Multimodality Cardiac Imaging in the Evaluation of Mitral Annular Caseous Calcification

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A 77-year-old man with severe aortic stenosis was referred for consideration of aortic valve replacement. He admitted to mild exertional dyspnea and was systemically well. His past medical history was remarkable for hypertension, hypercholesterolemia, and a previous transient ischemic attack. A transthoracic echocardiogram confirmed severe calcific aortic stenosis (peak gradient 95mm Hg, estimated orifice area 0.5 cm²) with normal left ventricular ejection fraction (70%). In addition, a well-circumscribed, echodense mass (25 mm in diameter) was observed in the posterolateral basal wall attached to the posterior mitral valve leaflet (Figures 1A and 1B). Although highly echogenic, the mass also contained central echolucent areas. The mitral valve was competent, with no functional stenosis. There was no pericardial effusion. Three-dimensional transesophageal echocardiography confirmed an encapsulated, heterogenous mass arising from the posterior mitral valve annulus, adherent to the posterior mitral valve leaflet P2 scallop, in close proximity to the left atrioventricular groove and circumflex artery (Figures 1C and 1D; online-only Data Supplement, Movie 1). The mass did not appear to be pedunculated or vascular in nature.

The patient was referred for cardiac magnetic resonance imaging to further characterize the cardiac mass. Cine imaging with a balanced steady state free-precession sequence showed an immobile, low-signal mass with a smooth contour located at the posterior mitral valve annulus (Figure 2A; online-only Data Supplement, Movie 2). There was no restriction of the posterior mitral valve leaflet motion or associated mitral regurgitation. On both multicontrast T1-weighted turbo spin-echo (Figure 2B) and short-tau inversion recovery T2-weighted images (Figure 2C), the mass was hypointense. After intravenous gadolinium contrast administration, first-pass perfusion images revealed no

Figure 1. Transthoracic echocardiography (A and B) shows a round, echogenic mass (arrow) with central echolucent areas, located at the posterolateral basal wall. Three-dimensional transesophageal echocardiography (C and D) confirmed the presence of a heterogenous mass (arrow) arising in the region of the posterior mitral valve annulus. The anterior (α) and posterior (β) mitral valve leaflets open (C) and close (D) well. RV indicates right ventricle; LV, left ventricle.
enhancement of the mass, in keeping with an avascular structure. On the early gadolinium-enhancement images, the mass remained hypointense (Figure 2D); however, the corresponding late gadolinium-enhancement images acquired 15 to 20 minutes later demonstrated a prominent rim of circumferential enhancement that surrounded a dark central core (Figures 2E and 2F). The characteristic low-signal intensity of the mass seen with multispectral cardiac magnetic resonance analysis and the strong peripheral late enhancement were consistent with a central area of caseous tissue surrounded by a fibrotic cap.

The anatomic location, heterogeneous echocardiographic appearances, and cardiac magnetic resonance signal characteristics of the mass were therefore thought to support a diagnosis of mitral annular caseous calcification. This diagnosis was confirmed by cardiac computed tomography, which revealed that the mass consisted of an isodense central area with a hyperdense rim of peripheral calcification (Figure 3). Of note, the aortic valve leaflets were also heavily calcified. The patient was recommended to undergo aortic valve replacement but subsequently declined surgery.

Caseous calcification of the mitral valve is a rare form of mitral annular calcification that typically affects the posterior annulus. The condition, also referred to as liquefaction necrosis, usually runs a benign course, although associated valvular dysfunction and cerebral embolization of necrotic material have both been reported. Appreciation of this unusual condition is important because the echocardiographic appearances are nonspecific and commonly misinterpreted. Differential diagnoses include cardiac tumors and myocardial abscess, with the result that accurate assessment of this pathology frequently requires complementary imaging techniques. To the best of our knowledge, this report is the first to comprehensively illustrate the characteristic appearances in a single case by use of the full spectrum of noninvasive cardiac imaging modalities (transthoracic and transesophageal echocardiography, cardiac magnetic resonance imaging, and cardiac computed tomography).

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