Caseous Mitral Annular Calcification Along the Anterolateral Annulus Causing Mild Mitral Regurgitation
– Multi-Modality Imaging and Diagnosis –
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Figure 1. (A) Cardiac computed tomography (CT) 2-chamber image showing a homogeneous, hyperdense mass with peripheral calcifications (arrow) anterolateral to the mitral valve. The left atrium and left ventricle are moderately enlarged. (B) Transesophageal echocardiogram (TEE) 2-chamber view showing a fixed, globular mass (arrow) with smooth, echogenic borders and central lucencies located along the anterolateral border of the mitral annulus. (C) Cardiac magnetic resonance imaging (MRI) 3-chamber steady-state free precession image and (D) TEE 4-chamber color Doppler image demonstrating eccentric, posteriorly directed, mitral regurgitation (arrowheads) secondary to involvement of the anterolateral annulus by the mass. Although the jet was not centrally-directed, proximal isovelocity surface area from the TEE indicated an effective regurgitant orifice of 0.17 cm² with a regurgitant volume of 25 ml, consistent with mild mitral regurgitation. (E) Cardiac MRI short-axis delayed-enhancement image shows only a thin rim of peripheral late enhancement (arrow) surrounding the hypointense mass. (F) Cardiac CT short-axis image again demonstrates a hyperdense mass with peripheral calcifications (arrow) along the anterolateral mitral annulus. (G) Cardiac MRI axial T1-weighted post-contrast image shows no enhancement of the mass (arrow). (H) Non-contrast axial image from positron emission tomography/CT shows a globular, homogeneous, hyperdense mass along the anterolateral mitral annulus. AoV, aortic valve; LA, left atrium; LV, left ventricle; MV, mitral valve.
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n asymptomatic 60-year-old Filipino man with a history of diabetes, hypertension, and carotid stenosis underwent urgent cardiac catheterization for electrocardiogram changes seen during a routine cardiology appointment. The coronary angiogram showed severe 3-vessel disease, but it also indicated a radiodense mass adjacent to the mitral valve.

Subsequent cardiac magnetic resonance imaging (MRI) for viability assessment showed an approximately 20-mm mass anterolateral to the mitral valve. The mass was hypointense to the myocardium and did not demonstrate enhancement (Figure 1G). Eccentric, posteriorly directed, mitral regurgitation was also noted (Figure 1C). Delayed enhancement images indicated an infarct in the anteroseptum involving >50% of the myocardial thickness. Additionally, a thin rim of peripheral late enhancement surrounding the hypointense mass was present (Figure 1E). Cardiac computed tomography (CT) with contrast was performed to further characterize the mass, which demonstrated a homogeneous, hyperdense mass with peripheral calcifications along the anterolateral mitral annulus (Figures 1A,F). A transesophageal echocardiogram was also performed, which confirmed a fixed, globular mass originating along the anterolateral mitral annulus (Figure 1B) and mild, eccentric, mitral regurgitation (Figure 1D). Due to suspicion for a cardiac neoplasm, non-contrast positron emission tomography/CT was done, which showed an ametabolic, hyperdense mass (Figure 1H) corresponding to the lesion seen on the other modalities.

The patient was ultimately taken to the operating room for 3-vessel coronary artery bypass grafting, as well as biopsy of the mass adjacent to the mitral valve. The contents of the mass were described as being the consistency of toothpaste. Frozen sections of the mass contents consisted of scant fragments of dense fibrous tissue with focal populations of fibroblasts (arrow), as well as extensive amounts of dense (dark purple, star) and fine (pale purple, diamond) calcification. (B) H&E-stained section (40× power) of the involved myocardium showing fine (pale purple, diamond) calcification of the myocytes, and fine and dense (dark purple, star) calcification on the surface of the muscle. H&E, hematoxylin and eosin.

Figure 2. (A) H&E-stained section (10× power) of the contents of the mass showing dense fibrous tissue with focal populations of fibroblasts (arrow), as well as extensive amounts of dense (dark purple, star) and fine (pale purple, diamond) calcification. (B) H&E-stained section (40× power) of the involved myocardium showing fine (pale purple, diamond) calcification of the myocytes, and fine and dense (dark purple, star) calcification on the surface of the muscle. H&E, hematoxylin and eosin.

Degeneration and calcification of the mitral valve annulus with age is a common occurrence, particularly in older women. MAC has been reported to occur in 10.6% of patients undergoing an echocardiogram. Caseous calcification of the mitral annulus is a rare variant, reported in only 0.06–0.07% of patients undergoing echocardiography. Caseous MAC consists of a mixture of calcium, fatty acids, and cholesterol, with the gross appearance resembling that of toothpaste. The specific factors that lead to caseous degeneration of the mitral annulus are unknown.

As with typical MAC, the caseous variant is usually an in-
cidental finding. As seen in the present case, the lesion may occasionally distort the mitral annulus, resulting in mitral regurgitation. More commonly, however, this structure may be misdiagnosed as a cardiac tumor, thrombus, or abscess, which may lead to unnecessary cardiac surgery.

Caseous MAC is typically located along the posterior mitral annulus. The present case was unusual in that it was located along the anterolateral annulus. Involvement of both the anterior and posterior annulus by a large caseous mass has been reported. To our knowledge, however, involvement of solely the anterior annulus has been reported in only 1 other instance.

On echocardiography, caseous MAC appears as a well-defined, round or semilunar, echodense mass with smooth borders, central areas of echolucency representing liquefaction, and no posterior acoustic shadowing. Cardiac MRI demonstrates a mass along the mitral annulus that appears hypointense to the myocardium on all sequences. Contrast enhancement is not seen, although peripheral late enhancement of a fibrous capsule may be observed. CT shows a homogeneous, hyperdense mass with denser peripheral calcifications. Once again, contrast enhancement is not present. Unless pre-contrast images are also acquired, however, the hyperattenuation may be mistaken for contrast enhancement, because the caseous material within the mass is very similar in attenuation to iodinated contrast.

The differential diagnosis for a calcified myocardial mass includes such entities as intramural myxoma, hydatid cyst, teratoma, angioma, rhabdomyoma, fibroma, and chronic hematoma or abscess. Misdiagnosis of caseous MAC as abscesses, tumors, and left atrial thrombi have been reported in the literature.

In conclusion, although caseous MAC is typically an incidental finding, accurate recognition is needed to avoid mistaking the lesion for a myocardial tumor or abscess, which may result in unnecessary cardiac surgery. Although usually located along the posterior annulus, an anterolateral location with characteristic imaging findings should still call to mind this diagnosis. Although this entity can be diagnosed on cardiac MRI, multi-modality imaging, particularly with non-contrast CT, allows for the confident, prospective, diagnosis of this rare entity.

Disclosures

No potential conflicts of interest to disclose for any of the authors.

References