CASE REPORTS

Preoperative Angiographic Diagnosis of Primary Sarcoma of the Pulmonary Artery

JOHN K. HYNES, M.D., HUGH C. SMITH, M.D., DAVID R. HOLMES, JR., M.D.,
WILLIAM D. EDWARDS, M.D., TITUS C. EVANS, JR., M.D., AND THOMAS A. ORSZULAK, M.D.

SUMMARY  Primary neoplasms of the pulmonary artery are extremely rare and usually are not recognized until autopsy. We present a case in which the correct preoperative diagnosis was established by cardiac catheterization. The hemodynamic and angiographic findings of sarcoma of the pulmonary artery are distinct and should not be overlooked in the evaluation of right ventricular outflow tract obstruction.

PRIMARY malignant lesions of the pulmonary artery are extremely rare; to our knowledge, only 68 cases have been reported previously.1-9 Most of these case reports were based on autopsy study, and in only 17 of these patients was the correct antemortem diagnosis suggested. We describe a patient with primary leiomyosarcoma of the pulmonary trunk in whom the preoperative diagnosis was correctly suggested by cardiac catheterization.

Case Report

A 51-year-old man was referred to the Mayo Clinic for evaluation and treatment of presumed massive pulmonary thromboembolism. The patient was an active farmer with a smoking history of 25 pack-years. He had no history of leg trauma, deep vein thrombosis or hospitalization. Twelve months before the current examination, the patient had noted the onset of progressive dyspnea on exertion. Six weeks before admission, the patient had complained of right pleuritic chest pain, scant hemoptyes, chronic cough, night sweats, anorexia and a weight loss of 9.1 kg. Immediately before referral, right-heart catheterization performed with injection of contrast medium into the right atrium demonstrated a large filling defect in the pulmonary trunk, interpreted to be a "saddle embolus."

Physical examination revealed that the patient was dyspneic at rest and had a blood pressure of 135/75 mm Hg and a regular pulse of 100 beats/min. There were prominent "V" waves in the jugular venous pulse. A sustained right ventricular lift was present. The pulmonary valve closure sound was accentuated. A grade 3 pulmonary ejection murmur and a grade 2 tricuspid insufficiency murmur were evident. The abdominal examination was unremarkable. No peripheral edema, adenopathy, cyanosis, clubbing or evidence of deep vein thrombosis was noted.

Laboratory data included the following: hemoglobin 11.7 g/dl, total iron-binding capacity 208 g/dl, serum iron 34 g/dl, and iron saturation 16%. Results of the 12-channel chemistry group testing, including hepatic and renal function tests, were normal. The ECG showed sinus tachycardia without evidence of right ventricular hypertrophy. Arterial blood gases (breathing room air) were PaO2, 73 mm Hg, PaCO2, 34 mm Hg, pH 7.49 and plasma HCO3, 25 mEq/l. Chest roentgenography and tomography of the right lung demonstrated an irregular, thick-walled, cavitary lesion in the superior segment of the right lower lobe (fig. 1). Sputum cultures for acid-fast bacilli, fungi and pathogenic bacteria were negative. Results of bronchoscopy with brushings and transbronchoscopic biopsy in the region of the lesion in the right lower lobe were nondiagnostic. A search for an occult malignant lesion, which included proctoscopic examination, excretory urography and complete gastrointestinal radiographic studies, yielded normal findings.

The patient’s clinical course deteriorated despite administration of full-dose i.v. anticoagulant therapy. Right-heart catheterization was repeated for better definition of the obstructing pulmonary arterial lesion before anticipated thrombolytic or surgical therapy.

Catheterization was performed through the right femoral vein. After cineangiography of the right iliac vein and inferior vena cava showed no evidence of thrombus formation, a catheter was advanced into the right-heart chambers. The preoperative hemodynamic data are shown in table 1. The right ventricular and pulmonary arterial pressures were moderately elevated. There was a significant pressure gradient between the pulmonary trunk and the proximal right and proximal left pulmonary artery. Before pulmonary cineangiography, the catheter was repeatedly passed beyond the area of obstruction without change in the pressure gradient. Pulmonary cineangiography demonstrated a striking, lobulated, 3-cm-diameter, spherical mass near the origin of the left pulmonary artery. During each cardiac cycle, this mass had a "to-and-fro" movement that resembled a pedunculated polyp tethered to the pulmonary artery in this region. There were numerous filling defects in the right pulmonary lobar arteries and virtually no flow to the right lower lobe. Biplane pulmonary angiography was then performed for better anatomic delineation of the mass lesion (fig. 2).

A presumptive diagnosis of primary sarcoma of the pulmonary artery was made on the basis of the unique

From the Division of Cardiovascular Diseases and Internal Medicine and the Section of Thoracic, Cardiovascular, Vascular and General Surgery, Mayo Clinic and Mayo Foundation, Rochester, Minnesota.
Address for correspondence: J.K. Hynes, M.D., c/o Section of Publications, Mayo Clinic, Rochester, Minnesota 55905.
Received October 23, 1981; revision accepted January 21, 1982.
angiographic appearance, striking motion abnormality of the mass lesion detected by cineangiography and reproducible pressure gradient demonstrated by manipulation of the catheter proximal and distal to the obstructing lesion.

At surgical exploration, the right pulmonary artery was described as "tense, full of solid material, and without pulsation," and the diagnosis of sarcoma of the pulmonary artery was confirmed. The histologic interpretation was grade 3 leiomyosarcoma of the pulmonary trunk. There was no evidence of hilar or mediastinal lymph node metastases. The neoplasm involved the entire length of the pulmonary trunk, but not the pulmonary valve, and extended into the entire right pulmonary artery and proximal left pulmonary artery. Resection of all involved portions of the pulmonary artery was not possible. Tumor extensions into the lobar arteries were removed with forceps, and a proximal pulmonary endarterectomy was performed (fig. 3).

The postoperative course was unremarkable. Right-heart catheterization was performed on the ninth postoperative day. The postoperative hemodynamics (table 1) showed essentially normal pulmonary pressures and a small residual gradient between the pulmonary trunk and distal left pulmonary artery. Repeat pulmonary angiography (fig. 4) revealed persistent occlusion of the right lower lobe artery and absence of flow to the right lower lobe.

The patient was dismissed from the hospital on the tenth postoperative day to recover more fully from his operation before consideration of adjuvant chemotherapy. One month later, the patient required abdominal exploration at his local hospital for obstruction of the small bowel; a small leiomyosarcoma was resected from the jejunum. There was no evidence of regional lymph node spread or hepatic metastasis. The patient declined further medical attention. Nine months after the initial thoracic operation, he had signs of respiratory insufficiency and right ventricular failure and died. Permission for autopsy was not granted.
The diagnosis of primary sarcoma of the pulmonary artery is seldom made before autopsy. Pulmonary angiography has been performed in 17 cases and has revealed a large mass in all instances. Most of the patients who underwent catheterization, however, were referred for thoracotomy, with a diagnosis of pulmonary embolism or metastatic neoplasm. We report a case of pulmonary artery leiomyosarcoma diagnosed at the time of catheterization. The unusual cineangiographic appearance of a large, mobile, lobulated mass in the proximal pulmonary arteries associated with a persistent pressure gradient obtained by repeated manipulation of the catheter past the obstruction without fragmentation of the lesion suggested a primary neoplasm of the pulmonary artery rather than a pulmonary thromboembolus or a tumor metastasis.

Although our patient had a small leiomyosarcoma of the jejunum, the pulmonary arterial lesion clearly resembled the "solid sarcomatous pulmonary artery" described by Jacques and Barclay. The bulky neoplasm was attached to the intima of the pulmonary trunk and had spread as an intraluminal mass. This location and appearance, to our knowledge, have not been reported previously in hematogenous metastasis of leiomyosarcoma. Kovacs et al. described a patient with a primary leiomyosarcoma of the pulmonary artery that had metastasized to the jejunum. In our patient, however, the absence of regional lymph node involvement or hepatic metastasis suggests that the jejunal and pulmonary arterial leiomyosarcomas are independent primary tumors.

Since the original description of primary pulmonary arterial sarcoma by Mandelstamm in 1923, 67 additional cases have been reported. Shmookler et al. and Bleisch and Kraus summarized the findings in 60 case reports. Our more recent search of the literature identified an additional eight reported cases. The female-to-male sex ratio is approximately 2:1, and the median age at the time of diagnosis is 52 years. Characteristically, these patients present with alarming symptoms suggestive of occult malignancy and signs of tumor emboli in the distal pulmonary artery from the larger and more proximal primary sarcoma. The mortality rate 1 year after the onset of symptoms exceeds 80%.

Of the 17 patients who have undergone thoracotomy and partial or complete resection of the primary neoplasm of the pulmonary artery, five have had recurrences. Chemotherapy has been administered in four patients and radiation therapy in four patients—all without response.

Sarcoma of the pulmonary artery may mimic several more common clinical problems, including pulmonary thromboembolism, pulmonary hypertension, fibrosing mediastinitis, tuberculosis, metastatic carcinoma and obstructive lung disease with cor pulmonale. Early diagnosis is rare because symptoms of systemic illness are often ignored by the patient, while the tumor grows insidiously until distal tumor embolism or severe pulmonary arterial obstruction occurs. The poor prognosis apparently results from the delay in clinical diagnosis and the subsequent inability to resect all segments of involved pulmonary artery. Cardiac catheterization and pulmonary angiography should offer definitive preoperative diagnosis. Use of noninvasive imaging techniques such as two-dimensional echocardiography may adequately demonstrate the proximal pulmonary artery and allow for an earlier diagnosis.

References
10. Shmookler BM, Marsh HB, Roberts WC: Primary sarcoma of the pulmonary trunk and/or left or right main pulmonary artery—a rare cause of obstruction to right ventricular outflow: report on two patients and analysis of 35 previously described patients. Am J Med 63: 263, 1977
Preoperative angiographic diagnosis of primary sarcoma of the pulmonary artery.
J K Hynes, H C Smith, D R Holmes, Jr, W D Edwards, T C Evans, Jr and T A Orszulak

Circulation. 1982;66:672-674
doi: 10.1161/01.CIR.66.3.672
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1982 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/66/3/672.citation

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation is online at:
http://circ.ahajournals.org//subscriptions/