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Successful repair of a variant of mitral arcade

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Video clip is available online.

Mitral arcade (MA) is a very rare mitral anomaly characterized by abnormal subvalvular apparatus development. The papillary muscles are usually directly attached to both leaflets. We present an unusual case in which the posterior mitral leaflet (PML) was solely affected, leading to tethering, malcoaptation, and regurgitation.

CLINICAL SUMMARY

A 17-year-old girl was seen at our clinic after a new diagnosis of a systolic cardiac murmur. She reported symptoms of fatigue but not dyspnea, palpitations, or chest pain. History was negative for rheumatic disease. Physical examination revealed a healthy-appearing young woman with a harsh 3/6 holosystolic murmur at the apex, radiating toward the axilla. Chest radiography showed mild cardiac enlargement but was otherwise unremarkable. A transthoracic echocardiogram revealed a moderately thickened mitral valve with a central coaptation gap

resulting from restricted PML motion. There was severe regurgitation, with a mean gradient of 8 mm Hg. The left heart chambers were severely enlarged. Calculated ejection fraction was 58%. Surgical treatment with repair or replacement of the mitral valve was recommended.

Preoperative transesophageal echocardiography confirmed limited excursion of the PML, which created an eccentric regurgitation jet toward the dome of the left atrium. The PML chordae tendineae were foreshortened and thickened; the papillary muscles were closely spaced in the configuration of a partial arcadelike structure (Figure 1, A, and Video 1). During direct inspection at the time of surgery, the PML was small, thick, and severely tethered. Mobility was limited. The leaflet was delaminated with a sharp incision and completely detached from the annulus except for most extreme medial and lateral portions. An oval-shaped bovine pericardium patch was used to augment the PML surface and sutured circumferentially. The posteromedial commissure was then plicated to achieve better competence (Figure 2, A-D). The valve appeared to be functioning well, and saline solution testing revealed valve competence. A postprocedural echocardiogram showed bowing of the midportion of the neo-PML with the leading edge coaptation margin intact, revealing trivial regurgitation and mild stenosis (Figure 1, B-D, and Videos 2 and 3). The patient's postoperative course was uneventful, and she was released on postoperative day 6.

DISCUSSION

MA is a very rare mitral anomaly.^{1,2} First described by Layman and Edwards¹ in 1967, MA is characterized by abnormal subvalvular apparatus development. The papillary muscles are usually directly attached to both leaflets. When

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FIGURE 1. A, Transesophageal transgastric view of the mitral valve. The posterior chordae (*arrow*) are short and thickened. B and C, Postbypass intraoperative transesophageal 85° midesophageal view of the mitral valve (*arrow*) in systole (B) and diastole (C). During systole, the augmented posterior mitral leaflet has a domed appearance. D, Continuous-wave Doppler through the mitral valve demonstrates a diastolic mean Doppler gradient of 5 mm Hg at a heart rate of 100 beats/min. LA, Left atrium; LV, left ventricle.

present, short and thick chords are typically distributed equally to both cusps. The anterior mitral leaflet is usually severely tethered. There may be relatively less involvement of the PML, and valve stenosis may coexist. The unusual aspect of our case was the fact that the PML was solely affected, leading to tethering, malcoaptation, and regurgitation. In the variant that we encountered, the anterior mitral leaflet appeared to be spared by the disease. The papillary muscles were fused together in a typical arcadelike conformation. Chords to the PML were particularly short. This resulted in severe apical displacement of the PML, with an eccentric systolic jet directed toward the septum.

MA is usually diagnosed in infancy with symptoms of congestive heart failure resulting from severe mitral insufficiency, stenosis, or both. Milder cases can go unrecognized until adulthood or incidental diagnosis. There have been reports of successful repair of complete MA,³ as well as of need for valve replacement.⁴ We chose to enlarge the PML to reduce the tethering and achieve better valve

coaptation. This technique has been shown to be useful in the selective treatment of other mitral anomalies but has not previously been described for this rare mitral disease. We do think that congenital or acquired mitral anomalies, resulting in a small tethered PML, could benefit from this technique. Various materials, such as pericardium or CorMatrix (CorMatrix Cardiovascular, Inc, Roswell, Ga), can be selectively used. It is our opinion that valve repair is always preferable in young patients, especially those with mild symptoms.

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FIGURE 2. Intraoperative images and surgical repair. From the surgeon's view, the posterior mitral leaflet (*PML*) appears tethered as a result of severely shortened chords (*A*, arrow). A pericardial patch (*star*) is sutured circumferentially along the posterior annulus to enlarge the posterior mitral leaflet (*B*). Final result shows the reconstructed posterior mitral leaflet (*C*). The repair concept is demonstrated in the sketch (*D*). *AML*, Anterior mitral leaflet.

Magnetic resonance imaging–based management of silent cardiac rupture

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A 45-year-old man, who was a smoker, presented after recurrences of severe epigastric pain during the 2 preceding days. His electrocardiogram (ECG) showed an inferior ST elevation. Coronary angiography revealed an occluded proximal right coronary artery, which was successfully