Recurrent malignant Solitary Fibrous tumor of the pleura: Case Report

Maun Feteih[1], Haneen Hussain Alabsi[2], Manal Mahmood Alsalmi[3], Anas Ahmad Sultan[4], Manal Mahmood Alsalmi[4], Mohamed Abduljabar[5], Mohammed Alaa Jamjoom[6], Meshari Alnijaidi[7]

[1]mfeteih@gmail.com,Consultant Pulmonary Medicine,Department of Internal Medicine King Faisal specialist Hospital & Research Centre,Jeddah - Saudi Arabia,Orchid: 0000-0001-8833-9081
[2]Haneen94alabsi@gmail.com,Fellow,Faculty of Medicine,Department of Medicine.King Faisal Specialist Hospital and Research Center,University: King Abdulaziz University,Jeddah, Saudi Arabia,ORCID: 0000-0002-2573-0144
[3]dr.manalalsalmi@windowslive.com,Fellow,Faculty of Medicine,Department of Medicine University: Ibn sina national college,Jeddah, Saudi Arabia,ORCID : 0009-0008-3091-5080
[4]dranas_74@hotmail.com,Assistant consultant pulmonary,Department of Medicine,King Faisal Specialist Hospital and Research Center,Jeddah, Saudi Arabia,ORCID: 0000-0001-9956-3193
[5]m.jabbar75@gmail.com,Assistant consultant Gastroenterology,Faculty of medicine,Department of medicine,king faisal specialist hospital & research center,University: Aleppo Syria,Jeddah, Saudi Arabia Orchid: 0009-0009-6212-0976
[6]mohammed-jamjoom@hotmail.com,Fellow,Faculty of Medicine,Department of Medicine King Faisal Specialist Hospital and Research Center,University: King Abdulaziz University,- Jeddah, Saudi Arabia,- ORCID: 0009-0005-0273-802X
[7]Malnijaidi@gmail.com,Fellow,Faculty of medicine,- Department: Department of medicine University: Umm Alqura,- Makkah, Saudi Arabia,Orcid: 0009-0001-7753-2385

KEYWORDS

ABSTRACT:

Solitary fibrous tumors (SFTs) of the pleura are rare neoplasms originating from the mesenchymal tissue of the pleura. We present a case report of a 64-year-old female patient who presented with shortness of breath, dizziness, and palpitations. She also experienced fever, productive cough with hemoptysis, pleuritic chest pain, weight loss, anorexia, and night sweats. Imaging revealed a large hyper vascular lobulated solid mass occupying the right middle and lower lobes of the lung, causing atelectasis. (Image) A biopsy confirmed the diagnosis of solitary fibrous tumor, which was surgically excised. Histopathology revealed a malignant solitary fibrous tumor confined to the pleura. Adjuvant radiation therapy was given. She was kept on surveillance imaging including PET scan for duration of 4 years without any evidence of disease recurrence. She developed radiation pneumonitis, requiring steroid treatment. After 4 years of surveillance, she developed local large recurrence of the tumor in the right lower chest extending to the right upper abdomen. She had successful resection of the recurrent fibrous tumor of the pleura which was done by thoracic and General surgery.

This case highlights the clinical presentation, diagnostic and management challenges for recurrent malignant fibrous tumor of the pleura, emphasizing the importance of a multidisciplinary approach in the management of such rare tumors.

Introduction:

Solitary Fibrous Tumor (SFT) of the pleura is a rare neoplasm originating from the mesenchymal tissue of the pleura. It was first described by Klemperer and Rabin in 1931 as a localized fibrous tumor arising from the pleura. [1] SFTs are generally slow-growing and asymptomatic, making them difficult to diagnose without appropriate imaging studies or histopathological analysis. [2] they have the potential to become locally invasive and recur after surgical resection. [3]

In this case report, we present a unique case of recurrent solitary fibrous tumor of the pleura. We discuss the diagnostic challenges encountered, the radiological and histopathological findings, and the management strategy employed.[4] By sharing this case, we aim to contribute to the existing literature on SFTs and emphasize the importance of early recognition , the need for long-term follow-up and appropriate management of this rare pleural neoplasm.[5]

Case Presentation:

A 64-year-old female patient presented to our hospital in July 2019, complaining of shortness of breath (NYHA class II) associated with episodes of dizziness and palpitation for the past 4 months. Additionally, she experienced intermittent subjective fever, productive cough with yellowish to greenish sputum, hemoptysis, pleuritic chest pain, significant weight loss (>10 kg), anorexia, and drenching night sweats. Physical examination revealed decreased air entry over the right lung, but no palpable lymph nodes. Chest X-ray showed elevated right hemidiaphragm with right middle and lower lobe atelectasis/infiltrate and a possible small pleural effusion. CT chest without contrast revealed a hypervascular lobulated solid mass occupying the right middle and lower lobes, measuring 16 x 9 x 15 cm, causing atelectasis.(IMAGE) (figure 1). Pulmonary function tests indicated severe restrictive lung disease with preserved DLCO, predominantly due to compression and collapse of the right lung by the mass.

The patient underwent an US-guided biopsy of the right lung mass, which was consistent with a solitary fibrous tumor. Immunohistochemistry showed diffuse positivity for CD34, Bcl2, and CD99. Subsequently, she underwent excision of the right pleural mass in August 2019. Histopathology confirmed a malignant solitary fibrous tumor confined to the pleura, measuring 16 cm. Due to tumor involvement at the inked margin, she was referred to radiation oncology and medical oncology for adjuvant therapy. She completed adjuvant radiation therapy to the tumor bed and the large residual lesion in January 2020. A repeat PET scan showed no signs of disease recurrence.

However, the patient developed chronic dry cough, chest tightness, and wheezing, which exacerbated with dust and strong fumes. She was diagnosed with radiation pneumonitis and required a course of steroids for management.

She had regular follow-up CT chest imaging post tumor resection for 4 years which showed stable right middle and lower lobe interlobular septal thickening with traction bronchiectasis and surrounding pleural thickening with calcified pleural plaques, likely postoperative and radiotherapy related. Figure 2

Additionally PET scan was done also to evaluate for pleural thickening, which came negative for any FDG avid lesion, supporting the diagnosis that pleural thickening is due to post operative and radiotherapy related.

. After 4 years of surveillance, she developed local large recurrence of the tumor in the right lower chest extending to the right upper abdomen. Figre 3 and 4.

Multi-disciplinary meeting including thoracic surgery, general surgery, Oncology, pulmonology was done, and since there is no evidence of distal metastasis, she was considered for surgery again.

She had successful resection of the recurrent fibrous tumor of the pleura which was done by thoracic and General surgery.

Again follow-up multiple CT chest and PET scan was done to evaluate for persistent or recurrent disease.

PET scan showed hydropneumothorax with rim of mild FDG uptake SUV maximum 5.4, mildly FDG avid pleural thickening in the right upper lobe with SUV max 5, no other FDG avid lesion.

Final conclusion that this mild FDG uptake surrounding right hydropneumothorax favor inflammatory etiology.

Given that above PET scan finding, we will keep the patient on close follow-up regular imaging given that prior history of cancer recurrence.

Discussion:

Solitary Fibrous Tumor of the Pleura (SFTP) is a rare neoplasm originating from mesenchymal cells of the pleura. Although it is considered a benign tumor, it can exhibit malignant behavior in certain cases. SFTPs contributes to about 5% of all pleural tumours, making them relatively uncommon tumours. With a small male predominance, it typically affects people between the ages of 50 and 70. [6] SFTs can exist in extra pleural locations like the lung, mediastinum, and other soft tissues, albeit they are more commonly detected in the pleura. Due to its rarity, little information is known about the true incidence and prevalence of SFTP. [6]
The differential diagnostic techniques for SFTP presents numerous difficulties. First off, the clinical picture is frequently non-specific that could be mistaken for those of other, more prevalent respiratory disorders, delaying diagnosis. Although imaging tests such as a chest radiograph, computed tomography (CT), and magnetic resonance imaging (MRI) may offer preliminary cues, a histopathological investigation confirms the diagnosis. [7]
The spindle-shaped form of tumour cells grouped in a model "without model," with alternating hypocellular and hypercellular patches, characterizes the histological examination of SFTP. In order to establish a diagnosis, immunohistochemistry is essential. CD34, CD99, and Bcl-2 are frequently seen. However, in addition to the diagnostic difficulty, these immunohistochemistry markers are not specific to SFTP and may also be expressed in other mesenchymal cancers. [8]
The core management of SFTP presents particular difficulties because of their rarity and potential for malignant activity. Surgical excision remains the gold standard of treatment for localized diseases. The extent of the surgical procedure, including whether plural decortication or maybe lung excision if necessary, is determined by the size, involvement, and location of the tumor as well as any adjacent tissues. [3]

Conclusion:

In cases where complete resection is unfeasible or in cases of metastatic disease, adjuvant treatment options such radiation therapy, chemotherapy, or targeted therapy may be considered. Further study is necessary because the effectiveness of various modalities in SFTP is not clearly proven. [9]
There are various possibility of local recurrence or distant metastasis even years after the initial treatment, our case had recurrence of disease after 4 years surveillance, this highlights that long-term follow-up of SFTP patients is essential. To have a look on the symptoms of infection and recurrence, Radiological imaging, post-Surveillance, and detailed clinical evaluation are required.

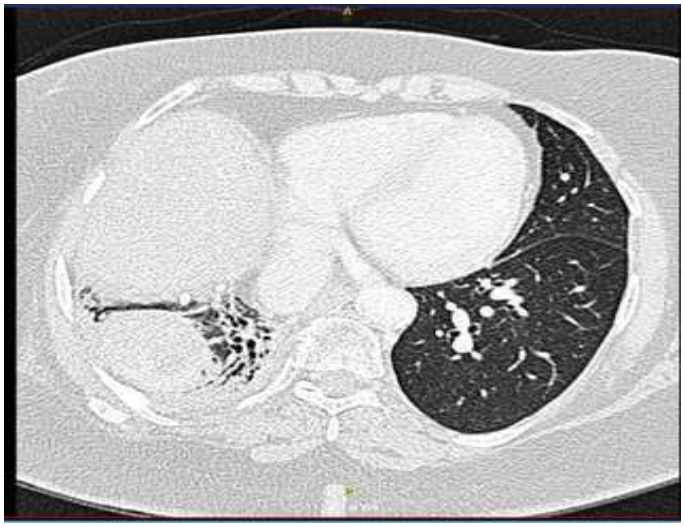
[10] The ideal protocol type and duration of follow-up have not been precisely determined because of the rarity of the tumor and the absence of established guidelines. [11]

References:

1. Fatimi S Hasnain, Inam H, Chagan FK, Choudry UK. Solitary fibrous pleural tumor. A rare and challenging case. PubMed Central (PMC) 2019. https://doi.org/10.1016/j.ijscr.2019.12.020.
2. Solitary fibrous tumour of the pleura: surgical treatment - PubMed. PubMed 2001. https://doi.org/10.1016/s1010-7940(00)00636-9.
3. Solitary fibrous tumors of the pleura - PubMed. PubMed 2002. https://doi.org/10.1016/s0003-4975(01)03374-4.
5. M. Mercer R, Wigston C. Management of solitary fibrous tumours of the pleura: a systematic review and meta-analysis. ERS Publications 2020. https://openres.ersjournals.com/content/6/3/00055-2020 (accessed May 8, 2023).
7. Solitary fibrous tumor of the pleura - PubMed. PubMed 2006. https://doi.org/10.1177/107327480601300403.
8. Solitary fibrous tumor of the pleura in a 22-year-old woman: a case report - PubMed. PubMed 2020. https://doi.org/10.1177/0300060520959495.
9. 3.Abodunrin FO, Killeen RB. Pleural Solitary Fibrous Tumors - Stat Pearls - NCBI Bookshelf. Pleural Solitary Fibrous Tumors - Stat Pearls - NCBI Bookshelf 2023. https://www.ncbi.nlm.nih.gov/books/NBK585038/#:~:text=Solitary%20pleural%20fibrous%20tumors%20represent,are%20mostly%20benign%20in%20nature
10. Solitary fibrous tumors of the pleura: clinical, radiological, surgical and pathological evaluation - PubMed. PubMed 2005. https://doi.org/10.1016/j.ejso.2004.09.017
11. Solitary fibrous pleural tumor. A rare and challenging case. Solitary Fibrous Pleural Tumor A Rare and Challenging Case - ScienceDirect 2019. https://doi.org/10.1016/j.ijscr.2019.12.020.

Figure 1: Large hypervascular solid mass in the right hemothorax causing atelectasis of middle and lower right lobe with significant volume loss of right hemithorax

Figured 2: Persistent right middle and lower lobe interlobular septal thickening with traction bronchiectasis and surrounding pleural thickening with calcified pleural plaques likely postoperative and radiotherapy related.



Figured 3: Right lower chest pleural recurrent soft tissue mass with adjacent pleural thickening, and extension to the upper abdomen

Figure 4: Fibrous tumor of the pleura with extension to the right abdominal cavity posteriorly inseparable from the right hepatic lobe.