

MAComa: Caseous calcifications presenting as intracardiac mass

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Abstract

Caseous liquefaction necrosis of mitral annular calcifications are rare. Rupture of its capsule may lead to systemic embolization. Surgical management entails Incision and Drainage with either mitral valve repair or replacement.

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Caseous calcifications of the mitral annulus(CCMA) have been reported with a 2.7% prevalence in a necropsy-series¹. However, the exact frequency is unknown, and it's rare in our own clinical experience. An echocardiographic-study estimated the prevalence in the general population as 0.06%–0.07%².

These intra-cardiac tumors are often incidental findings on Echocardiography(Figure1) in older-aged patients(e.g. diabetic-, hypertensive-, renal-failure patients), and can be mistaken for an abscess in the setting of questionable endocarditis. They are encapsulated, and rupture of its capsule can lead to chimney-like protrusions of a thick toothpaste-like material, with potential for severe embolic sequelae. The surgical approach entails Incision & Drainage of the lesion(Figure1), with either mitral valve repair or replacement. Our general approach is through the left atrium(Video1).

In this case the pathologic evaluation demonstrated calcifications with foci of lymphoplasmocytic infiltrates, with a mix of hemosiderin-laden neutrophils and macrophages. There were focal non-necrotic cardiomyocytes identified in the specimen. The microbiology evaluation was negative for an infectious process. Clinically, the mass did not cause stenosis and only mild regurgitation. However, the patient also required a single coronary-artery bypass.

Although previously referred to as CCMA, we want to propose a different name, which better describes the lesion as an intracardiac mass, which is derived from liquefaction of mitral annular calcifications(MAC): *a MAComa*. These lesions typically occur in the posterior mitral annulus, and only rarely involve the anterior annulus. The exact mechanism of liquefaction and caseation is not well understood but is thought to be related to an altered Calcium phosphate metabolism, as its prevalence is more common in end-stage renal failure patients³. Moreover, lipid-laden macrophages are implicated in the etiology of the liquefaction process, as these lesions more frequently occur with higher serum cholesterol levels. The condition can also be associated with bradyarrhythmias and/or atrioventricular blocks, due to the proximity of the mitral valve annulus to the conduction system³.

References

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Figures

Figure 1. A. Preoperative transesophageal Echo shows a 2cm x 1.3cm sub-valvular mitral valve mass, under the posterior mitral valve leaflet. **B.** Postoperative Echo. **C.** Intraoperative image demonstrating fullness underneath the posterior mitral valve leaflet. **D.** Caseous/toothpaste-like material underneath the mitral valve.

Videos

Video 1. MAComa: Caseous liquefaction necrosis of mitral annular calcifications.

