CHEST Postgraduate Education Corner
PULMONARY, CRITICAL CARE, AND SLEEP PEARLS

A Middle-Aged Man With Cough, Chest Pain, and Pulmonary Artery Filling Defect

Hidenobu Shigemitsu, MD, FCCP; Ngozi Orjioke, MD; Jabi E. Shriki, MD; John Varras, MD; and Michael N. Koss, MD

CHEST 2014; 145(2):407–410

A middle-aged man with no past medical history presented to the ED after 2 weeks of productive cough, rhinorrhea, high-grade fevers, and chills. He had been treated with ibuprofen, promethazine cough syrup, and albuterol as an outpatient without any relief. He had associated dull, nonradiating anterior chest wall pain and unintentional weight loss of 10 lb during the course of his illness. On evaluation, he was noted to speak in short sentences of four to five words with labored breathing.

Physical Examination Findings

His temperature was 36°C, and he had a heart rate 110 to 120/min, BP 138/89 mm Hg, and oxygen saturation of 90% on room air. The lung examination revealed occasional bilateral basilar crackles. The heart examination revealed tachycardia with a normal S1 and S2 with no visible jugular venous distention. The rest of the history and examination were noncontributory.

Diagnostic Studies

CBC count showed an anemia of 11.1 g/dL with normal WBC count and differential. The complete metabolic panel values and chest radiograph were normal. An ECG showed right axis deviation with tachycardia. A CT pulmonary angiogram was performed with findings shown in Figure 1. Transesophageal echocardiography revealed a large globular mass protruding into the right ventricular outflow tract (RVOT) across the pulmonary valve and extending into the main pulmonary artery. The right ventricular peak systolic pressure was 50 mm Hg.

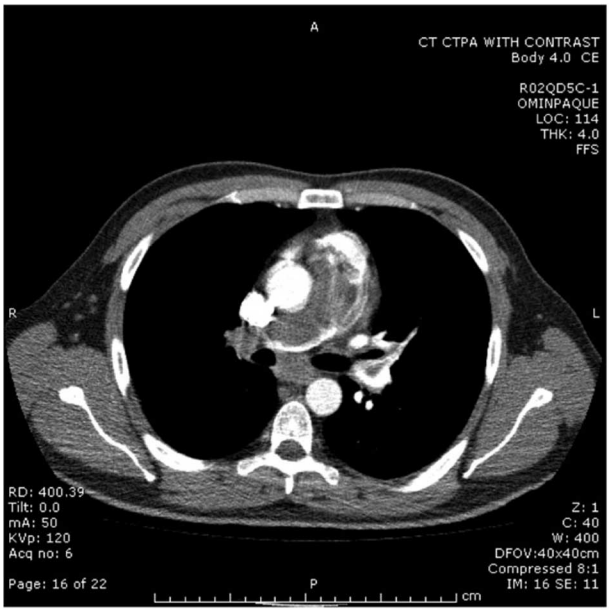

FIGURE 1. Initial chest CT scan at the level of the fifth thoracic vertebra.

What is the diagnosis?

Manuscript received June 27, 2013; revision accepted August 7, 2013.
Affiliations: From the Department of Medicine (Drs Shigemitsu and Varras), University of Nevada School of Medicine, Las Vegas, NV; and Division of Pulmonary and Critical Care Medicine (Drs Shigemitsu and Orjioke), Department of Radiology (Dr Shriki), and Department of Pathology (Dr Koss), Keck School of Medicine, University of Southern California, Los Angeles, CA.
Correspondence to: Hidenobu Shigemitsu, MD, FCCP, Division of Pulmonary and Critical Care Medicine, University of Nevada School of Medicine, 2040 W Charleston Blvd, Ste 300, Las Vegas, NV 89102; e-mail: hshigemi@gmail.com

DOI: 10.1378/chest.13-1488

Discussion

Primary pulmonary artery sarcoma is a rare disease first described on autopsy in 1923. It has been reported to occur between the ages of 45 and 55 years of age and equally affects men and women. Clinical signs and symptoms are often nonspecific, often resulting in delayed diagnosis. Cough, chest or back pain, dyspnea, and hemoptysis are common presenting symptoms. Signs suggestive of neoplasia such as anemia, unintentional weight loss, and elevated inflammatory markers may also be present.

Most primary pulmonary artery sarcomas originate in the pulmonary trunk or occasionally the RVOT, although the site of origin is often difficult to define. Tumors vary from soft and lobulated to firm and smooth macroscopically and by imaging studies. The presence of a pedunculated lesion arising from the RVOT, pulmonary valve, or trunk on MRI or echocardiography should raise the suspicion of pulmonary artery sarcoma. They can be classified as intraluminal or intramural, with intraluminal being more common. These intraluminal sarcomas are often poorly differentiated or undifferentiated and are also called intimal sarcomas.

Pulmonary artery sarcomas are commonly misdiagnosed as pulmonary emboli, and differentiation on initial imaging can often be difficult. However, several features, if identified correctly, should raise pulmonary artery sarcoma among diagnostic possibilities. First, a low attenuation filling defect occupying the entire luminal diameter of the main or proximal pulmonary artery branches with expansion of these vessels should raise the possibility of a pulmonary artery sarcoma. These findings are uncommon in pulmonary emboli. Second, extraluminal extension into the lung parenchyma is a common finding in pulmonary artery sarcomas, but should not be seen in pulmonary emboli. Third, although rarely performed, a noncontrast chest CT scan will generally demonstrate hyperattenuation in the setting of larger pulmonary emboli, whereas pulmonary artery sarcomas will display relatively low attenuation prior to contrast administration. Fourth, patients with pulmonary artery sarcomas tend to have more indolent symptoms and a relative paucity of symptoms, with clinical presentations disproportionately mild in comparison with the degree and size of filling defects within the pulmonary arteries. Finally, pulmonary emboli respond to anticoagulation therapy, thrombolytic therapy, or both, whereas pulmonary artery sarcomas do not.

Enhancement can also help differentiate pulmonary artery sarcomas from pulmonary emboli. Because CT pulmonary angiograms are typically obtained early after contrast administration, tumor enhancement may be difficult to perceive. For this reason, MRI/magnetic resonance angiogram is helpful in differentiating pulmonary artery sarcomas from pulmonary emboli. MRI affords the ability to obtain images in multiple, separate phases of contrast administration, without exposing the patient to added ionizing radiation. It should be noted that chronic pulmonary emboli may also show some enhancement, although this is typically mild compared with the enhancement seen with pulmonary artery sarcomas. Additionally, filling defects due to chronic pulmonary emboli will form obtuse angles with the walls of the pulmonary arteries, whereas filling defects from pulmonary artery sarcomas will form acute angles.

PET/CT scanning has also shown value in differentiating pulmonary artery sarcomas from pulmonary emboli. Usually sarcomas will show an avid uptake of radiopharmaceuticals (fluorodeoxyglucose), although this is somewhat variable, according to the histologic grade of the sarcoma.

Pathologic diagnosis can be achieved by endovascular catheter biopsy. The tumors typically reveal a heterogeneous hypercellular pattern intermixed with varying degrees of necrosis and mitotic activity. They often contain several mesenchymal features such as spindle cells and pleomorphic giant cells.

Immunohistochemical findings are based on the direction of differentiation. Routine immunohistochemical stains are performed for desmin, vimentin, cytokeratin, smooth muscle-specific actin, and muscle-specific actin. A proposed staging system based on local invasion and involvement of surrounding structures has been proposed (Table 1).

Given the rarity of the tumor and the absence of prospective trials, there are no validated treatment regimens, though surgical resection with curative intent is accepted as being the cornerstone of therapy. Patients considered for surgical resection should have adequate cardiopulmonary reserve, disease confined to the chest cavity, and adequate lung function reserve if pneumonectomy is considered. Complete resection of the tumor with clear margins by frozen specimen examination is recommended. If the tumor involves the RVOT and pulmonic valve, a homograft may be used for reconstruction. However, in select cases, patients may benefit from palliative surgical resection.

Table 1—*Proposed Staging System for Primary Pulmonary Artery Sarcoma*

Stage	Description
1	Tumor limited to the main pulmonary artery
2	Tumor involving one lung plus a main pulmonary artery
3	Bilateral lung involvement
4	Extrathoracic spread