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# Solitary Fibrous Tumors of the Chest: Case Series from a Portuguese Center

# Tumores Solitários Fibrosos do Tórax: Casuística de um Centro Português

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**ABSTRACT**

The solitary fibrous tumor is a rare mesenchymal tumor with predilection for the pleura. Most times it is an incidental diagnosis. Surgery is the recommended treatment. Most solitary fibrous tumors (SFTs) exhibit indolent behavior; however, 10% to 25% may experience recurrence. The aim of this retrospective analysis is to characterize their presentation, therapeutic approach and recurrence and was conducted at the Pulmonary Oncology Department in Unidade Local de Saúde Santa Maria, in Lisbon, Portugal, between May 2013 and December 2023. We identified nine patients, 66,7% male, median age 66 years. The average tumor size was 14.5 cm; 88.9% located in the pleura. Surgical intervention was proposed for eight patients, of whom seven underwent the procedure. Four patients had disease recurrence, three underwent new surgery and one died. One of these three patients underwent postoperative radiotherapy. Again, these three patients suffered a relapse, one underwent surgery and two started systemic therapy. At the time of cut-off, five patients were alive, and four were progression-free. The solitary fibrous tumor is a rare and underdiagnosed entity, which justifies the small number of patients, and its malignant potential is poorly recognized. Our findings are consistent with those reported in the existing literature; however, we observed a higher proportion of tumors exhibiting malignant characteristics. The high prevalence of recurrence is explained by the fact that only complex or previously recurrent cases are referred for discussion at our center.

**Keywords:** Solitary Fibrous Tumor, Pleural

**RESUMO**

O tumor fibroso solitário é um tumor mesenquimatoso raro com predileção pleural. Maioritariamente é um diagnóstico incidental. A cirurgia é o tratamento recomendado. Apesar da maioria dos tumores fibrosos solitários ter um comportamento indolente, 10% - 25% podem recidivar. Esta análise retrospetiva teve como objetivo caracterizar a apresentação, abordagem e recorrência dos tumores fibrosos solitários e foi realizada no Serviço de Pneumologia Oncológica da Unidade Local de Saúde Santa Maria, em Lisboa, Portugal, entre maio 2013 e dezembro 2023. Identificámos nove doentes, 66,7% do sexo masculino, com idade mediana de 66 anos. O tamanho médio dos tumores foi de 14,5 cm; 88,9% com localização pleural. Oito doentes foram propostos para cirurgia, sete realizaram o procedimento. Quatro doentes tiveram recidiva, três foram submetidos a uma nova cirurgia e um faleceu. Um desses três doentes foi submetido a radioterapia pós-operatória. Novamente, esses três doentes recidivaram, um foi submetido a cirurgia e dois iniciaram terapêutica sistémica. À data de *cut-off*, cinco doentes estavam vivos, incluindo quatro sem progressão da doença. O tumor fibroso solitário é uma entidade rara e subdiagnosticada, o que justifica o reduzido número de doentes, cujo potencial maligno é pouco conhecido. Os nossos dados são consistentes com a literatura prévia, mas existe maior proporção de tumores com características malignas. A alta recidiva é justificada pelo facto de apenas casos complexos ou recidivantes serem encaminhados para discussão no nosso centro.

**Palavras-chave:** Tumor Fibroso Solitário da Pleura

Solitary fibrous tumor (SFT) is a rare mesenchymal tumor, first described in 1931.[1] It corresponds to under 2% of soft-tissue masses,[2] with an annual incidence of one in a million.[3] It has a peak incidence in the fifth to sixth decades (older for pleural SFT) and sex is not a risk factor.[4,5]

Solitary fibrous tumors range in size from 1 to 40 cm, with a median diameter of 5 to 8 cm. They can arise in virtually any part of the body, although approximately 30% occur in the pleura. The term 'pleural SFT' (or SFTP) is the nomenclature endorsed by the World Health Organization (WHO),[6] although these tumors have also been referred to by other names, such as benign localized mesothelioma and hemangiopericytoma. Most SFTPs are incidental findings. Patients can present chest pain, cough, dyspnea or compression symptoms.[4,5] Less than 10% patients may have paraneoplastic syndromes (hypertrophic osteoarthropathy or paraneoplastic hypoglycemia).[3]

Pleural SFT/SFTP typically presents as a well-circumscribed lesion, characterized by a fibrous pseudocapsule, a dense collagenous stroma, and thin-walled capillaries.[4,5] Solitary fibrous tumors (SFTs) may exhibit a spectrum of malignant behavior. Prognostic factors associated with poorer outcomes include larger tumor size (typically greater than 10 cm), hypercellularity, necrosis, hemorrhage, nuclear pleomorphism, and increased mitotic activity. Immunohistochemical markers commonly expressed in SFTs include CD34, CD99, BCL-2, and STAT6. Surgical excision is the treatment of choice. Radiotherapy may be considered for recurrent disease or in patients who are not suitable candidates for surgery. There is no well-established standard of care for metastatic disease. Chemotherapy or anti-angiogenic agents may be used and there is evidence regarding anthracycline-based regimens, dacarbazine and temozolomide.[2,3,5]

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Most SFTPs have an indolent behavior but recurrence may occur (10-year recurrence of 10% - 25%).[5] A systematic review recommends imaging evaluation at three and six months and then yearly for 10 years.[7]

The malignant potential of SFTs is often underestimated, making these analyses essential for understanding their behavior and facilitating timely referral for treatment. The aim of this retrospective observational analysis is to characterize presentation, therapeutic approach and recurrence of SFT and was conducted between May 2013 and December 2023 at the Pulmonary Oncology Department in Unidade Local de Saúde Santa Maria, Portugal. Patients were identified through our department databases and data was obtained from medical files.

We identified nine patients with SFT, 66,7% male, median age 66 years (55 - 88 years). Performance status (PS) was 0 in 44.4%. The average size of the tumor was 14.5 cm (2 - 30 cm). The predominant location was the pleura (88.9%) and a pulmonary SFT was identified. Diagnosis was incidental in 55.6%. Symptoms included dyspnea, chest pain and cough. Three patients (33.3%) presented pleural effusion. Paraneoplastic hypoglycemia was observed in two patients. Histologically, two patients showed > 4 mitosis/10HPF and four had necrosis. Demographic and disease characteristics are displayed on Table 1.

Eight patients were proposed for resection (Fig. 1). The excluded patient had PS 2 and was not considered fit for surgery; radiotherapy was proposed, but he died of unrelated causes. One patient refused treatment and died of disease progression. Surgery was performed in seven patients (77,8%), lobectomy was necessary in four. R0 was achieved in five.

Four patients had disease recurrence (mean time to relapse 47 months), two of them with previous R1 resection (microscopic residual tumor) which were also the two biggest tumors. All four had at least one malignant feature. Three patients underwent new surgery, and one died of disease progression. This time, two procedures required resection of the adjacent muscles/costal arches. One patient underwent post-operative radiotherapy.

All three who were reoperated had a new relapse (mean time to relapse 9.3 months); one underwent surgery and two started systemic therapy. One of these last two underwent multiple lines of therapy (temozolomide-bevacizumab, doxorubicin, pazopanib, trabectedin) and died before initiating gemcitabine. The other was still doing first-line therapy (doxorubicin) at the time of cut-off.

At cut-off, five patients were alive, three of them for more than five years since the diagnosis. There was no evidence of progressive disease in four.

In light of the present data, SFT is a rare entity which justifies the small number of patients identified. Nevertheless, these results represent a sample of a Portuguese tertiary hospital. It is an underdiagnosed entity whose malignant potential is poorly recognized and should prompt quick

Table 1 – Demographic and disease characteristics

| Characteristic | Value, n = 9 |
|---|---|
| Age (years) | |
| Median (range) | 66 (55 - 88) |
| Male, n (%) | 6 (66.7) |
| Performance status, n (%) | |
| 0 | 4 (44.4) |
| 1 | 4 (44.4) |
| 2 | 1 (11.1) |
| Size (cm) | |
| Medium (range) | 14.5 (2 - 30) |
| Location, n (%) | |
| Pleura | 8 (88.9) |
| Lung | 1 (11.1) |
| Signs and symptoms, n (%) | |
| Chest pain | 1 (11.1) |
| Dyspnea | 1 (11.1) |
| Cough | 1 (11.1) |
| Pleural effusion | 3 (33.3) |
| Hypoglycemia | 2 (22.2) |
| Tumor characteristics, n (%) | |
| > 4 mitosis/ 10 HPF | 2 (22.2) |
| Necrosis | 4 (44.4) |
| High cellularity | 1 (11.1) |
| Cellular atypia | 2 (22.2) |
| Bcl-2 | 9 (100) |
| CD34 | 6 (66.7) |
| CD99 | 4 (44.4) |
| Vimentin | 3 (33.3) |
| Surgical procedure, n (%) | |
| Tumor excision | 2 (22.2) |
| Wedge resection | 1 (11.1) |
| Lobectomy | 4 (44.4) |
| Surgical margins, n (%) | |
| R0 | 5 (55.6) |
| R1 | 2 (22.2) |
| Outcome, n (%) | |
| Death due to disease progression | 3 (33.3) |
| Death due to other causes | 1 (22.2) |
| Alive and disease-free | 4 (44.4) |
| Alive with disease | 1 (11.1) |

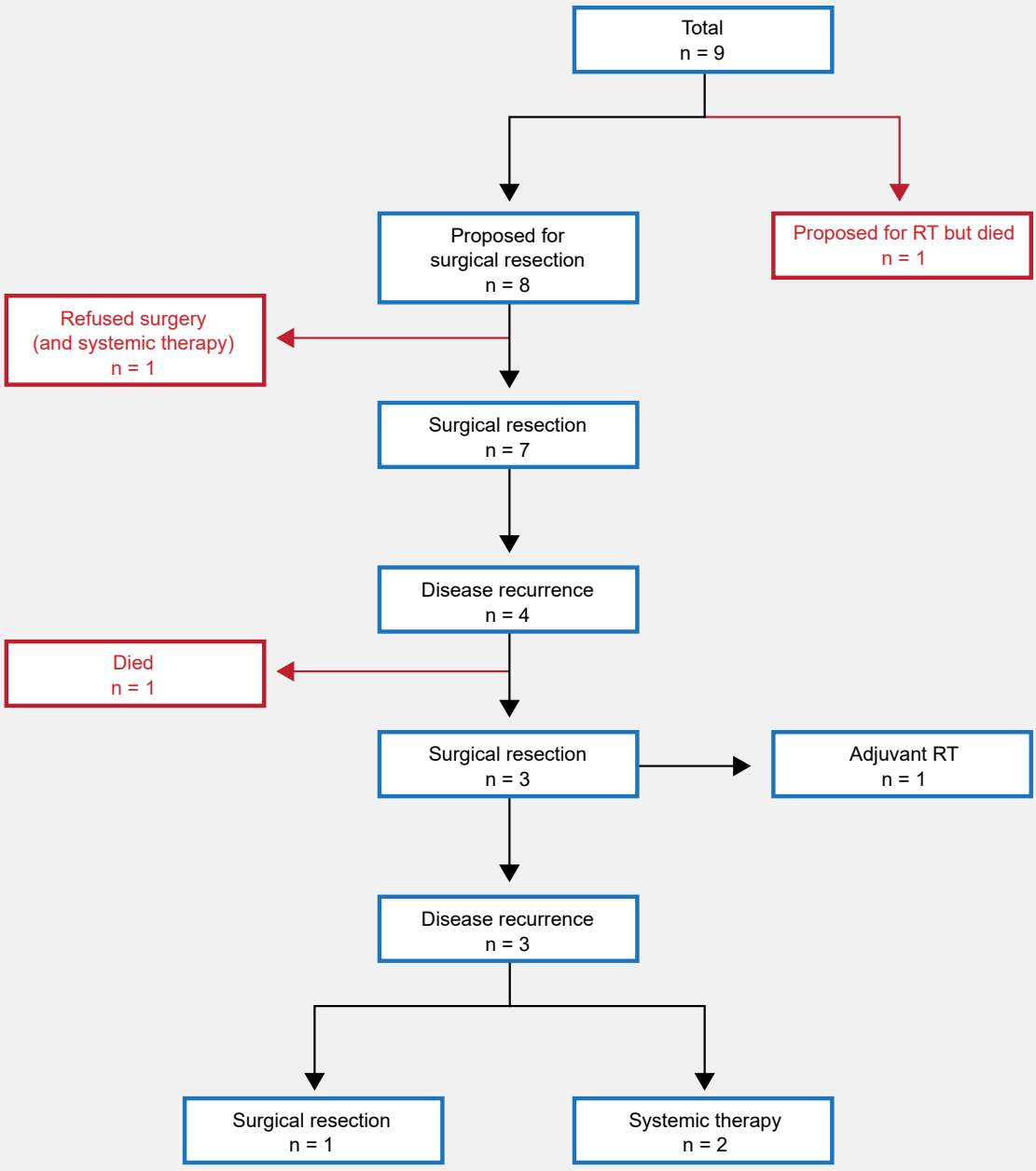


Figure 1 – Overview of the clinical course of patients

investigation and treatment. The average tumor size was 14.5 cm, which may be associated with a poorer prognosis.[8] The two largest tumors were observed in the patients who experienced multiple relapses and subsequently initiated systemic therapy. Most patients were asymptomatic; however, three presented with pleural effusion (33.3%), a slightly higher rate than reported in other studies,[8,9] and they all had at least one feature of malignant behavior. Paraneoplastic hypoglycemia was observed in two patients, indicating a higher prevalence than previously reported.[9,10]

Two patients exhibited a high mitotic rate, both of whom experienced disease recurrence. Necrosis was observed in four patients; among them, two had recurrent disease and one died after declining surgical intervention. Immunohistochemistry was consistent with the literature, with BCL-2 and CD34 having a higher positive rate. STAT6 is the most

sensitive and specific marker,[4,5] however, it was tested in only three patients.

Surgical excision is the recommended treatment. The two patients with R1 eventually had disease recurrence. Radiotherapy was used in a single patient with recurrence. Two patients received chemotherapy, with one still alive at the time of data cut-off. These small numbers do not allow for extrapolation, but most studies had unsatisfactory results.[5,9] A multidisciplinary approach is recommended. Recurrence rate was higher than expected (57.1% of patients undergoing surgery relapsed) but it is a small sample, and malignant characteristics were present in the majority of tumors.

Our study had the limitation of being a retrospective analysis with a small sample. Calculations of progression-free survival and overall survival would not provide statistically significant results. Another limitation is that some of these tumors may not reach our center, either due to the assumption that they are benign or because they are excised and subsequently managed through informal follow-up with the referring physician. Solitary fibrous tumors should be referred to and treated at specialized centers.

Future research should include larger and multicenter cohorts and focus on expanding prognostic markers to better guide therapeutic approaches.

In conclusion, our findings align with previously published data, but in our small sample the majority of tumors had malignant characteristics. Most patients were asymptomatic, but some had pleural effusion or paraneoplastic syndromes. Surgery is the recommended approach and should be performed by an experienced team, but relapse is still a possibility, especially in R1 resection.

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The authors have declared that no AI tools were used during the preparation of this work.

## AUTHOR CONTRIBUTIONS

MJS: Data collection and analysis, writing of the manuscript.

FF: Conceptualization, supervision, critical review of the manuscript.

AM, ASV, DH, PA: Supervision, critical review of the manuscript.

All authors approved the final version to be published.

## PROTECTION OF HUMANS AND ANIMALS

The authors declare that the procedures were followed according to the regulations established by the Clinical Research and Ethics Committee and to the Helsinki Declaration of the World Medical Association updated in October 2024.

## DATA CONFIDENTIALITY

The authors declare having followed the protocols in use at their working center regarding patients' data publication.

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