-ray (Figures 2A and 2B) that demonstrated a well-defined round-in-shape lesion located in the anterior mediastinal space (55 mm).

Fig.: 2A-B

**References:** Dipartimento di Scienze Cliniche e Biologiche, University of Torino, San Luigi Gonzaga - Orbassano (TO)/IT

Enhanced CT scans showed a well-defined mass, arising from the anterior mediastinum, with no signs of infiltration of adjacent structures (mediastinal vessels or lung parenchyma), characterized by homogeneous enhancement (Figures 2C and 2D).
Physical examination, routine hematochemical analysis and spirometry were normal.

Patient underwent transthoracic fine needle biopsy, but a definite diagnosis couldn’t be established preoperatively.

Wedge resection was performed: the lesion originated from the visceral pleura of the left upper lobe.

The Patient had an uncomplicated postoperative recovery, and remains well 5 years after surgery.

The histopathological diagnosis was that of a SFTP characterized by extended areas of sclerohyalinosis.

**CASE 3**

A 60-years-old woman, affected by multiple sclerosis, underwent an occasional chest X-ray that showed a well-defined, rounded mass occupying the left pulmonary apex without a cleavage plane with the mediastinal profile (Figures 3A and 3B).
Fig.: 3A-B

References: Dipartimento di Scienze Cliniche e Biologiche, University of Torino, San Luigi Gonzaga - Orbassano (TO)/IT

CT scans demonstrated a large lesion (110 x 80 mm) arising from mediastinal surface, that form a smooth tapering margin with the mediastinal pleura and that was characterized by homogeneous enhancement (Figure 3C).
Left upper lobectomy was performed: the lesion was growing up from the mediastinal pleura close to the origin of the subclavian artery.

The final histopathological diagnosis was that of SFTP.

Postoperative course was uneventful: after 2 years follow-up, the Patient is alive and well with no signs of recurrent disease.

**CASE 4**

A 82-years-old man with cough and dyspnea discovered, with a chest radiograph, a left upper hylum nodule (Figure 4A); during a 9-year follow-up this lesion increased in size (Figure 4B).

Pulmonary function tests pointed out an obstructive disease; fibreoptic bronchoscopy showed external compression of bronchial structures.

CT scans showed a large mass, with well-defined lobulated margins, without a cleavage plane from mediastinum (Figure 4C). Contrast-enhanced scans showed heterogeneous density with serpiginous branching linear areas of enhancement consistent with
intralesional vessels and a geographical patterns of low attenuation within the lesion because of round areas of necrosis (Figure 4D).

Fig.: 4C-D

References: Dipartimento di Scienze Cliniche e Biologiche, University of Torino, San Luigi Gonzaga - Orbassano (TO)/IT

A pre-operative diagnosis of SFTP was obtained by a transthoracic fine-needle biopsy.

At thoracotomy the mass (150 mm large) was occupying the whole left upper lobe and a portion of the lower and was characterized by nodules deepened into the mediastinum, however without signs of infiltration. Pneumonectomy was performed.

The Patient had an uncomplicated postoperative recovery, and remains well 3 years after surgery.

CASE 5

A 78-years-old woman, with no asbestos or other environmental pollutants exposure, underwent a chest X-ray because of fever, that showed a large mass originating from the anterior mediastinum and occupying partially the left lung (Figures 5A and 5B).
Enhanced CT scans demonstrated a well-defined oval lesion (100 x 90 mm), heterogeneous, without signs of infiltration of mediastinal vessels; an evident mass effect with mediastinal shift was pointed out (Figures 5C and 5D).
Left lateral thoracotomy with wedge resection was performed: the large mass was pedunculated, growing up from the lingular visceral pleura.

The final histopathological diagnosis was that of a SFTP.

Postoperative course was uneventful: after 3-years follow-up, the Patient is alive and well with no signs of recurrent disease.
Conclusion

The differential diagnosis of primary pleural tumours is relatively narrow and includes, in addition to SFTP, the rarer liposarcomas, synovial sarcomas and epithelioid haemangioendothelioma.

However, the differential diagnosis becomes particularly challenging when SFTP develops in unusual sites, thus increasing the number of possible diagnoses: SFTP that have a mediastinal pleural origin can mimic mediastinal neoplasms like thymic or dysembryogenetic tumours and the differentiation is often impossible.

In fact, on chest radiographs and CT scans, SFTP that have a mediastinal pleural origin and mediastinal neoplasms appear as lesions with their maximum diameter abutting the mediastinal pleural surface, with which they form obtuse, right or acute angles with a smooth tapering margin. These kinds of angles only allow us to confirm that the origin of the lesion is extra-parenchymal (pleural/mediastinal) rather than parenchymal.

A pathognomic finding in pedunculated SFTP is the mobility of the tumour with changes in Patient position, but also thymic neoplasm can present a pedicle. Moreover, the usefulness of this finding is limited by the size of the mass: the larger the SFTP, the more firmly it is attached to adjacent structures by adherences, which makes the mass less mobile.

So, analysis of the mediastinum structures is fundamental. In fact, in lesions of pleural origin, the thymic cavity 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).

With the advent of modern equipment and the use of multiplanar imaging that enables detailed study of the mediastinum, it has become easier to distinguish between masses of thymic origin and lesions of the mediastinal pleura, even though it should be recalled that SFTP may also arise in the mediastinum.

It is important to notice that in very large tumours, the discrepancy between the scarcity of clinical findings and the dramatic radiological picture is in itself indicative of a slow-growing lesion (lung or mediastinal tumours rarely grow so large without causing symptoms). Absence of lymph node involvement and preservation of cleavage planes with adjacent structures, despite the fact that these masses may occupy an entire hemithorax, provide confirmation of the benign nature of the lesion.
Finally, the differential diagnosis between mediastinal and pleural lesions is very important in order to plan the approach of the thoracic surgery. In fact, usually the approach for the exeresis of lesions that originate in the anterior mediastinum is made through sternothomy, while Patients with neoplasms arisen from the medium or the posterior mediastinum underwent thoracotomy.

In order to come to a precise preoperative diagnosis is recommended a CT-guided cutting-needle biopsy; if there is a broad area of contact with the chest wall, a US-guided biopsy can be performed.
References


