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Clinicopathological features and long-term outcomes of pleural solitary fibrous tumors

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ABSTRACT

Pleural solitary fibrous tumors (PSFT) are rare mesenchymal neoplasms with heterogeneous biological behavior. Although most tumors are histologically benign, recurrence and distant metastasis may occur, complicating prognostic stratification. This retrospective study evaluated the clinicopathological characteristics and long-term outcomes of 47 patients who underwent surgical resection for PSFT between January 2008 and June 2023, with particular focus on tumor size, positron emission tomography–computed tomography (PET-CT)–derived maximum standardized uptake value (SUVmax), and Ki-67 proliferation index. The mean tumor diameter was 8.4 cm, with 31.9% of the cases presenting a tumor size of ≥10 cm. Malignant histology was identified in 14 patients (29.7%). PET-CT was available in 28 patients; SUVmax ≥2.5 was observed in all malignant tumors and in 35.0% of benign tumors (p = 0.004). Ki-67 data were available in 18 patients, and Ki-67 ≥10% was significantly more frequent in malignant tumors than in benign tumors (p = 0.005). Complete R0 resection was achieved in all patients. The 5-year overall survival rate was 89.3%, with lower survival in malignant tumors compared with benign tumors (78.6% vs 93.9%). Local recurrence was observed in one patient with benign histology, while brain metastasis occurred in a patient with malignant histology. These findings suggest that the clinical behavior of PSFT cannot be reliably predicted based solely on histopathological features or tumor size. Instead, they indicate that metabolic and proliferative markers may provide valuable prognostic insights, highlighting the necessity for long-term, risk-stratified follow-up for all patients.

Keywords: pleural solitary fibrous tumors, malignancy, SUVmax, Ki-67 index, survival

INTRODUCTION

The World Health Organization (WHO) classifies solitary fibrous tumors (SFT) as rare fibroblastic mesenchymal neoplasms of intermediate (rarely metastasizing) biological behavior in its

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classification of soft tissue tumors.[1] The pleura is the most common site of involvement, with an estimated incidence of 2.8 per 100,000 population.[2] SFT typically originate from the visceral pleura and, less commonly, from the parietal pleura.[3] Although they generally exhibit benign histological features, approximately 12% of pleural solitary fibrous tumors (PSFT) demonstrate malignant behavior and may present with local recurrence or distant metastasis.[4]

Several clinicopathological features have been proposed as indicators of malignancy, including advanced age, larger tumor size (≥10 cm), increased cellularity, elevated mitotic activity (≥4 per 10 high-power fields or >2 mitoses per 2 mm^2), nuclear pleomorphism, tumor necrosis, and infiltrative growth patterns.[5] However, despite these established criteria, predicting biological behavior remains challenging, and aggressive clinical courses have occasionally been reported even in tumors classified as histologically benign.[6-8]

Complete surgical resection with negative margins (R0) remains the cornerstone of treatment for PSFT. The role of adjuvant radiotherapy or chemotherapy remains controversial and is generally reserved for unresectable or recurrent disease, as complete resection alone is associated with favorable long-term outcomes in most patients.[6]

Beyond conventional histopathological parameters, metabolic and proliferative markers such as maximum standardized uptake value (SUVmax) on positron emission tomography–computed tomography (PET-CT) and the Ki-67 proliferation index have been investigated as potential indicators of tumor aggressiveness in SFT.[9-11]

In this context, the present study aimed to evaluate the clinicopathological characteristics and long-term outcomes of patients with PSFT, with particular emphasis on the relationship between histological classification, tumor size, SUVmax, and Ki-67 proliferation index.

MATERIALS AND METHODS

Study design and patient selection

This single-center, retrospective observational study included 47 patients who underwent surgical resection for PSFT at the Department of Thoracic Surgery of our institution between January 2008 and June 2023. The study was approved by the institutional ethics committee (Republic of Turkey University of Health Sciences Istanbul Sureyyapasa Chest Diseases and Thoracic Surgery Research and Training Hospital Scientific Research Committee, approval number, July 25, 2022–300). All procedures were conducted in accordance with ethical principles of Declaration of Helsinki (1964) and its subsequent amendments, including the 2024 revision. Due to the retrospective nature of the study, the requirement for written informed consent was waived.

Patients' records were retrospectively reviewed to obtain demographic characteristics, presenting symptoms, tumor size and location, surgical approach, type of resection, tissue of origin, recurrence status, in-hospital mortality, follow-up duration, and survival outcomes.

PET-CT–derived SUVmax and Ki-67 proliferation index were analyzed in patients with available data. These parameters were not part of the mandatory inclusion criteria and were evaluated only when recorded in the medical files. Patients eligible for this study were those aged 18 years or older who had undergone surgery for primary PSFT with a confirmed pathological diagnosis. We excluded any cases that showed negative signal transducer and activator of transcription (STAT) 6 expression.

Preoperative diagnostic assessment

All patients underwent preoperative thorax CT to evaluate tumor size, anatomical location, and involvement of adjacent structures. Preoperative evaluation was primarily guided by thorax CT

findings. PET-CT scans were performed based on clinical indications at the time of diagnosis, particularly in cases where CT findings suggested a suspicion of primary lung malignancy; as such, PET-CT was not routinely utilized for the entire cohort.

Contrast-enhanced thorax CT was not routinely performed. In our institutional practice, it was selectively obtained when vascular or mediastinal invasion was suspected or when additional anatomical information was required for surgical planning. In the absence of suspected invasion, non-contrast thorax CT was considered sufficient for evaluating tumor size, location, and resectability.

In cases where radiological evaluation suggested a benign mesenchymal tumor, preoperative tissue sampling was typically avoided. As a result, transthoracic needle aspiration biopsy (TTNAB) was not implemented as a routine procedure. TTNAB was reserved for a select group of patients, specifically those where primary lung malignancy was suspected or those presenting with large-scale lesions. While imaging findings raised suspicion for PSFT in several instances, the definitive diagnosis and evaluation of malignancy could only be established after surgical resection.

Surgical management and histopathological evaluation

Complete resection with negative surgical margins was achieved in all patients. The surgical approaches, ranging from en bloc tumor excision and wedge resection to more extensive pulmonary or extrapulmonary resections, were tailored to the size and localization of the tumor. To ensure complete resection, neighboring structures, such as the lung or mediastinal components, were resected in cases where invasion or dense adhesions were encountered. Intraoperative frozen-section examination was not routinely performed for every patient but was used selectively when it could influence the surgical strategy. This approach was particularly preferred in cases of suspected malignancy, or when assessing the need for extended resection based on the tumor's relationship with adjacent structures. Additionally, it served to provide intraoperative confirmation of surgical margins in specific instances.

Tumor origin (visceral or parietal pleura) was determined intraoperatively based on the site of pleural attachment and was supported by pathological findings when required.

Histopathological classification into benign and malignant PSFT was based on routine morphological criteria. Tumors were classified as malignant when one or more of the following features were present: hypercellularity, mitotic activity ≥4 mitoses per 10 high-power fields, nuclear pleomorphism (atypia), tumor necrosis, or an infiltrative growth pattern.

All specimens were evaluated by three experienced pathologists. The Ki-67 proliferation index was assessed as an additional marker of proliferative activity but was not used as the sole determinant for malignant classification.

None of the patients received adjuvant chemotherapy or radiotherapy.

Follow-up

Follow-up and survival data were retrieved from routine postoperative clinical examinations and the hospital's electronic database. Recurrence, whether local or distant, was diagnosed through pathological confirmation whenever feasible. In cases where histological verification was not possible, recurrence was identified based on CT scans and clinical-radiological patterns, as adjudicated by a multidisciplinary tumor board.

Postoperative follow-up consisted of chest radiography at 1, 3, and 6 months during the first postoperative year, followed by annual thorax CT examinations.

Statistical analysis

All statistical analyses were performed using JASP software (version 0.19.3, JASP Team;

https://jasp-stats.org). Follow-up time, local recurrence, metastasis, and overall survival were calculated from the date of surgical resection.

Overall survival was defined as the time from surgery to death from any cause or the last follow-up. Survival curves were generated using the Kaplan–Meier method, and group comparisons were performed with the log-rank test.

Associations between continuous variables and binary outcomes were evaluated using the Mann–Whitney *U* test, while categorical variables were compared using Fisher's exact test. A *p*-value of <0.05 was considered statistically significant.

RESULTS

Demographic and clinical characteristics

The study included a total of 47 patients with a mean age of 53.5 ± 14.3 years (range, 25–76). Of the participants, 57.4% (n = 27) were male and 42.6% (n = 20) were female. The most common presenting symptoms were dyspnea (31.9%, n = 15) and chest pain (25.5%, n = 12), while 17.0% (n = 8) of patients were entirely asymptomatic. Other clinical presentations included cough (17.0%), hemoptysis (4.3%), fever (2.1%), and fatigue (2.1%). A history of malignancy was present in three patients.

Diagnostic evaluation

Preoperative fiberoptic bronchoscopy was performed in 25 patients (53.2%), revealing endobronchial tumor involvement in 3 cases. Representative thoracic CT images are shown in Figure 1.

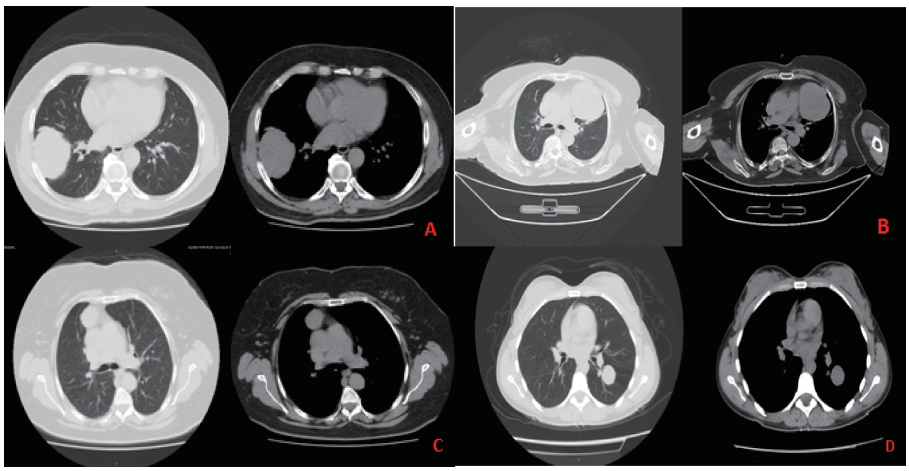

Fig. 1 Thoracic computed tomography (CT) findings of pleural solitary fibrous tumors
Axial CT images are shown in both lung window (left) and mediastinal window (right) settings.
Fig. 1A: A 70-year-old male patient with a well-defined solid mass broadly attached to the pleura in the right lower hemithorax, measuring 80 × 60 × 45 mm.
Fig. 1B: A 61-year-old female patient with an 80 × 80 × 100 mm mass in the lingular segment of the left upper lobe, appearing contiguous with and indistinguishable from the pericardium.
Fig. 1C: A 55-year-old female patient with a sharply demarcated, homogeneous soft-tissue density lesion measuring 41 × 32 mm located in the anterior mediastinum.
Fig. 1D: A 26-year-old female patient with a well-circumscribed nodule measuring 25.6 × 25 mm in the superior segment of the left lower lobe.

PET-CT imaging was available in 28 patients (59.5%). The mean SUVmax was 3.2 ± 4.8 (range, 0–22.1). Median and mean SUVmax values were 1.5 and 2.0 in benign tumors and 4.1 and 6.2 in malignant tumors, respectively. SUVmax values were significantly higher in the malignant group compared with the benign group (Mann–Whitney U test, $p = 0.004$).

Preoperative TTNAB was utilized selectively in only three patients. These individuals presented with lesions larger than 10 cm and imaging features suggestive of primary lung malignancy. In each case, the biopsy result was reported as compatible with a SFT.

Tumor characteristics and localization

Regarding tumor origin, 68.1% (n = 32) arose from the visceral pleura and 31.9% (n = 15) from the parietal pleura. The most common locations for visceral tumors were the right lower lobe (28.1%) and left lower lobe (28.1%). Regarding parietal pleural tumors, 28.1% were situated in the mediastinal pleura and 18.8% involved the chest wall.

The mean tumor diameter was 8.4 ± 5.2 cm, with a median value of 8 cm (range, 1.7–25 cm). Regarding size distribution, 68.1% of the tumors were smaller than 10 cm, while 31.9% were ≥10 cm in diameter (Table 1).

Table 1 Clinicopathological characteristics of patients with pleural solitary fibrous tumors (n = 47)

Variable	n	%
Demographic characteristic		
Male	27	57.4
Female	20	42.6
Age, mean ± SD (range), years	53.5 ± 14.3	25–76
Clinical presentation		
Dyspnea	15	31.9
Chest pain	12	25.5
Cough	8	17.0
Asymptomatic	8	17.0
Hemoptysis	2	4.3
Fever	1	2.1
Weakness	1	2.1
Tumor characteristic		
Visceral pleura origin	32	68.1
Parietal pleura origin	15	31.9
Tumor size <10 cm	32	68.1
Tumor size ≥10 cm	15	31.9
Tumor diameter, mean ± SD (range), cm	8.4 ± 5.2	1.7–25
Tumor localization		
Anatomical location (visceral, n = 32)		
Right upper lobe	7	21.9
Right middle lobe	1	3.1
Right lower lobe	9	28.1
Left upper lobe	6	18.8
Left lower lobe	9	28.1

Anatomical location (parietal, n = 15)		
Mediastinal pleura	9	28.1
Chest wall	6	18.8
Surgical approach		
Thoracotomy	34	72.3
VATS	12	25.5
Median sternotomy	1	2.1
Surgical procedure		
En bloc mass excision	31	65.9
Wedge resection	13	27.7
Left pneumonectomy	2	4.3
Left lower lobectomy	1	2.1
Pathological finding		
Benign	33	70.3
Malignant	14	29.7
Malignancy in tumors ≥10 cm	5	35.7
Outcomes		
Recurrence	2	4.3
Alive	42	89.3
Deceased	5	10.7
Overall survival, mean ± SD (range), months	108.2 ± 67.5	0.2–206

SD: standard deviation
VATS: video-assisted thoracoscopic surgery

The macroscopic appearances of the surgically resected PSFT are presented in Figure 2. Additionally, synchronous lung cancer was identified intraoperatively in two patients.

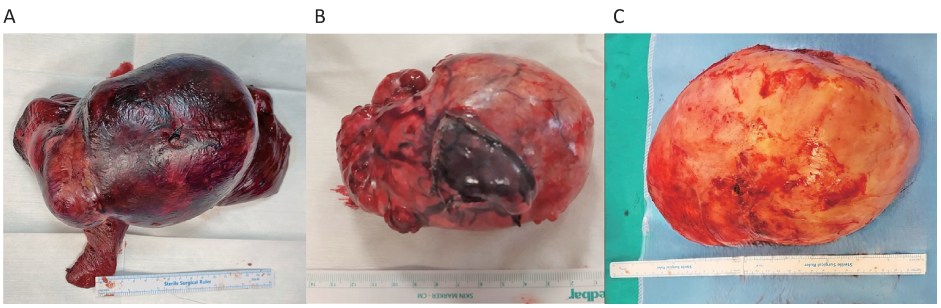

Fig. 2 Macroscopic appearance and clinicopathological characteristics of surgically resected solitary fibrous tumors

Fig. 2A: A 15-cm malignant solitary fibrous tumor in a 53-year-old male patient, presenting with dyspnea and originating from the left lower lobe and treated with left pneumonectomy.

Fig. 2B: A 10-cm benign solitary fibrous tumor in a 61-year-old female patient, presenting with back pain and located in the lingular segment of the left upper lobe and removed by wedge resection.

Fig. 2C: A 24-cm benign solitary fibrous tumor in a 44-year-old female patient presenting with dyspnea, located in the right lower lobe and treated with en bloc mass excision.

Surgical approach and procedures

Regarding the surgical approach, thoracotomy was performed in 72.3% (n = 34) of patients, while video-assisted thoracoscopic surgery was utilized in 25.5% (n = 12), and median sternotomy in 2.1% (n = 1). The most frequent surgical procedure was en-bloc mass excision (65.9%, n = 31), followed by wedge resection (27.7%, n = 13). Two patients (4.3%) underwent left pneumonectomy, and one patient (2.1%) underwent left lower lobectomy.

Intraoperative frozen section analysis was performed in 15 cases. In 10 of these patients, the analysis was requested to differentiate between benign and malignant lesions; in all such instances, the results were reported as consistent with a benign mesenchymal tumor. In the remaining five cases, frozen section evaluation was utilized to assess surgical margin status. Since negative margins were confirmed in all five, no additional or extended resection was required.

Pathological findings and malignancy analysis

Histopathological examination classified 70.3% (n = 33) of the patients as benign and 29.7% (n = 14) as malignant. The malignancy rate in tumors ≥10 cm was found to be 35.7%. When comparing benign and malignant cases, no statistically significant differences were observed regarding age, sex, tumor diameter, or pleural origin ($p > 0.05$). However, the mean SUVmax value was significantly higher in malignant tumors (6.2 ± 6.4) compared to benign tumors (2.0 ± 2.5; $p = 0.004$). Similarly, the Ki-67 proliferation index was significantly higher in the malignant group (16.6% ± 12.1%) than in the benign group (4.2% ± 4.1%; $p = 0.005$; Table 2).

Table 2 Comparison of clinicopathological characteristics between benign and malignant pleural solitary fibrous tumors

Variable	Benign (n = 33)	Malignant (n = 14)	*p* value
Age, mean ± SD, years	54.4 ± 13.8	51.3 ± 15.8	0.601
Male sex, n (%)	18 (54.5)	9 (64.3)	0.748
Tumor diameter, mean ± SD, cm	8.1 ± 4.7	9.1 ± 6.2	0.861
Tumor size ≥10 cm, n (%)	10 (30.3)	5 (35.7)	0.742
Visceral pleura origin, n (%)	22 (66.7)	10 (71.4)	1.000
SUVmax, mean ± SD	2.0 ± 2.5	6.2 ± 6.4	**0.004**
Ki-67, mean ± SD, %	4.2 ± 4.1	16.6 ± 12.1	**0.005**
Recurrence, n (%)	1 (3.0)	1 (7.1)	0.512
Follow-up duration, mean (range), months	42.5 ± 51.4 (0–178)	26.5 ± 25.8 (1–72)	0.798
Overall survival, mean ± SD, months	113.7 ± 66.1	95.2 ± 71.5	0.128

SD: standard deviation

SUVmax: maximum standardized uptake value

Statistical analysis: Mann–Whitney *U* test was used for continuous variables, Fisher's exact test for categorical variables, and the log-rank test for comparison of overall survival.

Follow-up and survival outcomes

The median follow-up period for the entire cohort was 37.7 months (range, 6 days–178 months). The mean follow-up duration was 42.5 ± 51.4 months (range, 6 days–178 months) in the benign group and 26.5 ± 25.8 months (range, 1–72) in the malignant group; no statistically significant difference was observed between the two groups regarding follow-up duration (p =

0.798). During the follow-up period, recurrence occurred in two patients (4.3%). Local recurrence developed at 32 months in one patient with benign histology (3.0%), while brain metastasis was identified in the third postoperative year in one patient with malignant histology (7.1%; p = 0.512; Table 2).

Postoperative complications were observed in two patients (4.3%), consisting of pneumonia and cardiac arrhythmia. No operative mortality occurred; however, in-hospital mortality was recorded in two patients (4.3%).

The estimated mean overall survival using the Kaplan–Meier method was 108.2 ± 67.5 months. The median overall survival (OS) was not reached during the follow-up period. Kaplan–Meier survival analysis showed no significant difference in OS between tumors <10 cm and ≥10 cm (log-rank p = 0.602; Figure 3).

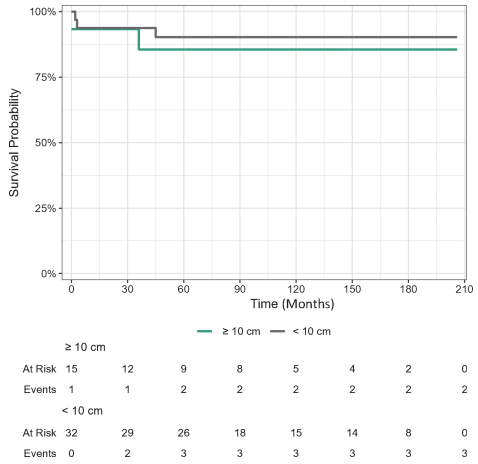

Fig. 3 Kaplan–Meier overall survival curves according to tumor size (<10 cm vs ≥10 cm)

Mean OS was found to be 113.7 ± 66.1 months in benign cases and 95.2 ± 71.5 months in malignant cases (p = 0.128; Figure 4), suggesting a trend toward lower survival in patients with malignant PSFT.

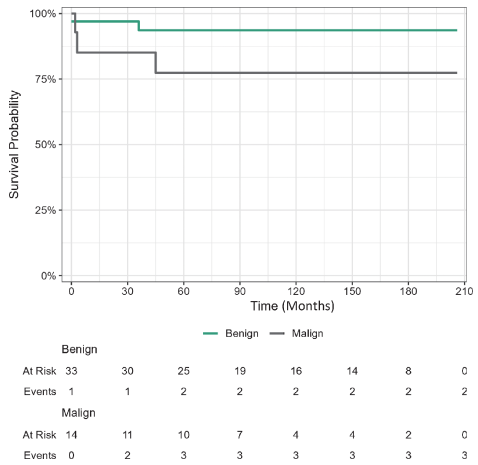

Fig. 4 Kaplan–Meier overall survival curves according to histopathological classification (benign versus malignant)

During the study period, 89.3% (n = 42) of the patients remained alive, while mortality was observed in 10.7% (n = 5). One patient died on the 6th postoperative day due to non-tumor-related postoperative complications. The remaining four deaths were considered tumor-related (80%): one patient due to distant metastatic disease, one following reoperation for local recurrence on the 8th postoperative day, and two patients with malignant PSFT due to disease progression at 2 and 3 months postoperatively, respectively.

Comparative analysis between benign and malignant tumors showed no significant differences in terms of age, gender distribution, tumor size, pleural origin, or overall survival. Conversely, SUVmax and Ki-67 values were significantly higher in malignant tumors ($p = 0.004$ and $p = 0.005$, respectively; Table 2).

DISCUSSION

PSFT are rare mesenchymal neoplasms characterized by a highly unpredictable clinical course. This study, based on a cohort of 47 patients, provides significant insights into the clinical behavior and long-term surgical outcomes of both benign and malignant variants. Although complete surgical resection is well-established as a primary factor for favorable long-term prognosis, the occurrence of recurrence, even in a case histologically classified as benign, highlights the persistent complexities and challenges that remain in the management of this entity.

In the literature, PSFT are typically described as gender-neutral neoplasms that manifest during the fifth and sixth decades of life.[4,12] Our study reflects these established demographic characteristics, showing age and gender distributions aligned with previous reports.[4,13] The primary clinical symptoms, most notably dyspnea and chest pain, are largely attributed to the mass effect and compression of adjacent structures by tumors that have reached substantial dimensions.[4] However, the occurrence of asymptomatic cases detected incidentally in our cohort highlights the insidious growth pattern inherent to these tumors.[4,14] This clinical feature explains why large tumor volumes are frequently identified at diagnosis, a finding that remains consistent with global literature and other single-center studies.[4,15]

A significant preoperative challenge is the difficulty in distinguishing between benign and malignant variants, as PSFT can radiologically mimic primary lung carcinomas.[14,16,17] Because definitive diagnosis is only achieved through comprehensive histopathological and immunohistochemical evaluation, surgical strategy is guided by tumor resectability and anatomical localization rather than preoperative tissue sampling.

Analysis of tumor characteristics reveals that while a majority of PSFT originate from the visceral pleura, parietal pleura localization remains a clinically significant consideration.[12,18,19] In our series, the predominance of visceral pleura-derived tumors reinforces this established anatomical trend. Furthermore, the average tumor diameter observed in our cohort confirms that these neoplasms frequently attain substantial macroscopic dimensions prior to detection.[15,18] Our finding of significantly elevated SUVmax values in malignant cases compared to benign ones underscores the role of PET-CT as a crucial adjunctive tool for malignancy stratification and predicting recurrence risk, aligning with current literature.[19,20]

Achieving a complete R0 resection remains the gold standard for curative therapy and is essential for minimizing the risk of recurrence in the management of PSFT.[6,7] In our series, while thoracotomy was the preferred approach in the majority of patients (72.3%), the utilization of video-assisted thoracoscopic surgery in 25.5% reflects the feasibility and safety of minimally invasive techniques in appropriately selected cases.[4,14]

Risk classification models proposed by Demicco et al are widely utilized to evaluate the

malignant potential and recurrence probability of PSFT.[11,18] These models identify parameters such as age, tumor size, and mitotic activity as the primary determinants of disease-specific mortality.[11,18,21] Although tumors larger than 10 cm are frequently associated with a higher likelihood of malignancy, our findings indicate that tumor size alone should not be regarded as an absolute prognostic marker. Consistent with existing literature, it is important to acknowledge that smaller tumors may follow an aggressive clinical course, whereas larger lesions can occasionally exhibit more indolent progression.[12,22] Consequently, these outcomes emphasize that comprehensive histopathological evaluation provides a more reliable prognostic assessment than relying solely on tumor size as an independent predictor.[22,23]

In pathological assessments, a Ki-67 proliferation index exceeding 10% is established in the literature as a significant independent risk factor for recurrence.[15] The findings from our pathological analysis, which showed significantly higher Ki-67 levels in malignant tumors compared to benign cases, underscore the critical role of proliferative activity in determining the biological aggressiveness of these neoplasms.[9,18,19]

Our recurrence and survival outcomes indicate that while PSFT generally follow an indolent clinical course, the risk of late recurrence and delayed adverse events remains a persistent concern. Although we observed a relatively low recurrence rate of 4.3%, the development of recurrence in a histologically benign case clearly demonstrates that initial pathology does not always guarantee an uneventful clinical trajectory.[18] Since the literature reports that recurrences can manifest as late as 10 or even 20 years after surgery, lifelong oncological surveillance is a vital necessity, particularly for cases with high malignant potential.[8,18,24,25]

SFT are recognized as biologically unified mesenchymal entities that exhibit comparable patterns of recurrence and metastasis regardless of their primary site.[21,26] This biological cohesion often results in a heterogeneous and unpredictable clinical behavior.[27,28] Ultimately, acknowledging SFT as a unified entity underscores the critical necessity for standardized risk stratification and vigilant, lifelong surveillance.[21]

The survival outcomes in our study highlight the significant impact of biological aggressiveness on the long-term oncological trajectory of PSFT. The high survival rates achieved following complete R0 resection align with the prevailing consensus that these tumors typically follow an indolent clinical course.[7,18] Although malignant cases exhibited a trend toward shorter survival durations compared to benign ones, the lack of statistical significance underscores the inherent unpredictability of the disease's clinical behavior.[4,18] Similarly, the observation that tumor size did not result in a significant survival disadvantage suggests that PSFT prognosis should be evaluated through multifactorial risk models rather than isolated morphological parameters.[18,23] The fact that the majority of observed deaths were directly linked to tumor-related causes emphasizes the persistent risk associated with the disease's malignant potential, regardless of initial surgical success.[18,24] However, given the limited number of tumor-specific events, these findings should be viewed as descriptive characterizations of our patient cohort, further reinforcing the necessity for lifelong oncological surveillance.[8,18,24,25]

Despite the significant insights provided by our study, several inherent limitations must be acknowledged. Its retrospective design and single-center experience naturally constrain the broader generalizability of our findings to the general population, a common challenge given the rarity of these tumors. The limited cohort size may have particularly affected the statistical power of clinicopathological comparisons between benign and malignant subgroups. Additionally, the lack of complete data for metabolic and proliferative markers, such as PET-CT and Ki-67 levels for every patient, introduces the possibility of selection bias in our available-case analyses. Furthermore, as these tumors were historically classified as benign lesions, some patients were lost to early follow-up, which resulted in a mean follow-up duration shorter than that reported

in some larger series. However, since the vast majority of our patients remain alive, we believe this limitation primarily impacts long-term recurrence data rather than overall survival outcomes, further highlighting the necessity for diligent, lifelong surveillance of this unpredictable entity.

In conclusion, PSFT are rare mesenchymal neoplasms characterized by complex and occasionally unpredictable biological behaviors. Complete surgical resection, particularly when R0 margins are achieved, remains the gold standard of treatment, ensuring favorable long-term survival for the vast majority of patients. While parameters such as tumor size, proliferation indices, and metabolic activity offer valuable prognostic insights, no single criterion can definitively predict the future clinical trajectory of these tumors. The occurrence of recurrences even in histologically benign cases decades later underscores the absolute necessity for standardized, multidisciplinary, and lifelong oncological surveillance. Future research involving larger multi-center cohorts and next-generation molecular markers is essential to more accurately forecast the behavior of these rare neoplasms.

CONFLICTS OF INTEREST

All authors have no conflict of interest to report in relation to this study.

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