**SAS Journal of Surgery**
Abbreviated Key Title: SAS J Surg
ISSN 2454-5104
Journal homepage: https://www.saspublishers.com

OPEN ACCESS

**Respiratory Diseases**

# Giant Pleural Solitary Fibrous Tumor Revealed by Superior Vena Cava Syndrome: A Case Report

K. Chaanoun[1*], N. Zaghba[1], H. Benjelloun[1], N. Yassine[1]

[1]University Hassan 2, Casablanca, Morocco

**DOI:** https://doi.org/10.36347/sasjs.2024.v10i12.010 | **Received:** 12.09.2024 | **Accepted:** 16.10.2024 | **Published:** 26.12.2024

***Corresponding author:** K. Chaanoun
University Hassan 2, Casablanca, Morocco

**Abstract** **Case Report**

***Introduction*:** Solitary fibrous tumors are rare mesenchymal tumors, often benign, of pluripotent fibroblastic or myofibroblastic origin. They usually develop at the expense of the pleura, but they can be localized to any organ. ***Clinical observation*:** we report the case of a 50-year-old patient who was admitted to the department for a superior vena cava syndrome revealing a giant thoracic tumor. The biopsy of the mass was in favor of a pleural solitary fibrous tumor. ***Discussion*:** The etiopathogenesis of the disease is unclear, but it could be related to accumulated somatic mutations, including the NAB2-STAT6 fusion, TP53 mutations and TERT promoter mutations. ***Conclusion*:** The diagnosis of a pleural solitary fibrous tumor is confirmed by histology after biopsy or surgical excision. The prognosis depends on the size of the mass, the mitotic index, and the presence or absence of necrosis.

**Keywords:** pleural tumor; solitary fibrous tumor; superior vena cava syndrome; thoracic mass.



## INTRODUCTION

Solitary fibrous pleural tumor (SFPT) is a rare entity of mesenchymal tumors, most often benign. Its discovery is most often fortuitous or occurs during non-specific respiratory signs or rarely hypoglycemia. A histopathological assessment is necessary to diagnose these tumors. And whose treatment of choice is based on surgical resection.

## OBSERVATION

We report the case of a 50-year-old patient, without toxic habits, diabetic for 16 years under oral antidiabetics. The patient consulted for dyspnea on exertion that had been developing for 6 months associated with a dry cough with a slight decline in general condition, the evolution was marked after by the appearance of dizziness, visual disturbances, slight edema of the eyelids, and a thickening of the supraclavicular fossa. The examination on admission found a syndrome of fluid effusion of the entire right hemithorax, digital clubbing and a left laterocervical mass that was mobile on swallowing. The chest X-ray showed a dense and homogeneous opacity occupying almost the entire right hemithorax, sparing only a strip of pulmonary parenchyma at the base (Figure 1). The chest CT scan showed a right thoracic mass of 17.7 cm/18.4 cm/11.4 cm attached to the parietal pleura, it is of thick inhomogeneous fluid density, the site of hemorrhagic changes, compressive and laminates the superior vena cava, the mass pushes back the lobar bronchi opposite with subsegmental ventilation disorders, we also note on the scanner the presence of a goiter with left endothoracic extension (Figure 2). The cervical ultrasound showed a heterogeneous multinodular goiter. The thyroid assessment was normal apart from a high thyroglobulin (over 500 ng /ml), Flexible bronchoscopy showed a reduced caliber trachea, the carina was misaligned and flared with extrinsic compression of the entire right bronchial tree. Transparietal biopsy of the mass demonstrated a fusiform tumor proliferation with features in favor of a TFSP. Immunohistochemistry showed cells expressing STAT6 and CD34, other markers (desmin, P-S100) were not expressed.

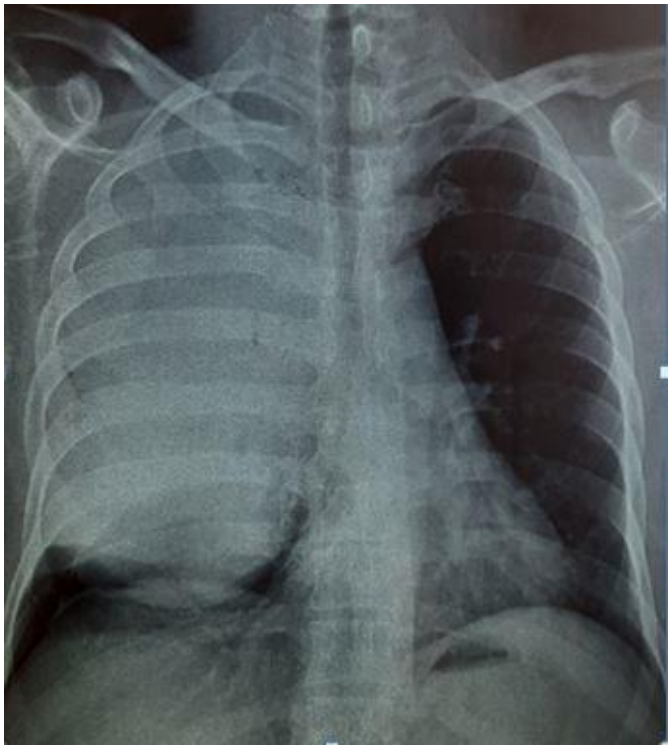

**Figure 1: Frontal chest radiograph showing an opacity occupying almost the entire right hemithorax**

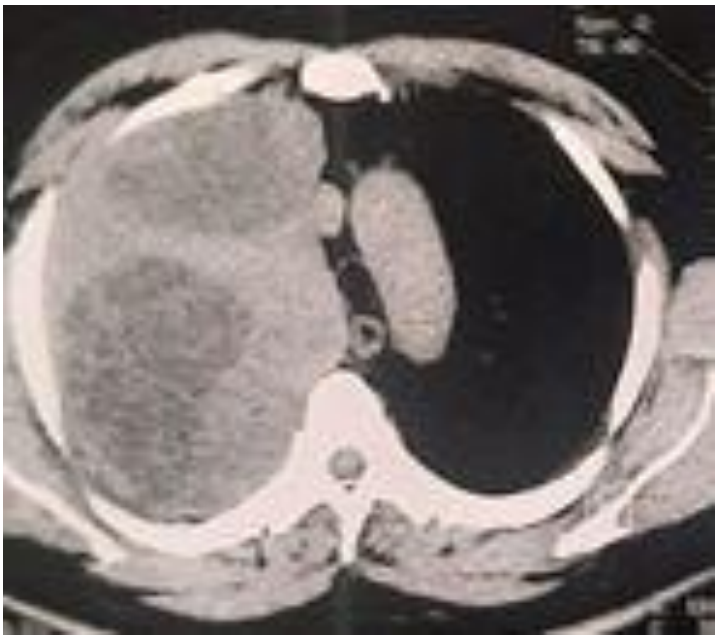

**Figure 2: Chest CT scan showing a massive right thoracic tumor mass.**

## DISCUSSION

Solitary fibrous tumors represent a rare entity of primary tumors originating from mesenchymal cells. They can involve any organ with a mesothelial lining. TFSPs represent less than 5% of primary pleural neoplasias [1]. The causes of TFSP are not known. Unlike other pleural malignancies such as mesothelioma associated with asbestos exposure, the risk factors for TFSP remain to be elucidated.

Patients are usually asymptomatic, but some nonspecific respiratory symptoms such as dyspnea, chest pain, or dry cough have been reported by up to 40–60% of patients. Paraneoplastic syndromes associated with this tumor are Pierre Marie and Foix syndrome (Hypertrophic Osteoarthropathy pneumopathy) probably due to abnormal production of hyaluronic acid or; Or a Doege-Potter syndrome which corresponds to refractory hypoglycemia related to the secretion by the tumor of an insulin-like growth factor. Diffuse arthralgia, galactorrhea or a deterioration in the general condition are also possible. These systemic effects generally

disappear after excision of the neoplasms, but can reappear during subsequent tumor recurrences.

Chest X-ray is a diagnostic step, most often showing a rounded, homogeneous and well-limited opacity, rarely associated with pleural effusion. However, for large tumors, with pulmonary compression as in our observation, the radiographic image is limited to a simple opacity of the entire pulmonary hemifield concerned associated with a significant mediastinal deviation. Chest CT scan is the key examination that confirms the pleural origin and the tissue nature of the tumor, as well as specifying its location and its relationships with the pleura and adjacent organs, which facilitates surgical planning. The tumor often appears well-limited and homogeneous, but a heterogeneous appearance can be observed when there are necrotic-hemorrhagic changes or myxoid degeneration. Calcifications and pleural effusions are rare. The tumor can enhance after injection of contrast agent and can be mobile.

Nuclear magnetic resonance imaging allows the fibrous nature of the tumor to be determined, especially in T1 sequence, which differentiates it from other mediastinal -pulmonary, parietal and diaphragmatic structures.

However, diagnostic certainty can only be provided by anatomopathological examination. Macroscopically, the tumor is often well circumscribed with a lobulated or smooth external surface, generally encapsulated, of variable size, which can be sessile or pedunculated [2]. Histologically, it is a more or less dense proliferation of spindle cells reminiscent of fibroblasts, dispersed in a more or less orderly manner and supported by a framework of collagen fibers of variable abundance [2]. The immunohistochemical study has been extremely useful in differentiating SFTP from mesotheliomas and other sarcomas, it classically shows a diffuse expression of vimentin and CD34, an expression of variable intensity of CD99 and the Bcl-2 protein and a negativity of epithelial markers (cytokeratin, EMA) and the S100 protein. Recently, an intrachromosomal NAB2- STAT6 genetic fusion has been identified as the defining genetic event of TFS [3]. This feature has allowed STAT6 to be a highly sensitive and almost perfectly specific immunohistochemical marker of TFS especially in CD34-negative cases, and useful to distinguish this type of tumor from histological mimics [3, 4].

The standard treatment for benign and malignant TFSP is complete resection of the tumor, which can sometimes extend to the lung parenchyma or wall in the event of invasion. Aggressive surgery is recommended due to the probability of recurrence or malignant transformation. However, chemotherapy and/or radiotherapy may be indicated for aggressive forms, in the event of recurrence or incomplete resection. Due to this difficult-to-evaluate evolutionary profile, long-term radio-clinical monitoring is recommended by some authors. The prognosis depends on the size of the mass, the mitotic index, and the presence or absence of necrosis [5].

## CONCLUSION

TFSPs are rare tumors, of which clinicians should be aware. The clinical presentation is nonspecific. Diagnosis can be suggested by imaging and confirmed by histology after biopsy or surgical excision. Prognosis depends on the nature and location. However, surgical resection continues to be the mainstay of treatment for TFSP. Surveillance at long-term is necessary to detect recurrences or malignant transformation.

## REFERENCE

1. Boothe, J. T., Budd, G. T., Smolkin, M. B., & Ma, P. C. (2018). Durable near-complete response to anti-PD-1 checkpoint immunotherapy in a refractory malignant solitary fibrous tumor of the pleura. *Case Reports in Oncology*, *10*(3), 998-1005.
2. England, D. M., Hochholzer, L., & McCarthy, M. J. (1989). Localized benign and malignant fibrous tumors of the pleura: a clinicopathologic review of 223 cases. *The American journal of surgical pathology*, *13*(8), 640-658.
3. Doyle, L. A., Vivero, M., Fletcher, C. D., Mertens, F., & Hornick, J. L. (2014). Nuclear expression of STAT6 distinguishes solitary fibrous tumor from histologic mimics. *Modern Pathology*, *27*(3), 390-395.
4. Yoshida, A., Tsuta, K., Ohno, M., Yoshida, M., Narita, Y., Kawai, A., ... & Kushima, R. (2014). STAT6 immunohistochemistry is helpful in the diagnosis of solitary fibrous tumors. *The American journal of surgical pathology*, *38*(4), 552-559.
5. Wushou, A., Jiang, Y. Z., Liu, Y. R., & Shao, Z. M. (2015). The demographic features, clinicopathologic characteristics, treatment outcome and disease-specific prognostic factors of solitary fibrous tumor: a population-based analysis. *Oncotarget*, *6*(39), 41875-41883.