Mitral Arcade

A Rare Cause of Fatigue in an 18-Year-Old Female

R. Thomas Collins II, MD; Michelle Ryan; Marie M. Gleason, MD

A previously healthy 18-year-old female presented for evaluation of a 3-month history of fatigue. Other notable findings in the history were progressively worsening dyspnea on exertion, 3-pillow orthopnea, a newly developed cough, and a 20-lb weight loss. The physical examination demonstrated tachypnea, with a respiratory rate of 24 breaths per minute, and the lungs were clear. The heart rate was 88 bpm and regular. There was a regular rhythm with a normal first heart sound and a second heart sound that was slightly accentuated and narrowly split. There was a 2/6 systolic regurgitant murmur at the left ventricular apex. There was no diastolic murmur. There was no jugular venous distension or hepatic engorgement.

A chest radiograph demonstrated fine, reticular prominence of the pulmonary interstitium of both lungs, increased pulmonary vascular markings, and mild prominence of the left atrial appendage (Figure 1). A nongated computed tomography scan of the chest demonstrated abnormal pulmonary parenchyma with diffuse ground-glass opacities (Figure 2). A 15-lead ECG was performed that demonstrated sinus rhythm, right-axis deviation, and biatrial enlargement (Figure 3).

A complete echocardiogram demonstrated multiple abnormalities. The right ventricular pressure was elevated at 70 mm Hg above the right atrial v wave. The left atrium was moderately dilated with moderate mitral regurgitation. The mitral valve orifice was of normal size; however, the mitral valve apparatus was abnormal, with thick and shortened chordal attachments of the anterior leaflet to the papillary muscle and nearly nonexistent spaces between the abnormal chordae, all of which are quintessential anatomic findings in mitral valve arcade (Figures 4A and 4B; Movie I in the online-only Data Supplement). Color Doppler interrogation of the mitral valve inflow demonstrated multiple jets of accelerated flow through the small orifices between the foreshortened mitral chordae (Figure 4C; Movies II and III in the online-only Data Supplement). The mean gradient across the mitral valve was measured at 22 mm Hg by continuous-wave Doppler (Figure 5). Ventricular function was normal, and there were no other anatomic abnormalities.

The patient was referred to surgery for mitral valve replacement with a 29-mm St Jude prosthetic valve. The perioperative course was unremarkable. On follow-up, the patient's exercise capacity had improved greatly, and her weight had returned to baseline.

Congenital mitral valve stenosis has been associated with abnormal pulmonary function testing and secondary pulmonary parenchymal disease on lung biopsies in pediatric patients.[1] Pulmonary parenchymal findings in mitral stenosis can include alveolar hemorrhage, pulmonary hemosiderosis, and pulmonary ossification, all of which can be demonstrated radiographically. Pulmonary hemosiderosis in the setting of mitral stenosis results from recurrent small alveolar hemorrhages that are related to elevated pulmonary capillary pressure. Pulmonary hemosiderosis is seen in 10% to 25% of patients with mitral stenosis and can present subtly as reticular changes on chest radiograph.[2] Furthermore, hemosiderosis may be seen as ground-glass opacities on computed tomography of the chest, as shown. More common radiographic findings seen in mitral stenosis, and demonstrated in the present chest radiograph, include left atrial enlargement; pulmonary vascular cephalization; interstitial, perivascular, or occasionally pulmonary edema; and a normal-to-small left ventricle.[2]

In 1967, Layman and Edwards[3] first described anomalous mitral arcade in 3 infants as a rare mechanism for congenital mitral insufficiency. The salient features of their description of an anomalous mitral valve arcade include the following: (1) An adequately sized mitral valve orifice; (2) short, thick, and poorly differentiated chordae with direct union of the papillary muscles to the anterior leaflet; (3) narrow or nearly nonexistent spaces between the abnormal chordae; and (4) greater differentiation of the chordae attached to the posterior papillary muscle.[3] Since that initial description, mitral arcade has been reported rarely and has also been reported as a "hammock valve." The exact developmental origin of mitral arcade is not known; however, it is suspected that it represents an arrest of mitral valve development at a stage after loss of muscle in chordae and leaflets but before the final attenuation and elongation of mitral chordae have occurred.[4]

The clinical course of isolated mitral arcade is varied depending on the presence and severity of mitral stenosis and regurgitation. The majority of the reported cases have been in

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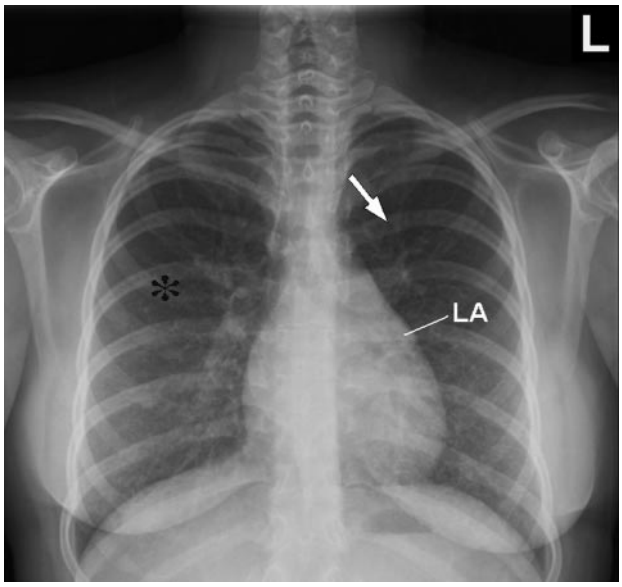

Figure 1. Chest radiograph in the posteroanterior projection demonstrates fine reticular prominence of the pulmonary interstitium of both lungs (asterisk), increased pulmonary vascular markings (arrow), and mild prominence of the left atrial appendage (LA) with an otherwise normal cardiac silhouette.

pediatric patients, and those cases had significant morbidity and mortality. There have been 3 reports of mitral arcade in adults, with the oldest report in a 65-year-old man.[4] The clinical course in those patients was associated with less morbidity than the reports in pediatric patients, most likely owing to a less severe degree of stenosis.

The present case is, to the best of our knowledge, only the fourth report in the English-language literature of a mitral arcade in an adult patient. The case illustrates some of the presenting findings that can be seen in severe mitral stenosis, and it highlights echocardiographically the sine qua non of an anomalous mitral arcade. Furthermore, the known significant impact that chronic mitral obstruction can have secondarily on the pulmonary parenchyma is demonstrated.

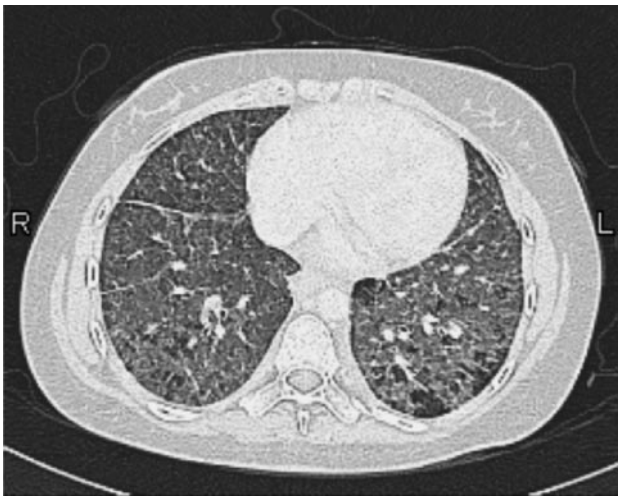

Figure 2. Axial computed tomographic image with pulmonary windowing demonstrates abnormal pulmonary parenchyma with diffuse ground-glass opacities. R indicates right; L, left.

Disclosures

None.

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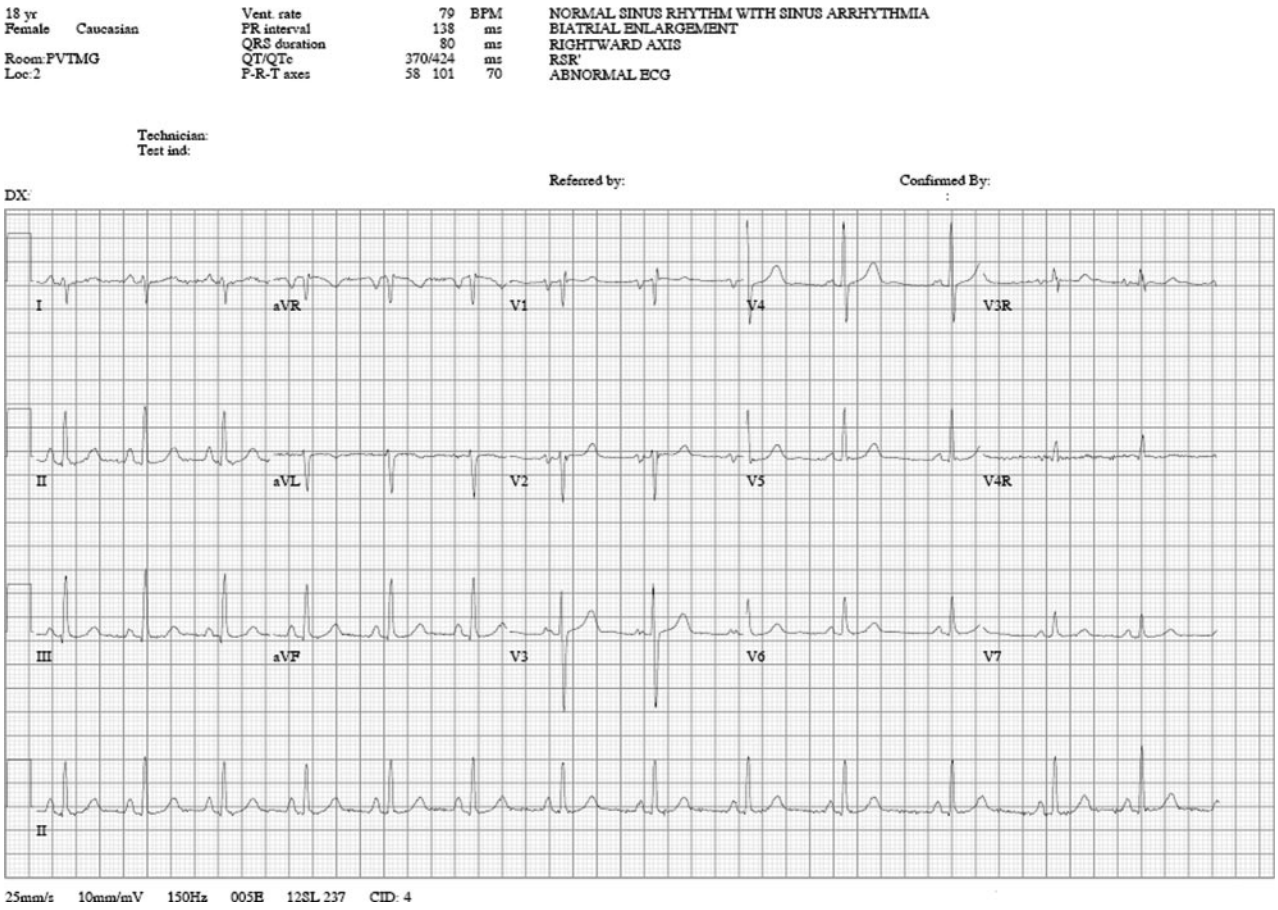

Figure 3. A 15-lead ECG showing sinus rhythm, right-axis deviation, and biatrial enlargement.

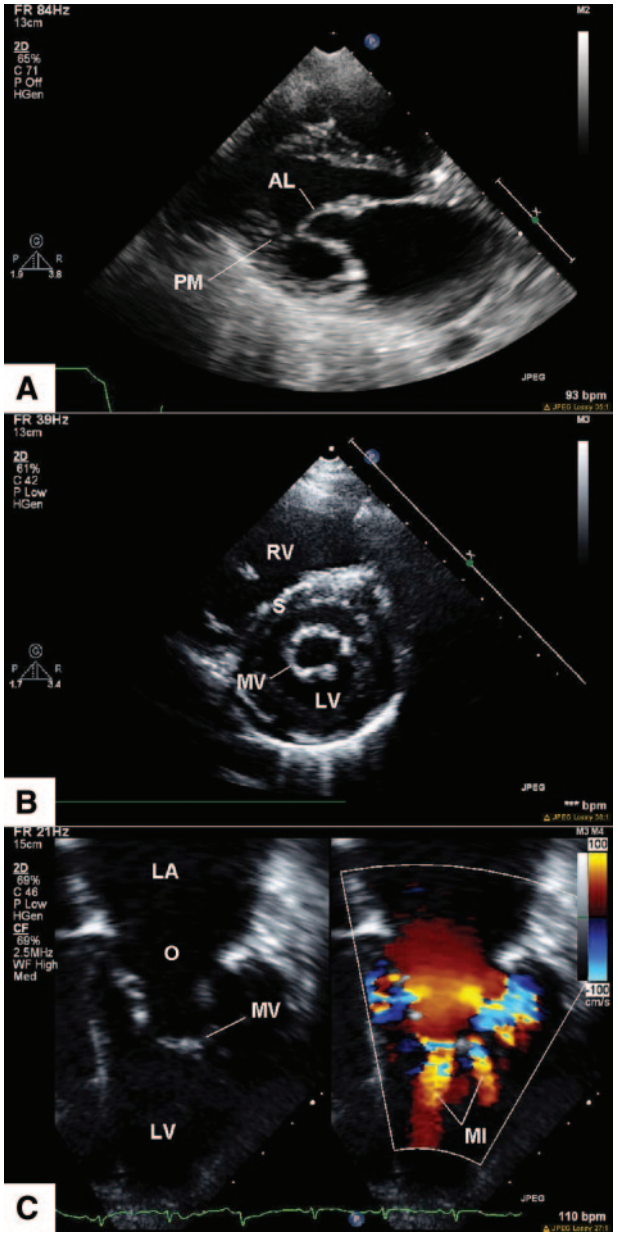

Figure 4. Typical echocardiographic images of mitral arcade. A, Two-dimensional parasternal long-axis view of adequately sized mitral valve orifice with direct attachment of anterior leaflet (AL) of mitral valve (MV) to papillary muscle (PM). B, Parasternal short-axis view of stenotic mitral valve with thickened chordae and small, nearly nonexistent orifices in the chordal apparatus. C, Apical 4-chamber view of moderately dilated left atrium (LA), normally sized mitral orifice (O), direct attachment of anterior leaflet to papillary muscle, and multiple splayed jets of accelerated mitral inflow (MI) through multiple small orifices. RV indicates right ventricle; S, ventricular septum; and LV, left ventricle.

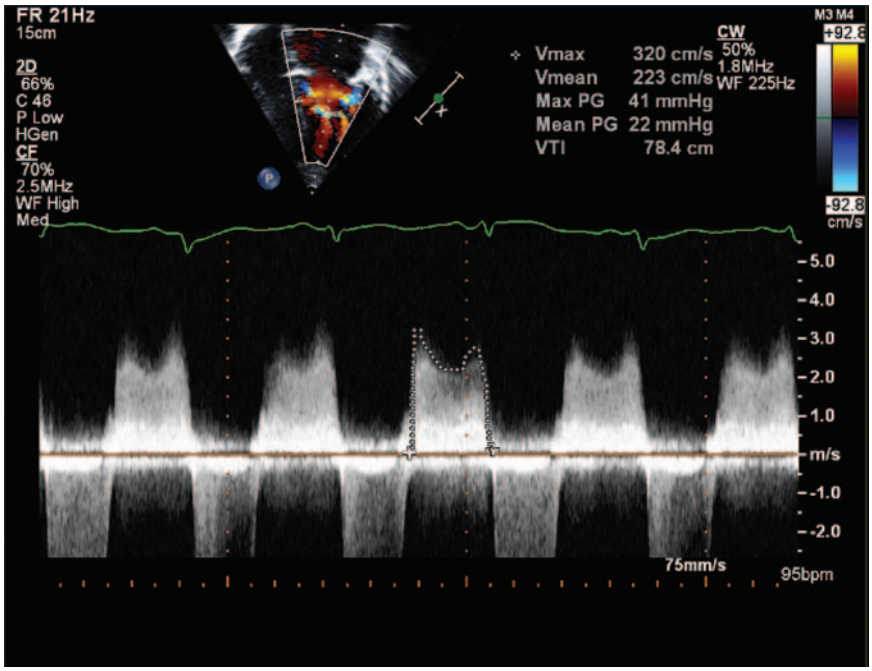

Figure 5. Continuous-wave Doppler interrogation across the mitral valve demonstrates a mean pressure gradient (PG) of 22 mm Hg, consistent with severe mitral stenosis. Vmax indicates maximum velocity; Vmean, mean velocity; Max, maximum; and VTI, velocity-time integral.

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