Pulmonary Artery Sarcoma*  
A Case Report of Surgical Cure and 5-Year Follow-up  

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Pulmonary artery sarcoma is a rare tumor that is frequently misdiagnosed as chronic pulmonary embolism. With heightened clinical awareness and advancement in technology, the diagnosis is now increasingly being made preoperatively. Previous literature has described the disease to be uniformly fatal, with surgical resection as the single most effective modality for short-term palliation. We present the case of a patient in whom pulmonary artery sarcoma was diagnosed preoperatively and who underwent surgical resection with no evidence of recurrence during long-term follow-up, suggesting that early identification and aggressive surgical intervention has the potential to be curative.

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Pulmonary artery sarcoma is a rare tumor of the cardiovascular system with only a few hundred cases reported in literature. The disease was first described at autopsy by Mandelstamm1 in 1923. Most of the subsequently reported cases have been diagnosed at autopsy. In the last decade, heightened clinical suspicion and improved diagnostic modalities have allowed the diagnosis to be made prior to surgery. The prognosis of pulmonary artery sarcoma is poor, with reported survival times of a few months to a few years regardless of therapy. We present the case of a patient with pulmonary artery sarcoma who, after preoperative confirmation of the diagnosis with pulmonary angioscopy, underwent successful surgical resection of the tumor followed by pneumonectomy. The patient has been observed for 5 years with no apparent recurrence of disease.

CASE REPORT

A 32-year-old woman with no significant medical history was evaluated in early 1996 for chronic cough, low-grade fever with sweats, and a 4.5-kg weight loss over a 6-month period. She also complained of intermittent chest tightness and shortness of breath. There was no history of exposure to tuberculosis. However, she worked as a teacher in a local school, where she was in contact with a large population of immigrant children. On examination, she appeared healthy. There was no palpable lymphadenopathy. Cardiovascular examination revealed a prominent pulmonary component of the second heart sound. Examination of the chest showed slight dullness to percussion at the left base. Digital clubbing was present in both hands.

Among the results of relevant laboratory studies, hemoglobin level was 10.2 g/dL and the erythrocyte sedimentation rate was 72 mm/h. A chest radiograph showed a left lower lobe cavitary lesion. Initially, the patient was empirically treated with antituberculous treatment that was discontinued after a negative result of a purified protein derivative test and the absence of acid-fast bacilli in sputum and bronchoscopic specimens. A week later, she presented with acute shortness of breath. A spiral CT scan with IV administration of contrast material showed a large filling defect in the left main pulmonary artery (Fig 1). An ill-defined cavity also was noted in the left lower lobe. Venous duplex examination of the lower extremities was negative for deep vein thrombosis. A presumptive diagnosis of pulmonary embolism was made, and the patient was treated with 100 mg tissue plasminogen activator followed by unfractionated heparin. A follow-up scan of the chest 24 h later failed to show any dissolution of the clot. The possibility of pulmonary artery sarcoma was entertained, and the patient was referred to the University of California at San Diego for further evaluation.

Evaluation at the University of California at San Diego included a ventilation scan, which demonstrated reduced ventilation to the left lung, and a perfusion scan, which showed an absence of perfusion to the same area. Both ventilation and perfusion of the right lung were normal. Transthoracic echocardiography demonstrated normal left and right ventricle chamber size and function with mild tricuspid regurgitation. Pulmonary angiography showed an intraluminal rounded filling defect causing near-total occlusion of the left main pulmonary artery. Pulmonary angioscopy confirmed the presence of a grayish intraluminal mass. In November 1996, the patient underwent pulmonary artery exploration and resection of the lobulated mass (Fig 2) from the left pulmonary artery with reconstruction of the artery using a bovine pericardial patch. The distal pulmonary artery seemed to be free of tumor. Histopathologic examination of the specimen was consistent with low-grade sarcoma (Fig 3).

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Figure 1. Chest CT scan with IV contrast showing a filling defect (arrow) in the left main pulmonary artery.
The immediate postoperative course was uncomplicated. A postoperative perfusion scan demonstrated only a small amount of flow to the left lower lobe.

On the initial follow-up, the patient seemed to be doing well. A few months later, however, she developed intermittent hemoptysis. A repeat CT scan showed persistence of the left lower lobe cavity. In addition, a fungal ball was visualized within the cavity (Fig 4) with evidence of pericavitary infiltrate. Specimens obtained from a diagnostic bronchoscopy were negative for Aspergillus. A repeat perfusion scan continued to demonstrate only minimal perfusion to the left lower lobe.

In February 1998, the patient underwent a left pneumonectomy. Pathology specimens confirmed the presence of invasive Aspergillus infection as well as a distal pulmonary arteriopathy. There was no evidence of recurrent tumor. The patient has been doing well during subsequent follow-up examinations, with no evidence of disease recurrence. On the last visit, the patient had successfully conceived and had an uncomplicated course in the second trimester of her pregnancy.

**Discussion**

Pulmonary artery sarcoma is a rare tumor of the cardiovascular system, and very few cases have been reported in the literature. The reported age at presentation has ranged from 13 to 86 years, with the majority of cases occurring in middle age. The etiology of these tumors is obscure. It has been suggested that they arise from the mesenchymal cells of the muscle anlage of the bulbus cordis. Histopathologically, most pulmonary artery sarcomas are leiomyosarcomas or "undifferentiated spindle cell sarcomas." Histopathologic classification, however, does not seem to be useful clinically or prognostically. Most pulmonary artery sarcomas arise from the dorsal area of the pulmonary trunk, although the tumors also may arise from the right and left pulmonary arteries, the pulmonary valve, and the right ventricular outflow tract.

Because of its rarity and insidious growth characteristics, pulmonary artery sarcoma is often misdiagnosed for pulmonary embolism, leading to inappropriate therapy such as prolonged anticoagulation or thrombolysis. The need for a precise preoperative diagnosis is crucial for the proper exploration of the pulmonary artery to ensure complete resection and reconstruction. Symptoms and signs such as weight loss, fever, anemia, and digital clubbing may be subtle clues to diagnosis. Other characteristics, such as the absence of risk factors for deep vein thrombosis, high sedimentation rate, nodular parenchymal infiltrates on CT scans, and lack of response to anticoagulation should raise the suspicion of a process other than pulmonary embolism. Unfortunately, none of the above features can exclude the possibility of chronic thromboembolic disease. Enhancement of an intraluminal filling defect with gadolinium-diethylenetriamine pentaacetic acid on MRI has been suggested as a sensitive way of differentiating a tumor mass from a thrombus. Multiplane transesophageal echocardiography and fluorodeoxyglucose positron emission tomography have been used as preoperative diagnostic tools. Positron emission tomography has the added advantage of diagnosing indeterminate hilar masses. Other novel diagnostic methods...
currently being refined include the use of a transvenous catheter suction biopsy\(^2\) and pulmonary angiography, which was performed in our case.

Previous literature on the disease has reported it to be uniformly fatal, with the longest survival time in one series reported to be 3.5 years\(^7\) despite surgical resection. Surgical resection offers the only chance of prolonged survival. The role of chemotherapy and radiation are still undefined. This is in contrast to pulmonary parenchymal sarcoma, which behaves less aggressively and can be cured by resection with or without adjuvant therapy.

Our case suggests the possibility of a cure with surgical resection if the disease is diagnosed early, before the occurrence of distal metastasis or involvement of adjacent mediastinal structures. The lack of reperfusion of the left lung after removal of the central mass appears to be the consequence of a distal arteriopathy, which has not been described previously in cases of pulmonary artery sarcoma. It has, however, been described in cases of unilateral thromboembolic obstruction.\(^{24}\)

REFERENCES


Successful Lung Volume Reduction Surgery in a Child With Severe Airflow Obstruction and Hyperinflation due to Constrictive Bronchiolitis* 

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Lung volume reduction surgery (LVRS) may improve pulmonary function in patients with severe emphysema. However, its effects in other types of obstructive lung disease are unknown. To delay the need for lung transplantation, we performed LVRS in a 14-