

Caseous calcification of mitral annulus

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Introduction

Caseous calcification of the mitral valve annulus (CCMA) is a rare variant of mitral annular calcification (MAC) (1). Because it is similar in appearance to lesions such as a cyst, a thrombus, a tumor, or an abscess, misdiagnoses and unnecessary surgical treatments have been reported. It is therefore important to consider CCMA in the differential diagnosis of other similarly appeared lesions. We report a 69-year-old man with CCMA detected incidentally by the intraoperative transesophageal echocardiography during coronary artery bypass grafting.

Case report

A 69-year-old man was referred for surgical coronary revascularization to our department. He had a long-time history of hypertension, type 2 diabetes mellitus, hyperlipidemia and hyperuricemia. He also had end-stage kidney failure and had been on continuous ambulatory peritoneal dialysis (CAPD) for 2 years. He had a long history of coronary artery disease with remote percutaneous coronary interventions (PCIs), first in the context of an acute myocardial infarction, and subsequently for angina recurred 6-years later. On referral, he described recurrent angina for approximately 6 months. Coronary angiography showed left main trunk disease and severe stenosis in the left anterior descending (LAD), and in the left circumflex (LCx) branches. He was referred for coronary bypass surgery.

Intraoperative transesophageal echocardiography (TEE) revealed an ovoid mass (short and long axis diameter 20 mm × 30 mm) with smooth surface and central echolucency, which

was located underneath the posterior mitral annulus. Based on its characteristic features, the mass was diagnosed as CCMA (*Figure 1*).

Because no significant mitral valvular dysfunction was detected (trace regurgitation and normal valvular opening) no surgical intervention for this lesion was performed. Routine CABG (both internal mammary arterial grafts to LAD and LCX, and saphenous vein grafts bridging between ascending aorta and diagonal branch) was done. Postoperative transthoracic echocardiography (TTE) showed no morphological change of CCMA with normal mitral valve function. Good patency of all bypass grafts was confirmed by multi-detector-row computed tomography (MDCT).

Retrospective review of the preoperative TTE and serial CT images confirmed the existence of CCMA, which had been erroneously diagnosed as MAC. Interestingly it was present in recent CT images but not in the film taken 7 years previously (*Figures 2,3*).

Discussion

Mitral annular calcification describes degenerative changes of cardiac fibrous skeleton, and mainly occurs in the space between the crest of the posterior left ventricular muscle and the posterior mitral annulus (2), although it sometimes occurs circumferentially. MAC is found in almost 10% of population older than 50. Caseous calcification of mitral valve annulus (CCMA) is a rare variant of MAC, constituting 0.6% of all MAC cases (1,3). But whether CCMA will sequentially develop into MAC is unknown, and vice versa.

A calcified rim is surrounding caseous material that

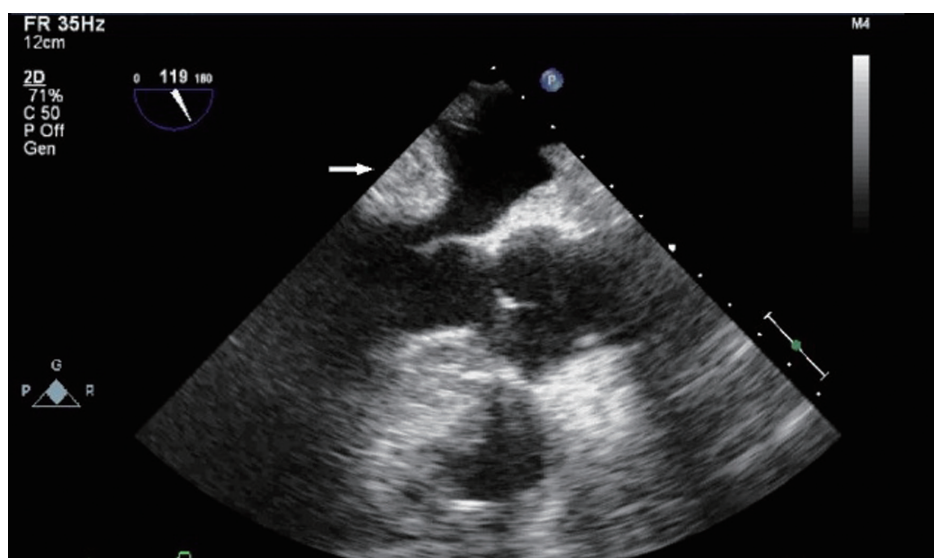


Figure 1 An intraoperative transesophageal echocardiogram. Ellipsoid mass (20 mm × 30 mm) was detected beneath the posterior mitral annulus. Its surface was smooth and the center was echolucent

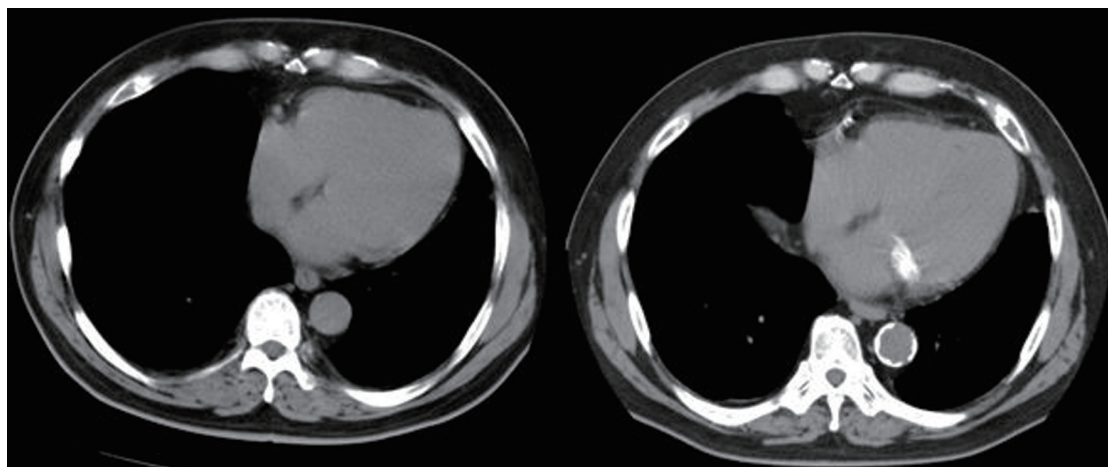


Figure 2 Serial computed tomograms taken 7 years previously (left) and immediately before CABG (right). During these 7 years calcification had progressed to form CCMA, which was misdiagnosed as simple MAC preoperatively

is composed of an admixture of calcium, fatty acids, and cholesterol with a toothpaste-like texture. Microscopic examination shows an amorphous, acellular, basophilic, and calcific structure, with a chronic inflammatory reaction manifested mainly by macrophages (4). Though the lesion is usually located on the posterior mitral annulus, a few reports of the anterior annular involvement have been described. CCMA has a strong relation to the history of hypertension (100%). Although the association with chronic renal failure or hemodialysis is comparatively less obvious (14%), implication of abnormal calcium metabolism in this

disease subset should be considered (3,5).

CCMA is usually a benign and asymptomatic lesion with no need of therapeutic intervention. Because misdiagnoses and unnecessary surgical treatment is not uncommon, CCMA should be considered in the differential diagnosis of similarly appeared lesions such as a cyst, thrombus, tumor, or abscess.

In this context, usefulness of several imaging modalities including TEE, magnetic resonance imaging (MRI), and CT has been reported. With echocardiography, CCMA usually appears as a large, round, echo-dense mass with

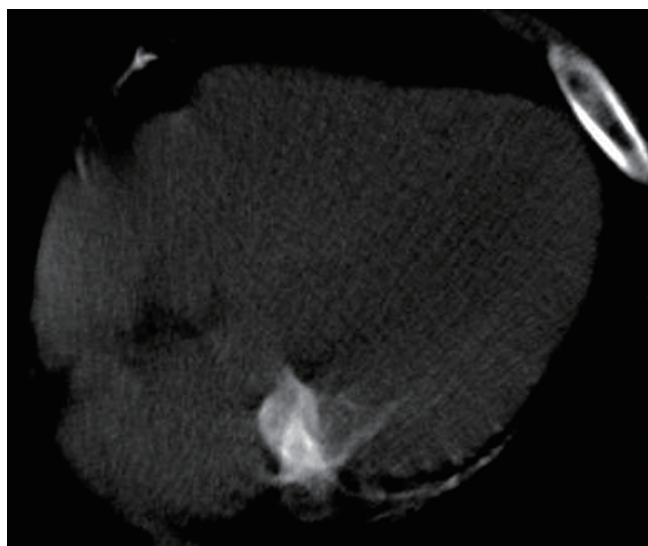


Figure 3 The peripheral calcification and central hyperdensity can be seen by changing the grayscale/window setting of the right panel of *Figure 2*

smooth borders and central echolucency. With cardiac MRI, the mass has a low-signal intensity (signal void) on T1 and T2 weighted imaging (6,7). CT scans typically show a round, smooth mass with central hyperdensity and peripheral dense calcifications. The lack of soft tissue attenuation and contrast enhancement is a clue to differentiate CCMA from a tumor. In our experience, combination of multiple imaging modalities is often necessary for correct diagnosis.

On rare occasion, if mitral dysfunction and/or conduction disturbance appear, surgical intervention should

be considered. In these patients, mass excision and valvular repair or replacement is the treatment of choice (5,8).

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