

## Intrathoracic Giant Solitary Fibrous Tumor: Case Report

Mindaye ET<sup>1\*</sup>, Tesfaye GK<sup>2</sup> and Aboye AG<sup>3</sup>

<sup>1</sup>Department of surgery, Saint Paul's Hospital Millennium Medical College, Swaziland Street 1271 Addis Ababa, Ethiopia

<sup>2</sup>Department of surgery, Saint Paul's Hospital Millennium Medical College, Addis Ababa, Ethiopia

<sup>3</sup>Department of Radiology, Addis Ababa University, Addis Ababa, Ethiopia

### \*Corresponding author:

Esubalew Taddese Mindaye,  
Department of surgery, Saint Paul's Hospital  
Millennium Medical College,  
Swaziland Street 1271 Addis Ababa,  
Ethiopia, E-mail: esubetad24@yahoo.com

Received: 07 Dec 2020

Accepted: 22 Dec 2020

Published: 26 Dec 2020

### Copyright:

©2020 Mindaye ET et al. This is an open access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and build upon your work non-commercially.

### Citation:

Mindaye ET, Intrathoracic Giant Solitary Fibrous Tumor: Case Report. Clinics of Surgery. 2020; 4(3): 1-4.

### Keywords:

Solitary Fibrous Tumor; Diaphragm, Pleura; Case report

### Abbreviations:

CD: Cluster of Differentiation; CT: Computerized Tomography; MRI: Magnetic Resonance Imaging; SPHMMC – St. Paul's Hospital Millennium Medical College; SFTP –Solitary Fibrous Tumor of the Pleura

## 1. Abstract

**1.1. Introduction:** Solitary Fibrous Tumor of the Pleura(SFTP) is exceedingly rare mesenchymal tumor commonly arising from the visceral pleura and accounts for < 5% of all pleural tumors. Although it commonly has benign histologic characteristics, the tumor behavior is often unpredictable and less understood due to limited number of cases.

**1.2. Case Presentation:** We present a rare case of Intrathoracic Giant SFTP in a 65 years old female who presented with a progressive worsening of shortness of breath of 1year duration associated with intermittent dry cough, low grade fever, easy fatigability and loss of appetite. Complete enbloc resection was done and she was discharged improved.

**1.3. Discussion:** Most patients with SFTP are asymptomatic and definitive diagnosis is often made after surgical exploration and histopathologic study. SFTP should be considered as differential diagnosis in patients with atypical or recurrent respiratory symptoms despite adequate medical treatment. Complete surgical excision is the main stay of treatment and meticulous post-operative follow up is mandatory as the risk of recurrence is higher and the tumor behavior is still less understood.

## 2. Introduction

Solitary Fibrous Tumor of the Pleura (SFTP) is exceedingly rare mesenchymal tumor and accounts for < 5% of all pleural tumors [1, 2]. Majority of SFTP are pedunculated with benign histologic characteristics and about 800 cases have been reported in literatures [3, 4]. It most commonly arises from visceral pleura and, the tumor's behavior is often unpredictable and doesn't always correlate with the histologic findings and mandates long term follow up [3, 5]. Due to rarity of the tumor, complete understanding of its pathogenic factors, clinical features, imaging characteristics, management options and prognostic analysis are very limited [6]. Most patient are asymptomatic especially those with small tumor; however, patients may present with respiratory symptoms [1]. Pre-operative diagnosis is usually challenging and definitive diagnosis is often made after operative exploration [7].

Although Chest Computerized Tomography(CT), Magnetic Resonance Imaging(MRI) and Doppler Ultrasound have lower specificity, they provide important clue to identify the tumor, it's local extent and invasion of adjacent organ, which is crucial in guiding surgery [7].

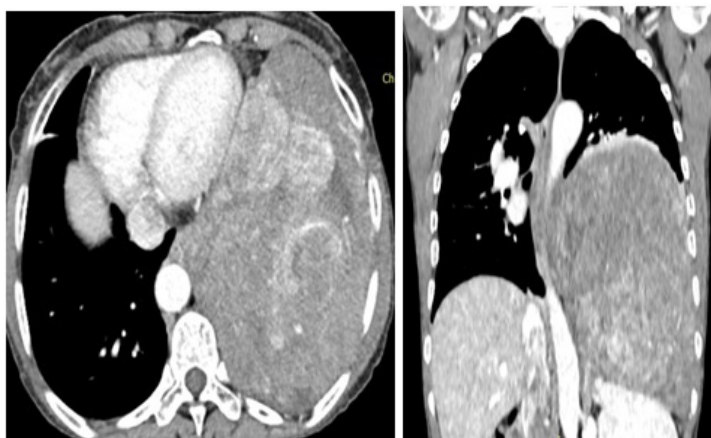
Complete Surgical resection is the main stay of treatment for SFTP

[5]. The case report has been reported in line with the SCARE 2018 criteria [8].

### 3. Case Presentation

Sixty-five years old female presented with a complaint of progressive worsening of shortness of breath of 1 year duration associated with intermittent dry cough, low grade fever, easy fatigability and loss of appetite. For these complaints she visited different hospitals and was treated for severe pneumonia with intravenous antibiotics and supported with oxygen. But her shortness of breath progressed, saturation of oxygen deteriorated and she became dependent on intranasal oxygen support. For this reason, she was referred to our tertiary care center, Saint Paul's Hospital Millennium Medical College, Addis Ababa, Ethiopia. She had no contact history with known pulmonary tuberculosis patients and have never smoked cigarettes. she has no history of drug allergy, self or family history of relevant medical or surgical illness. On presentation her oxygen saturation was 86 % with 3 liters of intranasal oxygen and respiratory rate was 28-34 breath/mint. She was emaciated and had digital clubbing. There was absent air entry over her left anterior and posterior lower two third lung field. Otherwise, there was no remarkable finding on other systems.

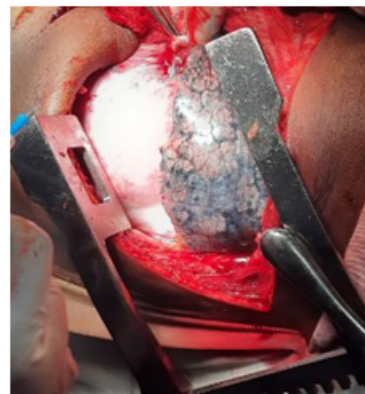
Her hemoglobin was 12.6gm/dl and serum albumin was 3.4gm/dl. Other parameters of complete blood count, renal function test and serum electrolytes were in the normal range. Chest computerized tomography (Chest CT) showed 20 cm x 13 cm x 15cm huge heterogeneously enhancing, lobulated, left intrathoracic mass that seem to arise from the left diaphragmatic pleura (Figure 1). It has compressed the left basal lung and thoracic aorta. Ultrasound guided tissue biopsy from the mass showed proliferative oval to spindle cells with mild to moderate nuclear pleomorphism in short and long fascicles with diagnostic impression of benign spindle cell lesion likely solitary fibrous tumor. Abdominal ultrasound didn't reveal any evidence of secondary.



**Figure 1:** Chest CT scan showing huge heterogeneously enhancing left intrathoracic mass compressing the thoracic aorta and left lung

With an impression of left intrathoracic mass likely arising from the left diaphragm the surgical team decided to do surgical ex-

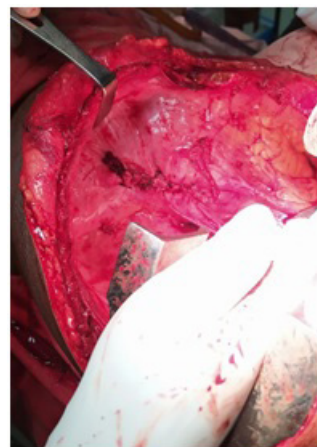
ploration and the patient was operated through left posterolateral thoracotomy after getting informed written consent. The intraoperative finding was a 20cm x 15 cm huge, highly vascular, firm, pedunculated left intra thoracic mass arising from the left diaphragmatic pleura with about 4cm stalk (Figure 2 and 3). The left lung was collapsed and had extensive adhesion with the mass. There was no mediastinal lymphadenopathy. With these findings en-bloc complete excision of the mass with the diaphragmatic stack was done and the diaphragmatic defect was closed in 2 layers (Figure 4). Left tube thoracotomy was left and thoracotomy wound was closed 2 in layers.



**Figure 2:** Intra operative picture showing huge left intrathoracic mass compressing the left lung



**Figure 3:** Post-operative picture showing 20 cm x 15cm firm mass weighing 3kg.



**Figure 4:** Intra operative picture showing diaphragmatic defect closed in 2 layers

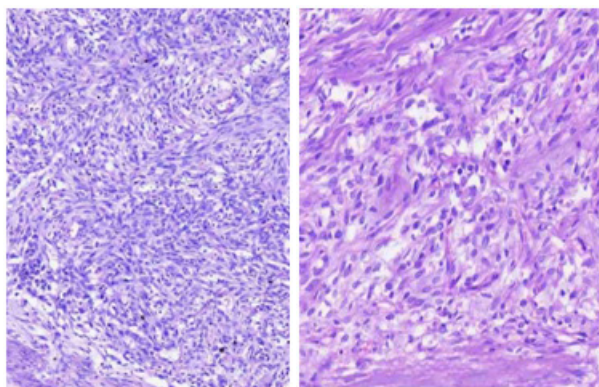
Post procedure, she was transferred to intensive care unit and put on oxygen support as well as epidural analgesics. Subsequently, she had smooth recovery and was transferred to surgical ward. The chest tube output was insignificant and minor bubbling decreased gradually. Control chest x ray taken on her 4th post op day showed well expanded left lung with no evidence of pneumothorax for which the chest tube was removed.

Tissue section of the mass showed lobulated, yellow-white appearance (Figure 5) and histopathologic study showed proliferative spindle to ovoid cells with ovoid nucleus and scanty cytoplasm in a collagenous stroma (Figure 6). There was no evidence of mitosis and necrosis the final diagnosis being Giant solitary fibrous tumor of the pleura.

Subsequently, the patient showed a remarkable improvement and was discharged from the hospital in a stable condition.



**Figure 5:** post-operative tissue cut section showing lobulated, yellow-white mass



**Figure 6:** Histologic section showing proliferative spindle to ovoid cells with ovoid nucleus and scanty cytoplasm

#### 4. Discussion

Solitary fibrous tumor is slow growing, rare, mesenchymal tumor accounting for < 2% of all soft tissue tumors [9]. About 50-70% of them are extra pleural involving the abdomen, pelvis, head and neck [10]. SFTP spreads locally to the chest wall, axilla and su-

praclavicular area, and distant metastasis to the liver, bone, brain and adrenal gland is unusual [2]. Hilar and mediastinal lymphatic metastasis occurs in <50% of patients [2]. Our patient has no imaging and intraoperative evidence of metastasis. When SFTP arise from the diaphragmatic pleura both the diaphragm and peritoneum may be involved [2] and we have resected part of the diaphragm attached to the stalk of the mass. Although 80% of SFTP arise from visceral pleura [2], the origin in our case was from the parietal pleura which is rare. About 12% of SFTP are malignant and are characterized by their larger size usually >8cm, arise in atypical locations like parietal and mediastinal pleura, and histological features of increased cellularity, pleomorphism and >4 mitosis/10 HPF [11]. In this regard, although our patient's tumor arises from the parietal pleura and have larger size the histological features go for benign SFTP. Benign SFTP may rapidly enlarge and transform into the malignant form [11]. For this reason, complete resection of all SFTP is mandatory.

Most SFTP are asymptomatic being diagnosed incidentally in 50% of cases [11] and those who are symptomatic are usually malignant and larger in size [4]. Symptoms include cough, chest pain, dyspnea, digital clubbing and clinical findings of pleural effusion. Rarely, SFTP may produce insulin-like growth factor 2 and cause refractory hypoglycemia (Doege-Potter syndrome) [4]. Although our patient was being treated for pneumonia which could be explained by obstructive pneumonitis in patients with SFTP, there was delay to reach at diagnosis of SFTP as its rarity hinders it from being considered into the differential diagnosis.

Although no specific computed tomographic features have been described, SFTP typically appear as well-defined, enhancing, heterogeneous or homogenous mass in contact with the pleural surface [4]. Like our case, definitive diagnosis is often made after surgical exploration and histopathologic study. Although we couldn't do immunohistochemistry study in our case, SFTP are positive for vimentine and CD34, and are negative for keratin [7].

The main stay of treatment for all SFTP is complete enbloc surgical excision with 2 cm tumor free margin [4]. Post-operative risk of recurrence is higher especially for malignant, sessile SFTP and usually occurs within the first 2years of follow up [4]. Although neo-adjuvant and adjuvant treatment with chemotherapy and radiotherapy is being used sporadically, its benefit is still unproved because of limited number of patients [4]. In this regard, our patient was not initiated on adjuvant treatment. We are following her with clinical evaluation and chest x-ray every 3 months, and have chest CT scan annually. She has marked improvement of her symptoms and is also happy with her treatment.

#### 5. Conclusion

Although SFTP is rare mesenchymal tumor, its behavior is unpredictable. Patients have no specific clinical presentation and are usually misdiagnosed. So, SFTP should be considered as differ-

ential diagnosis in patients with atypical or recurrent respiratory symptoms despite adequate medical treatment. Complete surgical excision is the main stay of treatment and meticulous post-operative follow up is mandatory as the risk of recurrence is higher and the tumor behavior is still less understood.

## References

1. Lateef HA. A Large Solitary Fibrous Tumor of the Pleura Associated with Massive Pleural Effusion: A Case Report and Review of the Literature. 2019; 1: 4.
2. Fiore F, Stoia V, Somma F. Surgical recurrence of solitary fibrous tumor of the pleura treated with microwave (MW) thermoablation: A case report. *Thorac Cancer*. 2020; 11: 443-6.
3. Sánchez-Mora N, Cebollero-Presmanes M, Monroy V, Carretero-Albiñana L, Herranz-Aladro M, Alvarez-Fernandez E. Clinicopathological features of solitary fibrous tumors of the pleura: a case series and literature review. *Arch Bronconeumol Engl Ed*. 2006; 42(2): 96–9.
4. de Perrot M, Fischer S, Sekine Y, Keshavjee S. Solitary Fibrous Tumors of the Pleura. *Ann Thorac Surg*. 2002; 74: 285-93.
5. Gengler C, Guillou L. Solitary fibrous tumour and haemangiopericytoma: evolution of a concept. *Histopathology*. 2006; 48(1): 63–74.
6. Liu D, Wang Y, Zheng Y, Zhang H-L, Wang Z-H. Massive malignant solitary fibrous tumor of the diaphragm. 2020; 5.
7. Ge W, Yu D-C, Chen G, Ding Y-T. Clinical analysis of 47 cases of solitary fibrous tumor. *Oncol Lett [Internet]*. 2016; 12(4): 2475–80.
8. Agha RA, Borrelli MR, Farwana R, Koshy K, Fowler AJ, Orgill DP, et al. The SCARE 2018 statement: updating consensus Surgical CAse REport (SCARE) guidelines. *Int J Surg*. 2018; 60: 132–6.
9. Gold JS, Antonescu CR, Hajdu C, Ferrone CR, Hussain M, Lewis JJ, et al. Clinicopathologic correlates of solitary fibrous tumors. *Cancer*. 2002; 94(4): 1057–68.
10. van Houdt WJ, Westerveld CM, Vrijenhoek JE, van Gorp J, van Coevorden F, Verhoef C, et al. Prognosis of solitary fibrous tumors: a multicenter study. *Ann Surg Oncol*. 2013; 20(13): 4090–5.
11. Abeeleh MA, Alsmady M, Ennab R, Saleh S. Solitary Fibrous Tumor Arising from the Mediastinal Pleura: Case Report. 2011; 45: 109-12.