Primary Pulmonary Artery Sarcoma*  
A Method of Resection

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Primary pulmonary artery sarcoma classically presents with symptoms and findings suggestive of acute pulmonary artery occlusion. An angiocentric mass or the finding of spindling neoplasm on needle biopsy should suggest this neoplasm. The rare unilateral location in the present case permitted resection by pneumonectomy. An endarterectomy technique was used to extract loosely adherent tumor-thrombus from the more proximal portions of the left pulmonary artery.  
(Chest 1990; 98:752-52)

D = diffusing capacity of carbon monoxide

Primary pulmonary artery sarcoma is an uncommon problem. Those cases that have been reported have, almost without exception, involved the right ventricular outflow tract and the main pulmonary trunk and often extended into both main pulmonary artery branches. Bleisch and Kraus1 reviewed 60 collected cases from the literature and cited only five incidents of unilateral pulmonary artery sarcomas arising more distally.‡‡ Only two of these were managed by resection.

The present report is of a patient with classic clinical evolution and findings of unilateral pulmonary artery sarcoma. This finding permitted total resection via an endarterectomy technique to clear the left pulmonary artery of proximal tumor extension.

Case Report

A 56-year-old woman presented to her physician in September 1987 because of the sudden onset of sharp left upper back pain and dyspnea. Her chest roentgenogram was normal. A ventilation-perfusion lung scan suggested left pulmonary embolus; a pulmonary angiogram showed some nonfilling peripheral pulmonary arteries on the left; and systemic anticoagulant therapy was initiated. The patient felt good until December 1987, when symptoms recurred.

A repeat angiogram demonstrated complete occlusion of the left main pulmonary artery. Investigations of a source of emboli were negative; venograms of the upper and lower extremities were normal, and an echocardiogram revealed no evidence of intracardiac tumor or thrombus. Computed tomographic scans of the chest, abdomen, and pelvis were normal. The patient was treated as an outpatient with subcutaneous heparin.

In June 1988, the patient was seen briefly because of intractable cough. In August 1988, she had episodes of hemoptysis and complained of night sweats. She had no pain or dyspnea. A chest roentgenogram at this time clearly demonstrated a left hilar mass. A repeat pulmonary angiogram in August 1988 (Fig 1) was said to be unchanged from that of December 1987. A CT-guided needle aspiration revealed "suspicious" cytology. Bronchoscopy and biopsy were carried out without diagnosis. A tuberculin skin test was negative, and the level of angiotensin-converting enzyme was within normal limits. The patient was referred to the Mayo Clinic for further evaluation.

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Figure 1. Pulmonary angiogram (Aug 29, 1988) demonstrated left hilar mass and complete occlusion of left pulmonary artery at its origin. Right lung and pulmonary artery vasculature were normal. Pulmonary outflow tract was uninvolved as well.

Figure 2. Computed tomography of thorax (Sept 29, 1988) demonstrated dense filling of pulmonary artery and its branches with tumor. Mediastinal settings were normal, without demonstrable lymphadenopathy. Sections through upper abdomen failed to reveal hepatic, adrenal, or other metastases.
of the lung, the pericardium was closed.

After an uneventful month of postoperative convalescence, a course of radiotherapy was initiated because of the proximity of tumor to the margins of resection. There has been little experience with adjunctive therapy for sarcoma at this site, but experience with sarcomas of the extremities suggests adjuvant radiotherapy may be of benefit after local excision.

The pathologic examination indicated a solid sarcomatous left pulmonary artery distally (Fig 3). The vessel was completely obstructed by a primary pleomorphic sarcoma (malignant mesenchymoma). The neoplasm was multinodular, localized to the hilum, and involved a zone 8 × 7 × 6 cm. Grossly, the neoplasm filled the distal left pulmonary artery and many of its distal branches. Distally, lung tumor nodules developed in an angiocentric fashion resembling, in gross cross-section, preoperative CT cuts. More proximal pulmonary artery was unaffected by tumor where the plug of tumor-thrombus had been extracted. The bronchial margin was widely free of neoplasm. There was no invasive neoplasm at the pulmonary resection margin. Multiple hilar, subcarinal, paratracheal, subaortic, inferior pulmonary ligament, and other intrapulmonary peribronchial lymph nodes were free of tumor.

**DISCUSSION**

The clinical presentation of our patient resembles that of the 51-year-old man described by Goldstein and Joubert, with recurring severe pleuritic pain, cough, and occasional hemoptysis. Investigations failed to demonstrate the mass initially; the presentation strongly suggested a pulmonary embolism. Even in the few cases that have been reported, the findings have been deceptive. Two cases presented as lung abscess, one with and one without an accompanying mass; three cases showed features of embolus, with pleuritic pain, dyspnea, and occasional hemoptysis. This suggests that when unilateral pulmonary artery occlusion has been demonstrated, especially if there is evidence of progression, as there was in our case, a strong suggestion of pulmonary artery neoplasm should be entertained. A CT scan or MR imaging can be helpful in differentiation. Decreased vascularity, which has been described, can be difficult to appreciate if the vascular tree is filled with tumor or tumor emboli. Few cases of primary pulmonary artery sarcoma have been reported; even fewer are amenable to resection when found. Early diagnosis, where possible, offers a chance for resection and potential tumor control. Use of "endarterectomy" technique can be helpful in complete resection when tumor-thrombus extends proximally within the pulmonary artery.

**REFERENCES**

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