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Clinical case / Caso clínico

# Solitary Fibrous Tumor of the Pleura: Clinical Case Report

## Tumor fibroso solitario de la pleura: presentación de un caso clínico

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**ABSTRACT**

**Introduction:** Solitary fibrous tumors (SFT) of the pleura are a rare neoplasm that originates from mesenchymal cells underlying the pleura. Its clinical course is usually indolent until it reaches large dimensions, producing symptoms due to compression. This clinical case provides valuable evidence on the effective diagnosis and management of this rare tumor. It also highlights the usefulness of molecular tools and oncological surveillance. **Clinical case:** We present a clinical case of a 53-year-old female patient diagnosed with SFT of the pleura, treated by surgical resection and followed up for 6 years. **Results:** The patient presented with cough and dyspnea. Chest computed tomography revealed a 23 cm left pleural mass. Histopathological study confirmed SFT with intermediate risk of recurrence. No recurrence was evident in subsequent follow-up. **Conclusions:** SFT of the pleura requires early diagnosis, complete surgical resection, and follow-up. Stratification of recurrence risk is essential for clinical management.

**Keywords:** Solitary fibrous tumor of the pleura, Pleural neoplasm, Thoracic surgery.

**RESUMEN**

**Introducción:** El tumor fibroso solitario pleural es una neoplasia rara que se origina de células mesenquimatosas subyacentes a la pleura. Su curso clínico suele ser indolente hasta alcanzar grandes dimensiones, y genera síntomas por compresión. El presente caso clínico aporta evidencia valiosa sobre el diagnóstico y manejo efectivo de este tumor raro. Además, resalta la utilidad de herramientas moleculares y la vigilancia oncológica. **Caso clínico:** Se presenta un caso clínico de una paciente femenina de 53 años con diagnóstico de tumor fibroso solitario pleural, tratada mediante resección quirúrgica y con seguimiento durante seis años. **Resultados:** La paciente presentó tos y disnea. La tomografía computarizada de tórax reveló una masa pleural izquierda de 23 cm. El estudio histopatológico confirmó un tumor fibroso solitario con riesgo intermedio de recurrencia. No se evidenció recidiva en controles posteriores. **Conclusiones:** El tumor fibroso solitario de pleura requiere diagnóstico temprano, resección quirúrgica completa y seguimiento. La estratificación del riesgo de recurrencia es esencial para el manejo clínico.

**Palabras clave:** tumor fibroso solitario de la pleura, neoplasia pleural, cirugía torácica.

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## 1. Introduction

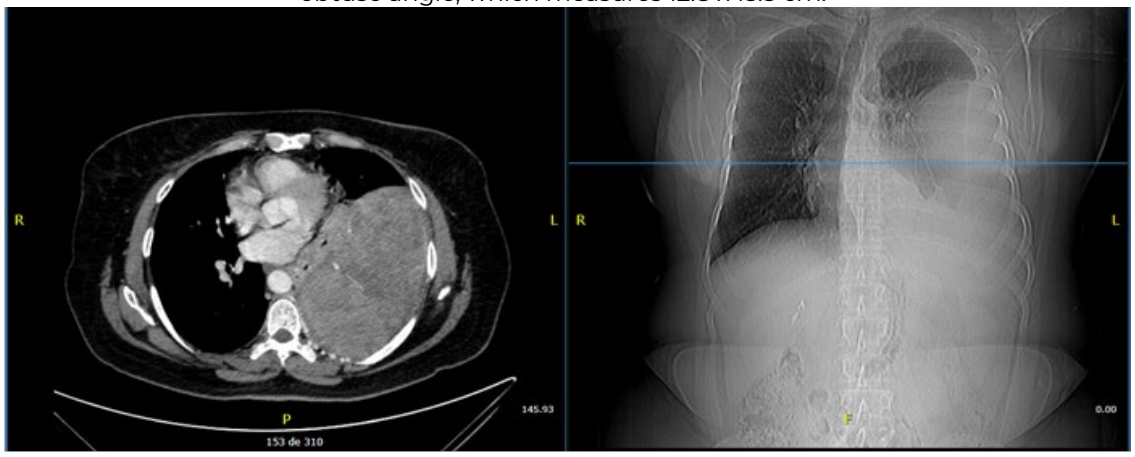
Solitary fibrous tumors (SFT) are a rare neoplasm of mesenchymal origin, accounting for less than 2% of all soft tissue tumors, with an estimated annual incidence of one case per million people. It occurs predominantly in adults between the fifth and sixth decades of life, with no gender preference [1]. The most common locations are deep somatic soft tissues and body cavities (especially the pleura and abdominal cavity) [2]. The World Health Organization's Classification of Tumors of Soft Tissue and Bone, fifth edition (WHO, 2020), categorizes it within tumors of fibroblastic origin, avoiding the terms "typical" or "malignant," given that even cases with benign-appearing histology can progress. The risk of recurrence or metastasis has been reported to range from 5% to 10%, typically to the lungs, liver, and bone [2]. The estimated 5- and 10-year overall survival rates are 89% and 73%, respectively, and the 5- and 10-year recurrence-free survival rates are 74% and 55%, highlighting the importance of long-term surveillance [2,3].

Through the presentation of a clinical case of a large pleural tumor, the application of histopathological and molecular criteria that confirm the diagnosis is addressed, and the inclusion of the mDemicco scale for metastasis risk stratification reinforces the importance of prolonged surveillance. In essence, this case provides evidence of the usefulness of these diagnostic and follow-up tools, which is of great value to the medical community, given the rarity of the disease.

## 2. Clinical case

A 53-year-old female patient presented with a history of hypothyroidism treated with levothyroxine 100 mcg daily and stage IA2 cervical cancer treated with radical hysterectomy in 2005. There was no known family history of cancer. Denied smoking or exposure to environmental toxins. She presented to the Pneumology Department in May 2019 with a nonproductive cough lasting more than three months and progressive dyspnea. Initial physical examination revealed a respiratory rate of 22 rpm, oxygen saturation of 95% on room air, hypoventilation in the left lung base on pulmonary auscultation, and dullness to percussion in the left basal region. Tumor marker results were as follows: CYFRA 21-1: 1.85 ng/ml; NSE: 19.51 ng/ml. Pulmonary function tests evidenced a moderate restrictive pattern with no response to bronchodilators. Contrast-enhanced chest CT scan showed a heterogeneous extrapulmonary mass measuring 12.8 x 15.3 cm with a broad base on the left parietal pleura and no adenomegaly (Figure 1).

**Figure 1.** Contrast-enhanced chest CT scan. Left extrapulmonary heterogeneous lesion with an obtuse angle, which measures 12.8 x 15.3 cm.



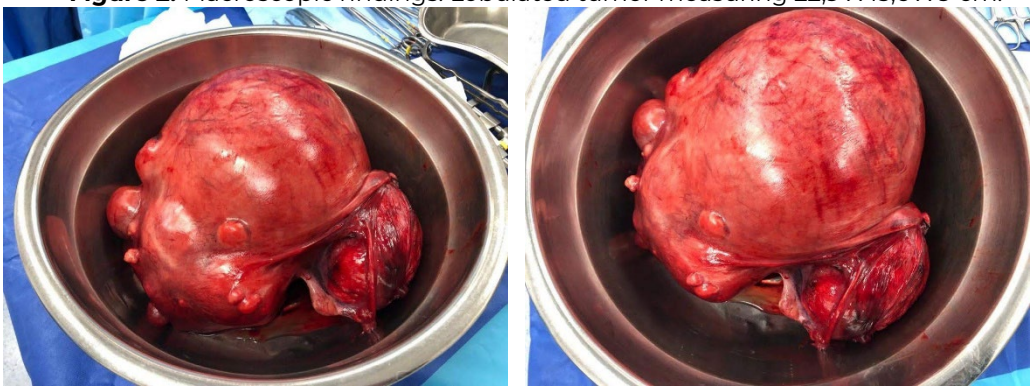
A CT-guided percutaneous biopsy was performed, which showed the presence of spindle cells arranged in a storiform pattern. This histological pattern is classic in soft tissue pathology and points the differential diagnosis mainly toward tumors such as solitary fibrous tumor, dermatofibrosarcoma protuberans, and undifferentiated pleomorphic sarcoma. Immunohistochemical analysis showed positivity for BCL-2 and CD34 markers. These results, together with the clinical-imaging correlation, confirmed the diagnosis of solitary fibrous tumor. Consequently, the proposed therapeutic approach was surgical resection. A left thoracotomy was performed with complete resection of a 23 cm lobulated tumor originating in the parietal pleura without pulmonary or bone invasion.

The pathological report revealed spindle cells arranged in a storiform pattern with minimal nuclear atypia and moderate mitotic activity (5/10 fields), which is a common pattern in SFT, as well as positivity for CD34 and BCL2 immunophenotypes of SFT. Although the absence of the STAT6 marker is a limitation of the study, the combination of the other histological findings and positivity for these markers, in the context of a pleural tumor, is highly suggestive. It is important to note that dermatofibrosarcoma protuberans (DFSP) was ruled out because, in addition to being a neoplasm that usually originates in the dermis and subcutaneous tissue, it presents an infiltrative honeycomb pattern and low mitotic activity, characteristics that were not observed in this case. Furthermore, DFSP usually expresses CD34 but lacks the histological pattern characteristic of SFT ("star" or "made of rag" pattern, hemangiopericytoma-like branching vessels, and areas of hyalinized collagen), which allowed it to be excluded in correlation with the pleural location of the tumor.

Pleomorphic undifferentiated sarcoma (formerly known as malignant fibrous histiocytoma) was ruled out based on the immunohistochemistry result, as this type of sarcoma typically does not express CD34 or BCL2, whereas in the present case, both proteins were positive, a finding characteristic of solitary fibrous tumor. Additionally, pleomorphic sarcoma usually shows marked cellular pleomorphism, abundant atypia, and a high mitotic index; features that were absent in the patient's histological evaluation.

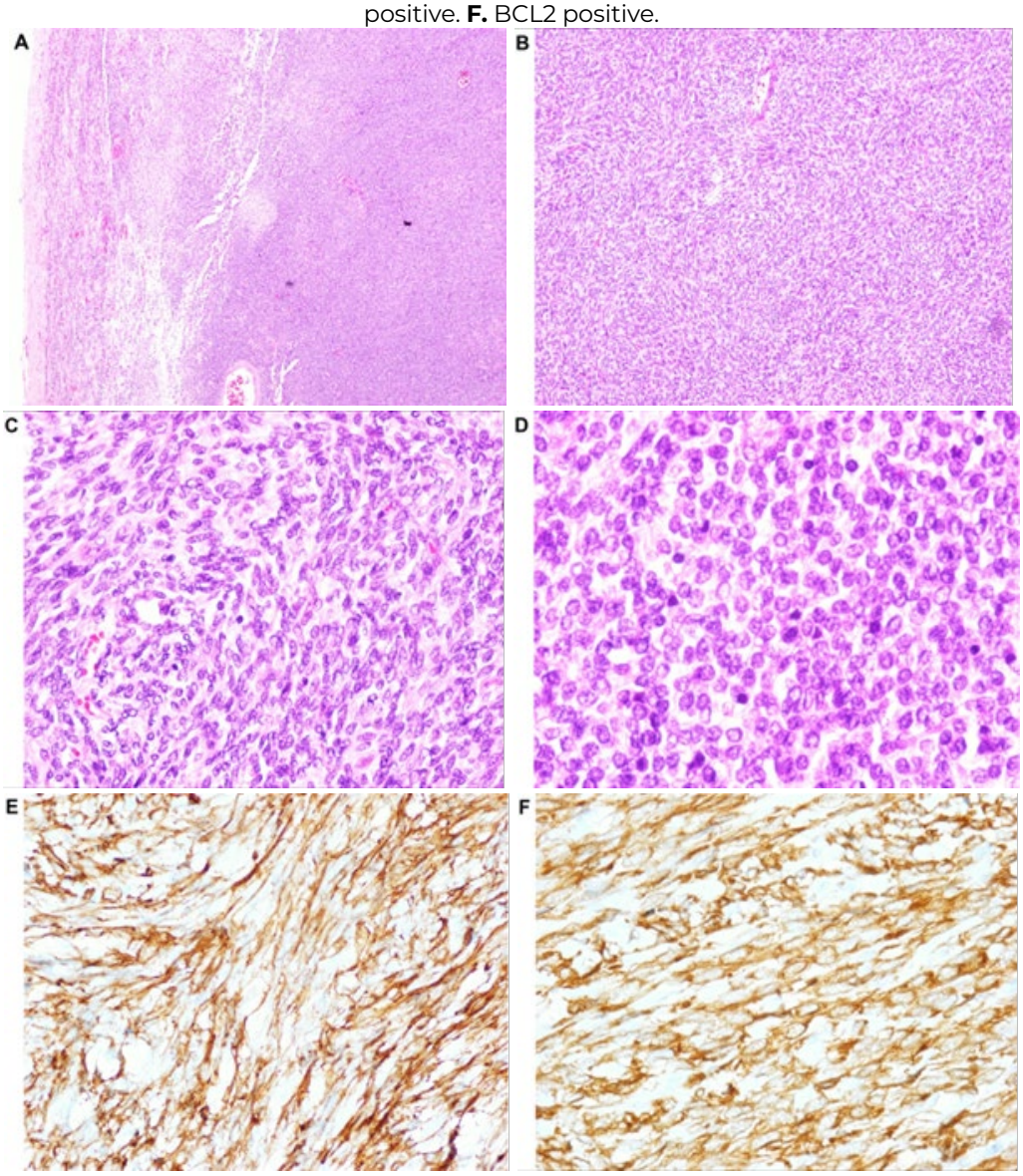
The pleural location, the exophytic size of 23 cm, and the resected block specimen without macroscopic infiltration favor a pleural SFT, consistent with the histological and immunohistochemical findings already described.

**Figure 2.** Macroscopic findings. Lobulated tumor measuring 22,9 X 15,6 X 9 cm.



Risk stratification for recurrence using the mDemicco scale yielded a score of 5, classifying the patient as intermediate risk. Adjuvant treatment was not indicated given the negative surgical margins and intermediate risk stratification. The patient remains under clinical follow-up with serial chest CT scans. At six years post-operatively, the patient remained asymptomatic with stable respiratory function tests and no evidence of recurrence. The patient demonstrated good adherence to follow-up and no adverse events related to surgical treatment were observed.

**Figure 3.** Histological findings. **A.** Highly fusocellular vascularized, solid neoplasm (H&E, 5x magnification). **B.** Cells distributed in short fascicles with a storiform pattern; no areas of necrosis were identified (H&E, 10x magnification). **C.** Vesicular nuclei with inconspicuous nucleoli and mild nuclear atypia (H&E, 20x magnification). **D.** Atypical mitoses, 5/10 fields at 40x (H&E, 60x magnification). **E.** CD34 positive. **F.** BCL2 positive.




## 3. Discussion

Pleural solitary fibrous tumors (SFT) are rare, usually slow-growing mesenchymal neoplasms that can reach large dimensions before producing clinical symptoms, which are generally compressive [4]. The etiology remains unknown, and no association with environmental factors such as tobacco use or asbestos exposure has been demonstrated [3]. Diagnosis requires imaging studies combined with histopathological and immunohistochemical analysis. Complete surgical resection with negative margins is the treatment of choice, and there is a better prognosis when complete tumor resection with clear surgical margins is achieved. However, some cases may have an unpredictable course; therefore, long-term follow-up is mandatory [5].

Classic histopathological findings include randomly arranged spindle-shaped to ovoid cells, with branching blood vessels, thin deer antler-shaped walls, and prominent stromal collagen, taking into account the mitotic count and the percentage of tumor necrosis [2]. Nuclear expression of STAT6 has been shown to be highly specific for SFT, which is a direct consequence of the NAB2-STAT6 gene fusion and is detectable by immunohistochemistry. In retrospective studies evaluating localized disease, the fusion between exon 4 of NAB2 and exon 2 of STAT6 was the most frequent variant, which has been correlated with a thoracic location, older patient age, lower mitotic count, and better prognosis. On the other hand, the fusion between exon 6 of NAB2 and exon 16 of STAT6, identified as the second most common fusion, is associated with extrathoracic locations, young patients, and a worse prognosis [6].

In some studies, CD209 overexpression has shown a significant correlation with an unfavorable clinical prognosis [6]. Additionally, evaluation of the Ki-67 proliferation index is useful for predicting prognosis [7]. Other markers, although nonspecific, such as CD34, BCL-2, and CD99 expression, may also be present; however, CD34 may be negative in approximately 10%, while CD99 and BCL2 are often expressed in other neoplasms [8].

To predict the risk of recurrence, the mDemicco scale is used, based on four parameters such as age (>55 years: 1 point), tumor size (5-10 cm: 1 point, 10-15 cm: 2 points, and ≥ 15 cm: 3 points), mitotic index (1 to 3 mitoses per 10 fields: 1 point, and ≥ 4 mitoses per 10 fields: 2 points), and percentage of tumor necrosis (≥ 10%: 1 point). Based on this score, the risk is classified as low (0–3 points), intermediate (4–5 points), or high (6–7 points) [7,9]. Complete surgical resection is the cornerstone of treatment for SFT. However, postoperative management should be tailored individually, considering surgical margins and risk stratification score. Although data are limited, neoadjuvant chemotherapy and radiotherapy may be considered in advanced cases. In cases of intermediate to high risk with positive margins and unresectable/inoperable tumors, adjuvant RT is a reasonable option [10]. Likewise, the use of the tyrosine kinase inhibitor (TKI) pazopanib could be considered as first-line systemic treatment in these cases [6]. Without a doubt, all treatment decisions should be made by a multidisciplinary team.

## 4. Conclusion

This report highlights that early diagnosis, complete resection, and a structured follow-up plan are crucial for achieving favorable clinical outcomes in solitary fibrous tumors of the pleura. Although the tumor was large, the postoperative course was uneventful, and long-term follow-up showed no evidence of recurrence. These findings underscore the key role of a multidisciplinary approach and close monitoring in the management of this entity.

## 5. Abbreviations

BCL-2: B-cell lymphoma 2

CD34: Cluster of differentiation 34

CYFRA: Cytokeratin fragments 19

DFSP: Dermatofibrosarcoma protuberans

IA2: Islet Antigen-2

NAB2: Gen NGFI-A Binding Protein 2

NGFI-A Binding Protein 2

NSE: Neuronal-Specific Enolase

STAT6: Signal transducer and activator of transcription 6.

SFT: Solitary fibrous tumor

TKI: Tyrosine kinase inhibitor

## 6. Administrative information

### 6.1 Additional files

None.

### 6.2 Acknowledgments

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### 6.3 Contributions of the authors

- Conceptualization: Fernando Salazar Reinoso
- Project management: Iliana Encalada Valdivieso
- Supervision: Tannia Rivera Rivera
- Writing - draft/original: Iliana Encalada Valdivieso, Edwin Ross Rodriguez
- Writing, review, and editing: Iliana Encalada Valdivieso, Tannia Rivera Rivera, Fernando Salazar Reinoso, Edwin Ross Rodriguez
- All authors read and approved the final version of the manuscript.

### 6.4 Funding

This study did not receive funding from any external source.

### 6.5 Availability of data and materials

The clinical data used in this study are available upon request to the corresponding author, respecting patient confidentiality.

### 6.6 Consent for publication

The patient gave her informed consent for the publication of her clinical case and the use of her information for scientific and academic purposes.

### 6.7 Conflict of interest

The authors declare that they have no conflicts of interest.

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