Circulation: Cardiovascular Imaging

CARDIOVASCULAR IMAGES

Caseous Calcification of the Mitral Annulus With Atrial and Ventricular Fistulization

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A 65-year-old women with past medical history of hypertrophic cardiomyopathy, prior myocardial infarction, hypertension, and transient ischemic attack presented in cardiology clinic with increased dyspnea (New York Heart Association class II) with no features to suggest endocarditis. The patient underwent a transthoracic echocardiogram, which showed features of hypertrophic cardiomyopathy and moderate mitral regurgitation. A cardiac magnetic resonance examination was performed for further evaluation. Cardiac magnetic resonance demonstrated features of hypertrophic cardiomyopathy, as well as a 4×3 cm ovoid cavity at the inferolateral aspect of the mitral annulus with signal intensity similar to the blood pool (Figure 1A and 1B). Communication between this ovoid cavity and the left atrium was seen (Figure 1A and 1B; Movies I and II in the Data Supplement). These findings were concerning for pseudoaneurysm versus caseous necrosis of mitral annulus calcification with rupture into the left atrium. Late gadolinium enhancement images identified peripheral enhancement of the residual mitral annulus with central cavitation (Figure 1C). Cardiac computed tomography and transesophageal echocardiogram were also performed to characterize the lesion better. Cardiac computed tomography demonstrated a contrast filled space between the posterior mitral valve and basal lateral myocardial wall with a peripheral rim of calcification (Figure 2A and 2B). In comparison, the previous computed tomographic scan from 8 years prior showed mitral annular calcifications (MACs) with no central contrast filled space (Figure 2C). Transesophageal echocardiogram confirmed the presence of the calcified mass with central echo lucent zone in the region of the posterior mitral annulus with systolic expansion (Figure 3A; Movie III in the Data Supplement). Communication between this cavity and the left atrium was seen, with a large eccentric color Doppler flow jet directed toward the interatrial septum (Figure 3B; Movie IV in the Data Supplement). Another communication between this cavity and the left ventricle was also seen with color Doppler flow jet directed into the lesion in systole (Figure 3B; Movie IV in the Data Supplement). In addition, mitral regurgitation was also demonstrated (Figure 3B; Movie IV in the Data Supplement). Because of progressive symptoms, and severe mitral regurgitation-like physiology, surgical management was advised, and the patient underwent mitral valve replacement. At the time of surgery, the posterior mitral valve leaflet was identified as a large mass of calcific material with cavitary change. Two posterior perforations and one anterior perforation were seen within this cavity. The entire posterior leaflet along with the subvalvular apparatus was completely excised and a 31 mm Edwards Perimount Plus valve was sewn in place. Overall, the imaging features were consistent with caseous calcification of mitral annulus with atrial and ventricular fistulization resulting in mitral regurgitation-like physiology. Assessment of excised mitral valve apparatus confirmed presence of diffuse leaflet thickening and fibrosis with calcific degeneration.

Key Words: dyspnea ■ echocardiography, transesophageal ■ ischemic attack, transient ■ magnetic resonance imaging ■ mitral regurgitation ■ mitral valve ■ multidetector computed tomography

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DISCUSSION

MAC occurs secondary to progressive calcification of the fibrous mitral annulus commonly affecting posterior mitral annulus with an estimated prevalence of ≈10%. Risk factors for the development of MAC include female sex, advanced age, chronic kidney disease, prior chest irradiation, and conditions that predispose to left ventricular hypertrophy.[1] Caseous calcification of the mitral annulus (CCMA) represents a rare variant of MAC seen in 0.63% of all patients with MAC.[2] It has been shown that CCMA is associated with higher prevalence of cardioembolic events compared with MAC. Imaging plays an important role in differentiating MAC from CCMA. On liquefaction of the central portion of the MAC it adopts a more ovoid mass-like appearance. Therefore, CCMA can often be confused with an intracardiac mass which is seldom the case with noncaseous MAC. On computed tomographic scan, CCMA typically appears as a sharply marginated ovoid mass at the posterolateral aspect of the mitral annulus with a hypodense or hyperdense center and a peripheral rim of calcification with no enhancement. Cardiac magnetic resonance may help in differentiating MAC from CCMA because of calcification of the entire lesion in MAC resulting in consistently low signal compared with presence of proteinaceous fluid in CCMA resulting in central high T1 signal. Low signal is typically seen on gradient echo, T2, and steady-state free precession sequences. Peripheral delayed contrast enhancement of the lesion has been reported.[3] Cardiac magnetic resonance may particularly be useful in differentiating CCMA from other causes,[4] such as malignant cardiac neoplasm or thrombi with the left chambers. Fistulization of the cavity into the left atrium or left ventricle is uncommon and may lead to cardioembolic stroke.[2] Fistulization may also lead to change in imaging characteristics of the cavity with central aspect resembling the blood pool. Flow acceleration in the area of fistulization may also assist in making the diagnosis. CCMA, a rarely encountered clinical entity, is important to recognize, particularly when complicated by fistulization, which may lead to cardioembolic and hemodynamic (mitral regurgitation-like physiology) consequences.

ARTICLE INFORMATION

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Disclosures

None.

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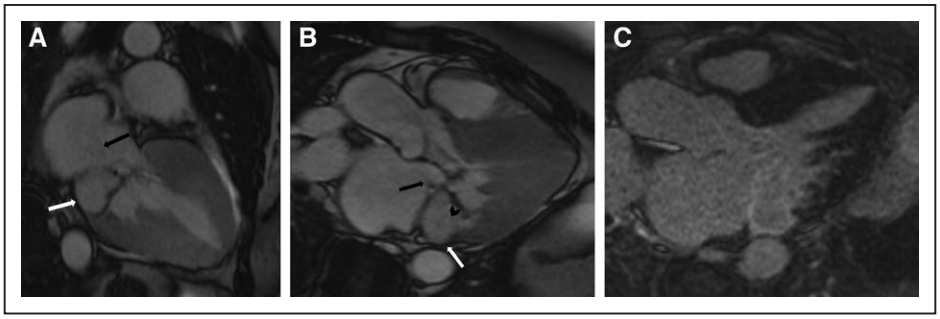

Figure 1. Cardiac magnetic resonance imaging.
A, Two-chamber steady-state free precession (SSFP) image shows an ovoid cavity (white arrow) at the posterolateral aspect of the mitral annulus with signal intensity similar to the blood pool. Flow acceleration (black arrow) demonstrates communication between this ovoid cavity and the left atrium. **B**, Three-chamber SSFP image shows an ovoid cavity (white arrow) at the posterolateral aspect of the mitral annulus with signal intensity similar to the blood pool. Flow acceleration (black arrow) demonstrates communication between this ovoid cavity and the left atrium. Another communication between the left ventricle and ovoid cavity is seen (black arrowhead). **C**, Three-chamber late gadolinium enhancement image shows peripheral enhancement of the residual mitral annulus with central ovoid cavitation.

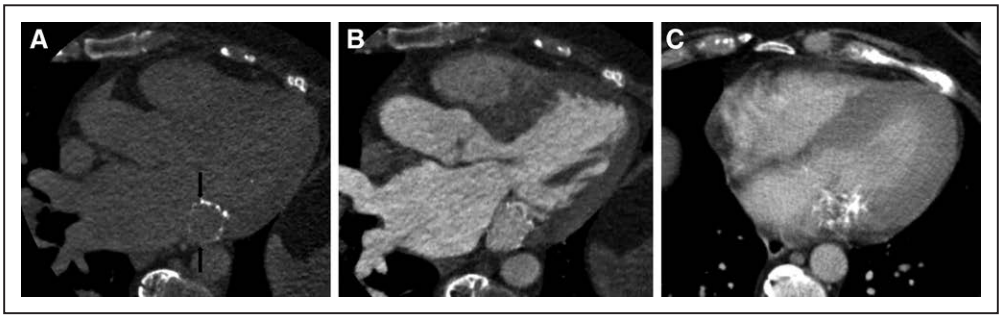

Figure 2. Cardiac computed tomography.
A, Three-chamber view of noncontrast cardiac computed tomography (CT) scan shows calcification at the periphery of the ovoid cavity at posterolateral aspect of the mitral annulus (black arrows). **B**, Three-chamber view of postcontrast cardiac CT scan demonstrates that the ovoid cavity at posterolateral aspect of the mitral annulus is filled with contrast. **C**, Four-chamber view of noncontrast cardiac CT scan from 2009 shows mitral annular calcification with no central contrast filled space.

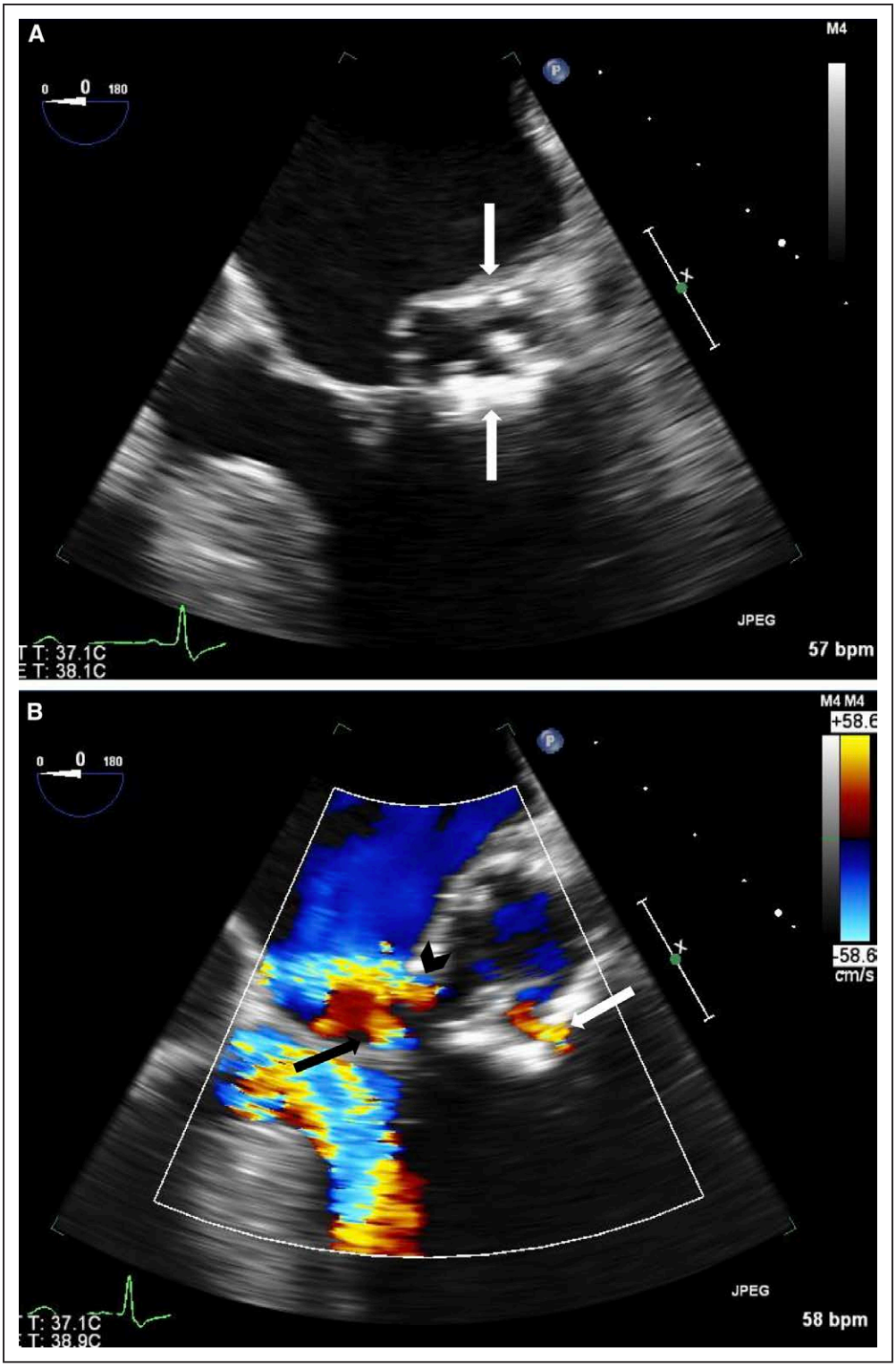

Figure 3. Transesophageal echocardiography.
A, Transesophageal echocardiogram (TEE) showing calcified mass with central echo lucent zone in the region of the posterior mitral annulus (white arrows). **B**, Color Doppler examination on TEE shows a communication between the ovoid cavity and the left atrium with eccentric color Doppler flow jet directed toward the interatrial septum (black arrowhead). Another communication between this cavity and left ventricle is seen with flow jet directed toward the lesion (white arrow). Flow jet because of mitral regurgitation (black arrow) is also seen.

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Video Legends

Video 1: Two chamber steady-state free precession (SSFP) cine shows an ovoid cavity at the posterolateral aspect of the mitral annulus with signal intensity similar to the blood pool. Flow acceleration demonstrates communication between this ovoid cavity and the left atrium.

Video 2: Three chamber steady-state free precession (SSFP) cine shows an ovoid cavity at the posterolateral aspect of the mitral annulus with signal intensity similar to the blood pool. Flow acceleration demonstrates communication between this ovoid cavity and the left atrium. Another communication between the left ventricle and ovoid cavity is seen.

Video 3: Transesophageal echocardiogram (mid-esophageal view) showing calcified mass with central echo lucent zone in the region of the posterior mitral annulus.

Video 4: Color Doppler examination on transesophageal echocardiogram (mid-esophageal view) shows a communication between the ovoid cavity and the left atrium with eccentric color Doppler flow jet directed towards the interatrial septum. Another communication between this cavity and left ventricle is seen with flow jet directed towards the lesion. Flow jet due to mitral regurgitation is also seen.