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Case Report

A Giant Synovial Sarcoma of the Left Lung

Georgi Yankov[1], Magdalena Alexieva[2], Silvia Ivanova[3], Stefka Yankova[1], Evgeni Mekov[1]

[1] *Department of Respiratory Diseases, St Ivan Rilski University Hospital, Medical University of Sofia, Sofia, Bulgaria*

[2] *St Sofia MHAT, Sofia, Bulgaria*

[3] *St Ivan Rilski University Hospital, Sofia, Bulgaria*

Corresponding author: Evgeni Mekov, Department of Respiratory Diseases, St Ivan Rilski University Hospital, Medical University of Sofia, Sofia, Bulgaria; Email: evgeni.mekov@gmail.com; Tel.: +359 888 320 476

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Abstract

Primary pulmonary synovial sarcoma is an extremely rare and aggressive neoplasm that primarily affects young people and has a poor prognosis. Establishing this diagnosis requires the exclusion of a wide number of other neoplasms with multimodal clinical, imaging, histological, immunohistochemical, and cytogenetic assessment. We present a case of synovial sarcoma of the left lung in a 44-year-old man, diagnosed immunohistochemically after left lower lobectomy with atypical resection of the 5th segment. Imaging, diagnostic workup, histological and immunohistochemical characteristics, surgical treatment, and prognosis are discussed.

Keywords

diagnostic imaging, lung, immunohistochemistry, synovial sarcoma, surgical treatment

INTRODUCTION

The majority of malignant lung tumors are of epithelial origin. Soft tissue tumors are far less common. Primary lung sarcoma is a rare and aggressive malignancy and lung metastases from extrapulmonary sarcomas are significantly more common than primary pulmonary synovial sarcoma (PPSS).

Because most mesenchymal malignancies have a benign analogue and certain epithelial tumors show sarcomatoid differentiation (e.g., renal cell carcinoma, melanoma), accurate histological diagnosis, including assessment of the size of the lesion, is critical.[1]

We present a case of synovial sarcoma of the left lung in a 44-year-old man, diagnosed immunohistochemically after left lower lobectomy with atypical resection of the 5th segment.

CASE REPORT

A 44-year-old male was admitted to the Department of Thoracic Surgery with pain in the back and the lateral part of the chest wall on the left, fatigue, and cough with mild hemoptysis for a month. He was a smoker and heavy drinker (100 ml daily). Physical examination was remarkable for decreased breath sounds on the left. The laboratory tests, pulmonary function tests, and arterial blood gas analysis showed no abnormalities.

On chest X-ray, a large heterogeneous mass in the left lower hemithorax was visualized. The computed tomography (CT) shows an ovoid lesion of 69×105×90 mm with heterodense structure up to 56 HE in the left lower lung with numerous gas deposits in the matrix, which was widely adhered to the chest wall without infiltrating it but with radicular and reticular interstitial changes around the lesion ***(Fig. 1)****.*

Fibrobronchoscopy (FBS) showed a deformed B8 segment with submucosal proliferation but was not diagnostic.

The surgical approach was through a left lateral thoracotomy in the 5th intercostal space ***(Fig. 2)****. A left lower lobectomy with atypical resection of the 5th segment, due to the proximity of the tumor to the major fissure, was performed. In the left lower lobe, a moderately dense tumor lesion of 100×70 mm was found. There was a presence of visibly*

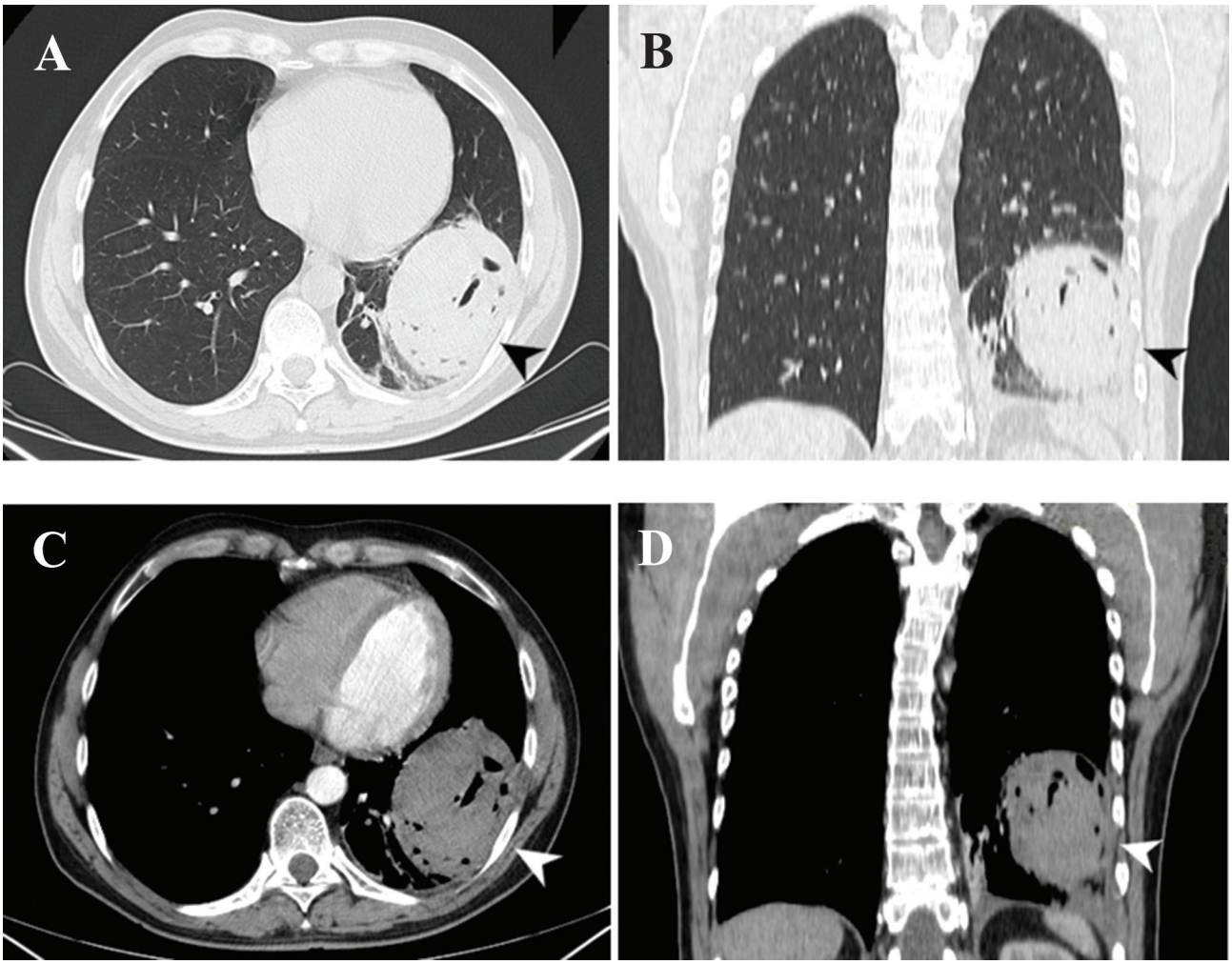

Figure 1. Axial and coronal CT images, visualizing left lower lobe synovial sarcoma.

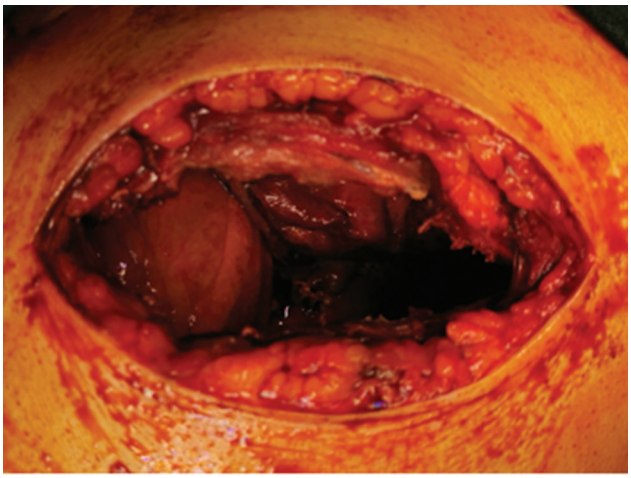

Figure 2. Intraoperative image of lateral muscle-sparing thoracotomy in the 5th intercostal space after left lower lobectomy and an atypical resection of the 5th segment.

pathological lymph nodes in the hilum and the pulmonary ligament, the first being located as a cuff around the hilum of the lower lobe. Thorough lymph dissection was performed at levels 5, 7, 9, and 10.

The specimen dissection showed cavitary tumor lesion which was filled with blood, and a large coagulum ***(Fig. 3)****. A pathohistological study demonstrated destruction of the lung parenchyma by tumor tissue (over 100 mm), represented by bundles of monotonous atypical spindle cells, focal myxoid stromal changes, areas of hemorrhages, extensive areas of necrosis, areas of cystic degeneration, and mitoses. The morphological finding refers to malignant intrapulmonary mesenchymal neoplasm. The bronchial stump margin was free of tumor infiltration. Seventeen lymph nodes were examined and all of them were without metastases.*

The immunohistochemical analysis showed positive expression in neoplastic cells for TLE-1 (1F5), CD99 (O13), and Bcl2 (bcl-2/100/D5); negative expression in tumor cells for AE1/AE3 (PANCK), desmin (DE-R-11), SOX-10; negative expression in neoplastic cells but positive in the vascular network in the tumor mass for CD34 (QBEnd/10), Actin (SMA) (αSMA-1) ***(Fig. 4)****.*

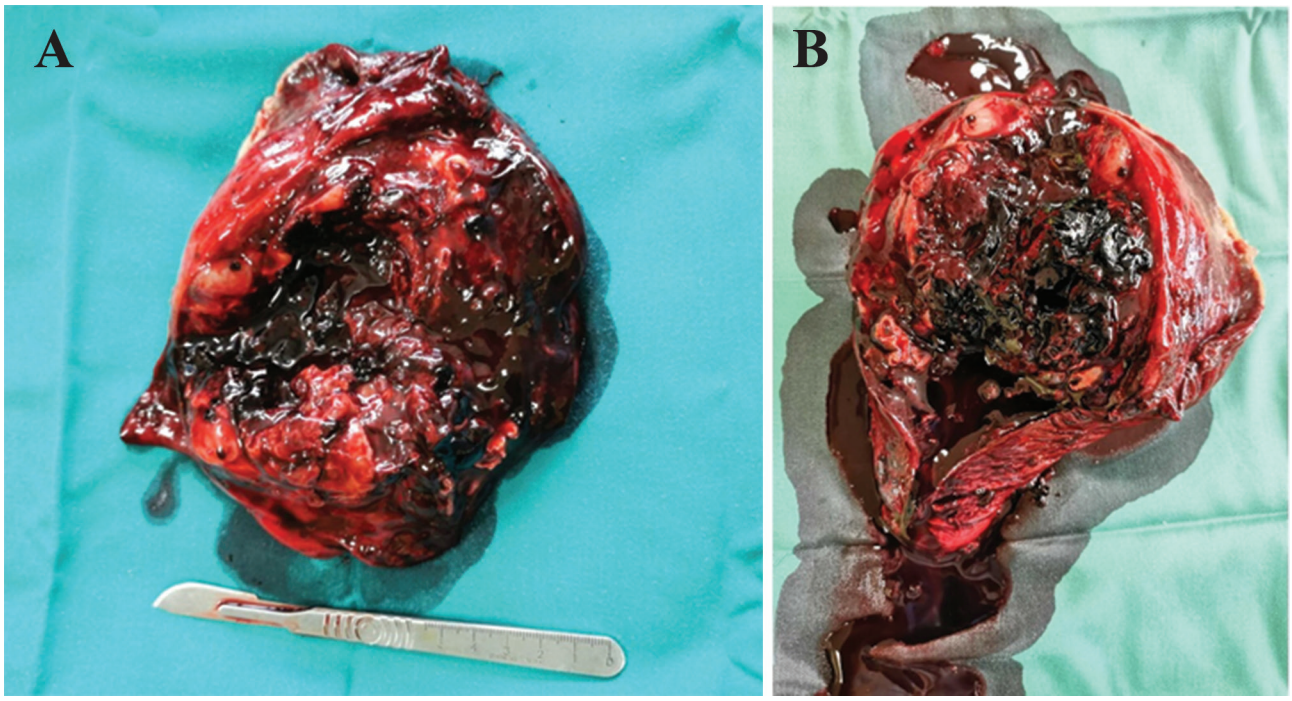

Figure 3. Macroscopic view of the tumor mass.

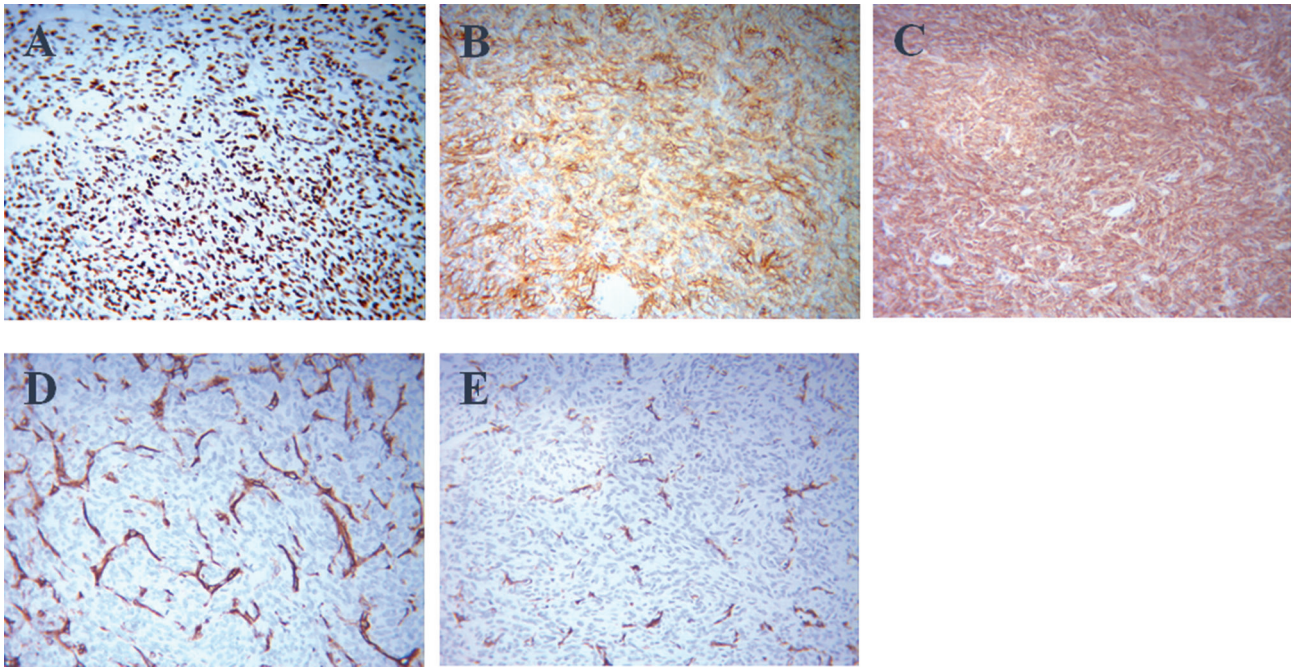

Figure 4. Photomicrograph of immunohistochemical staining, showing positive expression of neoplastic cells for TLE (**A**), CD99 (**B**), Bcl-2 (**C**), positive expression in the vascular network for CD34 (**D**), and Actin (**E**).

The immunoprofile of the neoplastic process in the lung favors synovial sarcoma (SS). Further fluorescent in situ hybridization (FISH) revealed monophasic fibrous (spindle cell) synovial sarcoma. According to the French Federation of Cancer Centers Sarcoma Group (FNCLCC) grading criteria, the tumor was graded as G3 (differentiation – score: 3; mitoses – score: 3; necrosis – score 2) and staged as T4N0M0R0.

The postoperative period was uneventful. The patient was discharged from the hospital on postoperative day 7 and was referred to an oncology clinic. One year after surgery, the patient is in excellent condition and without PET/CT data for recurrence.

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DISCUSSION

Primary lung sarcomas are uncommon and aggressive malignancies with similar treatment and prognosis compared to other soft tissue sarcomas.[2] Lung sarcomas account for only 0.1%–0.5% of all primary pulmonary malignancies[3] with a 5-year overall survival rate of 50%[4]. The most common variants of lung sarcoma are malignant fibrous histiocytoma and synovial sarcoma.[4]

Synovial sarcoma is a mesenchymal spindle cell tumor that accounts for 5%–10% of all soft tissue sarcomas and is defined by a pathognomonic chromosomal translocation t(X;18)(p11.2;q11.2).[3] It arises from immature mesenchymal elements rather than from the synovium.[4] SS is most common in the extremities (more than 90%), especially near large joints, but the head and neck, lungs, heart, mediastinum, and the abdominal wall could also be primary localizations.

PPSS usually affects young patients (median age between 31 and 50 years).[3] Patients with pulmonary synovial sarcoma usually report pleuritic chest pain (50.0%), cough (28.6%), palpable mass (7.1%), and dyspnea (7.1%).[5] The differential diagnosis is very wide, but primary lung cancer should be excluded. The absence of significant lymphadenopathy in a young adult with a relatively large ovoid tumor favors PPSS rather than primary lung cancer.[5]

PPSS presents on chest X-ray as a homogenous parenchymal formation with a well-circumscribed rounded or lobulated border, a pleural mass, and partial or complete opacification of the hemithorax, with dimensions often more than 7 cm^3. On CT scan, these tumors show commonly heterogeneous enhancement with necrotic or cystic areas, vessels in the lesion, calcifications, tumor rupture, pleural/chest wall extension, and pleural effusion.[5]

PPSS is aggressive, possibly due to its anatomic location and large tumor size generally resulting in insufficient resection and high proliferative activity.[6] Treatment of PPSS includes surgical resection followed by chemotherapy or radiation therapy. Surgical treatment is the main option as there are no other standardized approaches. Adjuvant chemotherapy improves the time to local recurrence and recurrence-free survival and tends to improve overall survival.[4] Due to the lack of preoperative morphological diagnosis and the giant size of the mass, we undertook open surgery through lateral muscle-sparing thoracotomy. A left lower lobectomy with atypical 5th segment resection was performed because of the lesion's size and proximity to the large interlobar fissure.

Primary pleuropulmonary SS seems to be with higher local aggressiveness than its soft-tissue counterpart, and this could be due to the difficulties in achieving wide surgical margins combined with a late presentation.[7] Patients with PPSS have a poor prognosis, with a 5-year overall survival rate of 50%.[4] Male gender, large tumor size, extensive tumor necrosis, neurovascular invasion, high histological grade, and mitotic rate are poor prognostic factors.[8]

CONCLUSIONS

PPSS is a rare and aggressive neoplasm that primarily affects young people and has a poor prognosis. Establishing this diagnosis requires the exclusion of a wide number of other neoplasms and a wide work-up plan. The method of choice for treatment is mainly surgical with subsequent adjuvant chemo- or radiation therapy. Due to the risk of recurrence, these patients should be carefully monitored.

Ethical Approval

Ethical approval was not necessary as this is not a clinical study. The research meets all applicable standards concerning the ethics of experimentation and research integrity, and the following is being certified/declared true. No identifiable images or information were used.

Informed Consent

Informed consent was obtained for all the procedures as a part of the patient's hospital stay.

Consent for publication

All authors read and agreed with the manuscript.

Availability of data and material

All figures and data are readily available.

Conflict of Interest

None of the authors of this paper has a financial or personal relationship with other people or organizations that could inappropriately influence or bias the content of the paper.

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Гигантская синовиальная саркома левого лёгкого

Георги Янков[1], Магдалена Алексиева[2], Силвия Иванова[3], Стефка Янкова[1], Евгени Меков[1]

[1] Кафедра респираторных заболеваний, УМБАЛ „Св. Иван Рилски“, Медицинский университет - София, София, Болгария
[2] МБАЛ „Св. София“, София, Болгария
[3] УМБАЛ «Св. Иван Рилски», София, Болгария

Адрес для корреспонденции: Евгени Меков, Кафедра респираторных заболеваний, УМБАЛ „Св. Иван Рилски“, Медицинский университет - София, София, Болгария; Email: Evgeni.mekov@gmail.com; тел.: +359 888320476

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Резюме

Первичная синовиальная саркома лёгких — крайне редкое и агрессивное новообразование, поражающее преимущественно молодых людей и имеющее плохой прогноз. Установление этого диагноза требует исключения широкого числа других новообразований с помощью мультимодальной клинической, визуализационной, гистологической, иммуногистохимической и цитогенетической оценки. Представлен случай синовиальной саркомы левого лёгкого у мужчины 44 лет, диагностированный иммуногистохимически после нижней лобэктомии слева с атипичной резекцией 5-го сегмента. Обсуждаются методы визуализации, диагностическое обследование, гистологические и иммуногистохимические характеристики, хирургическое лечение и прогноз.

Ключевые слова

диагностическая визуализация, лёгкие, иммуногистохимия, синовиальная саркома, хирургическое лечение