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 bialtrial 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. 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.

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
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. 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.

Disclosures

None.

References

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.
Mitral Arcade: A Rare Cause of Fatigue in an 18-Year-Old Female
R. Thomas Collins II, Michelle Ryan and Marie M. Gleason

Circulation. 2010;121:e379-e383
doi: 10.1161/CIR.0b013e3181db1ee4
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 2010 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the
World Wide Web at:
http://circ.ahajournals.org/content/121/15/e379

Data Supplement (unedited) at:
http://circ.ahajournals.org/content/suppl/2010/04/01/121.15.e379.DC1

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation is online at:
http://circ.ahajournals.org//subscriptions/