Primary Pulmonary Artery Sarcoma Diagnosed Noninvasively by Two-dimensional Echocardiography

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SUMMARY A 21-year-old white male was evaluated in our echocardiographic laboratory for presumed mitral valvular bacterial endocarditis. Electrocardiographic, physical, and initial two-dimensional echocardiographic findings suggested a left-to-right shunt at the atrial septal level. However, injection of contrast saline solution failed to demonstrate signs of an atrial septal defect. Continuation of the echocardiographic study led to the diagnosis of an unsuspected primary sarcoma of the pulmonary trunk, which was rapidly confirmed by computerized axial tomography. Therapeutic interventions were undertaken. This case highlights the usefulness of two-dimensional echocardiography.

PRIMARY SARCOMAS of the pulmonary trunk are very rare. A review of available literature revealed only 67 reported cases. All but 15 of these cases were diagnosed at autopsy. The 15 cases not diagnosed at autopsy were diagnosed at catheterization and angiography performed for a presumptive diagnosis other than tumor of the pulmonary trunk. We report a case of an unsuspected primary sarcoma of the pulmonary trunk diagnosed noninvasively by two-dimensional echocardiography.

Case Report
A 21-year-old white male steel worker was referred for evaluation of a heart murmur. Thirteen months previously, he had been successfully treated for right upper lobe pneumonia. Subsequently, he complained of intermittent cough and dyspnea on exertion. Six months before we saw him, he experienced the sudden onset of severe dyspnea and chest pain and a 30-second syncopal episode while running. He did not seek medical attention. Three weeks before referral, the patient was seen by his physician for chills and cough. An apparently new systolic murmur was noted and the patient was hospitalized for possible endocarditis. Evaluation failed to produce evidence of bacterial infection. An M-mode echocardiogram suggested a flail posterior mitral valve leaflet or mitral valve prolapse. The patient was discharged with a diagnosis of an upper respiratory illness, probably viral, and referred to the Cardiac Noninvasive Laboratory at the University of Virginia Medical Center for further evaluation of his murmur and mitral valve apparatus.

Physical examination revealed an extremely thin white male who was dyspneic on minimal exertion. There were no features of Marfan’s syndrome. His respiratory rate was 26 breaths/min and his pulse rate was 110 beats/min. The jugular veins were not distended and there was no pedal edema. His lungs were clear. Examination of the heart revealed a normally situated apical impulse and a right ventricular heave. The first heart sound was normal. The pulmonic component of the second heart sound was accentuated and widened physiologic splitting was present. An early to middiastolic click and a soft blowing mid- to late systolic murmur were present apically. Additionally, a high-pitched ejection murmur that increased in intensity with inspiration was present to the left of the sternum.

A two-dimensional echocardiogram was performed with the patient reclining at 30° elevation in the left lateral decubitus position. The left ventricle was normal in size, wall thickness and function, although the intraventricular septum moved paradoxically. The left atrium was of normal size. The mitral valve demonstrated systolic prolapse. The aortic valve was normal. The right atrium was markedly enlarged. The right ventricle was markedly dilated and hypokinetic. The tricuspid and pulmonic valves were normal. Because a left-to-right shunt was suspected, contrast saline solution was injected intravenously in the left arm. No negative contrast could be demonstrated at either the atrial or ventricular level. No microbubbles passed retrogradely into the inferior vena cava or hepatic veins, thereby excluding significant tricuspid regurgitation. Because the microbubbles cleared slowly from the right ventricle, the right ventricular outflow tract, pulmonary valve and main pulmonary trunk were examined more closely. Just beyond the normal pulmo-
nary valve, a large, granular mass substantially filled the lumen of the pulmonary trunk, but did not distort its external contour (fig. 1). This mass extended into the left and right main pulmonary arteries. On the basis of this study, the patient was admitted for further evaluation.

The admission chest radiograph demonstrated slight mediastinal displacement to the right and hypovascularity and patchy interstitial scarring of the right lung. The left lung and cardiac silhouette were normal. The ECG demonstrated right ventricular hypertrophy with strain. Computerized axial tomography of the thorax revealed a filling defect in the main pulmonary artery extending into the right pulmonary artery (fig. 2). There was a defect in the wall of the pulmonary trunk, which suggested that the mass arose from there. A lung scan demonstrated no perfusion of the right lung. Several perfusion defects were demonstrated in the left lung.

Cardiac catheterization and angiography was performed. The mean right atrial pressure was 7 mm Hg and right ventricular pressure was 72/7 mm Hg. The pulmonary artery pressure was 72/30 mm Hg with a simultaneous left ventricular pressure of 102/8 mm Hg. There was no gradient across the pulmonic valve. No attempt was made to advance the Swan-Ganz catheter beyond the proximal main pulmonary artery. Right-heart angiography was not performed. Thoracic supravalvular aortography demonstrated normal coronary arteries and hypertrophy of the bronchial arteries to the right lung. There was no evidence of a neovascular capillary pattern in the region of the roots of the great vessels.

Because the patient was in extreme respiratory distress and gravely ill, surgical resection was attempted. An exploratory median sternotomy was performed and the patient was placed on cardiopulmonary bypass. The main pulmonary artery was opened, exposing a large, glistening, pale tan-to-focal gray multilobular mass with the gross appearance of a myxoma (fig. 3). The tumor largely occluded the main pulmonary artery and extended into both the right and left main pulmonary arteries (fig. 4). Analysis of a frozen section of the tumor indicated a spindle-cell sarcoma. A cleavage plane was established between the tumor mass and the pulmonary artery wall in the pulmonary trunk and left main pulmonary artery. When the main pulmonary artery was mobilized, the tumor was seen to be growing through the posterior wall and involved adjacent lymph nodes. A cleavage plane could not be established in the right main pulmonary artery, and because the tumor extended so far distally, a right pneumonectomy was performed.

The microscopic appearance included spindle forms

![Figure 1. A stop-frame, short-axis echocardiographic view of the great vessels at the base of the heart. (A and A') A normal pulmonary artery (labeled and unlabeled, respectively). (B and B') The tumor mass (T) fills up the main pulmonary artery (MPA) and extends down into the right and left pulmonary arteries (RP and LP). The smooth, encapsulated tumor mass can be seen in the immediate supravalvular area of the pulmonary artery. The arrow in panel B indicates the pulmonary valve. a = anterior; ao = aorta; l = left; p = posterior; pv = pulmonic valve; r = right; rvo = right ventricular outflow tract.](image1)

![Figure 2. Computed axial tomography scan sectioned at the plane of the great vessels. The images were taken after injection of Renografin. The aorta (A) is well outlined. The tumor mass nearly occludes the main pulmonary artery (PA) and extends well into the right pulmonary artery. A small rim of dye is visible laterally.](image2)
were diagnosed by angiography antemortem. All of these 15 patients had a major filling defect in the main pulmonary artery.

The gross pathology reveals a multinodular tumor fixed at the level of the pulmonary artery valve or base of the pulmonary artery trunk. The tumor often fills the vessel as a solid mass and usually extends to and beyond the bifurcation of the main pulmonary artery. Multiple microscopic patterns have been described, including undifferentiated pleomorphic sarcoma, leiomyosarcoma, rhabdomyosarcoma, fibrosarcoma and myxosarcoma.1-10

Our patient is interesting for several reasons. He represents the youngest reported case of a primary pulmonary sarcoma involving the pulmonary trunk. He is also the first patient in whom a tumor of the pulmonary artery trunk was diagnosed noninvasively and confirmed with surgery and pathology. The noninvasive diagnosis led to rapid surgical and chemotherapeutic intervention, and the patient is still alive 16 months after the initial diagnosis, a longer duration of survival than most patients with primary pulmonary artery sarcoma.

The real advantage of two-dimensional echocardiography over M-mode echocardiography is the ability to investigate cardiac and great vessel anatomy tomographically. Excellent reviews of this technique, including image orientation and structure identification have been published.11, 13-15 The ability of two-dimensional echocardiography to tomographically visualize the heart systematically from many acoustical windows allows the technique to aid in a differential diagnosis of possible cardiac abnormalities suspected on the basis of a careful history and physical examination of the cardiovascular system.

In our patient, cardiac auscultation revealed a murmur suggestive of mitral regurgitation and an ejection murmur localized to the cardiac base that increased in intensity with inspiration. In view of the patient's right ventricular hypertrophy on the ECG, the differential diagnosis of pulmonary artery ejection murmur included valvular pulmonic stenosis, a flow murmur in con-

**FIGURE 3.** Operative photograph showing the tumor mass protruding through the incision of the main pulmonary artery, just distal to the pulmonary valve (tumor). RV = right ventricle.

and epithelioid-appearing cells. The tumor seemed to arise from the pulmonary artery wall and focally extend through the adventitia. Tumor was present in mediastinal soft tissue and one mediastinal lymph node. The tumor was diagnosed as a pleomorphic spindle-cell sarcoma. The patient subsequently underwent chemotherapy. He has returned to a nearly normal lifestyle and has had no recurrence of major symptoms.

**Discussion**

Primary sarcoma of the pulmonary arteries is very rare. Bleisch and Kraus's review provides a composite picture of this malignancy.1 Females outnumber males 2:1, and most patients are in their sixth decade. To date, no patient younger than 22 years old has been reported. The mean duration of symptoms is usually 10 months; chest pain, cough and hemoptysis are the most common complaints. In Bleisch and Kraus's series of 60 cases, 61% of the patients had a systolic murmur without definite localization.1 The chest radiograph is abnormal in the majority of patients; a hilar mass or cardiac enlargement is present in 83% of the patients. Some patients show peripheral pulmonary hypovascularity. One-third of the patients demonstrate right ventricular hypertrophy by ECG. Fifteen of 60 patients

**FIGURE 4.** A gross pathologic specimen of the tumor illustrates its size and shows the outline of the tumor involving the bifurcation of the pulmonary arteries.
junction with a left-to-right shunt, and a subvalvular or supravalvular obstruction to the right ventricular outflow tract. The possibility of a secundum atrial septal defect with mitral valvular regurgitation due to prolapse was our initial working hypothesis. The two-dimensional study excluded the presence of either a shunt or valvular pulmonic stenosis and led to the proper identification of the lesion in the pulmonary artery.

At echocardiography, the cause of the mass in the main pulmonary artery trunk was unclear. The site of origin and size of the mass were determined by computerized axial tomography. These two anatomic, noninvasive techniques often complement each other in accurately diagnosing cardiovascular pathologic states. The physicians involved in the patient’s care chose to have a limited heart catheterization performed, although it was not essential for planning therapeutic interventions.

References

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