CASE REPORT

A tale of 2 coexisting congenital mitral pathologies: Arcade and parachute

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CLINICAL SUMMARY

A 26-year-old woman presented with heart murmur since childhood and occasional palpitations. Physical examination was notable for soft pan-systolic murmur at the apex. Electrocardiogram was normal. Transthoracic echocardiography (TTE) revealed suspected parachute mitral valve (Figure 1) with moderate mitral stenosis with mild-moderate mitral regurgitation, normal biventricular systolic function, moderately dilated left atrium, and mild pulmonary hypertension. Detailed transesophageal echocardiogram (TEE) was performed, showing mitral valve parachute (dominant antero-lateral papillary muscle with majority of attachments) and arcade (shortened, thickened, and rudimentary chordae), both contributing to severe subvalvular mitral stenosis (gradients 26/13 mm Hg) with moderate mitral regurgitation. Cardiac computed tomography performed for coronary and thoracic anatomy evaluation, using Siemens Definition Force dual source 2 × 192-slice scanner showed similar mitral valve anatomy (Figure 2). Institutional review board approval was not required; the patient provided informed written consent for publication of her anonymized data for this case report.

Operative findings include that most of the chords were from the antero-lateral papillary muscle, the anterior head and the posterior head were fused together, and the posterior head was flat and arcade-like providing short chords to P1 and lateral P3 (Figure E1). The chords were individually identified and fenestrated to elongate and create a more natural papillary orientation. The anterior head was separated from the posterior head to increase mobility and opening. A secondary chord to the posterior head, which was limiting motion, was cut. Subsequently, the P1/P2 chords were separated to further improve mobility, and a Duran mitral valve annuloplasty ring (size #35) was placed to stabilize. The postoperative course was uncomplicated. Predischarge TTE showed stable mitral valve repair with unremarkable gradients (mitral valve gradients of 11/6 mm Hg) and trivial mitral regurgitation, along with normal biventricular size and systolic function.

DISCUSSION

Parachute mitral valve (PMV) is formed by specific malformations of the mitral leaflets per se, as well as various subvalvular structures, giving rise to a funnel configuration of the mitral valve. The chords are short and thick with a convergent papillary insertion, whether a single papillary muscle receives all chords (true PMV), or 2 papillary muscles with all chordae inserting into 1 muscle and the other being hypoplastic, and commissures barely distinguishable and frequently underdeveloped.1 Mitral arcade (MA), first described in 1967, is an extremely rare congenital anomaly of the mitral chordal apparatus, so diagnosis is often not

CENTRAL MESSAGE

Resolving a diagnostic dilemma of a rare mitral valve anomaly (arcade and parachute) by multimodality imaging, to guide further cardiothoracic surgical management as well as follow-up assessment.

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considered. It is characterized by an arc-like configuration of 2 papillary muscles due to an interconnecting band of fibrous tissue that runs along the line of closure of the mitral leaflets. There is severe shortening or complete absence of chordae tendineae such that the leaflets insert almost directly into the papillary muscle. When viewed from the atrial surface, the mitral valve appears like a hammock. The fibrous band interferes with normal valvular motion, which prevents the typical apposition of mitral leaflets, thereby resulting in mitral regurgitation. Additionally, mitral stenosis may coexist due to the presence of mechanical obstruction related to prominent papillary muscles blocking the subvalvular apparatus and inadequate opening of the mitral valve. Symptomatic patients present with worsening dyspnea and congestive heart failure from mitral valve dysfunction. An incidental cardiac murmur may be auscultated in asymptomatic people, years before clinical presentation, as seen in the index case.

In patients when etiology, severity, and/or need for intervention of significant mitral regurgitation are uncertain by TTE alone, other multimodality imaging techniques provide supplementary roles. TEE is critical in better characterizing these features. Contemporary 3-dimensional imaging techniques with multiplanar reconstruction on TEE can accurately delineate the precise cause, location, and severity of mitral regurgitation in the majority of cases. Exercise stress echocardiography can provide complementary information. Cardiac computed tomography angiography can also precisely assess the complete anatomical details of the mitral valve apparatus, including papillary muscles and chordae, as demonstrated in our case. Finally, cardiac magnetic resonance imaging can be useful to quantify mitral regurgitation severity by regurgitant volume and fraction, chamber quantification, and has tissue characterization abilities to assess myocardial scar and concomitant cardiomyopathy.

Adults with MA seem to have milder mitral valve dysfunction that can escape detection during early life and therefore present later with degenerative valvular apparatus over time. Medical treatment is limited to symptomatic relief of heart failure and arrhythmias if present. Intervening on the mitral valve is similar to other etiologies when there is severe mitral stenosis and/or regurgitation with concomitant symptoms or other secondary indications, and surgery remains the definitive strategy. Although mitral valve repair is typically preferred over replacement for mitral regurgitation, our case is among the few that has reported a
successful repair for MA and PMV. Replacement remains the mainstay, and transcatheter interventions have not currently been reported.\(^3\)

**Conflict of Interest Statement**

The authors reported no conflicts of interest.

The Journal policy requires editors and reviewers to disclose conflicts of interest and to decline handling manuscripts for which they may have a conflict of interest. The editors and reviewer of this article have no conflicts of interest.

**FIGURE 2.** Cardiac computed tomography evaluation of parachute and arcade mitral valve case. A, Apical 4-chamber view showing parachute mitral valve with prominent anterolateral papillary muscle attached to both leaflets (red arrow). B, Apical 2-chamber view showing arcade of subvalvular apparatus of the mitral valve (arcade morphology) (yellow arrow). Short and rudimentary chordae are seen (blue arrow). C, Left ventricular short axis view mitral valve level showing prominent leaflet and subvalvular muscle thickening with restricted opening (orange arrow). D, Left ventricular short axis papillary muscle level showing one prominent anterolateral papillary muscle (red arrow).

**References**


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FIGURE E1. Intraoperative image of the parachute and arcade mitral valve case from the left atrial aspect. It shows narrowed mitral valve orifice with arcade subvalvular apparatus.