right pleural effusion. Intrusion into the atrial septal defect and superior vena cava caused the facial swelling present on the day of her death. Finally, because blood entered her lungs via her left pulmonary artery, thromboemboli from the pulmonary artery aneurysm traveled exclusively to the right lung, causing infarction and contributing to the unilateral pleural effusion.

Surgical correction of pulmonary artery aneurysms has been attempted in patients with congenital left-to-right shunts and pulmonary hypertension. However, when pulmonary hypertension is irreversible, the long-term prognosis is poor.

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Figure 2. The massive pulmonary artery (PA) is occluded by thrombus. RV = right ventricle; Ao = aorta; LV = left ventricle; PV = pulmonic valve.

Sarcoma of the Pulmonary Artery

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A patient is described in whom clinical presentation, V/Q scan, and pulmonary angiogram were consistent with pulmonary embolus. No improvement occurred despite therapy with heparin. When sent for surgical embolectomy, sarcoma of the pulmonary artery was diagnosed. This entity, though rare, should be considered in the differential diagnosis of pulmonary vascular disease.

Sarcoma of the pulmonary artery is a rare entity. The case presented here describes a patient in whom both clinical and radiologic criteria were consistent with chronic pulmonary embolism. In view of the patient's failure to improve with anticoagulation, surgical embolectomy was attempted, at which time sarcoma of the pulmonary artery was diagnosed. Though previously reported, this rare entity is often overlooked in the differential diagnosis of pulmonary vascular disease, and should be considered.

CASE REPORT

A 45-year-old, non-smoking white woman with a two-year history of recurrent pneumonia began to complain of increasing exertional dyspnea associated with a dry cough, but denied chest pain, sputum production, or hemoptysis. Chest radiographs, which had shown normal findings in the past, now revealed left hilar enlargement and lobulation, as well as bilateral lower lobe infiltrates (Fig 1).

Physical examination was positive solely for an increase in the pulmonary second sound. Pulmonary function studies showed a restrictive defect with the FVC and TLC approximately 50 percent of predicted, and a normal FEV1/FVC ratio. The diffusing capacity was 55 percent of predicted. There was a 20 mm Hg decrease in Pao2 on maximal exercise: Pao2 was 63, and Paco2 26 on room air.

Bronchoscopy with transbronchial biopsy showed only interstitial inflammation with no evidence of infection or malignancy. Mediastinoscopy was negative for malignancy or granulomatous inflammation. Gallium scan, ACE level, ANA, and echocardiogram were all normal.

The patient was treated with steroids, without benefit, for possible interstitial lung disease. She was then referred for further evaluation. A V/Q scan and pulmonary angiogram were consistent with pulmonary embolism (Fig 2), and IV heparin was started. Right leg venogram showed old DVT in the popliteal vein and fresh thrombus in soleal veins of the right calf. Right ventricular pressure was 95/0 and PA pressure was 95/10 during angiography. Despite therapy with heparin, the patient did not improve, and was sent for surgical embolectomy.

At surgery, tumor rather than thrombus was found in both pulmonary arteries, with total occlusion of the right pulmonary artery. A frozen section was read as sarcoma. A bypass was placed from the main pulmonary artery to the right pulmonary artery, but the patient developed endobronchial bleeding and died.

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Autopsy showed leiomyosarcoma of the pulmonary trunk extending to both pulmonary arteries with occlusion on the right, and extension to the anterior mediastinum. There were metastases present in a mediastinal lymph node, lower lobe of the left lung, and site of previous mediastinoscopy. Organized thrombi were present in branches of the left pulmonary artery and there were sites of new and old infarction within lung parenchyma. There was no evidence of tumor elsewhere. Histology showed spindle cells, and bizarre cells and mitotic figures (Fig 3).

**DISCUSSION**

Sarcoma of the pulmonary artery is a rare entity, with approximately 40 cases reported in the world literature. There is a 2:1 female predominance with an average age of 50 years. Survival from diagnosis is from weeks to 3½ years. The diagnosis of sarcoma of the pulmonary artery is virtually never considered prior to surgery or autopsy. There should be no obvious site of primary sarcoma outside the pulmonary artery system.

**REFERENCES**