Case report

A series of two cases of rare tumors: Solitary fibrous tumor of the pleura

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\textbf{ABSTRACT}

Solitary fibrous tumors of the pleura (SFTPs) are rare neoplasms of mesenchymal origin most commonly arising in the pleura and have a benign biological behavior in the majority of cases. Most patients with SFTPs are asymptomatic; however, symptoms are mostly related to the mass effect within the thoracic cavity. Chest computed tomography (CT) scanning is the radiographic test of choice, but findings lack specificity. Surgical resection is the treatment of choice for most patients. Long-term survival after resection of benign SFTPs is excellent. In this case series, we present two females, one presented with shortness of breath (SOB) and non-productive cough and the other one was referred because of abnormal x-ray findings. The histological as well as the immunohistochemical examination revealed the mass to be a solitary fibrous tumor of the pleura.

1. Introduction

Solitary fibrous tumor of the pleura (SFTP) is a rare, slowly growing tumor that originates from mesenchymal cells of the submesothelial tissue of the pleura [1,2]. SFTPs can usually be distinguished from malignant mesothelioma by their radiographic features, gross appearance (often pedunculated), immunohistologic characteristics (negative for cytokeratin expression and positive for CD34), and ultrastructural characteristics [3].

2. Case number 1

A 57-year-old white female was referred from the outpatient clinic for abnormal chest x-ray, that showed 4.7 cm mass in the left upper lung lobe. The patient denied chest pain, cough, shortness of breath, and fever. A CT scan of chest without contrast showed smooth well-defined mass measures 3.5 × 4.4 × 2.8 cm in size at the anterior portion of the left first rib but there was no associated bone abnormality (Figs. 1–5). A positron emission tomography (PET) scan showed a 1.8 cm nodule at left upper lobe that is non-fluorodeoxyglucose (FDG) avid (Fig. 4). There was a no abnormal FDG uptake within the chest, abdomen, or pelvis.

The patient had a CT guided biopsy of the mass. The histopathology examination revealed benign solitary fibrous tumor (Microscopic examination, low power view showed uniform cellular tumor with intralesimal blood vessels (Fig. 5), higher power showed epithelioid cells and numerous angulated blood vessels (Fig. 6)). Tumor cells were positive immunohistochemical stain CD34 (Fig. 7), negative for cytokeratin AE1/AE3, high molecular weight cytokeratin, S100, desmin, and CD117 (Fig. 8), there was no evidence of increase in mitotic figures and no necrosis was noted. Based on this limited biopsy specimen. The patient underwent Video-assisted thoracoscopic surgery (VATS) with complete resection of left upper lobe mass. The patient followed up with serial CT scans of the chest for 2 years with no evidence of recurrence or new nodule.

3. Case number 2

A 72-year-old white female was admitted to the hospital for progressive SOB and nonproductive cough. Laboratory workup revealed positive respiratory syncytial virus (RSV) rapid antigen. CT scan of the chest with IV contrast was negative for pulmonary embolism (PE) but showed 2 cm low-attenuation mass at the superior aspect of left major fissure (Fig. 9). Patient was treated for RSV tracheobronchitis and was discharged home. A follow-up CT chest 3 months later revealed the same finding. The patient underwent CT guided biopsy of the mass; PET scan showed a 1.9 cm nodule adjacent to the left major fissure that is non-FDG avid (Fig. 10). Tracer distribution was physiologic. No
additional pulmonary nodule was noted.

Histopathology examination was consistent with benign solitary fibrous tumor (Microscopic examination, low power view showed cellular spindle cell neoplasm (Fig. 11), higher power showed monomorphic spindle cells (Fig. 12)). Immunohistochemical stains showed that tumor cells were strongly positive for CD34 (Fig. 13), negative for Cytokeratin AE1/AE3, high molecular weight cytokeratin, S100, desmin, and CD117 (Fig. 14).

Fig. 1. A CT scan of chest without contrast showed smooth well-defined mass measures 3.5 × 4.4 × 2.8 cm in size at the anterior portion of the left first rib.

Fig. 2. A coronal plane of the CT scan of chest showing a smooth well-defined mass at the anterior apical portion of the left lung.

Fig. 3. A sagittal plane of the CT scan of chest showing a smooth well-defined mass at the anterior apical portion of the left lung.

Fig. 4. A positron emission tomography (PET) scan showed a 1.8 cm nodule at left upper lobe that is non-fluorodeoxyglucose (FDG) avid.

Fig. 5. Low power view shows uniform cellular tumor with intralesional blood vessels.

Fig. 6. Higher power shows epithelioid cells and numerous angulated blood vessels.
The patient underwent left sided thoracotomy with wide wedge dissection of the upper lobe mass, and she did well since then. The patient is followed up with serial CT scans for 2 years. There was no recurrence or new nodule has found.

4. Discussion

SFTP is a rare neoplasm, with fewer than 800 cases reported in the literature [4]. The first report of a primary localized pleural tumor is attributed to Wagner in 1870 [5]. These tumors are frequently observed in middle-aged adults, greatest occurrence in the fourth to sixth
decades, with no sex predilection [6]. There is no evidence of the relation-
ship of such tumors with exposure to tobacco, asbestos, or any other
environmental agents [7,8]. SFTs most commonly arises from pleura, with uncommon inward growth into lung parenchyma. Two-
thirds of pleural SFTs occur in the visceral pleura and one-third occurs
in the parietal pleura, where the tumors are often larger, with a broad-
based attachment.

The main differential diagnoses of SFT include pleural mesothelioma, peripheral bronchial carcinoma, solitary pleural"metastasis,
empyema, neurogenic sarcoma, synovial sarcoma, fibrosarcoma, and
malignant fibrous histiocytoma [4,9]. The diagnostic approach to dif-
ferentiate SFT from these conditions include history and physical,
imaging include CXR, CT scan of the chest and PET scan, histopath-
ology and Immunohistochemistry as described below.

Solitary fibrous tumors of the pleura can present with various
clinical signs and symptoms such as dyspnea, chest pain, or hemoptysis.
More rarely, paraneoplastic syndrome, presents as a hypoinsulinemic
hypoglycemia (Doege–Potter syndrome), from the ectopic secretion of
insulin-like growth factor II from a solitary fibrous tumor [10] and
secondary hypertrophic osteoarthropathy (also known as, Bamberger-
Marie syndrome or Osteoarthropathia hypertrophicans) [4]. It has been
described in as many as 20% of the SFT cases, particularly in larger
tumors (e.g. > 7 cm) [11]. These paraneoplastic syndromes symptoms
gradually disappear after tumor resection. Furthermore, SFTs can pre-
sents with non-specific symptoms such as fever, weight loss, and fatigue
[12]. Although, approximately one-third of cases are found incidentally
during chest imaging.

Macroscopically, SFTs are usually large well circumscribed masses,
with a fibrous pseudocapsule or serosal lining [13]. Pleural tumors are
frequently associated with a pedicle; the pedicle typically contains large
feeder vessels for the tumor. The cut surface of SFT ranges from firm
and white for more fibrous tumors to a yellow-brown with homogenous
density for cellular variants [13]. Hemorrhage, necrosis, and calcifica-
tion can be seen [12], particularly in larger tumors.

The majority of SFTs are benign and behave in a slow clinical course
and very rarely to recur locally or distantly. However, malignant forms
of SFT which forms a minority of SFTs are defined as hypercellular,
nuclear pleomorphism mitotically active (> 4 mitoses/10 high-power
fields), tumor size > 10 cm, and finding tumor necrosis, or stromal/
vascular invasion [14,15].

The diagnosis of SFT may be suspected based upon imaging and
clinical features. However, a definitive diagnosis requires histologic
confirmation. Complete resection is required for full histopathologic
evaluation. Whereas fine needle aspiration biopsies are often in-
adequately cellular and are not recommended for diagnosis [16].

On imaging pleural mesotheliomas are nearly always present as
multiple pleural nodules or as a diffuse tumor that encases a portion
of the lung. However, SFT typically appear in contact with the pleural
surface and show displacement or invasion of the surrounding struc-
tures, and appear as homogeneous, well-delineated, and occasionally
lobulated mass of soft tissue attenuation [4]. In some cases where SFT
arises in an interlobar fissure makes it difficult to differentiate it from
an intraparenchymal mass, as the lesion appears to be surrounded by
lung parenchyma [4], as in our second case where the CT scan of the
chest showed 2 cm low-attenuation mass at the superior aspect of left
major fissure (Fig. 9).

There is no definite imaging features to differentiate benign from
malignant SFTP was found [8]. However, it can show the size and lo-
cation of the tumor clearly and help in surgical planning. Large size,
lobulate borders, presence of calcification, and ipsilateral pleural effu-
sion were the only CT features predictive of malignancy.

Neurogenic sarcoma, fibrosarcoma, Synovial sarcoma, and malig-
nant fibrous histiocytoma can be misdiagnosed as SFT because of the
dense, uniform spindle cell proliferation and the similar histologic
patterns seen in these tumors. However, malignant SFT characterized
by the presence of different patterns inside the same tumor and are not
histologically uniform [4].

Histologically, Benign fibrous pleural SFTs are hypocellular in
character. It is characterized by a dense collagenous background, often
with hyalized or thick collagen bands and haphazard (lack of pattern)
arrangement of monomorphic spindle cells with abundantropy col-
lagen mixed with tumor cells (18). Presence of hypercellular, pleo-
morphism, mitotic activity and necrosis may indicate aggressive be-
behavior [17].

Immunohistochemistry (IHC) is an extremely useful tool to differ-
entiate SFT from other tumors (including mesotheliomas and other
sarcomas). Conventional IHC markers of SFT include expression of
CD34, Bcl2, CD99, and vimentin in the absence of actin, desmin, S100
protein, or epithelial markers (epithelial membrane antigen [EMA], low
molecular weight cytokeratins) [18]. In our first case the microscopic
examination showed cellular spindle cell lesion resembling fibroblasts,
immunohistochemical stains was positive for CD34, negative for AE1/
AE3, high molecular weight cytokeratin, S100, desmin, and CD117
consistent with the diagnosis of SFTP.

Management of SFT should be discussed in a multidisciplinary
tumor board with specialists who have experience with the disease.
Surgical resection remains the mainstay of therapy for all localized SFTs
[12]. The choice of surgical approach is affected by the size and loca-
tion of tumors. There is no evidence that suggests that adjuvant

Fig. 13. Tumor cells are positive immunohistochemical stain CD34.

Fig. 14. Tumor cells are negative for immunohistochemical stain cytokeratin AE1/AE3.
chemotherapy is beneficial. By reviewing the literature, it was inconclusive with regards to the utility of postoperative radiotherapy (RT) for SFT treatment. Radiotherapy and chemotherapy have been used as adjuvant treatment when resection is incomplete or impossible. The median survival for patients with all types of SFT has been reported to be 24 years [8]. There is no clear evidence regarding the optimal interval of posttreatment surveillance, and no widely accepted guidelines. Careful long-term postoperative surveillance is recommended because local and distant relapse is possible, even with benign-appearing tumors. Our patients were followed for two years without recurrence.

5. Conclusion

In conclusion, SFT is a rare fibroblastic mesenchymal neoplasm that most likely arises from visceral pleura. Imaging can help in diagnoses, but definite diagnoses needs to be confirmed with histopathology, as well as immunohistochemistry. Surgical excision remains the mainstay of treatment. Long-term follow-up is recommended for early recurrence detection, particularly for tumors that have histological features for malignant behavior.

Data availability

All data are available in the manuscript.

Conflicts of interest

DR. Hazim Bukamur, DR. Emhemmid Karem, DR. Serag Fares, DR. Saroj Sigdel, DR. Emad Alkhankan, DR. Fuad Zeid have no conflict of interests to disclose related to the contents of the manuscript.

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Authors’ contributions

Hazim Bukamur, Emhemmid Karem, Serag Fares, Emad Alkhankan, participated in data collection and interpretation, and drafted the initial manuscript. Fuad Zeid critically revised the manuscript. Saroj Sigdel provided the slides and description of slides and revised the manuscript.

Consent for publication

Not applicable.

No consent needed as this is a case report without any patient identifiers.

Ethics approval and consent to participate

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Appendix A. Supplementary data

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References