

EC CLINICAL AND MEDICAL CASE REPORTS

Case Report

Solitary Fibrous Tumor of the Pleura (SFTP): A Rare Form of an Intrathoracic Space-Occupying Lesion

Dabeluchi C Ngwu^{1,2} and Nicholas A Kerna^{3,4*}

¹Cardiovascular and Thoracic Surgery Unit, Department of Surgery, Federal Medical Center, Umuahia, Nigeria

²Earthwide Surgical Missions, Nigeria

³Independent Global Medical Research Consortium

⁴First InterHealth Group, Thailand

*Corresponding Author: Nicholas A Kerna, (mailing address) POB47 Phatphong, Suriwongse Road, Bangkok, Thailand 10500. Contact: medpublab+drkerna@gmail.com.

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Abstract

Introduction: This paper reports on an elderly African female who presented with a right intrathoracic space-occupying lesion, which was subsequently discovered to be a giant benign solitary fibrous tumor of the pleura—with a discussion of the surgical intervention that followed.

Case Report: Presented is the case of a 75-year-old female, who was referred to the Cardiovascular and Thoracic Surgery Unit, Federal Medical Center, Umuahia, Nigeria, with complaints of vague chest discomfort and progressive shortness of breath over 18 months. The imaging investigations were reviewed, indicating the need for a right posterolateral thoracotomy for tumor resection. The surgical procedure lasted approximately 2 hours, followed by an uneventful postoperative recovery. After one week, the patient was discharged, is currently asymptomatic, and is being seen on an outpatient basis.

Discussion: Pleural SFTs are rare neoplasms, most benign. Pleural SFTs present with various and distinct symptoms. Chest CT scanning is the preferred imaging technique. Nevertheless, the radiological differentiation between the benign and malignant variants remains challenging. Surgery is the treatment of choice for a solitary fibrous tumor of the pleura (SFTP). Local recurrence can present in malignant cases but is infrequent in solitary benign tumors. Surgical resection's prognosis is generally favorable, even for a malignant SFTP.

Conclusion: Rapid preoperative diagnosis followed by expeditious surgical intervention is the goal in the surgical resection of a solitary fibrous tumor (SFT), resulting in rapid symptomatic relief and long-term survival in benign and most malignant SFTs. SFTs are rare tumors, challenging to diagnose definitively.

Keywords: Atelectasis; Chest Imaging; Immunohistochemical; Index Patient; Posterolateral Thoracotomy; Respiratory System Surgery

Introduction

According to Vejvodova., *et al.* (2017), the pleura's solitary fibrous tumors (SFTs) are rare, accounting for less than 5% of all primary pleural tumors, with an incidence of approximately 2.8 cases per 100,000 per year. Benign and malignant variants of these tumors have been reported [1].

Chest CT scan is the cornerstone of preoperative radiological assessment. Also, supplementary information is often obtained from other imaging techniques [2]. Complete surgical resection, whenever technically and medically feasible, is recommended for all cases of benign and localized malignant SFTs, typically resulting in long-term survival [3,4].

SFTs represent a heterogeneous group of primary pleural neoplasms with a low incidence rate. The biological origin, which consists of mesenchymal cells, is uncertain [1]. SFTs form about 5% of all pleural tumors [2]. SFTs' incidence is approximately 2.8 cases per 100,000 registered hospitalized patients, typically occurring in all age groups with a peak incidence in the fourth and sixth decades of life [3].

Due to the slow growth of these types of tumors, diagnoses are often incidental to routine chest imaging. Perrotta., *et al.* (2016) reported that the most frequent symptoms are cough, chest pain, and dyspnea—common to several respiratory diseases. Less frequent presentations are hemoptysis, obstructive pneumonia, atelectasis, and clubbing [6].

Harrison–Phipps., *et al.* (2009) noted that SFTs are distinct from diffuse malignant mesothelioma, which is associated with asbestos exposure and typically carries a poor prognosis [5]. Benign fibrous tumors can be defined as giants when their diameter is greater than 15cm or when tumors occupy more than 40% of the hemithorax [6]. Surgery remains the standard of treatment for SFTs and is performed with a wide margin to reduce the chance of recurrence [4,5].

The following case report focuses on an elderly African female with a large intrathoracic space-occupying mass—a giant solitary fibrous tumor of the pleura.

Case Report

Presented is the case of a 75-year-old female, who was referred to the Cardiovascular and Thoracic Surgery Unit, Federal Medical Center, Umuahia, Nigeria, with complaints of vague chest discomfort and progressive shortness of breath over 18 months. Various health care providers had treated the patient for multiple conditions, including asthma, pleural effusion, hydatid disease, and heart failure. However, chest imaging (chest radiograph and CT scan) revealed a large mass in the right hemithorax, necessitating referral for expert thoracic surgical care.

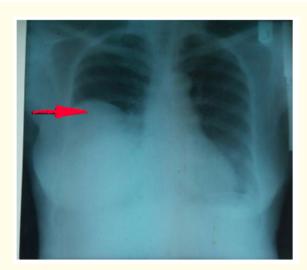


Figure 1: Benign solitary fibrous tumor of the lung.

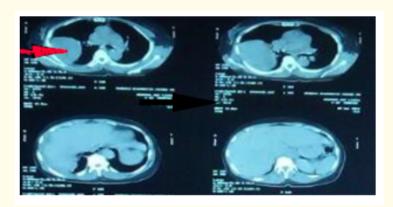


Figure 2: Preoperative CT chest showing a solid mass in the right hemithorax.

The imaging investigations were reviewed, and the previously-mentioned findings were noted. The patient had a formal right posterolateral thoracotomy for resection of the tumor. Intraoperatively, a large solid tumor was observed in the right hemithorax, occupying about 50% of the cavity. The tumor was seen to be pedunculated, arising from the lower lobe of the right lung and abutting the diaphragm, the mediastinum, and the chest wall.

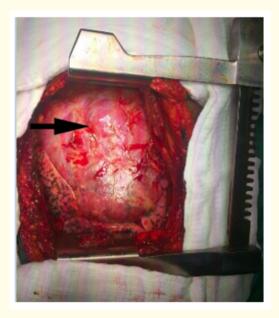


Figure 3: Intraoperative view of the tumor with a cuff of the uninvolved lung.

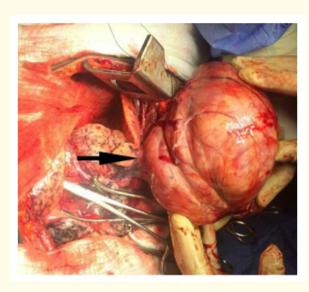
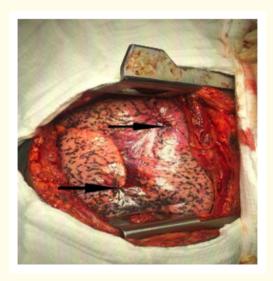


Figure 4: The tumor pedicle was exposed to show attachment to the underlying lung.



 $\textbf{\it Figure 5:} \textit{ After tumor excision, the defects in the lung parenchyma were repaired.}$



Figure 6: The tumor was excised and measured.



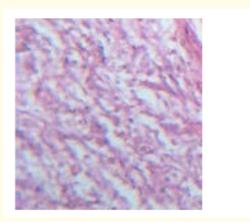
Figure 7: Cut sections of the tumor.



Figure 8: Postoperative chest radiograph: the previous preoperative right-sided shadow is absent.

Procedurally, the lower and middle lobes were collapsed. The surgery lasted about 2 hours, followed by an uneventful postoperative recovery. The patient was discharged home after 1 week, is asymptomatic, and is being followed up on an outpatient basis.

The gross appearance of the resected tumor, its histopathologic features, and the immunohistochemical study results were consistent with a diagnosis of a benign solitary fibrous tumor of the pleura.



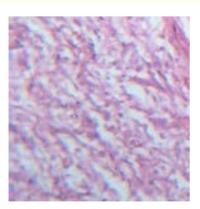


Figure 9 and 10: The histopathic features of the index patient's SFT.

Figure 9 (Left): Microscopic specimen of the tumor demonstrating spindle tumor cells (without patterns) intermingled with varying amounts of collagen and hyalinization of fibrous tissue (H&E ×200). Figure 10 (Right): Microscopic specimen of the tumor, depicting spindle tumor cells with sections of hyalinization and a seeming hemangiopericytoma vascular pattern (H&E ×200).

Discussion

Sung., et al. (2005) reported that pleural SFTs are a rare neoplasm with less than a thousand cases reported in decades [6]. Thus, this case report may be the first of its kind from Nigeria.

Although the majority of these neoplasms are benign, approximately 10–20% show a more aggressive biological behavior, with pathological findings of invasiveness, the appearance of distant metastases, and local relapse after resection [5]. Cardillo., *et al.* (2009) observed that SFTs with malignant morphology and behavior are generally larger than their benign counterpart [7]. However, as in the index patient, extensive tumors can also be benign.

Some researchers suggest that larger tumors are more likely to be malignant, having ample time to undergo genetic changes [8]. Pleural SFTs can present with various and distinct symptoms, such as intrathoracic symptoms (dyspnea, chest pain, and hemoptysis), systemic symptoms (hypoglycemia or hypertrophic osteoarthropathy), or nonspecific symptoms (fever, weight loss, and fatigue). In Sung., *et al.* (2005), the proportion of symptomatic patients ranged from 50–60% [6]. The index patient was symptomatic for longer than 18 months before specifying a definitive diagnosis.

Chest CT scanning is the test of choice, but the radiological differentiation between the benign and malignant variants remains challenging. However, specific findings could suggest malignancy, such as a diameter exceeding 10 cm, central necrosis, and ipsilateral pleural effusion, according to criteria described by Ferretti., et al. (1997) [9].

Surgery is the treatment of choice for a solitary fibrous tumor of the pleura (SFTP). Moreover, the resection complements represent the surgery's primary objective [5]. This result can usually be achieved via a formal thoracotomy, ensuring completeness of the resection.

Video-assisted thoracoscopic surgery (VATS) is indicated for lesions less than 5cm in diameter [4]. Local recurrence can present in malignant cases but is rare in solitary benign tumors. Such a recurrence may result from an incomplete or conservative surgery, lack of identification of a tumor during the operation, or growth of a synchronous neoplasm independent of that removed [4,10].

Long-term follow-up after complete resection is necessary because SFTPs can recur, particularly in malignant or incompletely resected benign SFTs. Prognosis is generally favorable with surgical resection, even for a malignant SFTP—having a reported overall 5-year survival of 81% [11].

Conclusion

Solitary fibrous tumors are rare tumors that can pose a diagnostic dilemma. Hence the need to maintain a high level of suspicion when evaluating a patient with an intrathoracic mass. Surgical resection is typically rewarded with rapid symptomatic relief, confirmation of the pathological diagnosis, and long-term survival in benign and most cases of malignant SFTs. A rapid preoperative diagnosis followed by expeditious surgical intervention is the objective when treating solitary fibrous tumor of the pleura.

Conflict of Interest Statement

The authors declare that this paper was written without any commercial or financial relationship that could be construed as a potential conflict of interest.

The manuscript was read and approved by both authors.

Supplementary Note

A previous abstract (appreciably updated herein) was presented at the Joint Association of Surgeons of Nigeria and the Nigerian Surgical Research Society Meeting, Umuahia, Nigeria, in July 2018 [12].

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