Case Reports

High-Grade, Nonmyogenic Pulmonary Artery Sarcoma

Rare Findings on Coronary Angiography

Vimal Ramjee, MD
Melinda M. Lewis, MD
Stephen D. Clements, Jr., MD

Pulmonary artery sarcomas are exceptionally rare, and they are often misdiagnosed as chronic pulmonary thromboemboli. Early and accurate diagnosis is crucial to the prognosis of patients who have pulmonary artery sarcomas.

Herein, we describe the case of a 74-year-old man who presented with dyspnea and was initially thought to have a pulmonary embolus. Anticoagulation with unfractionated heparin was ineffective. Rare angiographic findings during routine cardiac catheterization led to the diagnosis of a high-grade, nonmyogenic, primary pulmonary artery sarcoma. This case illustrates the usefulness of angiographic findings as an adjunct to conventional diagnostic methods in correctly identifying this rare, aggressive malignancy. ***(Tex Heart Inst J 2011;38(1):1-3)***

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From: *Department of Internal Medicine (Dr. Ramjee); Division of Cytopathology, Department of Pathology (Dr. Lewis); and Section of Cardiology, Department of Medicine (Dr. Clements), Emory University School of Medicine, Atlanta, Georgia 30303*

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Address for reprints:
Vimal Ramjee, MD, 870 Inman Village Pkwy. NE, #405, Atlanta, GA 30307

E-mail:
vramjee@emory.edu

Pulmonary artery (PA) sarcoma was first described by Mandelstamm in 1923.[1] Since then, fewer than 200 PA sarcomas have been reported.[2] Pulmonary artery sarcomas are commonly misdiagnosed as chronic pulmonary thromboemboli.[3] Early diagnosis is challenging but essential to the prognosis of patients with this aggressive malignancy.[4] Investigators have typically emphasized that the chief indicator of PA sarcoma is the absence of venous thromboembolic risk factors in the presence of suspicious chest images. Herein, we present the case of a patient in whom rare findings during cardiac catheterization heightened the suspicion of PA sarcoma.

Case Report

In July 2005, a 74-year-old man presented at our institution with a 2-month history of worsening shortness of breath that was associated with a sensation of increased pressure in the left side of the chest. His medical history included stage 1 hypertension, mild mitral regurgitation, and minimal aortic regurgitation. On physical examination, he had mild cervical lymphadenopathy and a grade 2/6 systolic ejection murmur at the apex of the heart with distant heart sounds.

A computed tomographic angiogram of the thorax showed multiple hilar nodules bilaterally and nearly complete obstruction of the left PA (Fig. 1). In view of these findings and the patient's occupational history of long, uninterrupted automobile trips, a pulmonary embolus was suspected. Although Doppler ultrasonography of the lower extremities revealed no evidence of deep vein thrombosis, the patient was started on unfractionated heparin.

The anticoagulation therapy resulted in no improvement, so further studies were performed to characterize the lesion. An echocardiogram showed a left ventricular ejection fraction of 0.60, a dilated right ventricle, PA pressures of 90 to 100 mmHg, an abbreviated pulmonary acceleration time, and minimal flow in the left main PA. Coronary angiography revealed markedly elevated left PA pressures and a filling defect that was consistent with nearly complete obstruction of the left PA (Fig. 2). The bulky, proximal defect appeared to be irregular, and large pressure gradients were detected across the lesion, increasing the suspicion of malignancy.

A whole-body 18-Fluorodeoxyglucose positron-emission tomographic scan showed high uptake of contrast medium in the left PA and pulmonary nodules that were consistent with primary PA sarcoma and secondary local metastases. Histopathologic analysis of specimens obtained through an intrapulmonary artery brush biopsy and an excisional micro-biopsy showed a high-grade malignant neoplasm with sarcomatoid features (Fig. 3). Immunostaining of the PA specimen was strongly posi-

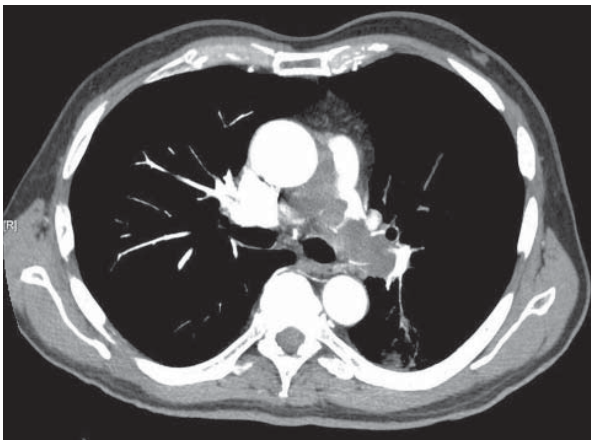

Fig. 1 *Contrast-enhanced computed tomogram of the chest shows pulmonary nodules and a large, enhanced lesion in the left pulmonary artery.*

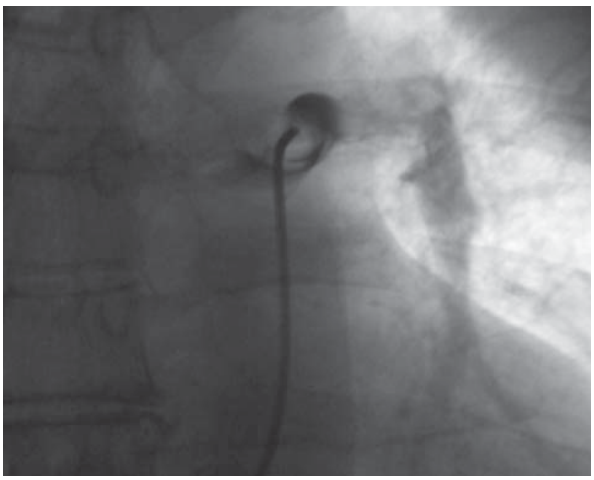

Fig. 2 *Angiographic image during balloon-inflated Swan-Ganz catheterization shows a nearly complete filling defect of the left main pulmonary artery proximal to its branches.*

Real-time motion image is available at www.texasheart.org/journal.

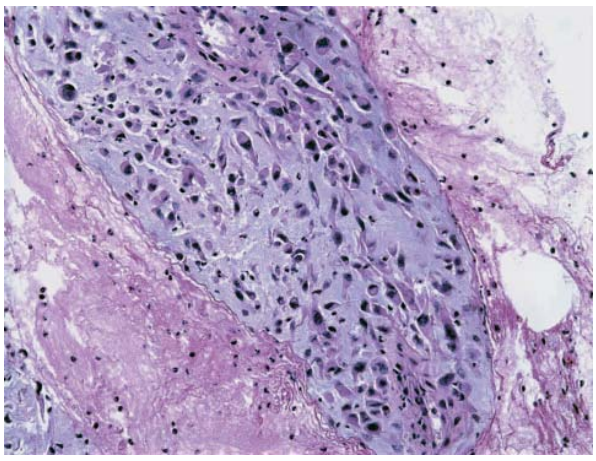

Fig. 3 *Photomicrograph of left intrapulmonary artery biopsy specimen shows features of a high-grade, nonmyogenic sarcoma (H & E, orig. ×10).*

tive for vimentin and negative for smooth-muscle actin, CD31, CD34, factor VIII, desmin, and pancytokeratin (AE1/AE3). Analyses of morphologic and immunohistochemical stains were consistent with a high-grade, nonmyogenic PA sarcoma.

The patient, who was a poor candidate to undergo surgical excision, could not tolerate more than 2 cycles of chemotherapy. He was then lost to follow-up. It was learned that he died of disease progression months after chemotherapy was halted.

Discussion

Fewer than 200 PA sarcomas have been reported, and the estimated incidence is 0.001% to 0.03% in the general population.[2] These neoplasms affect both sexes equally, but 1 study reported a predominance in women up to twice that of men.[2] A review of 138 PA sarcomas determined the average age at diagnosis to be 49.3 years (range, 13–86 yr).[3]

Vascular sarcomas are initially classified as intimal or mural on the basis of their growth pattern. In particular, nonmyogenic sarcomas have a tendency toward intraluminal growth in the PAs, where advanced neoplasms cause death from local tumor effects.[5] Immunostaining of arterial sarcomas helps to distinguish leiomyosarcomas (positive for desmin, actin, and vimentin) from angiosarcomas (positive for factor VIII, CD34, and vimentin).[4,5] However, the largest institutional review to date reported the successful histologic classification of only 35.3% of arterial sarcomas, demonstrating the limited usefulness of histopathologic stratification in directing clinical management.[5]

Clinically, patients present with symptoms related to right ventricular outflow tract obstruction, local mass effect, or pulmonary emboli; the most common symptoms are dyspnea, chest pain, cough, hemoptysis, and weight loss.[3-5] Examination can reveal systolic ejection murmurs, lower-extremity edema, hepatomegaly, and jugular venous distention as the neoplasm extends into the PA.[3,5]

Chest radiography can reveal PA enlargement, hilar nodules, an enlarged cardiac contour, and decreased pulmonary vascularity.[3,5] On chest computed tomography, most patients have nonspecific abnormalities in the pulmonary vasculature that are consequent to the primary lesion and independent of coexisting intimal sarcomatosis.[3,5] Findings that suggest a neoplastic process include heterogeneous tumor attenuation, intermittent enhancement of the mass, and extraluminal lesion extension.[5] Notably, angiography can reveal several differentiating features of PA sarcoma that may lead the physician to consider malignancy despite an otherwise low degree of suspicion. Key findings are proximal filling defects that are irregular and bulky in a polypoid or lobulated manner. In addition, a neoplastic process may

be present if there is total or nearly complete PA obstruction in the setting of a lesion that does not smoothly and gradually taper (as would be typical in chronic thromboembolic disease).

The exceptionally low incidence of PA sarcoma can result in late diagnosis. Because the mean survival duration after diagnosis is less than 2 months without surgery and 23 months after surgery,[5] early identification and treatment are essential to improving the prognosis.[4,5] Cox and colleagues[3] reported average survival rates at 1, 2, and 5 years in patients who had PA sarcomas. In the surgically untreated group, those survival rates were 5%, 3%, and 0, respectively. In the group that underwent surgery and chemoradiation treatment, the survival rates were 58%, 39%, and 0, respectively. These findings underscore the substantial benefit of surgical intervention in eligible patients over the earlier years, although long-term survival rates are unaffected. As an adjunct to computed tomographic and magnetic resonance imaging, angiographic findings during cardiac catheterization can enable earlier diagnosis and an improved prognosis through early surgical intervention.

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