A 44-year-old man was hospitalized after a syncopal episode. A thrill and loud systolic ejection murmur were present over the left border of the sternum. A 12-lead ECG and chest x-rays were normal. A 2D echocardiogram and Doppler studies documented normal cardiac chambers and a peak systolic pulmonary transvalvular gradient of 80 mm Hg. Biplane right ventriculography revealed a large mobile mass in the main pulmonary artery extending into the right ventricular outflow tract (Figure 1). Cardiac MRI showed the mass within the same area (Figure 2 and Figure I, which can be found at www.circulationaha.org). The patient underwent surgical resection of a large tumor in the main pulmonary artery (Figure 3). The tumor extended into the right pulmonary artery. The pulmonary valve was replaced with a homograft, and an endarterectomy of the main and right pulmonary arteries was accomplished. Histological examination revealed a spindle cell sarcoma (Figures 4 and 5). The patient remained asymptomatic and had no cardiac murmurs 8 months after surgery.1

Reference

Figure 1. Biplane right ventriculography reveals a large mobile mass in the main pulmonary artery that extends into the right ventricular outflow tract (arrow).

Figure 2. MRI taken before (left) and after (right) administration of gadolinium. Images were obtained in the right ventricular outflow tract projection. A large, enhancing mass (arrows) is present in the main pulmonary artery and upper outflow tract.
Figure 3. Surgical resection of the large tumor in the main and right pulmonary arteries.

Figure 4. The tumor shows features of highly pleomorphic cell malignant neoplasm, with numerous mitotic figures, some of which are atypical (arrow). Movat’s Pentachrome stain, ×360.

Figure 5. The tumor occupies the lumen of the pulmonary artery and does not invade its wall. The internal elastic lamina (arrow) is not interrupted. Movat’s Pentachrome stain, ×27.
Syncope in a Patient With Spindle Cell Sarcoma of the Main Pulmonary Artery
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Circulation. 2001;103:e99-e100
doi: 10.1161/01.CIR.103.20.e99

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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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:
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