Mitral Valve Aneurysm: Clinical Features, Echocardiographic-Pathologic Correlations

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Aneurysm of the mitral valve occurs most commonly in association with infective endocarditis of the aortic valve. The probable mechanism of its formation is destruction of the aortic valve which results in a regurgitant jet that strikes the anterior leaflet of the mitral valve, creating a secondary site of infection leading to the development of an aneurysm. Perforation of these aneurysms may occur, resulting in mitral regurgitation and pulmonary edema from a ventricle already volume overloaded from aortic regurgitation.

This report describes the clinical and echocardiographic-pathologic findings in five patients with pathologically proven aneurysm of the mitral valve. There are no clinical features that appear specific for this abnormality. The two-dimensional echocardiographic feature that is helpful in the diagnosis is a bulge of the mitral valve leaflet toward the left atrium that persists throughout the cardiac cycle. Preoperative diagnosis is important because a mitral valve aneurysm may produce serious complications and is frequently overlooked during surgery. Repair of the aneurysm may be feasible; otherwise, valve replacement becomes necessary. Careful two-dimensional echocardiographic examination should be done in patients with left-sided infective endocarditis to detect an aneurysm of the mitral valve.

Results

Five patients seen between 1974 and 1982 who had mitral valve aneurysm with pathologic confirmation are described (Table 1). Four patients had surgical intervention, and three underwent echocardiographic examination.

Clinical features. All patients were male ranging in age from 19 to 79 years (mean 46) and all had infective endocarditis of the aortic valve. The duration of symptoms ranged from 1 to 8 weeks in three patients and was unknown in one patient (Case 4). One died suddenly 18 months after aortic valve replacement. Four patients had symptoms and signs of congestive heart failure on admission to the hospital, and the patient who died suddenly had autopsy evidence of severe pulmonary edema. A history of intravenous drug abuse was obtained in three patients (Cases 2 to 4).

The clinical diagnosis of aortic and mitral regurgitation was made in all five patients at the time of initial examination. In the patient who died suddenly, the diagnosis of valvular regurgitation was made during the previous hospital admission. Chest X-ray films showed increased cardiac size in the four patients in whom they were available and pulmonary venous congestion in two. Left ventricular hypertrophy was present on the electrocardiogram in three patients.

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Table 1. Clinical Characteristics in Five Patients With Mitral Valve Aneurysm

<table>
<thead>
<tr>
<th>Case</th>
<th>Age (yr) &amp; Sex</th>
<th>Duration of Symptoms (wk)</th>
<th>Drug Abuse</th>
<th>CHF</th>
<th>AR</th>
<th>MR</th>
<th>Chest X-ray Film</th>
<th>ECG</th>
<th>Infecting Organism</th>
<th>Clinical Course</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>79M</td>
<td>1</td>
<td>-</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>Cardiomegaly: pulmonary venous congestion</td>
<td>LVH</td>
<td>α-streptococcus</td>
<td>AVR; died 5 hours postoperatively</td>
</tr>
<tr>
<td>2</td>
<td>19M</td>
<td>8</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>Cardiomegaly: pulmonary venous congestion</td>
<td>1° AVB</td>
<td>α-streptococcus</td>
<td>AVR, MVR; died 2 days postoperatively</td>
</tr>
<tr>
<td>3</td>
<td>30M</td>
<td>2</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>Cardiomegaly</td>
<td>LVH</td>
<td>Serratia marcescens</td>
<td>AVR, repair MVA; died 5 days postoperatively</td>
</tr>
<tr>
<td>4</td>
<td>36M</td>
<td>UK</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>Cardiomegaly</td>
<td>LVH</td>
<td>Candida parapsilosis</td>
<td>AVR 18 mo before sudden death</td>
</tr>
<tr>
<td>5</td>
<td>65M</td>
<td>UK</td>
<td>-</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>NA</td>
<td>NA</td>
<td>α-streptococcus</td>
<td>Died before surgery</td>
</tr>
</tbody>
</table>

* Autopsy examination  † Previous hospital admission

AR = aortic regurgitation; AVB = atrioventricular block; AVR = aortic valve replacement; CHF = congestive heart failure; ECG = electrocardiogram; LVH = left ventricular hypertrophy; M = male; MR = mitral regurgitation; MVA = mitral valve aneurysm; MVR = mitral valve replacement; NA = not available; UK = unknown; + = present; − = absent

Findings at surgery. Four patients had aortic valve replacement for severe valvular regurgitation and congestive heart failure caused by their infective endocarditis; one patient (Case 5) died shortly after hospital admission from severe congestive heart failure before surgery could be done. The aortic valves in all four patients had vegetations with extensive destruction of the valve leaflets. The aortic valve was replaced with a porcine prosthesis in three patients and a mechanical prosthesis in one. Three patients died within 10 days of surgery; the fourth patient died suddenly 18 months after replacement of his aortic valve.

The mitral valve aneurysms were not noted at surgery. In one patient who died from postoperative bleeding (Case 1), the mitral valve had been visualized through an atriotomy and, although the mitral valve was noted to be mildly incompetent, no structural abnormality was found. In another patient (Case 3), repair of a mitral valve perforation that had been visualized through an aortotomy was performed. The aneurysm was not diagnosed at surgery and this patient subsequently died postoperatively from intractable shock. The one patient (Case 2) who had mitral valve replacement did well hemodynamically but died suddenly 5 days postoperatively from rupture of a mycotic intracranial aneurysm; the mitral valve had been replaced because of severe regurgitation.

Pathologic findings. The mitral valve aneurysm, present in all five patients, involved the anterior valve leaflet; in no case was an aneurysm or vegetation noted on the posterior leaflet. The aneurysms were saccular masses filled with friable, necrotic material. They ranged in size from 1.0 × 0.7 cm to 2.0 × 2.0 cm at the base of the aneurysm and extended up to 1.9 cm into the left atrium. The base of the aneurysm was typically surrounded by multiple, pedunculated vegetations suggesting that the aneurysm had formed, at least in part, as a response to erosion by the infection. In one patient (Case 3), there was direct extension of the infection from the aortic valve onto the anterior leaflet of the mitral valve. In three patients, a perforation was noted in the area of the aneurysm (Cases 3, 4 and 5). In the patient (Case 3) who had had repair of a mitral valve perforation through an aortotomy, a large conical aneurysm protruding from the atrial surface of the anterior mitral valve leaflet was found with a perforation at the base of the aneurysm (Fig. 1). In four of the patients, there was no evidence of leaflet fusion or chordal thickening suggestive of intrinsic mitral valve abnormality. One patient (Case 2) had slight thickening of the valve and mild shortening of the chordae tendineae; histologic examination showed acute inflammation, increased vascularity and calcification of the mitral valve.

The aortic valve had vegetations with extensive destruction or perforation of all valve cusps in the five patients. The prosthetic valve was examined at autopsy in the four patients who had had surgery; in three patients, the prosthetic valve was normal. The fourth patient (Case 4), who died suddenly 18 months postoperatively, had an approximately 80% reduction of the porcine aortic valve orifice due to large fungal vegetations.

Echocardiographic findings. Echocardiograms had been obtained from three patients (Cases 1, 2 and 5). All three had M-mode studies, and two (Cases 1 and 2) also had two-dimensional echocardiography.

M-mode echocardiograms. All three patients had abnormal dense echoes in the left ventricular outflow tract during diastole, suggestive of prolapse of an aortic cusp or vegetation. The left ventricle was dilated and showed in-
Increased motion of the posterior wall and interventricular septum in these cases. In two patients, the aortic valve showed dense echoes with erratic motion during diastole, which were believed to represent vegetations. Aortic leaflets appeared normal during systole. The aortic valve echogram of the third patient showed thickened leaflets with reduced excursion during systole but no definite evidence of a vegetation.

*Fine diastolic fluttering of the mitral valve suggested aortic regurgitation in all three patients.* Premature mitral valve closure was not present. There were multiple dense echoes at the base of the valve suggestive of a vegetation or an aneurysm (Fig. 2) in two patients; the tip of the mitral valve in both cases was normal. The left atrium was mildly enlarged in all three cases.

*Two-dimensional echocardiograms.* Large vegetations that prolapsed into the left ventricular outflow tract during diastole could be seen on the aortic leaflets of the two patients studied (Cases 1 and 2). The aortic valve two-dimensional echogram in one of the patients (Case 1) confirmed the M-mode echocardiographic findings of valve thickening and decreased excursion; however, two-dimensional echocardiography also demonstrated definite evidence of a vegetation. The left ventricle was dilated and wall motion was normal in both patients.

The mitral valve echogram showed an unusual abnormality of the anterior leaflet in both patients. The base of the anterior leaflet of the mitral valve demonstrated a persistent bulge that protruded toward the left atrium throughout systole and diastole (Fig. 3 and 4, left panels). The abnormal bulge was best visualized in the parasternal long-axis view and corresponded to the location and shape of the mitral valve aneurysm observed on pathologic examination (Fig. 3 and 4, right panels). A vegetation was seen on the anterior leaflet in one patient (Case 2); the posterior leaflet was normal in both cases.

**Discussion**

Mitral valve aneurysm occurs in association with infective endocarditis of the aortic and mitral valves. Destruction of the aortic valve leaflets produces aortic regurgitation; the regurgitant jet strikes the anterior leaflet of the mitral valve and may produce secondary infection. The mitral valve infection may then lead to the development of an aneurysm (5). In our five patients, the aortic valve was presumed to be the primary site of infection. In one case, the infection of the aortic valve and the anterior mitral valve leaflet was confluent, but in the other four patients the involvement of the mitral valve appeared to result from a jet lesion. Mitral regurgitation in these patients can result from perforation of the aneurysm or valve leaflet, rupture of infected chordae tendineae, associated disease of the mitral valve such as mitral valve prolapse or rheumatic mitral valve disease, or a combination of these abnormalities. In three of our patients, perforation of the mitral valve aneurysm was noted pathologically.

*Surgical management.* There are extensive pathologic descriptions of this abnormality, and the diagnosis is usually made at surgery or autopsy (1–4). The presence of the mitral valve aneurysm was overlooked at the time of surgery in three of our patients, all of whom died in the immediate postoperative period. The most likely reason the aneurysm was missed during surgery is that the valve was examined through the aortotomy, which is not adequate because it does not allow inspection of the atrial surface of the mitral valve leaflets. Preoperatively, the presence of the mitral valve aneurysm was not suspected in any of the five patients because there are no specific clinical findings of this abnormality. In addition, three patients did not have preoperative cardiac catheterization because the physicians caring for the patients either were confident of the diagnosis or believed that the patient was too ill to undergo such studies.

In patients who have a perforation resulting in moderate to severe mitral regurgitation, repair or replacement of the mitral valve should be done at the time of aortic valve replacement (6). However, the ideal management of patients who have a small aneurysm that clinically has not ruptured or is not associated with significant regurgitation and who do not require surgery for other reasons has not been clarified.
Echocardiographic abnormalities associated with mitral valve aneurysm. Two-dimensional echocardiography in two of our patients showed an unusual abnormality of the mitral valve that subsequently correlated with the pathologic findings; in one patient we were able to make the diagnosis preoperatively. Specifically, the abnormality was a localized bulge of the anterior mitral valve leaflet toward the left atrium that persisted throughout the cardiac cycle. Mitral valve prolapse and flail mitral valve may show bulging of the leaflet into the left atrium during systole; when the leaflets straighten during diastole, the bulge disappears. The echocardiographic findings in our patients are in agreement with those of the single case report by DeLuca and Colonna (7). Although not specific, M-mode echocardiography can be helpful in suggesting an abnormality of the mitral valve by showing thickening of the base of the leaflet with sparing of the mitral valve tips (5).

Figure 3. Case 2. Left panel, Two-dimensional echocardiogram (parasternal long-axis view) taken during diastole. A large aortic vegetation (V) is seen prolapsing into the left ventricle. The anterior leaflet of the mitral valve shows an aneurysm (An) that bulges toward the left atrium (LA). Right panel, Surgical specimen of the anterior mitral valve leaflet viewed from the atrial surface corresponding to the echocardiogram shown in the left panel. The aneurysm appears in the center of the mitral valve as a polypoid structure that measures 0.8 cm in depth. The chordae appear thickened and shortened.
Clinical implications. There are no specific clinical findings of mitral valve aneurysm; the features are those of mitral regurgitation and of associated aortic regurgitation caused by infection of the aortic valve. Perforation of the aneurysm may result in or worsen mitral regurgitation and should be suspected in patients with left-sided endocarditis who suddenly manifest hemodynamic deterioration. The findings on two-dimensional echocardiography appear to be helpful and allow one to make the diagnosis before surgery. Mitral valve aneurysm may be missed at surgery if the mitral valve is evaluated only on its ventricular side through an aortotomy. Therefore, the mitral valve should be carefully examined by two-dimensional echocardiography in all patients with left-sided endocarditis. In patients undergoing aortic valve replacement, repair of the aneurysm may be feasible. If the valve is severely damaged, mitral valve replacement may be necessary. The role of surgery for small aneurysms that have not perforated or are not associated with severe mitral regurgitation has not been clarified.

References