Images in Cardiovascular Medicine

Severe Mitral Annular Calcification:

Insights from Multimodality Imaging

Benoy N. Shah, MBBS Sonya Babu-Narayan, PhD Wei Li, PhD Michael Rubens, FRCR Tom Wong, MD uring transthoracic echocardiography (TTE), a 77-year-old woman with exertional dyspnea was found to have large, well-defined masses adjacent to the mitral valve (Fig. 1). Spectral Doppler echocardiography revealed a mean transmitral gradient of 7 mmHg, a mitral velocity—time integral of 45 cm, and a pressure half-time of 130 ms. There was moderate mitral regurgitation, although the exact mechanism was unclear on TTE. Transesophageal echocardiography revealed that the posterior mass prevented complete leaflet apposition, resulting in regurgitation (Fig. 2A), whereas the anterior mass was not affecting leaflet motion—so no mitral regurgitation was seen there (Fig. 2B). The echocardiographic appearances seemed to be consistent with mitral annular calcification (MAC), but alternative causes (specifically tumor) were not excluded.

During cardiac magnetic resonance (CMR), cine imaging with balanced steadystate free-precession sequences showed immobile, low-signal masses in the mitral annulus. After intravenous gadolinium contrast administration, first-pass perfusion images revealed no enhancement of the mass, in keeping with an avascular structure. On the early gadolinium-enhancement images, the mass remained hypointense, but the corresponding late gadolinium-enhancement images revealed a rim of circum-

Section Editor:

Raymond F. Stainback, MD, Department of Adult Cardiology, Texas Heart Institute, 6624 Fannin St., Suite 2480, Houston, TX 77030

From: Departments of Echocardiography (Drs. Li and Shah), Cardiovascular Magnetic Resonance (Dr. Babu-Narayan), Cardiovascular Computed Tomography (Dr. Rubens), and Cardiology (Dr. Wong), Royal Brompton Hospital, London SW3 6NP; and National Heart and Lung Institute (Drs. Babu-Narayan, Shah, and Wong), Imperial College, London SW7 2AZ; United Kingdom

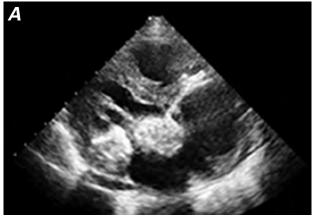
Address for reprints: Tom Wong, MD, Depart-

ment of Cardiology, Royal Brompton Hospital, Sydney St., London SW3 6NP, UK

E-mail:

tom.wong@imperial.ac.uk

© 2014 by the Texas Heart® Institute, Houston



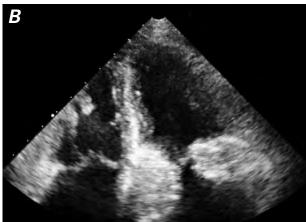


Fig. 1 Transthoracic echocardiograms show large intracardiac echo-bright masses in A) parasternal long-axis and B) apical 4-chamber views.

Supplemental motion images are available for Figures 1A and 1B.

ferential enhancement surrounding a dark central core (Fig. 3A). Non-contrast cardiac computed tomograms showed severe MAC, confirmed the absence of calcification in the valve leaflets and myocardium itself, and also showed a partially calcified rim with a lower attenuation signal located more centrally within the annulus (Fig. 3B). Therefore, both CMR and computed tomographic imaging features were consistent with caseous MAC, a variant of MAC in which the calcium undergoes liquefaction. Decalcification of the mitral annulus is a recognized surgical challenge¹; after extensive multidisciplinary discussion, we adopted a conservative management approach, and the patient remained under monitoring.

Comment

Mitral annular calcification is a chronic, noninflammatory process characterized by calcium deposition in the annular fiber of the mitral valve. It is more common in

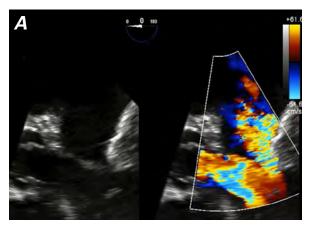
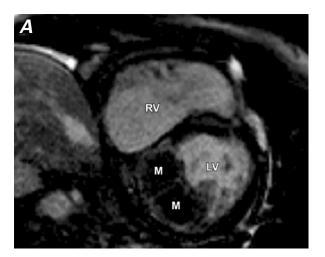




Fig. 2 Transesophageal color-flow Doppler echocardiograms (midesophageal 4-chamber views). A) Mitral regurgitation occurs because the posterior annular mass prevents normal leaflet apposition. B) Mitral regurgitation does not occur, because the anterior annular mass does not prevent normal leaflet apposition.

Supplemental motion images are available for Figures 2A and 2B.



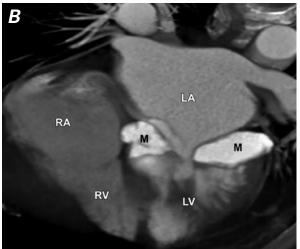


Fig. 3 A) Bright-blood cardiac magnetic resonance image shows a hypointense mass with hyperenhancement in the rim during delayed inversion recovery imaging, 10 minutes after gadolinium contrast administration. B) Cardiac computed tomogram shows the extent of mitral annular calcification and the absence of calcification in the mitral leaflets.

LA = left atrium; LV = left ventricle; M = mass; RA = right atrium; RV = right ventricle

the posterior than the anterior annulus and is usually mild—therefore of no hemodynamic significance. Potential complications include functional mitral stenosis, mitral regurgitation, infective endocarditis, and, rarely, valvular thrombosis.² Liquefaction necrosis, also known as caseous MAC, is a less frequent variant of MAC in which the core of the mass comprises amorphous eosinophilic fluid, and the surrounding rim contains multiple areas of calcification and inflammatory cell infiltration.³

Because of the saddle shape of the mitral annulus, comprehensive evaluation by echocardiography alone—particularly the exclusion of alternative causes such as tumor, thrombosed coronary sinus, and mitral annular abscess—can be challenging. Cardiac magnetic reso-

nance enables accurate tissue characterization; the unusual combination of a hypointense signal on T1- and T2-weighted images (pre- and post-contrast) strongly suggests high calcium content.3 Pre-contrast CMR imaging can also help differentiate caseous from noncaseous MAC: noncaseous MAC generates a low signal on T1 imaging, whereas the calcium salts and proteinaceous fluid within caseous MAC can generate a high signal on T1 images.4 Finally, cardiac computed tomography is the optimal imaging technique for calcium and for that reason best confirms or excludes involvement of the mitral leaflets and extension of calcification into the myocardium itself.

In conclusion, these images are typical of caseous MAC, a characteristic condition in which multimodality imaging is helpful in excluding alternative causes of intracardiac masses and in reassuring those who might be unfamiliar with such characteristics. Surgical annular decalcification is a highly challenging operation and is usually avoided.

References

- 1. Dietrich M, Doss M, Aybek T, Martens S, Scherer M, Wimmer-Greinecker G, Moritz A. Decalcification of the mitral annulus: surgical experience in 81 patients. Thorac Cardiovasc Surg 2006;54(7):464-7.
- 2. Adler Y, Fink N, Spector D, Wiser I, Sagie A. Mitral annulus calcification -- a window to diffuse atherosclerosis of the vascular system. Atherosclerosis 2001;155(1):1-8.
- 3. Di Bella G, Masci PG, Ganame J, Dymarkowski S, Bogaert J. Images in cardiovascular medicine. Liquefaction necrosis of mitral annulus calcification: detection and characterization with cardiac magnetic resonance imaging. Circulation 2008; 117(12):e292-4.
- 4. Shriki J, Rongey C, Ghosh B, Daneshvar S, Colletti PM, Farvid A, Wilcox A. Caseous mitral annular calcifications: multimodality imaging characteristics. World J Radiol 2010;2(4): 143-7.