CASE REPORT

Multimodality imaging assessment of a caseous calcification of the mitral valve annulus

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Caseous calcification of the mitral annulus (CCMA) is a rare echocardiographic finding. It is commonly misdiagnosed as an abscess, tumor or infective vegetation on the mitral valve. Since it is a benign process, differentiating it from malignant intra-cardiac mass is primordial to avoid unnecessary surgery. Various imaging modalities can be complimentary for definitive diagnosis. We present a case of CCMA in a 71-year-old female patient. Her medical history revealed hypertension, diabetes mellitus, hyperlipidaemia and coronary artery disease. She was referred to our department for coronary catheterization because of angina symptoms upon minimal exertion. The lesion was detected during echocardiography and was defined as a mass of heterogeneous content with calcification points, located at the posterior side of the mitral valve annulus. Restricted motion of the posterior leaflet and the mass effect caused only minimal mitral regurgitation. To establish the correct diagnosis, we performed the full spectrum of noninvasive cardiac imaging modalities. Transesophageal echocardiography identified well-organized, composite lesion with regular edges, markedly calcified margins and more echolucent central portion. A computed tomography (CT) was performed, showing a hyperdense mass with hypodense center and a calcified peripheral rim located at the posterior mitral ring. Cardiac magnetic resonance imaging (MRI) showed that the mass was hypointense with respect to the myocardium in the T1 and T2-weighted sequences and only presented late-phase enhancement in the surrounding capsule. Based on the CT and MRI findings, the diagnosis of CCMA was established. The patient was managed conservatively.

Keywords: Mitral valve, Imaging, Echocardiograpgy, CMR, Calcification

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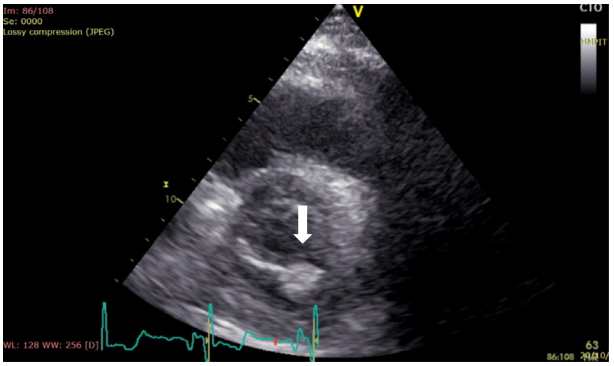
Introduction

Caseous calcification of the mitral annulus (CCMA) is a rare variant of mitral annular calcification that may be easily misdiagnosed or confused with an abscess, tumor, or infective vegetation. Multiple imaging modalities can be complementary to establish the correct diagnosis in suspicious cases. Due to it asymptomatic course, only monitoring of the patient's progress is recommended in most cases of CCMA. Surgery should be reserved for patients with significant valvular dysfunction or an uncertain diagnosis.

Case report

We present a case of CCMA in a 71-year-old female patient. Her medical history revealed hypertension, diabetes mellitus, hyperlipidemia, and coronary artery disease. She was referred to our department for coronary catheterization because of angina symptoms upon minimal exertion. She did not show any signs of infective endocarditis (fever, anorexia, weight loss, or night sweats). Physical examination was unremarkable. Laboratory tests showed no significant abnormalities, especially markers of inflammation (erythrocyte sedimentation rate and C-reactive protein were within normal limits). Transthoracic echocardiography revealed a mass of heterogeneous content with calcification points, located at the posterior side of the mitral valve annulus, not exerting a significant effect on mitral valve function (Fig. 1A and B). Transesophageal echocardiography (TEE) was performed to better delineate the mass, and identified a rounded immobile mass (17 mm × 16 mm) with regular edges, markedly calcified margins, and a more echolucent central portion, localized in the posterior part of the mitral annulus (Fig. 2). The valve geometry was distorted with an anteriorly displaced posterior leaflet resulting in mild regurgitation. Stenosis was absent. TEE did not show any thrombus or relevant spontaneous echo contrast in the left atrium and atrial appendage. No images suggestive of vegetations were observed in the aortic, tricuspid, or pulmonary valves. In computed tomography (CT), the entity appeared as a round mass with a hyperdense center and calcified peripheral rim. Cardiac magnetic resonance (CMR) cine and dark blood images demonstrated a hypointense mass involving a mitral valve annulus. Perfusion images revealed no enhancement of the mass compared with normal myocardium,

A



B

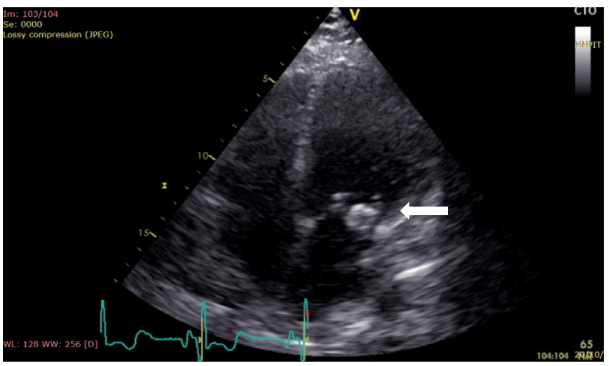


Figure 1. Transthoracic two-dimensional visualization of the mass. (A) Parasternal short axis view and (B) apical four-chamber view.

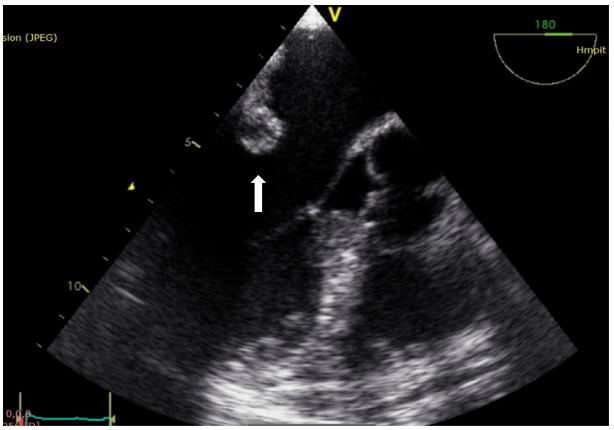


Figure 2. Transesophageal echocardiography showing a rounded, calcified mass with interior content of lower echogenicity.

consistent with an avascular mass. Additionally, a thin rim of peripheral late enhancement surrounding the hypointense mass was present (Fig. 3). Given the CT and CMR imaging findings and the absence of mitral valve dysfunction, surgery was deferred and conservative management was chosen. The echocardiographic follow up at 6 months showed no significant changes.

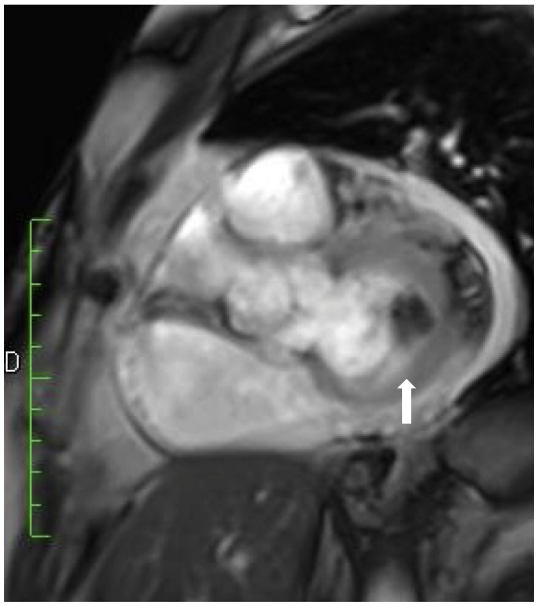


Figure 3. Basal short axis cine image shows hypointense round mass-like lesion (arrows). Delayed postcontrast cardiac magnetic resonance image shows peripheral enhancement (arrow).

Discussion

CCMA is a less-known and rarely described entity representing a variant of mitral annular calcification, which is typically located in the posterior mitral annulus. It represents <1% of all mitral calcifications, with a prevalence of 0.067% in the general population [1]. CCMA consists of a mixture of calcium, fatty acids, and cholesterol, with the gross appearance resembling that of toothpaste. The pathogenetic mechanism of CCMA has not yet been defined, but as an expression of atherosclerosis, it has identical risk factors as those of cardiovascular disease [2]. Incidental finding is the most common clinical presentation of CCMA, however some patients report palpitations, dyspnea, and rarely syncope. CCMA can cause obstruction of the transmitral left ventricular inflow tract or lead to prolapse of a mitral leaflet causing mitral regurgitation [3]. Systemic embolization has also been reported. No relationship has been described between CCMA and occurrence of atrial dysrhythmias.

Caseous calcification is characterized echocardiographically as large, round, tumor-like calcifications, with well-defined edges, echodense periphery, and a central echolucent area resembling liquefaction. It is less reflective than common calcification of mitral annulus, and usually there are no posterior acoustic shadowing artifacts behind it [4]. The use of transesophageal study enables one to define better the location, consistency, and aspect of the mass mainly in patients with limited acoustic window. Because CCMA is a rare condition, cardiac imagers may not be familiar with this incidental calcific mass, and it is often misdiagnosed as a cardiac tumor, thrombus, abscess, or even hydatid cyst, which may lead to unnecessary interventions [5].

In the present case, the localization, calcifications in the borders, and absence of peripheral extension made the diagnosis of cardiac tumor unlikely. Concerning endocarditis, the clinical context was essential to differentiate CCMA from vegetation.

Multiple imaging modalities can be complementary in differentiating between caseoma, tumor, and abscess. In this instance, correlating the echocardiographic findings with imaging on CT and CMR enabled an appropriate diagnosis without invasive studies [6]. On CT scan, CCMA appears as a well-defined oval or crescent-shaped hyperdense mass with peripheral calcification, usually along the posterior mitral annulus, has a high number of Hounsfield units, and lacks contrast enhancement [7].

CMR is considered to be the technique of choice in doubtful cases. CCMA is seen as a hypointense lesion both in T1- and T2-weighted sequences [8]. In postcontrast studies, contrast enhancement on first pass sequences is not expected; however, it may exhibit peripheral enhancement during delayed postcontrast sequences.

This case illustrates the complementary role of different imaging modalities. Although echocardiography was used to initially detect the mass and to assess its functional significance, the CMR was used to exclude other potential etiology. Late gadolinium enhancement is particularly interesting in the diagnosis as it shows unique peripheral enhancement of the mass. Conventional CT imaging confirmed the calcified nature of the mass [9].

Currently, there is no consensus on the optimal management for CCMA. The current data suggest conservative medical management for CCMA when the diagnosis is certain and there is no valvular dysfunction.

CCMA usually carries a benign prognosis, but patients so diagnosed require careful follow-up observation if they develop new symptoms or have a high propensity for tissue calcium deposition (as in chronic renal failure). Anticoagulation should be considered in patients who present with embolic manifestations.

The indications for surgical intervention include mitral valvular dysfunction, embolic manifestations, or when it is impossible to rule out the

possibility of a tumor. Mitral valve replacement is preferred to mitral valve repair. Wide debridement and excision of CCMA may result in perioperative embolic stroke. Minimal incision, drainage, closure, and suture obliteration of the cavity offers a safe and reproducible technique for surgical management CCMA [10]. Given the absence of mitral valve dysfunction and embolic events, conservative management was chosen for our patient.

In summary, caseous calcification represents a rare, underappreciated variant of mitral annular calcification that should be differentiated from an abscess or tumor. Although echocardiography is the mainstay imaging modality for diagnosis of CCMA, multimodality imaging, including TEE, cardiac CT, and CMR, can be used to confirm the diagnosis and avoid unnecessary surgery.

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