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Primary Sarcoma of the Lung – Prognostic Value of Clinicopathological Characteristics of 26 Cases

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Abstract. *Background/Aim: Primary sarcomas of the lung (PSL) represent a rare, largely unknown entity. We herein present a retrospective study of 26 patients diagnosed with PSL. Patients and Methods: For a period of 10 years, the records of patients from 5 centers were gathered and analyzed. Results: Median age at diagnosis was 61.96 years (range=31-75 years). Eight patients (33.33%) had mediastinal node invasion (MNI). From 17 patients (70.83%) with localized disease, 11 patients (64.70%) underwent surgery. Recurrence rate was 72.72%. Median disease-free interval was 15 months. The median overall survival (OS) of patients with metastatic disease was 4 months and 10 months for the whole population. Only surgery had an impact on survival. Conclusion: Prognosis of PSL is somber. The high proportion of patients with MNI at diagnosis may serve as an indication for surgical evaluation of mediastinum and raises the question whether patients with locoregional PSL should be treated with a more aggressive approach.*

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Primary sarcomas of the lung (PSL) constitute a heterogeneous group of rare malignant tumors, accounting for less than 0.5% of malignant lung tumors (1). They arise from mesenchymal tissue of the bronchial wall vessels or pulmonary stroma (2) and usually present in middle-aged to elderly individuals. Pathophysiological mechanisms and molecular pathways underlying PSL are largely unknown. One study has reported association with smoking, environmental pollution, industrial toxins, and chronic bronchitis (3). The most common histological subtypes described are synovial sarcoma, epithelioid hemangio-endothelioma, leiomyosarcoma, and malignant peripheral nerve sheath tumor (4) followed by undifferentiated pleomorphic sarcoma, liposarcoma, and rhabdomyosarcoma (5-7).

Diagnosis of PSL is always challenging due to the lack of clinical suspicion since primary lung carcinomas are 700 times more common than sarcomas (8). Symptoms are non-specific and the main diagnostic dilemma is whether the tumor is primary or metastatic, since sarcomas very commonly spread to the lungs. In most cases, the morphology with a carefully selected immuno-histochemical panel and molecular testing, will suffice in differentiating PSL from their histological mimickers (sarcomatoid lung carcinoma, lymphoma, and benign mesenchymal tumors).

In this study, we retrospectively analyzed the clinicopathological features of 25 cases of PSL diagnosed and managed in five different sarcoma treating centers.

Table I. *Patients characteristics and initial management.*

Nr	Gender	Age at diagnosis	Histotype	Stage at diagnosis	Neoadjuvant chemotherapy	Surgery	Margin status	Adjuvant chemotherapy
1	M	73	SS	Localized	N	Y	R0	N
2	M	73	SS	Localized	N	Y	R0	N
3	M	69	ES	Localized	N	Y	R0	N
4	M	75	AS	Metastatic	NR	Y	R1	N
5	M	34	SS	Localized	N	Y	R0	N
6	M	71	SS	Localized	N	Y	R0	N
7	F	71	SMARC4-deficient	Metastatic	NA	NA	NA	NA
8	M	62	SS	Localized	Y (AI)	N	NR	NR
9	F	71	UPS	Localized	N	Y	R0	Y
10	M	68	SS	Metastatic	NR	N	NR	NR
11	F	69	UPS	Localized	N	Y	R0	Y
12	F	48	UPS	Metastatic	NR	N	NR	NR
13	F	31	SS	Localized	N	Y	R0	N
14	M	64	SS	Localized	N	N	NR	NR
15	M	53	UPS	Metastatic	NR	N	NR	NR
16	F	47	UPS	Metastatic	NR	N	NR	NR
17	M	72	UPS	Localized	N	N	NR	NR
18	M	55	SS	Localized	N	N	NR	NR
19	M	70	UPS	Metastatic	NR	Y	R0	N
20	F	72	FBS	Metastatic	NR	N	NR	NR
21	M	48	UPS	Localized	N	N	NR	NR
22	M	68	UPS	Localized	N	Y	R0	Y
23	M	63	IFT	Localized	N	Y	R0	N
24	M	64	SS	Localized	N	N	NR	NR
25	M	58	UPS	Localized	N	Y	R2	N

M: Male; F: female; SS: synovial sarcoma; ES: epithelioid sarcoma; AS: angiosarcoma; UPS: undifferentiated pleomorphic sarcoma; IFT: inflammatory myofibroblastic tumor; N: no; NR: not related; NA: not available; Y: yes; AI: adriamycin/ifosfamide.

Patients and Methods

Selection of patients. Records of all patients 16 years old and above with a histological diagnosis of primary sarcoma of the lung in the five participating institutions, between June 2008 and December 2018, were retrospectively reviewed. We only included cases in which a primary location other than the lung was eliminated by imaging studies [computed tomography (CT) of the brain, thorax and abdomen]. The methods of this study comply with the current laws of Greece and the EU General Data Protection Regulation (GDPR).

Data collection. We collected clinical data of all patients including history, physical examination, hematological and biochemical tests and imaging studies [CT scan of the chest and CT or magnetic resonance imaging (MRI) scan of the abdomen and brain].

All systemic therapies were recorded in a pseudoanonymised database, including chemotherapy regimen, other systemic treatment and number of cycles. In addition, we recovered data on pathology, surgery including surgical margin status, radiation therapy, metastatic sites and clinical outcome. Tumor responses were recovered from the radiology reports of CT or MRI scans based on RECIST criteria version 1.1 (9).

Statistical analysis. Descriptive statistics were performed to evaluate patients' characteristics. For survival analysis, the Kaplan–Meier analysis and life-tables were used. Overall survival (OS) was defined as the time interval between the date of PSL diagnosis and the date of death or last follow-up. For patients with disease limited to the thorax at presentation who underwent surgical resection of their primary tumor, disease-free interval (DFI) was calculated from the date of surgery to the first known date of recurrence. Patients who were still alive were censored on the date of their last known follow-up visit. The log-rank test was applied for comparison between groups. Analysis was performed using SPSS v.24 (IBM Corp., Armonk, NY, USA).

Results

Clinical presentation. A total of 25 cases were identified from the files of the pathology, thoracic surgery or medical oncology department of each institution. The clinical features and the initial management are listed in Table I. Mean age at diagnosis (AD) was 61.96±12.26 years (range=31-75 years). The skewness and kurtosis for age at diagnosis distribution is –1.187 and 0.651, respectively. The majority of patients were males, with a sex ratio=18:7. From 17 patients with disease limited to the lung at initial presentation, primary tumor size was ≤5 cm in 9 patients (52.9%) and >5 cm in 8 patients (47.1%). Metastatic disease spread to the lungs (12), peritoneum (1), bone (4), pleura (4) and lymph nodes (1).

Staging of tumor at diagnosis was revised and patients were restaged according to the 8th edition of the American Joint Committee on Cancer (AJCC) system for lung cancer (10).

Tumor samples and histologic subtypes. Diagnostic tissue samples were obtained by surgical resection in 11 patients and by bronchial biopsy or CT-guided biopsy from the pulmonary lesion in the rest. Histological subtypes were synovial sarcoma (40%), undifferentiated pleomorphic sarcoma (40%), epithelial sarcoma (4%), angiosarcoma (4%), fibrosarcoma (4%), inflammatory fibroblastic tumor (4%) and SMARCA4-deficient sarcoma (4%).

Treatment and outcome. From the 17 patients (68%) with localized disease, 11 (70.6%) underwent surgery and six patients had extensive disease not surgically amenable. Surgical resection of the primary tumor was qualified as R0 (negative margins) in 9 cases and R2 (macroscopic residual tumor) in one case. Two patients with oligometastatic disease (confined to the lung) underwent surgery; R0 in one case and R1 in the other one. The patient with R2 resection exhibited progressive disease at first post-surgical evaluation and was excluded from recurrence analysis. With a median follow-up of 12 months (range=1-56 months) the recurrence rate was 72.72% (8/11). Median disease-free interval (DFI) was 15 months (95%CI=11.257-18.743), as shown in Figure 1A.

Actuarial cancer-specific survival at 3 years was 42% and median overall survival (OS) was 15 months (95%CI=11.056-18.944).

Three patients received adjuvant chemotherapy. One patient received neoadjuvant chemotherapy without subsequent surgical treatment due to progressive disease. All the remaining patients with recurrence received systemic therapies. From the four patients without relapse, one died due to surgical complications 2 months after surgery and a second patient died from causes unrelated to cancer 8 months after surgery.

Survival analysis data for the whole population and per stage, with a median follow-up of 10 (range=1-56 months), were available for 24 patients and are summarized in Table II.

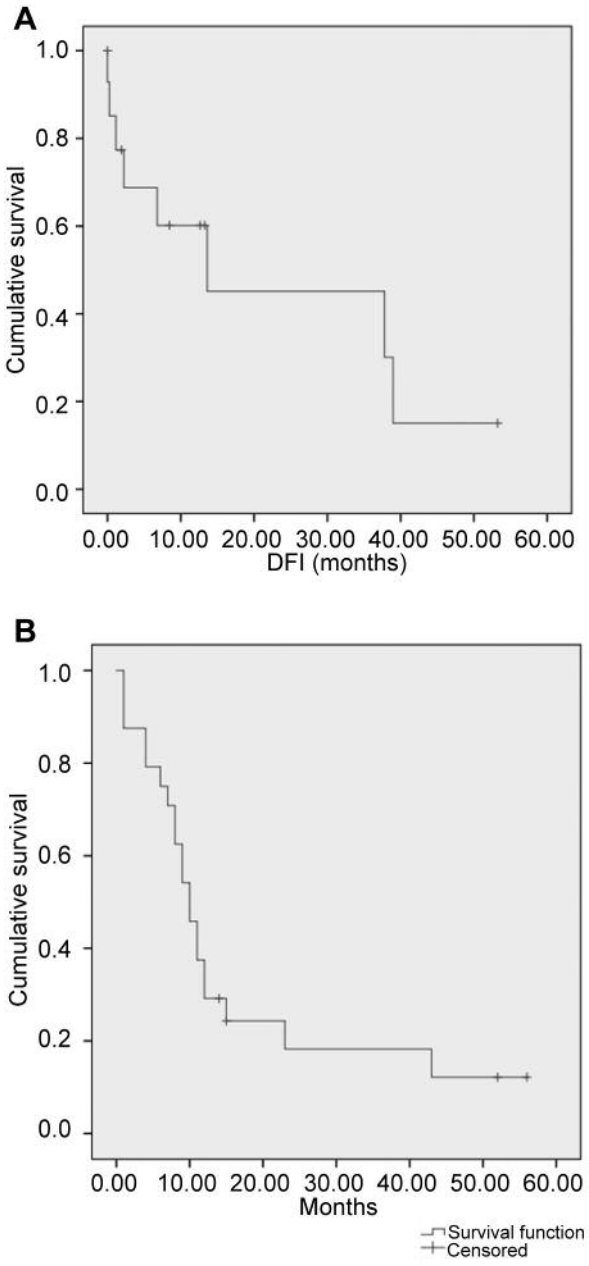

Figure 1. *Survival curves for overall survival (OS) and disease-free survival (DFS). A. Survival curve for disease free interval (DFI) of patients with resected localized disease. Median DFI: 15 months (95%CI=11.257-18.743). B. Survival curve for overall survival of the whole population. Median OS: 10 months (95%CI=7.608-12.392).*

Discussion

Soft tissue sarcomas (STS) are malignant neoplasms of mesenchymal origin that can arise in every part of the body. PSL are extremely rare since they account for less than 0.5% of all lung tumors (11). Due to their rarity, data on diagnosis and management are limited to case reports and series of cases (1-4, 8, 12-20). Therefore, no specific management guidelines have been established.

AD has been repeatedly reported to vary from 18 to 85 years, with a mean value of 45 to 63 years according to different series (2, 3, 15, 20). The mean AD of our series was 62 years (range=31-75 years) and the majority of patients were male, with a sex ratio of 2.57:1. This differs considerably from the ratios reported by earlier series. Repetitively until now, reported sex ratios indicated a weak trend to male sex of up to 1.8 (4). The analysis from the SEER database sets indicated a sex ratio of 1.25:1, while

other smaller series (including the most recent one of Golota *et al*. in 2018) (2) found a trend to the female sex.

Histopathological characterization of sarcomas has evolved through the last decades and the World Health Organization (WHO) Classification of Tumours of the Soft Tissue and Bone is continuously being updated (21). This evolution may explain the notorious incongruence in different histotype frequencies between the series. Leiomyosarcomas and myofibroblastic tumors are the most frequent histotypes reported (16% and 15%, respectively) while synovial sarcoma (12%) is the third most common subtype (18). However, synovial sarcoma has been found to be much rarer in earlier studies, and only Keel *et al*. (14) have found a higher rate of 23% of synovial sarcoma (6 cases out of 26). Unexpectedly, in our data set no leiomyosarcomas were found, whereas synovial sarcoma and undifferentiated pleomorphic sarcoma were the most frequent types (40% each).

Since STS metastasize to the lung, earlier studies have systematically excluded patients with metastatic disease from their analyses. Our series comprises eight patients with metastatic disease (stage IV) at diagnosis. Stage IV was described as a sole primary lung lesion with metastases to at least one organ other than the lung itself. Six patients with locally or locoregionally advanced disease, not amenable to surgical excision, presented metastatic progression with lung metastases (6), brain metastases (2) and pleural metastases (1), while seven patients with resected disease relapsed to the lung (7), pleura (1) and mediastinal lymph nodes (1). All patients with relapsed disease presented lung metastases, indicating that PLS retain lung tropism of metastasis, similarly to STS of other locations. However, locations such as the brain, mediastinal lymph nodes and pleura are indicative of metastatic bronchogenic carcinomas rather than sarcomas, suggesting that sarcomas may acquire a similar dissemination pattern according to their primary site.

Lymph-node involvement in STS has traditionally been reported at low rates. A recent, large series analyzing this issue have reported a rate of 1.1% of nodal disease from STS of the extremities, as well as a 3.1% from intrathoracic STS, although no differentiation of "intrathoracic sarcomas" (i.e. mediastinal, cardiac, lung or pleural sarcoma) was made (20). Conversely, high rates of MNI have been reported in previous studies of PSL, from 16%(18) to 25% (15). In our series, eight patients (33.33%) had MNI, which represents the highest rate.

A 25% reduction in the 5-year OS of patients with node involvement has been reported ($p<0.0001$) (18). Patients with MNI in our study had an inferior OS compared to those without MNI, although the difference was not statistically significant (Table II). More accurately, there were no statistically significant differences in OS between patients with stage II, stage III and stage IV lung cancer, in line with the data presented by Golota *et al*. (2) (Figure 2). This fact indicates that, even if PSL may spread as lung cancer, staging of patients as for lung cancer may not provide predictive information. Yet, due to the high rate of MNI, mediastinum should be surgically evaluated in all patients operated for PSL regardless of histotype.

Table II. *Overall survival*.

	mOS (months)	95%CI	*p*-Value
Whole population (N=24)	10	7.608-12.392	-
Whole metastatic population (N=18)	4	0.882-7.118	-
No surgery (N=11)	8	2.605-13.395	0.012
Surgery (N=12)	12	6.967-17.033	
Patients with MNI (N=8)	9	7.152-10.848	0.415
Patients without MNI (N=9)	12	0.313-23.687	
Adjuvant CT (N=3)	8	6.400-9.600	0.337
No Adjuvant CT (N=9)	15	7.679-22.321	
Tumor size >5 cm (N=8)	8	3.842-12.138	0.113
Tumor size ≤5 cm (N=9)	12	9.078-14.922	
Stage II (N=11)	12	0.00-42.897	0.052
Stage III (N=6)	8	4.399-11.601	
Stage IV (N=7)	11	5.868-16.132	
Localized disease (N=17)	10	7.349-12.651	0.513
Metastatic disease (N=7)	11	5.868-16.123	
Patients with MNI (N=8)	9	7.152-10.848	0.802
Metastatic disease (N=7)	11	5.868-16.123	

mOS: Median overall survival; CI: confidence interval; MNI: mediastinal node invasion; CT: chemotherapy. p: Statistical significance.

Tumor size has been suggested as a prognostic factor of utmost impact on survival (12, 13, 18). In our series, despite the trend towards a longer OS in patients with tumors <5 cm, no statistically significant differences were found. As our series included patients with metastatic and unresectable disease, analysis was also performed only for patients (10) with completely resected disease. The small sample size and the unevenly weighted groups (only one patient with tumor size >5 cm had a complete resection) did not allow conclusions to be drawn. However, Regnard and Porte, with 24 (15) and 18 (16) patients respectively, did not find statistically significant differences in OS between patients with tumors greater or smaller than 5 cm. These inconsistent data suggest that size is probably not a prognostic factor as robust as in STS of the extremities.

Thirteen patients in our study underwent surgical resection, including 11 patients with localized disease and two patients with metastatic disease. Complete surgical resection has been indicated as one of the most important prognostic factors in several series (2, 3, 13, 16-18). In our study, surgery was significantly related to survival (Figure 3), although this result may be jeopardized by the inclusion of patients with metastatic disease in this analysis. Since stereotactic radiotherapy (SBRT)

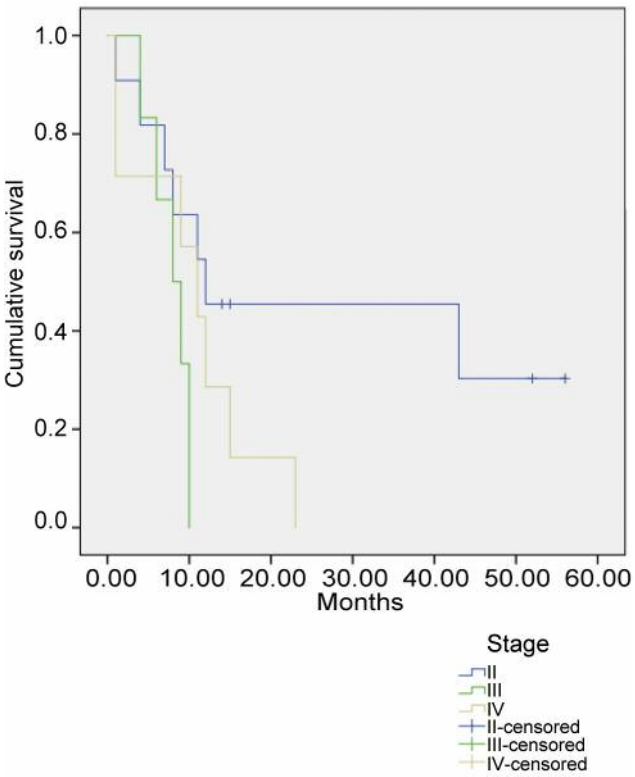

Figure 2. *Kaplan–Meier survival estimates for lung cancer staging. Median OS for Stage II: 12 months (95%CI=0.00-42.897). Median OS for Stage III: 8 months (95%CI=4.399-11.601).Median OS for Stage IV: 11 months (95%CI=5.868-16.132). p=0.052.*

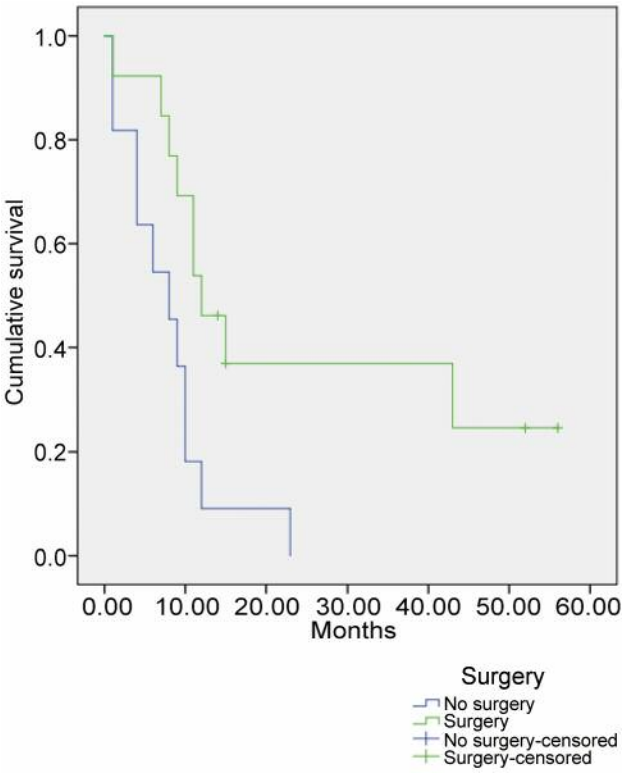

Figure 3. *Kaplan–Meier survival estimates according to surgery. Median OS for patients who did not undergo surgery: 8 months (95%CI=2.605-13.395). Median OS for patients who underwent surgery: 12 months (95%CI=6.967-17.033). p=0.012.*

is effective in the treatment of metastases from STS (22), the question whether this strategy could also be valid for PSL arises and should be investigated in prospective series.

Neoadjuvant chemotherapy has been reported only in two patients due to tumor size with important tumor shrinkage, followed by complete resection (16). In our study, one patient underwent induction chemotherapy without response, so surgery was dismissed. None of the patients with MNI were treated with neoadjuvant chemotherapy. As indicated by Spraker *et al*., even if MNI in PSL should not be regarded as bronchogenic carcinomas, better surgical outcomes and local control could still be achieved with neoadjuvant chemotherapy or chemoradiation (18).

Adjuvant anthracycline-based chemotherapy (AC) was applied in three patients without evident benefit in OS. Again, the uneven distribution of groups (3 with AC *versus* 9 without AC) does not allow any conclusion. Earlier series have not studied the effect of adjuvant chemotherapy due to the same problem (17).

The median OS for the whole population of our series was 10 months (Figure 1B), which differs remarkably from the one reported in earlier studies. This may be due to the higher percentage of patients with metastatic and unresectable disease in our group, as previous series comprised exclusively patients with tumors limited to the lung and in most cases resected. However, when analyzed separately, patients who underwent surgical resection present a median OS of 15 months, which is approximately 10 months less than the previously reported OS. As Golota *et al*. have reported (2), male sex could adversely affect survival of PSL, as also indicated in our series. Earlier studies have suggested that size and stage of lung carcinoma may be important prognostic factors. It should be noted that the population of our study has an even distribution as for size >5 cm and <5 cm (47% and 53%, respectively) and MNI (47% with involvement and 53% without involvement). It is unlikely that tumor size affects the survival of our series, due to other concurrent prognostic factors such as metastasis. Although MNI has also been reported to statistically affect OS, our study does not indicate that it is a poor prognostic factor.

Due to its retrospective design, our study was constrained by non-available data. Important unreported variables such as extension of surgery or radiotherapy could possibly

confound the analysis. Histopathological diagnosis could not be revised by a central pathologist because of logistic issues and this is a major limitation, as the dates of diagnosis range from 2008 to 2018, while WHO classification was modified in 2013 (21). Another limitation is the small sample size, which makes impossible to draw robust conclusions from the subgroup analyses. Since most of the limitations are recurrent in all series reported, it would be expected that a prospective analysis of these neoplasms could more reliably answer important questions such as the prognostic value of lymph node involvement and tumor size or the role of adjuvant chemotherapy.

Conclusion

The data of patients diagnosed with PLS differ significantly from earlier reported series. Histologic distribution is altered, with predomination of synovial sarcoma and undifferentiated pleomorphic sarcoma. Previously documented prognostic factors could not be confirmed in our study. The high proportion of patients with MNI at diagnosis may serve as an indication for surgical evaluation of mediastinum in all cases and raises the question if patients with locoregional PSL should be treated or not with a more aggressive approach as in bronchogenic cancer.

Conflicts of Interest

The Authors report no conflict of interests related to this study.

Authors' Contributions

Jose Duran Moreno, Stefania Kokkali and Maria Salomidou had the idea for the concept of the study. Jose Duran Moreno and Stefania Kokkali performed the literature search, the analysis and data curation and drafted the original article. Jose Duran Moreno, Stefania Kokkali, Maria Salomidou, Vasileios Ramfidis, Antonia Digklia, Anna Koumarianou, Periklis Tomos and Konstantinos Syrigos: Provision of resources. Nektarios Koufopoulos, Ioannis Vamvakaris, Eleni Psychogiou: Performed histologic diagnosis. All Authors performed reviewing and editing, and Konstantinos Syrigos supervised all work.

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