A 60-year-old white woman presented with a history of exertional shortness of breath rapidly progressing to orthopnea and episodes of paroxysmal nocturnal dyspnea over a period of 4 weeks. Her past medical history included hypertension; sick sinus syndrome, for which she had received a pacemaker; and a left carotid endarterectomy. Physical examination revealed normal vital signs, jugular venous distension of 5 cm, bibasilar rales, and a II/VI middiastolic murmur heard at the apex of the heart without an opening snap.

A transthoracic echocardiogram revealed a markedly thickened anterior leaflet of the mitral valve with limited diastolic excursion (Figure 1A). A possible mass hugging the atrial side of the anterior leaflet could not be excluded. The posterior leaflet had preserved motion. Doppler echocardiography across the mitral valve obtained a mean gradient of 20 mm Hg, with a peak gradient of 47 mm Hg and a calculated valve area of 1.57 cm² by the pressure half-time method (Figure 1B). The transesophageal echocardiogram delineated a mass involving the anterior leaflet of the mitral valve and extending to the interatrial septum, with consequent obstruction of the valve (Figure 1C and 1D). The left atrium was otherwise normal.

The patient was treated symptomatically for pulmonary edema, and after a coronary angiogram, which revealed single-vessel disease, she underwent exploratory cardiac surgery. During surgery, a large, multilobar, myxomatous mass 4 cm in diameter was seen on the anterior leaflet of the mitral valve. The mass had no distinct stalk, and it extended into the posterior commissure and the interatrial septum (Figure 2A). In addition, islands of fibrinous, gelatinous material that were histologically similar to the mass were also visualized in the left atrium. The patient underwent excision of the mass, mitral valve replacement with a 27-mm St Jude’s valve, and a right coronary artery bypass graft.

Histologically, the mass consisted of a malignant proliferation of spindle cells arranged in storiform pattern with focal myxoid changes. The cells displayed marked nuclear pleomorphism with occasional bizarre cells and frequent mitotic figures, including abnormal forms (Figure 2B and 2C). Immunohistochemistry revealed negative staining for desmin and smooth muscle actin. Tumor cells were strongly immunoreactive for CD-68 (Figure 2D) and focally with desmin. Electron microscopy demonstrated features consistent with a fibrohistiocytic differentiation (Figure 2E). The diagnosis was primary endocardiac malignant fibrous histiocytoma. The patient underwent adjuvant chemotherapy and is alive and well 1 year after surgery.
Figure 1. A, Parasternal long-axis view revealing thickened anterior leaflet of mitral valve (MV) and a normal left atrial (LA) size; B, pressure half-time (PHT) calculation showing mitral valve area of 1.57 cm²; C, transesophageal echocardiogram revealing mass on anterior leaflet of mitral valve; and D, turbulent flow across valve signifying severe obstruction. LV indicates left ventricle; LVOT, LV outflow tract; and PL, posterior leaflet.
Figure 2. A, Grossly, tumor is polypoid, lobulated, and myxomatous; B, low-power micrograph showing atypical spindle cells in myxoid background (hematoxylin-eosin [H&E] stain, magnification ×100); C, micrograph of more cellular areas with pleomorphic spindle cells and abnormal mitotic figure (H&E, ×400); D, tumor cells show intense immunoreactivity for CD68 (brown stain), confirming histiocytic differentiation; and E, electron micrograph demonstrates histiocytic differentiation as evidenced by presence of lysosomes, lipid inclusions, cytoplasmic membranes with blunt pseudopodia, and convoluted nuclei (×6050).
Primary Cardiac Malignancy Masquerading as Mitral Valve Stenosis
Vijay D. Subbarao, Richard G. Sheahan, Vincent R. Conti, Eduardo Eyzaquirre and Masood Ahmad

_Circulation_. 1999;99:2342-2344
doi: 10.1161/01.CIR.99.17.2342

_Circulation_ is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1999 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/99/17/2342

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/