

Primary Pulmonary Synovial Sarcoma

Francis J. Podbielski, MS, MD¹, Tracy E. Sambo, MD²,
Arsalan Salamat, MD², Matthew J. Blecha, MD²,
and Mark M. Connolly, MD²

PLEURA
Volume 3: 1-2
© The Author(s) 2016
Reprints and permission:
sagepub.com/journalsPermissions.nav
DOI: 10.1177/2373997516632755
plr.sagepub.com



Abstract

Primary sarcoma of the lung and/or pleural cavity is a rare finding accounting for <0.5% of all thoracic malignancies. Histologically, mesothelioma predominates in this subcategory of neoplasms; its relationship to asbestos exposure and cigarette smoking is well documented. Other tumor types include fibrosarcoma, malignant fibrous histiocytoma, angiosarcoma, leiomyosarcoma, synovial sarcoma, and Ewing sarcoma. Standard evaluation with bronchoscopy and radiographically guided biopsy are frequently nondiagnostic and require open biopsy to provide an adequate tissue sample.

Keywords

pleural images, pleural malignancy

Case Study

A 39-year-old man employed as a housepainter presented with fevers, weight loss, and a nonproductive cough. He reported having a positive tuberculin skin test and received Bacillus Calmette-Guérin vaccine as a youth. Current sputum cultures and cytology were negative for malignancy. Computed tomography of the chest showed a large right pleural effusion and 3 pleural-based nodules (Figure 1). Bronchoscopy demonstrated no endobronchial lesions and a negative aspirate cytology. Thoracentesis and pleural biopsy yielded only pleural fibrosis.

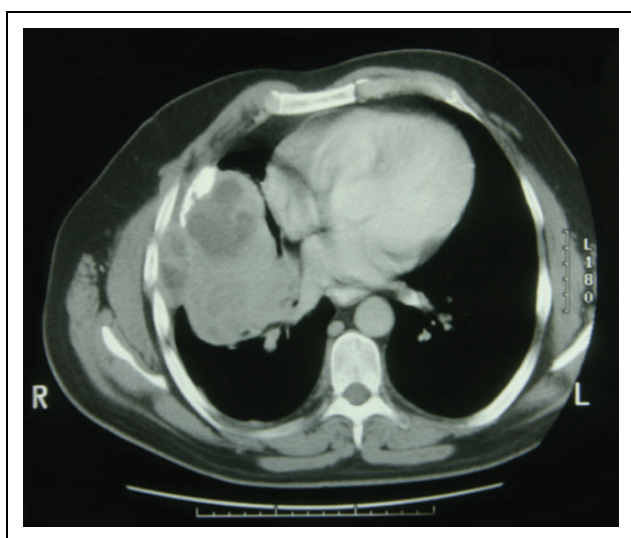


Figure 1. Computed tomography of the chest illustrating a large, irregular, pleural-based mass.

Thoracoscopic exploration of the chest revealed a longitudinally spreading, thick, friable tumor mass along the parietal pleural surface of the lung. Light microscopy showed a high-grade spindle cell neoplasm (Figure 2;

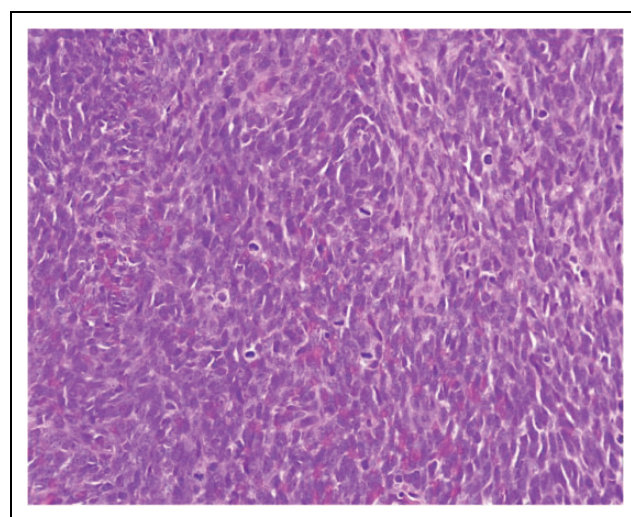


Figure 2. Hematoxylin and eosin stain of the tumor at $\times 40$ magnification demonstrating a spindle cell malignancy.

¹ University of Illinois at Chicago, Riverside, IL, USA

² Presence St. Joseph Hospital—Chicago, Chicago, IL, USA

Submitted: January 9, 2016. Accepted: January 25, 2016.

Corresponding Author:

Francis J. Podbielski, University of Illinois at Chicago, 212 Maplewood Road, Riverside, IL 60546-1846, USA.

Email: fjp@moqua.com



hematoxylin and eosin stain at $\times 40$). Further investigation demonstrated a tumor that was vimentin (+), cytokeratin (–), and epithelial membrane antigen (–), thus suggestive of a sarcoma, but with calretinin (+). Electron microscopic studies failed to show the long, thin microvilli characteristic of mesothelioma. Chromosomal studies with the reverse transcription-polymerase chain reaction test for the t(X;18) translocation confirmed the diagnosis of a synovial sarcoma.¹ Further workup revealed no evidence of extrathoracic disease. Given the presence of multiple nodules and extensive pleural tumor involvement, no further surgery was planned and the patient treated with adriamycin and ifosfamide. He is alive with disease at 10 months postdiagnosis and has asymptomatic progression of his tumor as seen on computed tomography scan.

Discussion

A paucity of literature exists on primary pulmonary synovial sarcoma. Most cases of intrathoracic synovial sarcoma are secondary to metastatic disease with a clinically obvious primary tumor adjacent to the joint capsule in a limb. As noted by Nascimento et al,² metastatic disease from these cancers generally results in multiple, rather than isolated, pleural nodules.

In contrast to a series of patients reported by Gaertner et al,³ this patient's tumor appeared to be monophasic with a relatively small epithelial contribution to the tumor cell population. All tumors in the series were pleural-based masses that were excised at surgical exploration. The median survival of the 5 study patients was 18 months, with 4 of the 5 patients dead of their disease by 30 months. Histological analysis of recurrent tumors showed a monophasic spindle cell pattern.

In the study of primary intrathoracic sarcomas by Régnard and colleagues,⁴ an actuarial survival of 48% at 5 years was noted if the primary tumor could be resected completely, and a 0% survival at 2 years if only incomplete resection was

achieved. Further, they found a significant prognostic difference between T₂ and T₃ tumors, with a lesser contribution of nodal status to overall survival.

The absence of an extrathoracic disease in this case supports the notion by Rys et al⁵ that primary synovial sarcomas are derived from immature mesenchymal elements and not from the synovium within the joint capsule. Differential subpopulations' cell growth probably plays an integral role in determining tumor recurrence and ultimately patient survival.

Declaration of Conflicting Interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Funding

The author(s) received no financial support for the research, authorship, and/or publication of this article.

References

1. Kaplan MA, Goodman MD, Satish J, Bhagavan BS, Travis WD. Primary pulmonary sarcoma with morphologic features of monophasic synovial sarcoma and chromosome translocation t(X;18). *Am J Clin Pathol*. 1996;105(2):195-199.
2. Nascimento AG, Unni KU, Bernatz PE. Sarcomas of the lung. *Mayo Clin Proc*. 1982;57:355-359.
3. Gaertner E, Zeren EH, Fleming MV, Colby TV, Travis WD. Biphasic synovial sarcomas arising in the pleural cavity. *Am J Surg Pathol*. 1996;20(1):36-45.
4. Régnard JF, Icard P, Guibert L, de Montpreville VT, Magdeleinat P, Levasseur P. Prognostic factors and results after surgical treatment of primary sarcomas of the lung. *Ann Thorac Surg*. 1999;68:227-231.
5. Rys J, Donna A, Betta PG, et al. Synovial sarcoma: a new approach to histological classification and a comparison with malignant mesothelioma. Immunohistochemical study with anti-mesothelial cell serum. *Patol Pol*. 1992;43:161-163.