Myxoma of the mitral valve associated with *Hemophilus parainfluenza* bacteremia

FREIDOON GHAZI, MD; RICHARD STERBA, MD; DOUGLAS S. MOODIE, MD; CARL C. GILL, MD; NORMAN B. RATLIFF, MD

■ A 17-year-old girl who presented with signs and symptoms of bacterial endocarditis did not respond clinically to antibiotic therapy. Cerebral embolic episodes subsequently occurred. Serial echocardiography disclosed an enlarging vegetation attached to the posterior leaflet of the mitral valve. During open heart surgery, the vegetation was removed, which on histological examination proved to be a myxoma covered by a layer of septic thrombus. Blood cultures obtained preoperatively eventually grew *Hemophilus parainfluenza*. Removal of the tumor, as well as mitral valve annuloplasty, resulted in complete cure.

□ INDEX TERMS: MITRAL VALVE NEOPLASMS; MYXOMA □ CLEVE CLIN J MED 1988; 55:470–472

MYXOMAS are by far the most common primary tumors of the heart, constituting 70% of all primary tumors in adults.[1] In 75% of cases, these tumors are attached to the intra-atrial septum and protrude into the left atrium. Among 49 cardiac myxomas mentioned by Prichard,[2] one developed on a valve. The following case describes a myxoma that developed on the mitral valve and, unlike most such tumors, became infected. To our knowledge, this case of an infected mitral valve myxoma is the first reported in the English literature. The role of echocardiography in the evaluation of the patient and ultimately successful therapy is discussed.

CASE REPORT

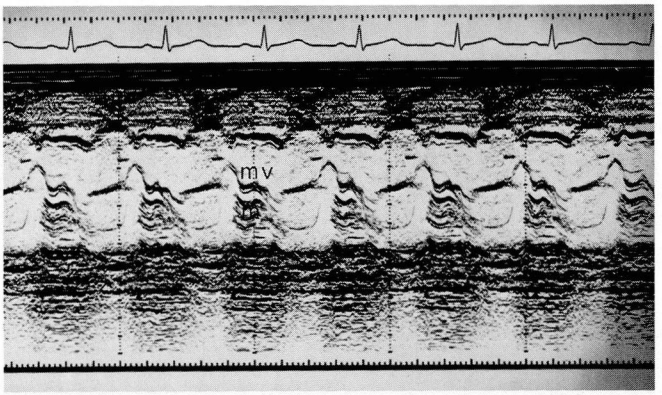
A 17-year-old white girl was admitted to the Cleveland Clinic because of fever of unknown origin. She had been in good health until three weeks prior to admission when she started to have chills, fever, night sweats, and malaise, followed by nausea, vomiting, diarrhea, and left-lower abdominal pain. Her medical history was unremarkable, except for a dental checkup and cleaning three months prior to admission.

Physical examination on admission showed an ill-looking young girl. Blood pressure was 100/80; pulse, 110 and regular; and temperature, 39° C. The pharyngeal mucosa appeared erythematous, with white ulcers on the soft palate. The neck was supple and no significant adenopathy was apparent. The lungs were clear. The heart examination revealed a regular rhythm with normal S1 and S2. A grade II/VI mid to late systolic murmur was heard at the apex with radiation to the axilla. There were no S3, S4, or diastolic murmurs. The rest of the physical examination was unremarkable.

Laboratory studies disclosed the following: hemoglobin, 8.3 g/100 mL; hematocrit, 25.4%; WBC, 13,000 with 71% polymorphonuclear leukocytes, 6% bands, and 19% lymphocytes; erythrocyte sedimentation rate, 50 mm/hour; SMA-18, within normal limits; urinalysis, unremarkable; and cytomegalovirus, Epstein-Barr virus, and mono spot tests, negative. The chest radiograph was

From the Departments of Cardiology (F.G., R.S., D.S.M.) Cardiothoracic Surgery (C.C.G.), and Pathology (N.B.R.), The Cleveland Clinic Foundation. Submitted for publication June 1986; revision accepted Mar 1988.

A



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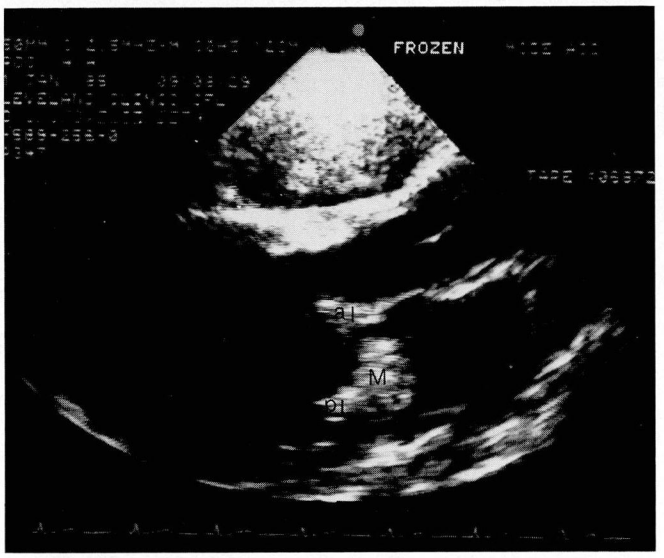


FIGURE 1A. M-mode echocardiogram obtained at the level of the mitral valve. A large echodense mass (arrows) is seen attached to the posterior leaflet of the mitral valve. FIGURE 1B. Two-dimensional echocardiogram (long-axis parasternal view). A large echodense mass is attached to the posterior leaflet of the mitral valve. *m* = mass, *lv* = left ventricle, *mv* = mitral valve, *al* = anterior leaflet, and *pl* = posterior leaflet.

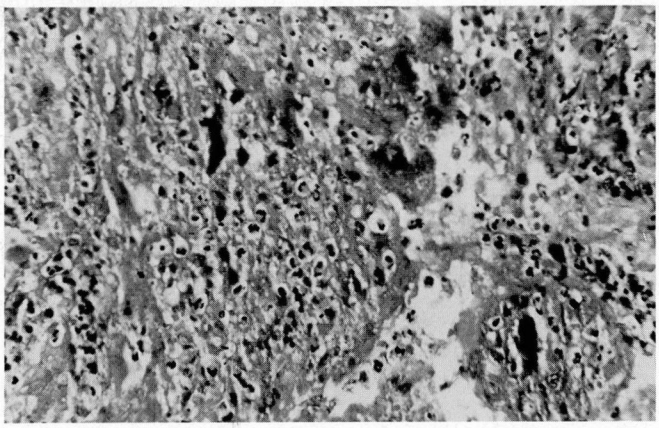
normal. An echocardiogram obtained on admission showed mild mitral valve prolapse without vegetation. Initial blood cultures were negative. A repeat echocardiogram showed some thickening of the anterior leaflet of the mitral valve with no vegetation.

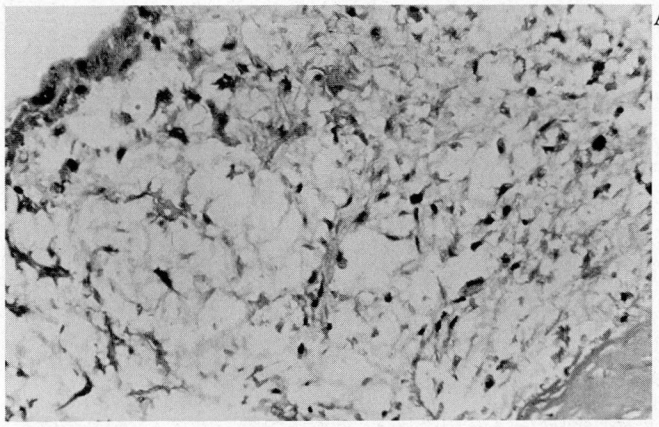
She continued to have fever spikes up to 39° C. On her 10th day in the hospital, a telangiectatic lesion was noted in the left upper chest, as well as petechial lesions on the right lower eyelid. Blood cultures remained negative. She was given vancomycin and gentamicin to treat the culture-negative endocarditis. She continued to experience fever spikes, however. An echocardiogram obtained five days later identified a large echodense mass attached to the posterior leaflet of the mitral valve that was thought to be a large vegetation (*Figure 1*). She continued to do poorly and the systolic heart murmur, consistent with mitral regurgitation, became louder. Two days later, numbness suddenly developed in the left side of her face and left arm down to her fingers, and her speech became labored. This lasted for 10 minutes and never recurred. Another echocardiogram obtained on the same day showed no change in the size of the vegetation. The next day, the patient underwent debridement and partial excision of the posterior leaflet of the mitral valve and annuloplasty with a Carpentier ring. No fibrosis, inflammation, or evidence of destruction of the underlying valves was apparent.

The resected specimen was a round (1 cm in diameter), nonpedunculated mass covered with a shaggy red-yellow exudate. A cross-section of the mass revealed the typical gray-white appearance of a myxoma. Frozen and subsequent permanent sections revealed a typical myxoma composed of branching and anastomosing thin-walled vascular channels in an abundant, afibrillar, slightly basophilic background. The myxoid tissue contained numerous stellate myxoma cells but only occasional polyhedral lepidic and multinucleate tumor cells. Perivascular cuffing and cell clusters were not observed. Microscopic hemorrhage and inflammatory cells, except at the surface, were scant. There was no fibrosis, no inflammation, and no evidence of destrucion of the underlying valve. The relative paucity of multinucleate cells, perivascular cuffing, and cells clusters is in accord with the observation that these features are less prominent in valvular than in atrial myxomas.[3] Covering the myxoma was a layer of thrombus containing multiple pockets of polymorphonuclear leukocytes mixed with reactive debris and gram-positive cocci (*Figure 2*). Tumor cultures were negative.

The postoperative period was uneventful until the second week when a febrile illness developed and lasted approximately three days, with symptoms of upper respiratory infection. (Coinicidentally, reports on the blood cultures that had been drawn during the first week of hospitalization revealed growth of *Hemophilus parainfluenza* [one out of two bottles].) The patient was treated with gentamicin and ampicillin for three weeks. The patient sub sequently recovered and did well throughout the rest of her hospitalization.

Nine months later, an echocardiogram showed no tumor recurrence. She has been enjoying a physically active life since discharge.





A, B

FIGURE 2A. Multiple pockets of polymorphonuclear leukocytes and cellular debris are embedded in a meshwork of fibrin (hematoxylin-eosin, x400). FIGURE 2B. The surface of the myxoma (at top left) is composed of a single layer of cuboidal cells. The body of the myxoma consists of spindle and stellate cells embedded in a loose myxoid stroma (hematoxylin-eosin, x100).

DISCUSSION

Infected atrial myxomas with organisms demonstrated in the tumor and/or systemic circulation are extremely rare and usually are diagnosed at autopsy.[4] Only a few cases of infected atrial myxomas have been reported.[4–6] This case is even more unusual because the tumor was attached to the mitral valve. Sandrasagra et al[7] reported a mitral valve myxoma, but it was not infected.

Clinical manifestations of myxomas of the heart are often similar to those of bacterial endocarditis, and large vegetations attached to the mitral valve leaflets in bacterial endocarditis may resemble left atrial myxomas.[8] Weight loss, fever and malaise, swelling, anemia, leukocytosis, and an increased erythrocyte sedimentation rate, often noted in patients with cardiac myxoma, were all present in our patient. Such signs and symptoms have been explained as a nonspecific response to necrotizing tumor tissues or as an immunologic response to the foreign protein represented by the myxoma.[9] However, these signs and symptoms are also typical of infectious endocarditis and make diagnosis difficult.

The nature of the infecting organism in this case, however, remains uncertain. A single positive blood culture for *Hemophilus parainfluenza* was obtained prior to surgical removal of the tumor, but gram-positive cocci were identified in the tissue itself.

Echocardiography remains the easiest, widely available diagnostic modality for detecting masses within the heart. This case emphasizes the importance of obtaining serial two-dimensional and M-mode echocardiograms of patients suspected of having bacterial endocarditis.

FREIDOON GHAZI, MD
Department of Cardiology
Good Samaritan Hospital
Dixsmyth Avenue
Cincinnati, Ohio 45220

ACKNOWLEDGMENT

We appreciate the assistance of Beverly Kane in the preparation of this manuscript.

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