

Solitary fibrous tumours: unusual aspects of a rare disease

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Abstract:

Background: In literature there are only a few descriptions of the typical presentation of solitary fibrous tumours (SFT) and only a few case reports showing its unusual clinical and radiological features.

Methods: We retrospectively evaluated the computed tomography scans of 36 patients presenting with a histological diagnosis of SFT between 1998 and 2008.

Results: We present five cases of SFT with an atypical clinical presentation and radiological features.

Conclusions: SFT can occasionally present with unusual radiological features making a differential diagnosis difficult. Even though imaging plays a fundamental role in the initial diagnostic approach, final diagnosis is only confirmed by biopsy and histology. Hippokratia. 2012; 16 (3): 269-274

Key words: solitary fibrous tumours of the pleura (SFTP), synchronous pleural and renal fibrous tumours, atypical presentation of fibrous tumours.

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Solitary Fibrous Tumours of the Pleura (SFTP), have been described by a variety of terms such as benign mesothelioma, localized mesothelioma, and subpleural mesothelioma¹⁻³, reflecting the controversy over these tumors. It is a rare tumour with a prevalence of 2,8 per 100.000 cases (less than 5% of growing pleural masses)².

Usually SFTP arise within the pleural cavity with most cases originating from the visceral pleura⁴⁻⁶. The latter are larger than those originating from the parietal pleura. SFTP originating from the pleura need to be distinguished from mesotheliomas. The differential diagnosis between these two tumours is important because SFTP is usually a benign tumour with a good prognosis whereas on the other hand, mesothelioma is typically an aggressive tumour with a poor prognosis. The majority of SFTP are benign (80%) however rare cases of malignancy have been reported in literature^{2,4}.

Mesothelioma cells originate from the mesothelial pleural cells. As some authors have previously described SFTP cells originate from the myofibrillary mesenchymal cells located in the submesothelial connective tissue¹, suggested by the histological and immuno-histochemical features⁷ (for example SFTP is CD34 positive⁸ as opposed to mesothelioma which is CD34 negative).

In rare cases this type of tumour does not originate from the pleura but rather from other sites within the thorax⁹⁻¹². In these cases the Solitary Fibrous Tumour (SFT) needs to be distinguished from other pathologies

such as neurogenic tumours, thymic tumours, teratoma, germ cells tumour, lipomas, sarcoma and epithelioid hemangioendothelioma^{1,4,7,11-14}. An extrathoracic presentation of the SFT is extremely rare^{1,7,15-19}.

Usually SFTP is discovered incidentally as a single intra-thoracic mass discovered on chest X-Ray requested for other reasons^{1,4,5}.

The majority of patients affected by this disease are asymptomatic. Symptomatic patients may present with cough, chest pain and dyspnea, while hemoptysis and obstructive pneumonitis are rare presentations¹⁻⁴. Patients with tumour localized on the diaphragmatic pleura can present with vague complaints localized in the right hypochondrium; occasionally SFTP can also present with a paraneoplastic syndrome (digital clubbing, hypertrophic osteoarthropathy and refractory hypoglycemia), even though this is rare^{1,2,5,20}.

The aim of this article is to present a case series of SFT having an atypical clinical presentation and radiological findings.

The aim of this report is to make the radiologist aware of the possible diagnosis of SFT when these have atypical clinical and radiological presentations.

Materials and Methods

We retrospectively evaluated the computed tomography (CT) scans of 36 patients from our pathological archive who presented with a histological diagnosis of SFT between 1998 and 2008.

We selected 5 out of the 36 cases, showing an atypical radiological and clinical presentation, all of whom had a complete radiological staging performed by CT scan (GE Lightspeed 16 slice CT scanner General Electric Healthcare; Milwaukee, WI).

In 4 out of 5 patients a PET/CT scan was performed, and in 1 out of 5 an additional ultrasound examination.

The histological diagnosis was based on the England criteria²¹.

Case 1

A 68 year old woman who presented due to refractory cough underwent chest x-ray and CT scan.

The chest X-ray showed a bulky 6 cm mass in the left lower lobe, originating from the pleural sheets and showing expansive behaviour.

The CT scan confirmed the presence of the mass with an inhomogeneous density after contrast medium administration (140 H.U) without infiltration of the surrounding tissues and no lymphadenopathy (figure 1). As a collateral finding an expansive mass of 2.5 cm was identified between the spleen and the left kidney (figure 2). This was further studied with an abdominal CT scan which showed a mild hyperdensity (55 H.U) on the basal CT scan, with progres-

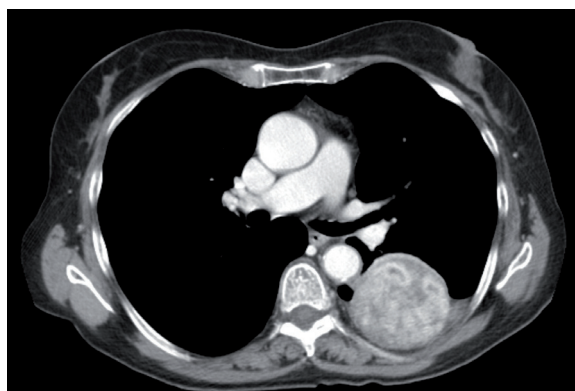


Figure 1: Case 1 - Axial CT scan of a 68 year-old woman showing a bulky mass in the left lower lobe with an inhomogeneous density after contrast medium administration (140 H.U)

sive contrast enhancement after contrast medium, reaching a value of 158 H.U. in the portal venous phase and complete contrast medium wash out at equilibrium (figure 2).

An ¹⁸FDG PET/CT scan showed uptake at the level of the thoracic mass (SUV max of 6.04), however no uptake was seen in the abdominal mass.

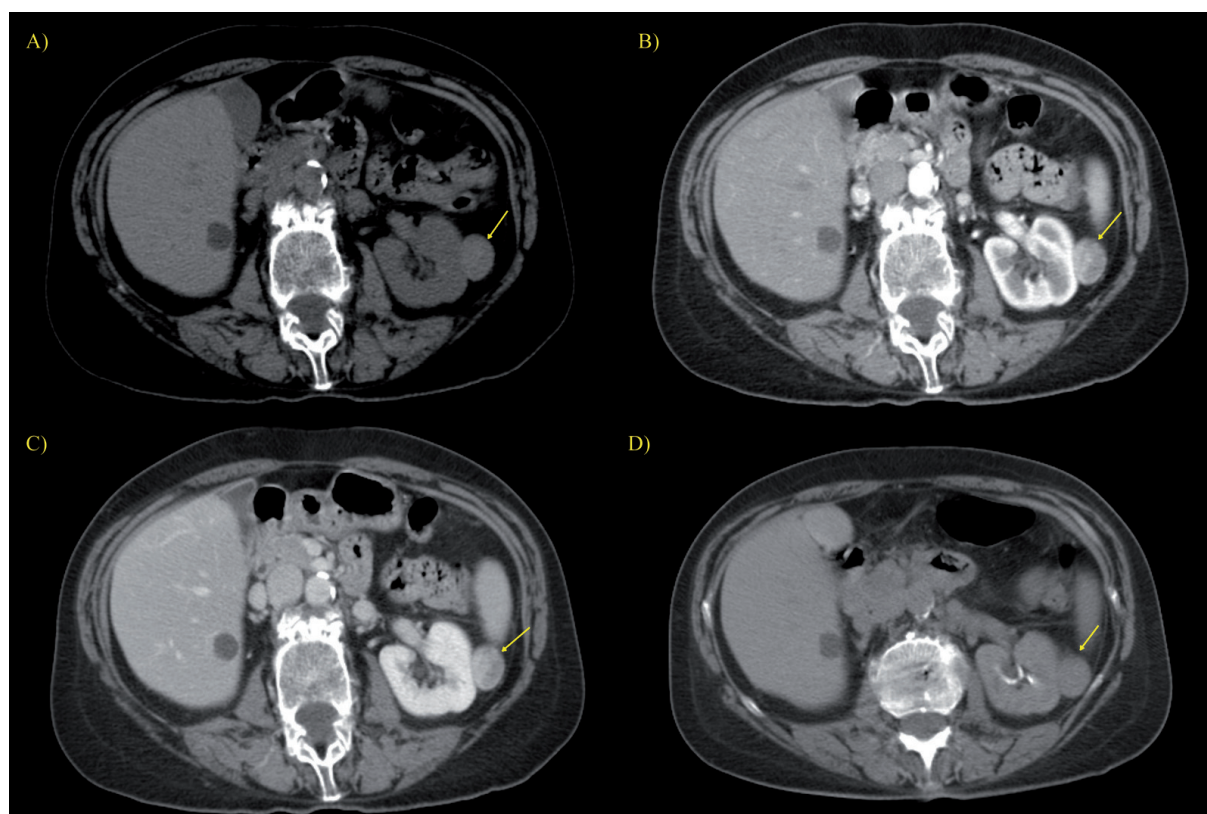


Figure 2: Case 1 - Axial CT scans of the renal mass showing the pre and post contrast behaviour of the renal lesion (SFT) mimicking renal cell carcinoma:

- A) pre-contrast phase: mild hyperdensity (55 H.U).
- B) arterial phase: rapid progressive uptake.
- C) portal phase: reaching maximal uptake (158 H.U).
- D) equilibrium phase with a complete contrast medium wash-out.

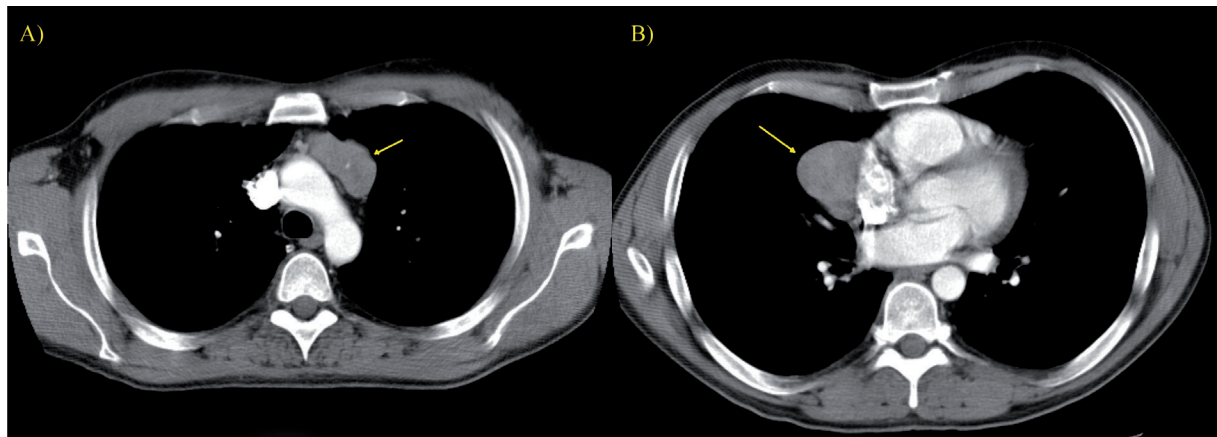


Figure 3: Case 2 - Axial CT scan of a 35 year old man showing two solid well defined masses in the mediastinum:
a) inhomogeneous contrast enhancement with hypodense areas with miliary calcified spots (Thymoma).
b) inhomogeneous contrast enhancement mass with hypodense areas without calcifications (SFT).



Figure 4: Case 3 - Axial CT scan of a 67 year old woman showing two masses both with oval shape and well defined margins with an expansive pattern of growth. The difference in contrast enhancement behaviour is due to the lipid content of the left mass (neuroma) which is richer in lipid content than SFTP.

In order to characterize the lesions we performed biopsies of the two masses.

For the lung mass we performed a CT-guided biopsy using a semi-automatic Gallini needle (18G) whereas for the left hypocondrial mass we performed an ultrasound guided biopsy using an automatic needle (18G). Both masses were confirmed to be a benign SFT at histology.

Following a multidisciplinary team meeting the decision was to surgically remove the thoracic lesion and perform regular follow-up of the abdominal mass.

After 4 years of follow-up the abdominal mass remains identical in both size and density at CT.

Case 2

A 35 year old male patient presented to our attention

with a six week history of asthenia and diffuse itching not responsive to pharmacological therapy.

A chest X-Ray showed bulging of the mediastinal borders, confirmed by thoracic CT. CT also showed two solid well defined masses in the mediastinum: one in the right paracardiac area and another left sided mass just anterior to the aortic arch.

Both mediastinal masses showed an inhomogeneous contrast enhancement with hypodense areas. The left sided mass showed miliary calcified spots (figure 3).

The patient underwent surgery to remove both masses with a final histopathologic diagnosis of :

- thymoma type B2 according to the WHO classification for the left sided mass,
- benign SFT with an intrathymic origin for the right sided mass.

The patient was diagnosed with myasthenia gravis a few months after surgery. The patient performed an annual follow-up CT and remains disease-free during a follow-up period of 4 years.

Case 3

A 67 year old woman had a surgical excision of the 5th and 6th left rib at another institution with a histological diagnosis of a lipoma-like sarcoma.

During the pre-surgical radiological assessment the CT scan demonstrated two other masses in the thorax.

One was identified between the IIIrd and IVth intercostal space, of probable pleural origin showing oval morphology, and well defined margins with an expansive pattern of growth. The other mass showing similar morphology was located in the posterior mediastinum behind the trachea. Both masses showed homogeneous contrast enhancement, however this was greater for the right sided lesion (figure 4).

CT-guided biopsies of the lesions with a semi-automatic Gallini needle (18G) were non-diagnostic of disease. A PET/CT scan did not show significant areas of pathological uptake. Two further surgical biopsies per-

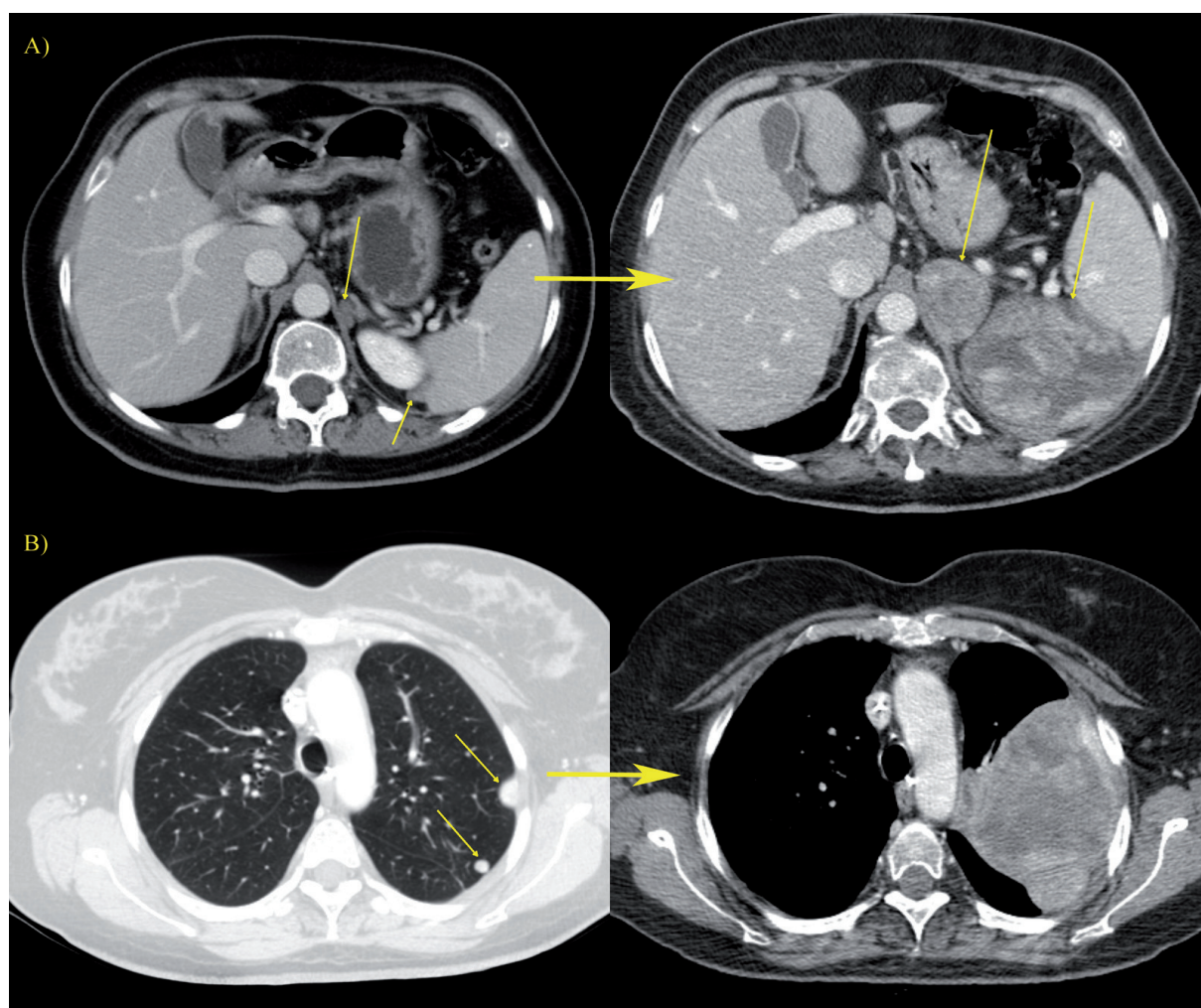


Figure 5: Case 4 - Axial CT scan of a 47 year old woman showing the progressive growth pattern typical of malignant lesions: two retroperitoneal masses with markedly inhomogeneous contrast medium uptake (A) as shown in the two pleural masses (B).

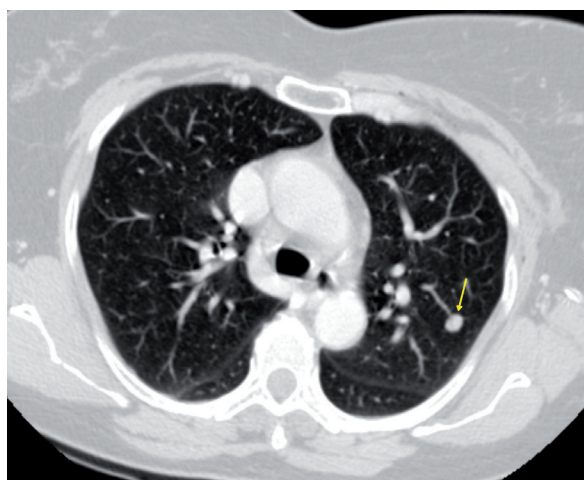


Figure 6: Case 5 - Axial CT scan of a 60 year old woman showing an entirely intrapulmonary solitary fibrous tumour with round shape and well defined margins. These could be typical features of a pulmonary metastasis, and therefore could potentially be a pitfall.

formed at different sittings revealed a benign SFTP for the right sided lesion and a Schwannoma for the left sided lesion.

The next two surgical approaches to the lesions, performed in different sessions gave two different diagnosis: benign SFTP for the right sided mass and a Schwannoma for the left sided mass.

Case 4

A 47 year old woman presented with a history of malignant Fibrous Tumours of the Pleura (FTP) of the left diaphragm; she had previously undergone several surgical procedures to remove several intrathoracic masses. The patient came to our attention after years later with multiple bilateral lung recurrences.

Baseline staging of the disease was performed using CT and PET scan.

The first CT scan performed at our Institute demonstrated distinct signs of tumour liquification with an inhomogeneous contrast enhancement, with hypodense areas correlating with areas of lower ^{18}F FDG uptake within the

lesions.

Follow-up examinations with CT and PET/CT showed disease relapse with several new lung metastasis. These were then treated with multiple surgical procedures as well as thermoablation and radiotherapy in view of the reduced Total Lung Capacity following several pulmonary procedures. A follow-up CT scan of the thorax, showed disease progression with pleuro-pulmonary metastases. A PET/CT detected homogeneously increased ¹⁸FDG uptake in the major tumour areas in the thorax and abdominal cavity which were consequently treated with chemotherapy. Several months later, a new relapse was diagnosed in both abdomen and thorax (figure 5).

Case 5

A 60 year old woman, performing a routine follow-up chest X-ray for a previous ductal breast carcinoma, was seen to have a radiopaque rounded lesion in the left pulmonary apex. This was further studied performing a thoracic CT scan and PET/CT.

Both techniques confirmed the suspicious secondary nature of the lesion:

At CT scan examination the nodule had a homogeneous contrast enhancement, well defined margins situated in the central parenchyma with no connections to the pleura or mediastinum (figure 6).

PET/CT showed an intermediate uptake of ¹⁸FDG.

Considering the oncological history of the patient, and radiological characteristics of the lesion could be attributed to metastases, a wedge surgical excision of the single lesion was performed. The histological diagnosis was an intraparenchymal SFT with a benign pattern. Considering the benign nature of the lesion an annual thoracic CT scan was scheduled with no evidence of disease relapse after 4 years.

Discussion

In literature there are only a few studies on SFT. Our aim was to present radiological features of some selected cases of this rare disease.

All the radiological features, particularly those seen on CT, showed the typical morphological aspects of an expansive mass (round shape and well defined margins), with an increased contrast enhancement, and an increasingly inhomogeneous enhancement directly proportional to the size of lesions (due to central necrosis, haemorrhage and cystic changes with a patchy pattern)^{2,15,20,23}. There were no obvious pathognomonic signs of SFT^{1,24}.

The gold standard for final diagnosis of SFT remains the surgical approach. In literature there are many papers describing SFT as a prevalently benign lesion^{1,2,4,5} but few papers report a malignant type of SFTP with local and distant metastasis¹⁵.

In cases 1 and 4 we showed how SFT presented with distant metastasis; a typical sign of malignancy. For example in case 1 we present a patient with a single pulmonary and a synchronous renal lesion thought to be two primary lesions due to the radiological features. This could potentially lead to a difficult diagnosis as the re-

nal mass may mimic a renal cell carcinoma^{25,26}. There are rare cases of primary renal SFT.

Case 4 was a malignant FTP with multiple relapsing metastasis in the abdomen diagnosed at surgery. These lesions were not responsive to therapy (surgery, radiofrequency and chemotherapy). SFTP, as reported in literature^{1,2,4,5}, originates from the parietal pleura when the lesions are larger in size. On the other hand the smaller SFTP originate from the visceral pleura.

Case 2 describes a patient with myasthenia gravis with 2 separate masses originating from the mediastinal pleura consisting of thymic tissue. The presence of myasthenia gravis relates to the presence of the thymoma; additionally one of these two masses also presented an intrinsic SFT. This example showed that SFT can potentially present within the mediastinal pleura¹². There are no standard features to differentiate an SFT from a thymoma²⁷; in this case we can't consider the presence of intrinsic calcifications a radiological sign of an SFTP as described in literature^{1,2,24} (the intrathymic SFT did not show intrinsic calcifications as opposed to the contralateral mass). When considering only the radiological features (such as calcifications) one could be lead into making an erroneous diagnosis.

In case 3 we demonstrate how the differential diagnosis of SFTP can also include neuroma. CT features can be helpful to differentiate these two pathologies. In particular the neuroma is richer in lipid content compared to SFTP, and is more likely to be hypodense. This could also explain the differences in contrast enhancement.

In case 5 we showed a rare case of SFT which did not originate from the pleura but originated in central lung parenchyma^{11,28}. The imaging characteristics (new, single lesion, round shape and well defined margins) and the positive uptake of ¹⁸FDG of the mass made it highly suspicious for a breast cancer metastasis due to the clinical history of the patient (previous surgery for invasive ductal breast carcinoma).

When encountering such cases we suggest considering the possibility of an fibrous tumour, because even though it is rare, the prognosis and treatment changes significantly. According to the limited literature available there is no evidence of malignancy for whole intrapulmonary solitary fibrous tumours, however owing to the limited experience follow-up must be performed^{4,11}.

In conclusion in this case series we presented a number of selected cases with an atypical clinical and radiological presentation. Imaging techniques are still the first fundamental tools to obtain an initial diagnosis based on the morphological and functional features of the fibrous tumour but the final diagnosis still remains the interventional approach.

Conflict of interest

The authors have no conflict of interest.

References:

1. Robinson LA. Solitary fibrous tumor of the pleura. *Cancer Control*. 2006; 13: 264-269.

2. Rosado-de-Christenson ML, Abbott GF, McAdams HP, Franks TJ, Galvin JR. From the archives of the AFIP: Localized fibrous tumor of the pleura. *Radiographics*. 2003; 23: 759-783.
3. Begin fibrous tumor of the pleura. In: text of Pleural Disease. RW Light and G Lee Hobber Stoughton Ltd 2008.
4. Dalton W.T, Zolliker A.S, McCaughey W.T.E. Localized primary tumors of the pleura: an analysis of 40 cases. *Cancer*. 1979; 44: 1465-1475.
5. Briselli M, Mark EI, Dickersin GR. Solitary fibrous tumors of the pleura: eight new cases and review of 360 cases in the literature. *Cancer*. 1981; 47: 2678-2689.
6. Cardillo G, Carbone L, Carleo F, Masala N, Graziano P, Bray A, et al. Solitary fibrous tumors of the pleura: an analysis of 110 patients treated in a single institution. *Ann Thorac Surg*. 2009; 88: 1632-1637.
7. Yuri T, Kanematsu S, Lei YC, Kuwata M, Oishi M, Tsubura A. Solitary fibrous tumor of soft tissue: a case report and immunohistochemical study. *Med Mol Morphol*. 2010; 43: 60-64.
8. Van de Rijn M, Lombard CM, Rouse RV. Expression of CD34 by solitary fibrous tumors of the pleura, mediastinum, and lung. *Am J Surg Pathol*. 1994; 18: 814-820.
9. Yousem SA, Flynn SD. Intrapulmonary localized fibrous tumor. Intraparenchymal so-called localized fibrous mesothelioma. *Am J Clin Pathol*. 1988; 89: 365-369.
10. Aufiero TX, McGary SA, Campbell DB, Phillips PP. Intrapulmonary benign fibrous tumor of the pleura. *J Thorac Cardiovasc Surg*. 1995; 110: 549-551.
11. Sakurai H, Tanaka W, Kaji M, Yamazaki K, Suemasu K. Intrapulmonary localized fibrous tumor of the lung: a very unusual presentation. *Ann Thorac Surg*. 2008; 86: 1360-1362.
12. Suehisa H, Yamashita M, Komori E, Sawada S, Teramoto N. Solitary fibrous tumor of the mediastinum. *Gen Thorac Cardiovasc Surg*. 2010; 58: 205-208.
13. Meade JB, Whitwell F, Bickford BJ, Waddington JK. Primary haemangiopericytoma of lung. *Thorax*. 1974; 29: 1-15.
14. Szydlowski GW, Cohn HE, Steiner RM, Edie RN. Rounded atelectasis: a pulmonary pseudotumor. *Ann Thorac Surg*. 1992; 53: 817-821.
15. Musyoki FN, Nahal A, Powell TI. Solitary fibrous tumor: an update on the spectrum of extrapleural manifestations. *Skeletal Radiol*. 2012; 41: 5-13.
16. Zubor P, Kajo K, Szunyogh N, Danko J. A solitary fibrous tumor in the broad ligament of the uterus. *Pathol Res Pract*. 2007; 203: 555-560.
17. Hong JP, Chung YK, Kim SW, Kim TH, Lee KG. Solitary fibrous tumour of the face: a rare case report. *Br J Plast Surg*. 2002; 55: 75-77.
18. Vargas PA, Alves FA, Lopes MA, Siqueira SA, Menezes LF, Aldred VL, et al. Solitary fibrous tumour of the mouth: report of two cases involving the tongue and cheek *Oral Diseases*. 2002; 8: 111-115.
19. Vallat-Decouvelaere AV, Dry SM, Fletcher SD. Atypical and malignant solitary fibrous tumors in extrathoracic locations: evidence of their comparability to intra-thoracic tumors. *Am J Surg Pathol*. 1998; 22: 1501-1511.
20. Cardinale L, Cortese G, Familiari U, Perna M, Solitro F, Fava C. Fibrous tumour of the pleura (SFTP): a proteiform disease. Clinical, histological and atypical radiological patterns selected among our cases. *Radiol Med*. 2009; 114: 204-215.
21. England DM, Hochholzer L, McCarthy MJ. Localized benign and malignant fibrous tumors of the pleura. A clinicopathologic review of 223 cases. *Am J Surg Pathol*. 1989; 13: 640-658.
22. De Pas T, Toffalorio F, Colombo P, Trifiro G, Pelosi G, Vigna PD, et al. Brief report: activity of imatinib in a patient with platelet-derived-growth-factor receptor positive malignant solitary fibrous tumor of the pleura. *J Thorac Oncol*. 2008; 3: 938-941.
23. Guo J, Chu X, Sun YE, Zhang L, Zhou N. Giant solitary fibrous tumor of the pleura: an analysis of five patients. *World J Surg*. 2010; 34: 2553-2557.
24. Wignall OJ, Moskovic EC, Thway K, Thomas JM. Solitary fibrous tumors of the soft tissues: review of the imaging and clinical features with histopathologic correlation. *AJR*. 2010; 195: W55-62.
25. Magro G, Cavallaro V, Torrisi A, Lopes M, Dell'Albani M, Lanzafame S. Intrarenal solitary fibrous tumor of the kidney report of a case with emphasis on the differential diagnosis in the wide spectrum of monomorphous spindle cell tumors of the kidney. *Pathol Res Pract*. 2002; 198: 37-43.
26. Taxa L, Huanca L, Meza L, Pow Sang M. Solitary fibrous tumor of the kidney (a case report). *Actas Urol Esp*. 2010; 34: 568-70.
27. Eguchi T, Ito N, Makiuchi A, Yoshida K. A solitary fibrous tumor arising from the thymus. *Interact Cardiovasc Thorac Surg*. 2010; 11: 362-363.
28. Pak PS, Yanagawa J, Abtin F, Wallace WD, Holmes EC, Lee JM. Surgical management of endobronchial solitary fibrous tumors. *Ann Thorac Surg*. 2010; 90: 659-661.