

A rare case of caseous necrosis of mitral valve



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ABSTRACT

Caseous calcification of the mitral annulus, an uncommon kind of intracardiac mass, is a form of degenerative disease that afflicts the mitral valve causing liquefactive or caseous necrosis. Caseous calcification of mitral annulus is rarely encountered in practice; however, it should be on the list of differential diagnosis of any intracardiac masses located close to the mitral annulus.

Key words: Caseous necrosis; Mitral annulus; Calcification; Intracardiac masses

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INTRODUCTION

Caseous calcification of the mitral annulus, an uncommon kind of intra-cardiac mass, is a form of degenerative disease that afflicts the mitral valve causing liquefactive or caseous necrosis. It is a benign condition, however, with a possibility of embolic consequences. It is, therefore important to be familiar with its imaging characteristics and differentials to avoid misdiagnosis and unnecessary surgical interventions. We herein present a case of caseous calcification of mitral annulus (CCMA) in an elderly woman.

CASE PRESENTATION

A 71-years-old female patient came to our hospital with a history of incidentally detected mass in the left ventricle on echocardiography. The patient was referred for contrast-enhanced Cardiac magnetic resonance imaging (MRI) for evaluation and characterization of the cardiac mass.

A cardiac MRI was done in Siemens Avanto 1.5 T MRI scanner with anterior body coil and posterior spine array. The following sequences were done:

- TrueFISP for anatomic definition
- Dynamic cine imaging (SSFP) for cardiac chamber and wall-motion analysis
- T1-weighted turbo spin echo images for morphologic analysis
- T2-weighted fat saturation sequences for the assessment of myocardial edema
- Dynamic perfusion scan
- T1 VIBE pre and post contrast
- Gadolinium-chelate was infused intravenously via a left arm antecubital IV at a dose of 0.2 mmol/kg at a rate of 4 ml/sec followed by 30 mL of normal saline at the same rate
- PSIR sequences for evaluation of late gadolinium enhancement, preceded by a TI scout to evaluate the optimal nulling time of the LV myocardium

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- Scan was acquired, post-processed, and reported in accordance with the guidelines of the Society for Cardiovascular Magnetic Resonance.

Patient information: Height 153 cm; Weight 75 kg; BSA = 1.73 m², eGFR-WNL.

Cardiac MRI revealed a non-enhancing well-defined hypointense mass in the posterior mitral annular region with an epicenter in the posterior mitral leaflet and broad based towards the annulus causing mild mitral stenosis and regurgitation, indenting on the basal inferolateral and inferior segments (Figure 1a-d). Correlative computed tomography (CT) (Figure 2a-c) and radiograph of chest were done which showed a dense calcified lobulated lesion in the posterior mitral region (Figure 3a and b). The final diagnosis was given as CCMA/posterior leaflet of the mitral valve.

REVIEW OF LITERATURE

The prevalence of echocardiography is 0.64% in patients with mitral annular calcification (MAC) and 0.068% in all studies in large series of patients of all ages.¹ However, the prevalence in an autopsy series was 2.7%,² which indicates that this condition is yet under-recognized.

It is an atheroma-like lesion, which might be a part of multiple atherosclerotic lesions in other vascular beds.^{3,4}

The exact underlying pathogenesis, although not well understood; elevated levels of blood calcium in patients with chronic renal disease undergoing dialysis has been proposed as an etiological factor.⁵ Microscopically, caseous calcification is composed of basophilic areas with surrounding fibrous tissue. Cartilage tissue may also be rarely encountered.⁶

Extrinsic coronary artery compression is a rare disease entity. We report a rather peculiar cause of extrinsic artery compression: CMAC inducing significant mitral stenosis and compressing the circumflex artery leading to myocardial infarction. To the best of our knowledge, this is the first case of extrinsic artery compression caused by CMAC.⁷

CMAC is often an asymptomatic condition but patients can present with conduction abnormalities or with embolization of caseous material. CMAC is a dynamic process, can grow in size and infiltrate the adjacent myocardium. The center of CMAC contains liquefied calcifications, cholesterol, and fatty acids.

CCMA is a rare variant of MAC, most frequently recognized in the elderly, particularly women. Although

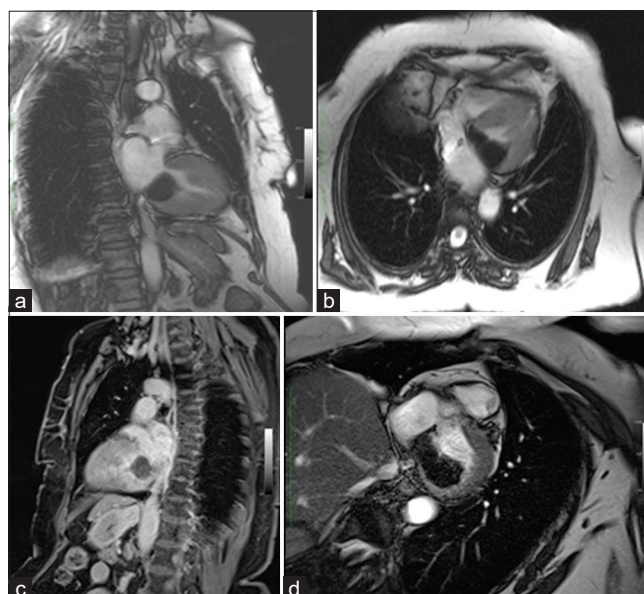


Figure 1: (a-d) Magnetic resonance imaging showing a well-circumscribed, lobulated hypointense non-enhancing lesion in the posterior mitral annulus

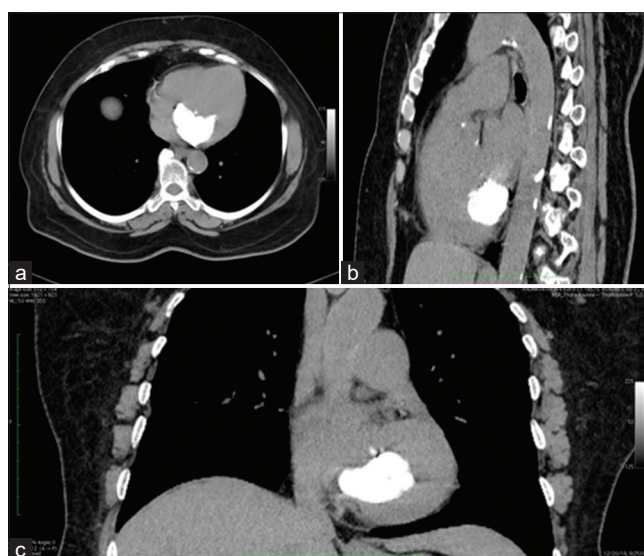


Figure 2: (a-c) Axial, sagittal, and coronal non-contrast computed tomography scan of the thorax showing densely calcified lobulated lesion in the mitral valve

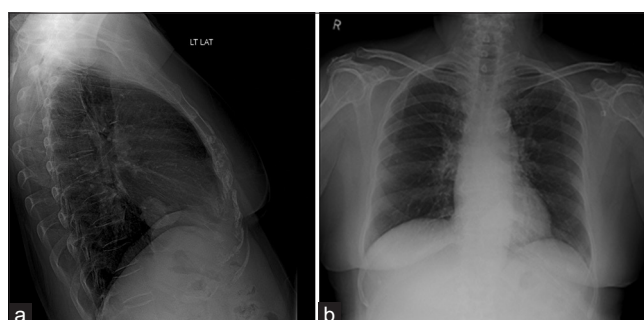


Figure 3: (a and b) Chest X-ray PA and lateral views show a well-defined lobulated calcific opacity in the region of the mitral valve

it is believed to be benign, CCMA may lead to abnormal flow across the mitral valve, resulting in chronic mitral regurgitation or less likely mitral stenosis.⁸

DISCUSSION

Caseous necrosis of the mitral valve is a rare form of MAC. This is brought on by atherosclerosis with a central “caseous” necrosis. The condition has gained significance in imaging as it can resemble a tumor. It is more commonly seen in the posterior atrioventricular groove of the mitral annulus.

An association has also been found with old age, the female gender, and hypertension. However, altered calcium-phosphate metabolism is seen to have the strongest association with the same.

MAC, in general, since recent reports note that calcification of the mitral annulus is related to atherosclerosis in other vascular beds including atherosclerotic coronary artery disease.^{4,9}

Patients with CCMA are usually asymptomatic or present with non-specific clinical symptoms. Spontaneous resolution has also been reported in the literature with a resolution of symptoms if any.¹⁰

Although it is generally thought to represent a benign entity, it has occasionally been associated with embolic phenomena, and surgery may be indicated in rare cases.

Diagnosis is usually made on the basis of a clinico-radiological picture. Echocardiography is the usually the first line of investigation. Typically, these lesions appear as a well-defined echogenic, round, or semilunar mass located on the posterior mitral annulus. A relatively hypoechoic central area may be present indicative of liquefaction.¹¹

Cardiac CT and MRI also add value to the confirmation of diagnosis. On multi-slice CT, it is a diffusely calcified mass. A cardiac protocol need not be followed in this case as a non-contrast CT of the thorax with a few slices encompassing the mitral annulus region would be sufficient for a definitive diagnosis. It does not enhance after IV contrast administration.¹²

Cardiac MRI is extremely helpful in equivocal cases. CCMA is a hypointense lesion on both T1- and T2-weighted sequences.¹³ In post-contrast studies, contrast enhancement on first-pass sequences is not seen; however, it may enhance peripherally enhancement on a delayed sequence.

Main differential diagnoses of CCMA include cardiac myxoma and other neoplasms, valvular endocarditic

vegetations, abscesses, and thrombi. Heterogeneous mass lesions with local invasion and metastases favor the diagnosis of cardiac malignant neoplasm. In a setting of sepsis or widespread infection, the presence of a mobile mass should raise the suspicion of a vegetation or abscess.¹⁴ Myxoma is a mobile enhancing soft tissue lesion which does not contain extensive calcification.

CONCLUSION

CCMA is rarely encountered in practice; however, it should be on the list of differential diagnoses of any intracardiac masses located close to the mitral annulus. The increased incidence of embolic events signifies the reporting of characteristics of the lesions such as size and mobility. Echocardiography is the first-line imaging modality for diagnosis of CCMA. However, confirmation of the diagnosis is by Cardiac MRI.

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