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Case Reports

Primary Sarcoma of the Pulmonary Trunk: Successful Surgical Intervention and Prolonged Survival

W. P. KLINKE, M.D., F.A.C.P., F.R.C.P.(C), F.A.C.C.,* E. T. GELFAND, M.D., M.S., F.R.C.S.(C), F.A.C.C., F.C.C.P.,†
L. BARON, M.D.†

*Royal Alexandra Hospital and †University of Alberta Hospital, Edmonton, Alberta, Canada

Summary: We present a rare case of pulmonary trunk sarcoma in a young male in which the diagnosis was made antemortem. Total surgical removal of the tumor, including replacement of the pulmonary valve, was undertaken. This patient is the longest reported survivor of pulmonary trunk sarcoma and remains free of any signs or symptoms of recurrence. No adjuvant chemotherapy was required.

Key words: pulmonary trunk, sarcoma, cardiac surgery

Introduction

Primary sarcoma of the pulmonary artery is a rare tumor. We report a case in which we were able to make a diagnosis soon after the onset of symptoms. Successful resection of the tumor involved replacement of the pulmonary valve and a right ventricular (RV) outflow tract pericardial patch.

Address for reprints:

W. P. Klinke, M.D.
Division of Cardiology
Royal Alexandra Hospital
10240 Kingsway Avenue
Edmonton, Alberta
T5H 3V9 Canada

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Case Report

A 39-year-old male was seen for assessment of dizziness of approximately 4 months duration and two syncopal episodes occurring with exertion. He had no complaints of angina pectoris nor of symptoms of congestive heart failure or cardiac arrhythmias. Two months prior to the onset of symptoms the patient was noted to have a new heart murmur by his own physician.

Physical examination revealed a young acyanotic male in no acute distress. Respiratory rate was 20/beats/min and resting heart rate was 76/beats/min and regular. Blood pressure was 100/70 mmHg in both arms. There was no jugular venous distention nor any peripheral edema. Palpation of the precordium failed to reveal any significant thrills or heaves. Auscultation revealed a normal first heart sound. A notably diminished second heart sound was detected as well as an S_4 gallop along the sternal border. A grade III/VI systolic murmur was identified maximally along the left sternal border with radiation toward the left shoulder. No diastolic murmur was heard. The lungs were clear.

The electrocardiogram demonstrated right ventricular hypertrophy with strain. A chest x-ray showed mild cardiomegaly. Routine blood values were within normal limits.

An M-mode echocardiogram was interpreted as showing normal left ventricular (LV) function but an enlarged RV measuring 31 mm in diameter. The mitral and aortic valves were normal. A two-dimensional echocardiogram study suggested that a mass arose from the septal area and extended into the RV outflow tract. The pulmonary valve and the pulmonary artery (PA) could not be clearly visualized.

Cardiac catheterization with right and left ventriculography was performed. Mean right atrial pressure was 5 mmHg and RV pressure was 94/14 mmHg. The PA was

cannulated with a great deal of difficulty and the PA pressure was 13/8 mmHg. Right ventricular angiography demonstrated an enlarged and trabeculated chamber with posterior displacement of the LV. An ovid mass was seen to arise from the septum relatively high on the posterior wall. The pulmonary valve was not clearly identified, but a lobulated mass was seen in the midportion of the pulmonary trunk (Fig. 1). Left heart catheterization and coronary angiography were normal. A presumptive diagnosis of pulmonary trunk sarcoma was made and the patient was referred for immediate surgery.

The patient underwent surgical resection utilizing a median sternotomy and total cardiopulmonary bypass. Palpation fo the PA revealed a hard mass within the lumen. The PA overlying the mass was opened, revealing a lobulated tumor attached to the anterior cusp of the pulmonary valve (Fig. 2). There were two main portions to the mass, and therefore, the PA incision was extended into the RV outflow tract across the valve. Excision of the tumor involved removal of the anterior leaflet of the pulmonary valve and a corner of an additional leaflet, thereby necessitating a pulmonary valve replacement. A #25 Carpentier-Edwards porcine prosthesis was chosen and sutured into place. A pericardial patch was sutured in place beginning at the apex of the RV outflow incision and continuing onto the PA, and the suture lines were then completed. The patient recovered without incident.

On pathologic examination the tumor measured 3.5 × 3.0 × 2.5 cm and was glistening pale yellow in appearance (Fig. 3). The section revealed a soft yellow to pale grey-white tissue and a small focus of hemorrhage within the tumor. Light microscopy showed a polypoid spindle-cell sarcoma which contained bizarre cells with large hyperchromatic nucleii among the spindle-cell element. Heavy chronic inflammatory cell infiltrate was noted. No invasion of adjacent myocardium was seen. The tumor was diagnosed as a polypoid sarcoma of the pulmonary trunk.

Upon discharge from the hospital, the patient was investigated at a local cancer clinic where further investigations revealed neither any other potential sites of primary sarcoma nor any evidence of metastases. Adjuvant chemotherapy and radiotherapy were not considered necessary. Repeat cardiac catheterization 6 weeks later demonstrated an RV pressure of 34/3 mmHg and a PA pressure of 18/7 mmHg, the resulting gradient due to the prosthetic valve. No angiographic evidence of residual tumor was present. The patient has done well without evidence of recurrence of tumor 18 months postoperatively.

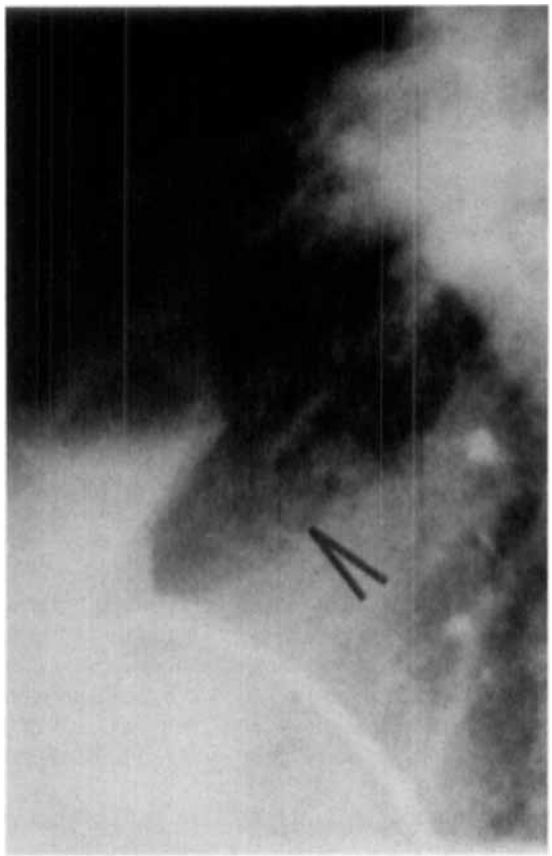

FIG. 1 Lobulated mass filling lumen of main pulmonary artery.

Discussion

Polypoid sarcoma of the pulmonary trunk is an exceedingly rare tumor (Bleisch and Kraus, 1980). In most instances, diagnosis has been at postmortem with only 19 of 70 patients diagnosed antemortem. Successful diagnosis and therapy has been uncommonly accomplished (Hynes *et al.*, 1982; Shmookler *et al.*, 1977; Wright *et al.*, 1983).

The mortality rate of primary sarcoma of the pulmonary trunk after onset of symptoms exceeds 80% at one year. This poor prognosis relates to both the critical location of the tumor mass and to its malignant nature. Delay in diagnosis and subsequent inability to resect all segments of the tumor is associated with recurrence of the tumor and poor prognosis.

In this patient the development of exertional syncope in the presence of a new cardiac murmur led to further investigation with two-dimensional echocardiography and cardiac catheterization and angiography. Findings of these investigations were considered to be typical for sarcoma of the pulmonary trunk.

Complete removal on cardiopulmonary bypass included excision of the pulmonic valve, a sizable portion of the RV outflow tract and PA, necessitating an outflow patch, and replacement with a prosthetic pulmonary valve. Grossly and histologically, it was felt that complete extirpation of the tumor had been accomplished. Unlike

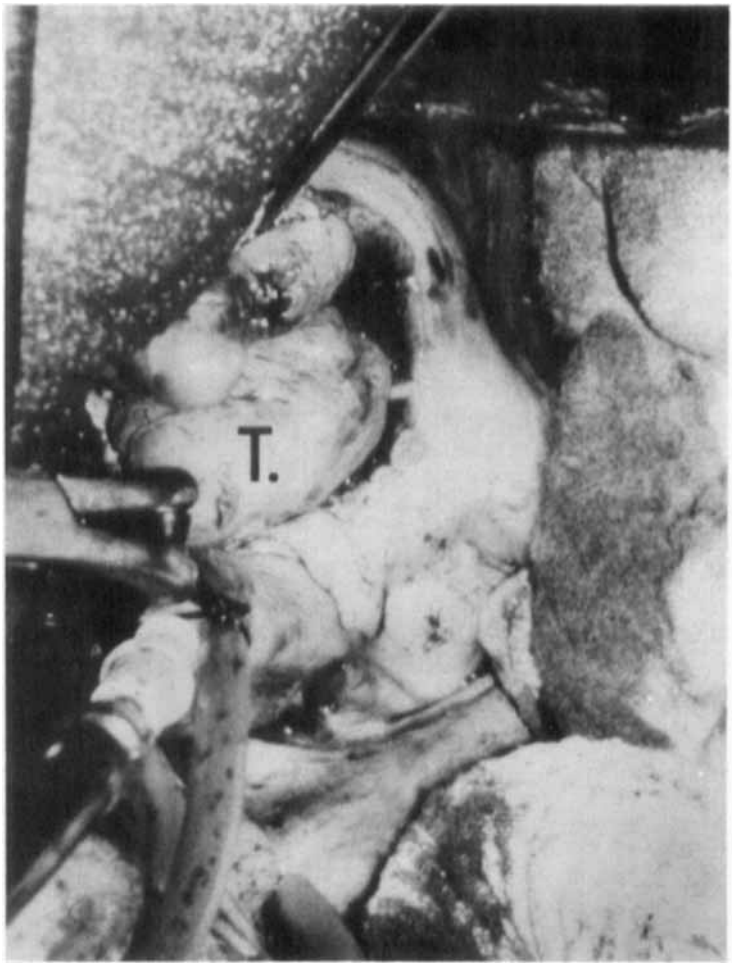

FIG. 2 Operative photograph showing opened right ventricular outflow tract with bulging tumor (T).

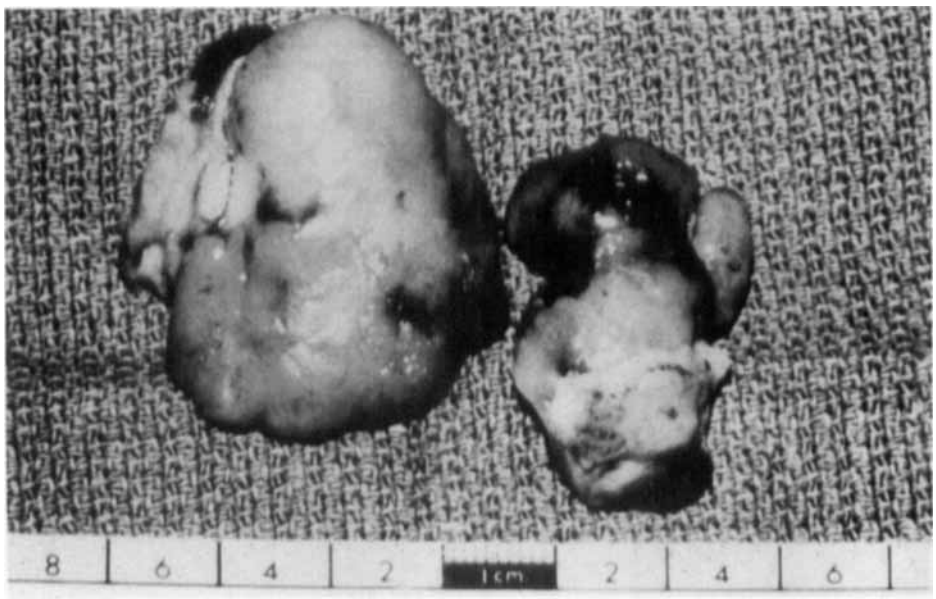

FIG. 3 Excised tumor specimen.

previously reported cases, our patient has not had a recurrence of tumor or required additional therapy. Our patient is the longest reported survivor of pulmonary trunk sarcoma.

Addendum

The patient was readmitted to the hospital 24 months postsurgery for evaluation of symptoms of shortness of breath and presyncope which had reoccurred in the preceding month. Clinical examination revealed a very loud systolic ejection murmur in the pulmonic area. Repeat right heart angiography demonstrated a recurrence of the pulmonary sarcoma in the main pulmonary artery and extending up into the right pulmonary artery. Another operation was undertaken with complete excision of a recurrent tumor which appeared to be arising from the aortic side of the pulmonary artery near the pulmonary valve prosthesis. The tumor mass measured 2.5×2.5 cm and proved to be a spindle-cell tumor with low mitotic figures. A Dacron graft prosthesis was sutured in place. The CT scans of the thorax and liver and bone scans failed to demonstrate any evidence of metastatic disease. In an effort to prevent future recurrence, the patient was referred to the Berkeley-Lawrence Laboratory at Stanford for heavy ion radiation therapy.

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