The Heart Surgery Forum #2009-1123
13 (1), 2010 [Epub February 2010]
doi: 10.1532/HSF98.20091123

Online address: http://cardenjennings.metapress.com

A Rare Reason for Pulmonary Hypertension: Primary Sarcoma of the Pulmonary Artery

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ABSTRACT

Primary sarcoma of the pulmonary artery (PSPA) is an extremely rare tumor of the cardiovascular system. The prognosis is very poor. The clinical symptoms and imaging findings imitate those of pulmonary emboli, causing delays in diagnosis. In this case report, we describe a 73-year-old man with PSPA who initially was admitted with exertional shortness of breath. Transthoracic echocardiographic evaluation revealed 2 masses in the pulmonary artery causing pulmonary hypertension. The patient underwent operation, but he could not be weaned off cardiopulmonary bypass at the end of the operation and died. Pathologic examination of the masses revealed pulmonary sarcoma. Although this patient was admitted to our clinic only 2 weeks after the initial symptoms, he already had distal metastases.

INTRODUCTION

Primary sarcoma of the pulmonary artery (PSPA) is an extremely rare tumor of the cardiovascular system. These tumors generally originate from the main or proximal pulmonary arteries [Delany 1993]. They grow inside the lumen of the pulmonary artery, and the symptoms appear when the tumor obstructs the pulmonary artery or its branches. Usually, the symptoms are dyspnea, chest pain, cough, or hemoptysis. The clinical symptoms and imaging findings imitate those of pulmonary emboli, causing delays in diagnosis. The prognosis is very poor, and the survival time without surgical intervention is 1½ months [Kotooka 2003]. In this case report, we describe a 73-year-old man with PSPA who initially was admitted with exertional shortness of breath.

CASE REPORT

A 73-year-old man with a medical history of type 2 diabetes mellitus, hypertension, and Parkinson disease presented with symptoms of progressive exertional shortness of breath of 2 weeks' duration. On admission, the patient's blood pressure was 140/90 mm Hg, his heart rate was 78 beats/min, and a systolic murmur was heard in the pulmonary area. The erythrocyte sedimentation rate was 17 mm/h, and the results of a complete blood count, hepatic function tests, and a d-dimer test were normal. The patient had compensated chronic renal failure. His electrocardiography examination revealed marked right atrial and ventricular enlargement, suggesting increased pressure in a pulmonary artery. A transthoracic echocardiography evaluation revealed left ventricular hypertrophy, right atrial and ventricular dilatation, moderate tricuspid insufficiency, and mild pericardial effusion. The maximum pulmonary artery pressure was 104 mm Hg. Two hyperechogenic masses causing significant obstruction were demonstrated in the pulmonary artery (Figure 1). A contrast-enhanced computed tomography (CE-CT) examination revealed 2 polypoid masses in the main and right pulmonary artery. Anticoagulant therapy was started upon patient admission. During the follow-up, the pulmonary artery pressure did not decrease, and a venous Doppler examination of the lower extremities revealed no findings of deep venous thromboembolism.

Biopsy and surgery procedures were planned on the basis of these data. A coronary angiogram showed 3-vessel

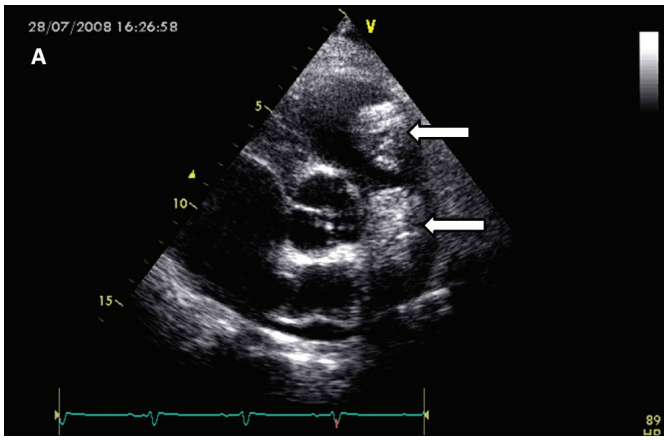

Figure 1. Transthoracic echocardiogram demonstrating 2 masses in the pulmonary artery (arrows), causing pulmonary hypertension.

Received September 4, 2009; accepted September 25, 2009.

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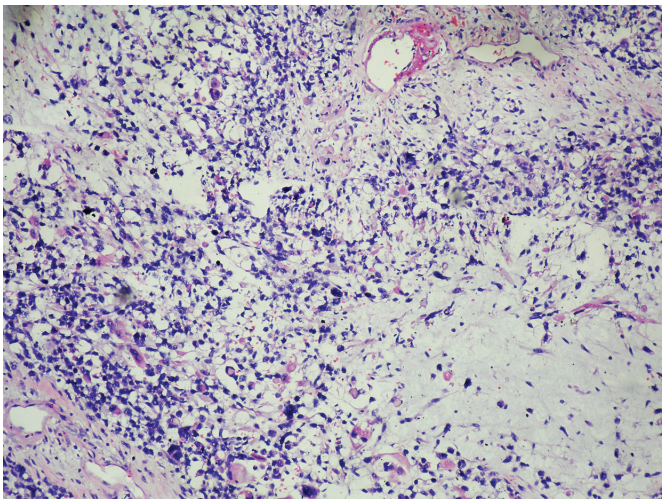

Figure 2. Low-power view of rhabdomyosarcoma with alternating cellular and myxoid areas. This tumor was composed predominantly of primitive ovoid cells with scattered rhabdomyoblasts. The rhabdomyoblasts in this case had eccentric vesicular nuclei and abundant densely eosinophilic cytoplasm.

disease. Reversible ischemia was established in the apex and anteroseptal wall of the left ventricle with dobutamine stress echocardiography. Catheterization of the right heart was not performed because of the risk of pulmonary embolism. The operation was performed via a median sternotomy with aorto-bicaval cannulation and the standard technique of cardiopulmonary bypass with moderate hypothermia (30°C-32°C). Myocardial protection was provided with anterograde intermittent crystalloid or cold blood cardioplegia, combined with topical cooling. A solid tumor invading the pulmonary valve, as well as other tumors in the left and right pulmonary arteries, was found in the main pulmonary artery. Because a frozen section suggested sarcoma, the mass in the main pulmonary artery was resected along with the pulmonary valve; the other tumors were excised from the left and right pulmonary arteries. The pulmonary artery incision was extended to the distal segment of the right and left pulmonary arteries. Exploration of the pulmonary artery revealed a number of small masses. The right ventricular outflow tract was repaired with a composite prosthetic graft. The right and left pulmonary arteries were enlarged with a pericardial patch. Before the replacement of the composite prosthetic graft, coronary artery bypass grafting was performed for 3 vessels. During the operation, the patient's pulmonary artery pressure was higher than the systemic arterial pressure. Because of right ventricular failure, the patient could not be weaned off cardiopulmonary bypass at the end of the operation and died.

The pathologic features of the resected masses were characterized. The pathologic examination of the resected masses revealed multiple fragments of soft gray brown tissue, measuring 7.0 × 4.0 × 2.0 cm in aggregate. Cut surfaces revealed gray yellow-brown, focally irregular solid areas. Microscopically, the tumor showed alternating cellular and myxoid areas predominantly composed of primitive ovoid cells with scattered rare rhabdomyoblasts (Figure 2). This tumor also displayed foci of chondroid and osseous metaplasia. Immunohistochemical studies demonstrated the tumor cells with diffuse labeling of desmin and focal labeling of smooth muscle actin. The labeling of Ki-67 was high. The final diagnosis given by our pathology department was PSPA, rhabdomyosarcoma being very likely.

DISCUSSION

PSPA is a very rare tumor of the cardiovascular system, with an incidence of 0.001% to 0.003% [Miura 2005]. The age range is 13 to 86 years, and the majority of patients are between 45 and 55 years of age [Matotoo 2002]. Most pulmonary artery sarcomas arise from the dorsal area of the pulmonary trunk, although such tumors also may arise from the right and left pulmonary arteries, the pulmonary valve, and the right ventricular outflow tract. The symptoms are usually related to right ventricular outflow obstruction and can include hemoptysis, dyspnea, cough, chest pain, swelling of the lower extremities, and loss of consciousness. Our patient presented with exertional shortness of breath and severe pulmonary hypertension. The clinical symptoms and radiologic findings are similar to those of pulmonary emboli, which consequently can cause delays in diagnosis [Çakır 2005]. A lack of the predisposing factors for thromboemboli, the persistence and recurrence of symptoms despite adequate anticoagulation therapy, a hilar mass in the chest radiograph, and unilateral distribution of a massive perfusion defect can aid in the differential diagnosis.

A diagnostic algorithm is still not clear for pulmonary masses. Echocardiographic evaluation was chosen as the first diagnostic modality in this patient, and the second was CE-CT. Recent reports have emphasized the importance and reliability of noninvasive techniques in the detection of intracardiac masses [Nanda 1977; Arslan 1996]. The patient underwent operation on the basis of the diagnostic data. Three mass lesions were excised from the main, right, and left pulmonary arteries. Coronary artery bypass grafting was performed for 3 vessels. At the end of the operation, the patient's pulmonary artery pressure was higher than the systemic arterial pressure. The patient could not be weaned off cardiopulmonary bypass at the end of the operation and died.

Tumor embolization from the pulmonary artery is a commonly recognized phenomenon and presents as distal metastases and with compromised blood flow to distal branches of the pulmonary artery and structures [Sandhu 2008]. The PSPAs enlarge throughout the lumen of the vessel. When the mass enlarges and obstructs the pulmonary artery, it can produce manifest symptoms. Although this patient was admitted to our clinic 2 weeks after the initial symptoms, he already had distal metastases. In conclusion, both misdiagnosis of the disease as pulmonary emboli and the late manifestation of symptoms contribute to the poor prognosis for PSPA patients.

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