An 83-Year-Old Man With Bilateral Spontaneous Pneumothoraces and Multiple Cysts

Shambhu Aryal, MD
Chau Chu, MD
R. Scott Morehead, MD, FCCP

From the Division of Pulmonary and Critical Care Medicine, University of Kentucky Medical Center, Lexington, KY

*Correspondence to: Shambhu Aryal, MD, Division of Pulmonary and Critical Care Medicine, University of Kentucky, 740 S Limestone St, Ste L543, Lexington, KY 40536

E-mail address: sar222@uky.edu

Manuscript received November 12, 2010, accepted December 6, 2010

Reproduction of this article is prohibited without written permission from the American College of Chest Physicians (http://www.chestpubs.org/site/misc/reprints.xhtml).

PII S0012-3692(11)60317-6

An 83-year-old man was transferred to our medical center with bilateral pneumothoraces. A biopsy of a nonhealing scalp lesion was done 4 weeks previously, and the specimen showed high-grade angiosarcoma. During the subsequent staging evaluation a week later, he was found to have asymptomatic bilateral pneumothoraces and multiple pulmonary cysts that were not metabolically active on 18 F-fluorodeoxyglucose (FDG)-PET scan. These findings were initially believed to be secondary to emphysema, and because of his absence of symptoms, the patient was sent home without intervention. However, several days later he developed a nonproductive cough with pleuritic chest pain and went to an outside ED. A chest radiograph again showed bilateral pneumothoraces, which resulted in placement of bilateral chest tubes with the subsequent transfer. His medical history was otherwise only remarkable for benign prostatic hypertrophy and cataracts. He admitted to smoking cigarettes remotely but denied fever, chills, leg swelling, palpitations, and syncope. He denied relevant occupational exposures, including asbestos.

Physical Examination

The patient was a thin, elderly man with bilateral chest tubes; vital signs, including oxygen saturation, were normal. The midskcalp had an incision from a prior biopsy with intact sutures. The chest was hyperexpanded with diminished breath sounds. The cardiac examination was normal. The left shin had an erythematous plaque. There was no cyanosis or clubbing of the extremities. The remainder of the examination was unremarkable.

Laboratory and Radiologic Findings
A hemogram and a comprehensive metabolic panel were within normal limits. The chest CT scan performed for staging of the scalp angiosarcoma is shown in [Figure 1], [Figure 2]. It demonstrated bilateral pneumothoraces and multiple thin-walled pulmonary cysts that were of varying sizes and contained no internal fluid or debris. In addition, there were several nodular and scattered ground-glass opacities. A lung biopsy was not performed.

![Figure 1](image1.png)  **Figure 1**  Axial CT scan of the chest showing multiple bilateral thin-walled cysts. Also seen are a large right pneumothorax and a smaller left pneumothorax.

![Figure 2](image2.png)  **Figure 2**  A different section from the same axial CT scan of the chest showing more thin-walled cysts and the bilateral pneumothoraces.

**What is the diagnosis?**

**Diagnosis:** Metastatic pulmonary angiosarcoma with bilateral secondary spontaneous pneumothoraces

**Discussion**

Angiosarcomas are very rare and highly malignant sarcomas of vascular endothelial origin, characterized by angiomatous areas of freely anastomosing channels lined by atypical endothelial cells alternating with Kaposi-like spindle cell areas. The mean age at presentation is about 60 years, and there is a slight male
preponderance. The most common primary site of involvement is the skin of the head and neck, but other reported sites include the heart, adrenal gland, ovary, prostate, vagina, and maxillary sinus. Factors associated with angiosarcomas include exposure to polyvinyl chloride, thorium dioxide, trauma, and radiation. Reports of association with foreign body, neurofibromatosis, tuberous sclerosis, and meningiomas have been reported. Regardless of their site of origin, angiosarcomas have a striking propensity for lung metastasis. Other frequent sites of metastatic disease include the bone, liver, and lymph nodes.

Metastatic pulmonary angiosarcoma manifests in various ways, with hemoptysis and weight loss being the most common symptoms, but cough, chest pain, dyspnea, or unexplained fever may be present. Some patients may be incidentally discovered to have metastatic disease on staging studies, as in the present patient. Although most patients have no lung examination findings, crackles, clubbing, chest tenderness, and arm swelling have been noted. The most common radiographic finding is multiple nodules, although linear infiltrates, pleural effusion, and diffuse alveolar infiltrates can be seen. On CT scans, multiple solid nodular lesions are the most common pattern, followed by multiple thin-walled cysts and solitary nodules. Pneumothorax is uncommonly reported but appears to be more common in patients with the primary site of disease in the scalp.

There seems to be a clear distinction between a pulmonary cyst and a cavitary pulmonary nodule. The differential diagnosis for pulmonary cysts include congenital cystic lung lesions, such as congenital cystic adenomatoid malformation, bronchogenic cysts, pneumatoceles, pulmonary Langerhans cell histiocytosis, lymphangiomatosis, and confusion with emphysematous changes (not true cysts). On the other hand, cavitary pulmonary nodules occur with metastatic lung disease, infections with mycobacteria and certain endemic fungi, septic emboli, and noninfectious inflammatory conditions such as Wegner granulomatosis and rheumatoid arthritis. Cavitation in malignancy is most commonly associated with metastatic squamous cell carcinoma from the head and neck or cervix of the uterus. Although a thin-walled cavitary lesion is usually not considered to be a malignancy, angiosarcomas seem to be an exception to this rule.

The pathogenesis of thin-walled cysts in pulmonary angiosarcoma is not well understood. Three possible mechanisms for the development of malignant cysts have been described in the past: (1) excavation of a nodular tumor through the discharge of the necrotic material inside, (2) infiltration of malignant cells into the walls of a preexisting benign pulmonary bulla, and (3) infiltration of malignant cells into the walls of air sacs formed by cystic distension of small airways through the ball-valve effect of the tumor. Regardless of the mechanism of formation, rupture of subpleural cysts can cause hemopneumothorax, but bilateral simultaneous pneumothoraces as the presentation of metastatic angiosarcomas seems to be very rare.

Although FDG-PET scan is well known as a functional imaging technique in the evaluation of various malignant tumors, to our knowledge there are no published reports of PET scan features of pulmonary angiosarcomas. Case reports have described PET scan positivity in angiosarcomas of the skin, breast, liver, and spleen, but whether pulmonary angiosarcomas behave in the same way is not known. It has been noted that a negative PET scan in the presence of suspicious CT scan findings in the chest cannot reliably exclude pulmonary metastasis from osseous and soft tissue sarcomas. This case demonstrates that the same may be true for angiosarcomas. As such, this entity, when presenting as multiple thin-walled cysts as in the present patient, could easily be confused with benign cysts, and recognition of this occurrence is important.

The prognosis of metastatic pulmonary angiosarcoma is generally poor, with median survival of no more than several months. There is no established systemic treatment of the malignancy; surgery, chemotherapy, and radiation have all been tried without uniform results. Therefore, pulmonary complications are usually managed symptomatically.

**Clinical Course**

In the present patient, findings on the first imaging were erroneously ascribed to emphysema. Consequently, there was a delay in the management. At the subsequent presentation, a diagnosis of metastatic pulmonary angiosarcoma was made on the basis of underlying high-grade scalp angiosarcoma and the characteristic radiologic appearance of the cysts in the absence of other known causes of cystic lung disease. Although the hospital course was complicated by respiratory failure due to hospital-acquired pneumonia with a loculated right pleural effusion, he ultimately underwent bilateral chemical pleurodesis and was weaned from mechanical ventilation. He was determined to be a poor candidate for systemic chemotherapy because of his functional status and died 3 months after initial diagnosis. A postmortem examination of the lungs was refused
by the family. Although lung pathology was unavailable for definitive confirmation, the clinical course and radiographic finding strongly supported the final diagnosis.

**Clinical Pearls**

1. Finding of simultaneous bilateral spontaneous pneumothoraces may indicate a serious parenchymal lung disorder.

2. Cavitary thin-walled lesions are generally considered to be nonmalignant, but angiosarcomas defy this rule.

3. Metastatic pulmonary angiosarcoma can be FDG-PET scan-negative.

4. The prognosis of metastatic pulmonary angiosarcoma is usually poor, with median survival being only a few months after diagnosis. Treatment is usually symptomatic.

**Acknowledgments**

**Financial/nonfinancial disclosures:** The authors have reported to CHEST that no potential conflicts of interest exist with any companies/organizations whose products or services may be discussed in this article.

**Other contributions:** We thank Anil K. Attili, MD, from the Division of Radiology, University of Kentucky Medical Center, Lexington, Kentucky, for helping with selection and reproduction of the images. The manuscript was prepared at the Division of Pulmonary and Critical Care Medicine, University of Kentucky, Lexington, Kentucky.

**SUGGESTED READINGS:**


Iagaru et al., 2006 Iagaru A, Chawla S, Menendez L, Conti PS: 18F-FDG PET and PET/CT for detection of pulmonary metastases from musculoskeletal sarcomas. Nucl Med Commun 27. (10): 795-802.2006;

