Aortic Stenosis and Rupture of Mitral Chordae Tendineae

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Ten patients with aortic stenosis and ruptured mitral chordae tendineae constituted 8% of 125 consecutive surgical cases of chordal rupture. Their ages ranged from 54 to 87 years (mean 68). Six patients presented with acute onset of congestive heart failure, and eight were in New York Heart Association functional class III or IV at the time of cardiac catheterization. Extensive mitral anulus calcification was observed by fluoroscopy in seven patients. The mean aortic valve area index was 0.4 cm$^2$/m$^2$ and nine patients had moderate to severe mitral regurgitation by angiography. Calcific aortic stenosis affected a tricuspid valve in nine cases and a bicuspid valve in one case. One patient had a rheumatic mitral valve and one a redundant myxomatous mitral valve; the remaining eight had no abnormality of the mitral apparatus commonly regarded as predisposing to chordal rupture. Mitral anulus calcification and ventricular anatomic and hemodynamic alterations in aortic stenosis may contribute to rupture of the mitral chordae tendineae.

Rupture of the mitral chordae tendineae, first described by Corvisart (1) in 1806, is now recognized as a common cause of isolated mitral regurgitation. Chordal rupture was usually attributed to rheumatic heart disease or infective endocarditis in earlier case series (2–5). Rupture of the mitral chordae without an identified cause, first noted by Frothingham and Hass (6), has emerged as an important and often major subgroup in more recent reports (7–14). Mitral valve prolapse is a less frequently associated condition (15,16).

Aortic valve disease is not commonly associated with rupture of the mitral chordae tendineae. Cases of combined aortic regurgitation and chordal rupture have been described (4,14,17,18), but aortic stenosis with mitral chordal rupture has rarely been observed (11,14,18). This report describes 10 patients with aortic stenosis and mitral regurgitation due to ruptured mitral chordae tendineae. In eight, there was no intrinsic abnormality of the mitral apparatus known to predispose to chordal rupture.

Patients and Methods

Ten patients with the diagnoses of aortic stenosis and mitral regurgitation with ruptured mitral chordae tendineae who underwent valvular surgery at Stanford Medical Center during 1973 through 1982 were identified with the assistance of a computer-based databank. Medical records were reviewed for presenting clinical features, physical findings, electrocardiographic and echocardiographic findings, hemodynamic and angiographic data and type of valvular operation. Preoperative congestive heart failure was graded according to the New York Heart Association functional classification. Intraoperative observations and the gross pathology of excised cardiac valves were noted. Valves and chordae tendineae were examined for evidence of healed infective endocarditis, rheumatic changes or gross evidence of myxomatous degeneration; histologic examination was not performed. Follow-up information on functional status and survival was obtained from the medical records and by communication with the patient or primary physician.

Results

The 10 patients with aortic stenosis and ruptured mitral chordae tendineae are part of a larger series of 1,908 consecutive patients who underwent aortic, mitral or combined aortic and mitral valve replacement with porcine heterografts during the 10 year study period. Within this surgical series, the 10 patients constitute 1.4% of 702 patients with aortic stenosis, 7% of 141 patients with aortic stenosis and mitral regurgitation of any origin and 8% of 125 patients with ruptured mitral chordae tendineae.

Clinical characteristics (Table 1). There were five men and five women ranging in age from 54 to 87 years (mean 68). No patients reported a history of rheumatic fever or infective endocarditis. Six patients experienced the acute onset of congestive heart failure. Of these six, one had an initial episode of syncope and one presented with cardiac arrest due to ventricular fibrillation. The other four patients noted a more gradual progression of symptoms over a period of 6 months to 3 years. At the time of preoperative cardiac
<table>
<thead>
<tr>
<th>Case</th>
<th>Age (yr) &amp; Sex</th>
<th>Clinical Presentation</th>
<th>NYHA Class</th>
<th>Murmurs (intensity 1 to 6)</th>
<th>ECG</th>
<th>Pressures (mm Hg)</th>
<th>Cardiac Index (m²/m²)</th>
<th>Aortic Valve Area Index (cm²/m²)</th>
<th>Ejection Fraction (%)</th>
<th>Angiographic Grade (0 to 4)</th>
<th>Coronary Arteriogram</th>
<th>Extensive Mitral Anulus Calcification</th>
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AFB = atrial fibrillation; AR = aortic regurgitation; CAD = coronary artery disease. DEC = decrescendo diastolic murmur. 1st AVB = first degree atrioventricular block. ECG = electrocardiogram. HSM = holosystolic murmur. LAA = left atrial abnormality. LBBB = left bundle branch block. LVED = left ventricular end-diastolic. LVH = left ventricular hypertrophy; MR = mitral regurgitation. NSR = normal sinus rhythm. NYHA Class = New York Heart Association functional class. PA = mean pulmonary artery. PAW = mean pulmonary artery wedge. RBBB = right bundle branch block. SEM = systolic ejection murmur.
catheterization, 8 of 10 patients had moderate to severe symptoms of heart failure (New York Heart Association functional class III or IV) despite medical therapy. The examining physicians distinguished two cardiac murmurs suggestive of aortic stenosis and mitral regurgitation in seven patients, and noted a single loud murmur in three patients.

Eight patients had electrocardiographic findings of left ventricular hypertrophy and the other two patients had bundle branch block. M-mode echocardiography was performed in seven patients; it revealed normal-appearing mitral leaflets in five patients, increased echoes of the anterior mitral leaflet in one patient and flail leaflet motion in one patient. All seven patients showed increased echoes of the aortic valve leaflets consistent with fibrosis and calcification and six patients met echocardiographic criteria for left ventricular hypertrophy (left ventricular end-diastolic posterior wall dimension greater than 1.3 cm).

Preoperative hemodynamic findings (Table 1). The mean pulmonary artery pressure was greater than 30 mm Hg in five patients, and the pulmonary artery wedge V wave was greater than 40 mm Hg in four patients. Left ventricular end-diastolic pressure was elevated in nine patients and greater than 30 mm Hg in four. Nine of the 10 patients had a reduced cardiac index with a group mean value of 2.0 liters/min per m². Left ventricular chamber dilation was observed by angiography in seven patients and was absent in three. Angiographic ejection fraction, determined in nine patients, averaged 37%.

The mean aortic valve area index was 0.4 cm²/m² (range 0.2 to 0.9). Aortic valve calcification was observed by fluoroscopy in all patients. Supravalvular aortography was not performed in three patients who lacked auscultatory evidence of aortic regurgitation. Aortic regurgitation in the remaining seven patients was either absent or mild (grade 1/4 or 2/4). Aortic stenosis was regarded as the predominant hemodynamic lesion of the aortic valve in the 10 cases.

Seven patients had fluoroscopic evidence of mitral anulus calcification. Mitral regurgitation was graded by angiography as moderately severe or severe (grade 3/4 or 4/4) in eight patients. Coronary arteriography was performed in nine patients and revealed coronary artery disease in five. One patient (Case 4) had segmental dyskinesia on left ventriculography consistent with prior inferior myocardial infarction.

One patient (Case 5) had a normal cardiac index and no mitral regurgitation by angiography, but moderately severe mitral regurgitation with ruptured chordae tendineae was observed at the time of operation 5 days later. The chordal rupture was presumed to have occurred during the interim period.

Pathologic anatomy, surgical procedures and follow-up data (Table 2). Ruptured chordae tendineae had detached from the anterior mitral leaflet in six cases, from the posterior leaflet in two cases and from both leaflets in one case. The affected leaflet was not specified in one case. Operative reports generally did not indicate which papillary muscle was associated with the ruptured chordae or which chordal division was affected. In one patient, fusion of the mitral commissures and chordal thickening suggested prior rheumatic valvulitis. The anterior mitral leaflet of one patient (Case 1) with ruptured anterior chordae was highly redundant and had the gross appearance of myxomatous degeneration. The chordae in this patient were long and attenuated. The mitral valve in the remaining eight patients displayed normal or slightly thickened leaflets and no additional abnormalities of the chordae. Focal mitral leaflet calcifications were present in five cases, most prominent at the leaflet base. There was no evidence of active or healed endocarditis. In the patient with an inferolateral ventricular aneurysm (Case 4), the papillary muscles were not involved and appeared normal. The excised or debrided aortic valves were all noted to be heavily calcified and without commissural fusion suggestive of rheumatic disease. Nine valves were tricuspid and one was bicuspid.

Valve replacement operations were performed with Hancock heterograft bioprostheses. Seven patients had replacement of both the aortic and mitral valves. Three patients had mitral valve replacement and aortic valvuloplasty. Concurrent aortocoronary bypass operations were performed in four subjects, one of whom also had a left ventricular aneurysmectomy.

There were no intraoperative deaths. Two patients died of pulmonary complications during the early postoperative period. Four patients died 2 months to 3 years after operation; two died suddenly, and two died of noncardiac causes. Four patients are alive and asymptomatic 9 months to 6 years after surgical treatment.

Discussion

Incidence. The combination of aortic stenosis and mitral regurgitation due to ruptured mitral chordae tendineae is a rare occurrence. The present group of 10 patients is the largest reported series of such cases and is identified within the extensive valvular surgical series at this medical center. These 10 patients constitute a small but significant proportion of cardiac surgical patients with combined aortic stenosis and mitral regurgitation (7%) and patients with ruptured chordae tendineae (8%).

Four patients with calcific aortic stenosis were reported in a series of 122 cases of ruptured mitral chordae tendineae (14). Two other patients with mixed aortic regurgitation and stenosis were included in group case reports (11,18). The severity of aortic stenosis was not described in those cases. In the current patient group, aortic stenosis was usually moderate or severe and was considered the predominant aortic valve lesion.

In contrast to the paucity of reported cases with combined
Aortic stenosis and ruptured mitral chordae, several patients with aortic regurgitation and mitral chordal rupture have been described (4,14,17,18). Infective endocarditis or rheumatic valvulitis contributed to chordal rupture in some of these cases, but most of the patients had no apparent predisposing abnormality of the mitral apparatus.

Clinical and hemodynamic findings. The clinical characteristics and catheterization results of our patients can be compared with observations in patients with isolated mitral chordal rupture. Their mean age of 68 years is greater than that described for isolated rupture (4,12, 14). This corresponds to the older age profile of patients with calcific aortic stenosis. As with isolated rupture of the mitral chordae, symptoms developed either acutely or showed a gradual progression, and moderately severe or severe symptoms of congestive heart failure (functional class III or IV) persisted despite medical therapy. Although the systolic murmur of isolated mitral chordal rupture is often loud or loudest at the right upper sternal border, two murmurs were usually distinguished in the patients with combined aortic stenosis and chordal rupture, leading to suspicion of both hemodynamic lesions. Although the left ventricular ejection fraction is often normal with isolated chordal rupture (13), most of our patients with combined lesions had a diminished ejection fraction.

Causal factors. Conditions generally recognized as predisposing to rupture of the mitral chordae tendineae were present in 2 of these 10 patients. One patient had evidence of antecedent rheumatic valvulitis and one had a redundant myxomatous mitral valve. It is possible that subtle changes of rheumatic or healed infective endocarditis could have been missed in the other cases on pathologic examination. Myocardial infarction not involving the papillary muscles was present in one patient, but the role of ischemic disease in chordal rupture is less certain (12).

Although a causal association of calcific aortic stenosis with rupture of the mitral chordae is not demonstrated in this patient group, potentially related factors can be considered.
Extensive calcification of the mitral anulus, present in 7 of these 10 patients, commonly accompanies aortic stenosis in older patients (19,20). The aortic and mitral anuli are anatomically contiguous and may share in a localized pathologic or degenerative process (21). Calcification and fibrosis may reduce the sphincter motion of the mitral anulus (22), possibly altering the distribution and degree of stress on the mitral apparatus. Calcification can also extend into the mitral leaflet tissue.

Changes in ventricular anatomy concomitant with the hypertrophic response to aortic stenosis may alter forces on the mitral chordae tendineae. Postmortem examination of ventricles with concentric hypertrophy shows tilting of the mitral ring to a plane more nearly parallel to that of the aortic ring and elongation of the anterior mitral leaflet and subvalvular apparatus (23). Supervention of ventricular dilation due to cardiac failure or the presence of a regurgitant lesion leads to a more spherical chamber shape (24) and can further distort the normal relation between components of the mitral apparatus. Moreover, the excessive intraventricular pressure of aortic stenosis may contribute to chordal rupture. The role of all of these factors, however, remains speculative.

Clinical implications. Severe aortic stenosis and severe mitral regurgitation in the absence of significant aortic regurgitation or mitral stenosis are an uncommon combination of hemodynamic lesions. No such cases were noted in a consecutive series of 100 combined aortic and mitral valve replacements (25). Obstruction of aortic outflow exacerates the degree of mitral regurgitation (26,27). Concurrently, the presence of mitral regurgitation counters the advantages of the augmented preload needed to sustain forward left ventricular output in aortic stenosis. These combined pressure and volume lesions can lead to particularly unfavorable conditions of ventricular wall stress and cardiac work, most likely contributing to the chamber dilation and diminished ejection fraction observed in patients with these findings.

Despite the poor level of ventricular performance before operation, symptomatic congestive heart failure did not persist in these patients after surgical treatment. Their variable postoperative survival may relate to advanced age, the presence of coronary artery disease and arrhythmias associated with residual left ventricular dysfunction. As part of a surgical series, these patients were selected as survivors of the initial chordal rupture. Their degree of hemodynamic compromise and the presentation of one patient with ventricular fibrillation suggest that rupture of mitral chordae tendineae may also contribute to the incidence of sudden death in patients with aortic stenosis.

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References

