A Report of Two Cases with Caseous Annular Calcification of the Mitral Valve

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Abstract

Introduction: Caseous calcification of the mitral annulus (CCMA) is a rare variant of mitral annulus calcification, a chronic and degenerative mitral valve fibrous ring process. It usually carries a benign prognosis. The prevalence of CCMA is about 0.06%-0.07% of the population. CCMA is generally diagnosed incidentally and can be confused with other intracardiac masses such as cardiac tumors, abscesses, vegetation, or calcified thrombi. Multimodality imaging, including TEE, cardiac computed tomography, and cardiac magnetic nuclear resonance, can easily differentiate CCMA from other masses and help avoid unnecessary surgery. CCMA is typically located in the basal area of the posterior mitral valve, and the calcification seems like a round, large, soft mass with a central echo-dense location. CCMA may have a benign course, but it may sometimes be complicated with mitral valvular dysfunction, systemic embolization, or conduction abnormalities in the scenarios mentioned above, as well as when the diagnosis is unclear, surgery is indicated. Mitral valve replacement should be preferred compared to mitral valve repair.

Conclusions: It is important to note that the decision for mitral valve replacement, including CAC cases, should be individualized based on various factors, including the patient’s clinical condition, symptoms, severity of valve disease, and associated comorbidities.

As scientific understanding and research progress, there may be ongoing developments and refinements in the conservative and surgical management of CCMA.

Keywords: Caseous calcification, mitral annulus, surgical management

Introduction

Caseous calcification of the mitral annulus (CCMA) is a rare variant of mitral annulus calcification, a chronic and degenerative mitral valve fibrous ring process. It seems familiar in older adults, particularly in women. It usually carries a benign prognosis, and current data suggest conservative therapy and follow-up unless complicated with mitral valve dysfunction (mitral valve stenosis/mitral regurgitation) or systemic embolization. Also, it can resolve spontaneously, so the critical point of managing this entity is avoiding unnecessary surgery. In our cases, the indications to operate were valvular dysfunction, specifically mitral valve regurgitation.

Case 1

A 56-year-old female patient with CMAC was diagnosed with trans-thoracic and confirmed by histologic examination after surgery. The patient complained about chest discomfort and dyspnea for several weeks. The trans-thoracic echocardiogram showed a rounded mass in the posterior...
mitral annulus with a faint central echo-lucent area. The posterior mitral valve leaflet seemed to be retracted.

This prompted the need for trans-esophageal echocardiography, which revealed moderate to severe mitral regurgitation and the exact characteristics of the mass located in the posterior annulus. The patient underwent resection of the group, closure of the cavity with a pericardial patch, and mitral valve replacement. The postoperative period was uneventful. (fig. 1)

![Figure 1. Intraoperative toothpaste-like view of the caseous calcification](image)

**Case 2**

A 72-year-old female patient is admitted to our clinic with a mass in the posterior mitral valve ring, probably a caseoma. For months, she has complained of dyspnea and severe fatigue, which have progressed in the last two or three months. Trans-esophageal echocardiography revealed a slightly thickened mitral valve with severe mitral regurgitation, calcified mitral annulus, and a 3.5 cm mass with noted prolapse in the left ventricle starting from the annulus, probably caseous calcification. The patient underwent resection of the mass and mitral valve replacement with a biological prosthesis no. 29 Epic Supra.

It was observed that the entire mitral ring was significantly damaged, the mass prolapsed mostly at the P2 level, and calcification extended far down to the posterior wall of the left ventricle. The content was caseous. The cavity was drained, and aggressive debridement was performed. The procedure resulted in the patient suffering a complete A-V block. Therefore, a definitive pacemaker was placed on the fourth postoperative day with no further complications.

**Discussions**

The prevalence of CCMA is about 0.06%-0.07% of the population [1]. Although no standard protocol exists for the optimal management of CCMA, current data suggest conservative medical management for CCMA when there is especially no mitral valvular dysfunction or embolic manifestations. Valve dysfunction (valve stenosis and mitral regurgitation) [2], systemic embolization [3], abnormalities in conduction [4], and uncertain diagnosis are indications for curative surgery. Although there are successful examples of both mitral valve repair and replacement, mitral valve replacement (MVR) is the most appropriate choice compared to mitral valve repair [5]. Complications may arise due to aggressive debridement, such as an increased risk of left ventricular perforation, complete A-V blocks, etc. In cases of left ventricular perforation risk, mitral valve replacement could support the area in question, which is why most surgeons prefer valve replacement to mitral repair in cases of aggressive debridement.

Also, other international case reports show that the mass may extend into the left atrial wall or the ventricular endocardium during surgery. Thus, unroofing of the cavity may continue to expose necrotic debris to the systemic blood flow. MVR could support this area and prevent ventricular rupture [6, 7, 8].

While most patients are instead treated conservatively, and spontaneous resolution or transformation back to typical calcification has been described, there are also cases where CCMA recurs even after surgical excision[9, 10]. Therefore, a thorough follow-up is advised even after surgery.

**Conclusions**

It is important to note that the decision for mitral valve replacement, including CAC cases, should be individualized based on various factors, including the patient’s clinical condition, symptoms, severity of valve disease, and associated comorbidities.

As scientific understanding and research progress, there may be ongoing developments and refinements in the conservative and surgical management of CCMA.

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**References**


