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Case Report

Multimodality imaging in assessment of mitral valve tumors: An unusual papillary fibroelastoma? ☆

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ABSTRACT

Mitral valve masses are uncommon. These tumors and tumor-like lesions may have similar morphological and clinical characteristics, but different outcomes. Unlike valvular tumors, caseous calcification of the mitral annulus (CCMA) is a benign degenerative disorder, commonly misdiagnosed, thus differentiating it from other mitral valve masses is important to avoid unnecessary surgery. Multimodality imaging can prove a valuable tool for definitive diagnosis. We present a case of a 72-year-old female patient, with coronary artery disease, referred for angina symptoms. Echocardiography detected a mass in the mitral valve annulus. Cardiac magnetic resonance imaging showed a mobile mass respecting the myocardium suggesting an atypical papillary fibroelastoma and surgery was indicated. However, the definitive diagnosis, after histological examination, was CCMA. The aim of this case report is to illustrate the difficulty in differentiating between mitral valve masses and the repercussion on the subsequent management, emphasizing the pivotal role of multimodality imaging.

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Introduction

Mitral valvular masses comprise a heterogeneous group of disorders. Autoimmune diseases, thrombotic lesions, extracardiac malignancies, and primary cardiac tumors account for the majority of cases, creating a diagnostic challenge. Unlike valvular tumors, mitral annular calcification (MAC) is a chronic degenerative disorder of the mitral valve fibrous ring, commonly seen in the elderly, particularly in women [1]. Caseous calcification of the mitral annulus (CCMA) is a less known, rarely described variant, typically located in the posterior mitral annulus [2]. It is often misdiagnosed as a cardiac tumor, thrombus, or abscess leading to unnecessary cardiac surgery [3].

In this report, we document a case of mitral valve mass casually found in the echocardiographic examination.

Case report

A 72-year-old female with high cardiovascular risk (arterial hypertension, dyslipidemia, and diabetes mellitus) and

history of coronary artery disease with multiple percutaneous coronary interventions presented with angina symptoms upon minimal exercise evolving for 2 months. The physical examination was completely normal; blood pressure was 130/70 mmHg and heart rate 63 beats per minute. The electrocardiogram was normal. Laboratory tests results showed no anomalies.

Transthoracic echocardiography (TTE) revealed an echo dense spherical mobile, tumor-like mass (21×15 mm, area 3.15 cm²) found in the periannular posterior region close to the ventricular side of the posterior mitral leaflet (Fig. 1). The mass was sharp and had distinct borders with a different and bright echogenicity in comparison to the adjacent myocardium. The posterior leaflet was stretched and arched over the mass, while the anterior leaflet was normal. On Doppler color-flow mapping, a mild regurgitation was seen without stenosis. The left ventricle was mildly hypertrophic with normal segmental wall motion and the left atrium was slightly enlarged. Coronary angiography showed no modifications compared to the previous one, but a calcific mass attached to the mitral annulus could be observed (Fig. 2).

Cardiac magnetic resonance (CMR) imaging was performed for a more precise evaluation. It showed a hypointense mass in the posterior region of the mitral annulus in both T2 se-

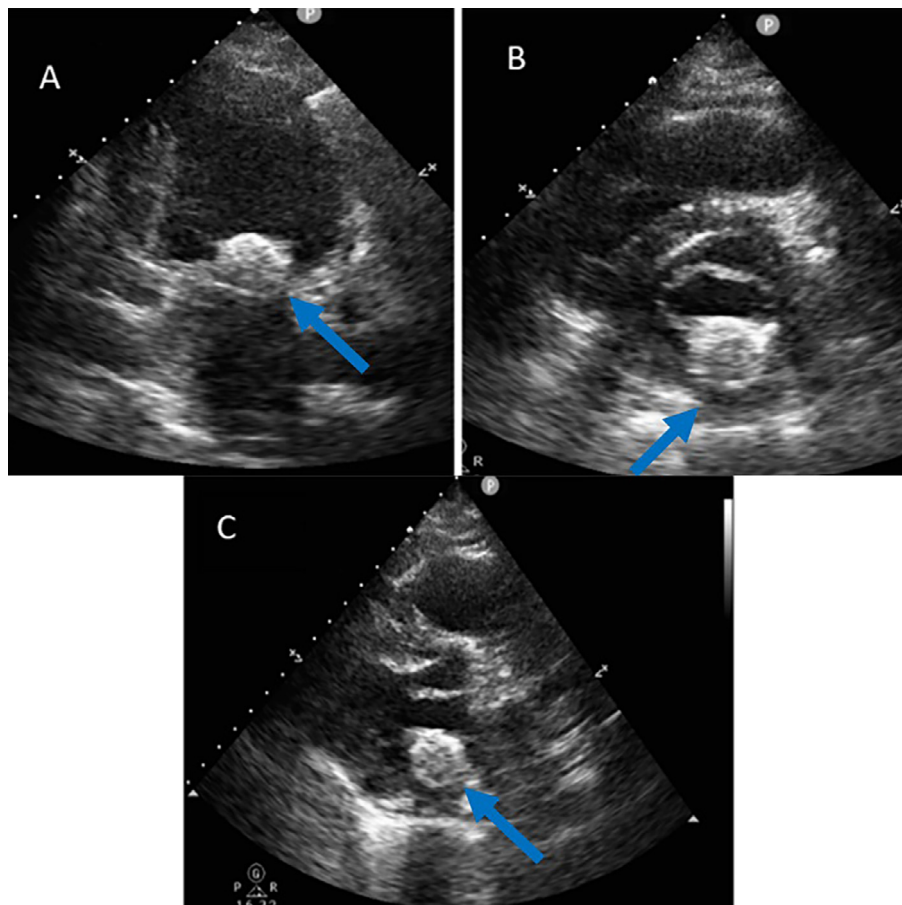


Fig. 1 – Transthoracic echocardiogram. Depicting echo dense spherical, tumor-like mass in the periannular posterior region close to the ventricular side of the posterior mitral leaflet (blue arrows). (A) A 2-chamber view; (B) parasternal short axis view; (C) parasternal long axis view.



Fig. 2 – Coronary angiogram. Left anterior oblique view, showing stable coronary lesions and a 21 × 15 mm calcific mass attached to the mitral annulus (blue arrow).

quences, perfusion and cine with no late-phase gadolinium enhancement acquired 15 to 20 minutes later (Figs. 3–5). Based on the above findings, a presumptive diagnosis of an atypical papillary fibroelastoma was made and open-heart surgery

was indicated. It revealed a small yellow tumor of about 2 × 1.5 cm on the posterior mitral annulus, which was excised (Fig. 6). The diagnosis of CCMA was confirmed by histological examination, showing calcifications and necrotic material, as well as inflammatory cells such as eosinophils, histiocytes, and lymphocytes. The patient had an uneventful postoperative course and was discharged shortly after.

Discussion

MAC is a chronic and degenerative process of the mitral valve fibrous ring, involving the posterior annulus [1]. Commonly seen in the elderly, especially women [4]. CCMA is a less known entity, rarely described. It is a soft peri annular extensive calcification, resembling a tumor, composed of an admixture of fatty acids, calcium, and cholesterol with a toothpaste-like texture [1]. The precise mechanisms involved are still a mystery. CCMA is fairly rare; its exact prevalence is unknown, in some studies as low as 0.06%-0.07% [5]. However, it is largely underestimated due to the asymptomatic course of the condition. Some patients may present with palpitations, dyspnea, and syncope (a consequence of atrioventricular blocks) [4].

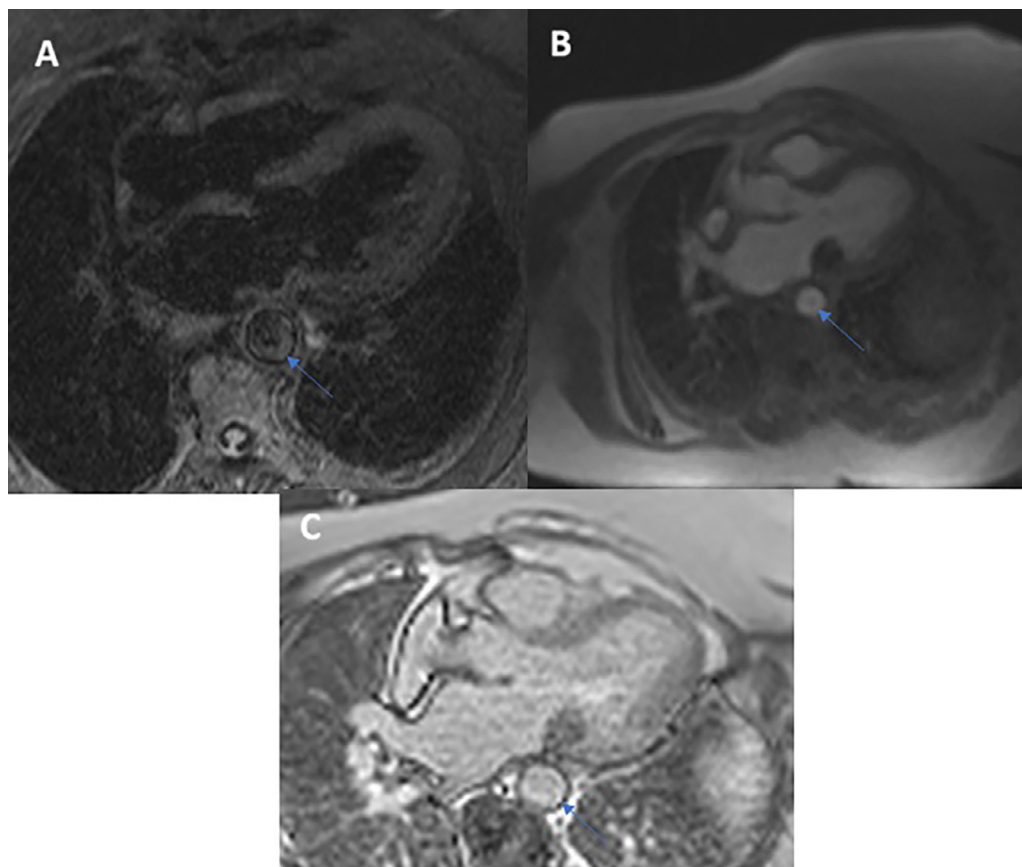


Fig. 3 – Cardiac magnetic resonance. Long axial sequence showing a hypointense mass (blue arrows) in the posterior region of the mitral annulus in both T2 (A) and cine (B) sequences with no late-phase gadolinium enhancement 20 minutes later (C).

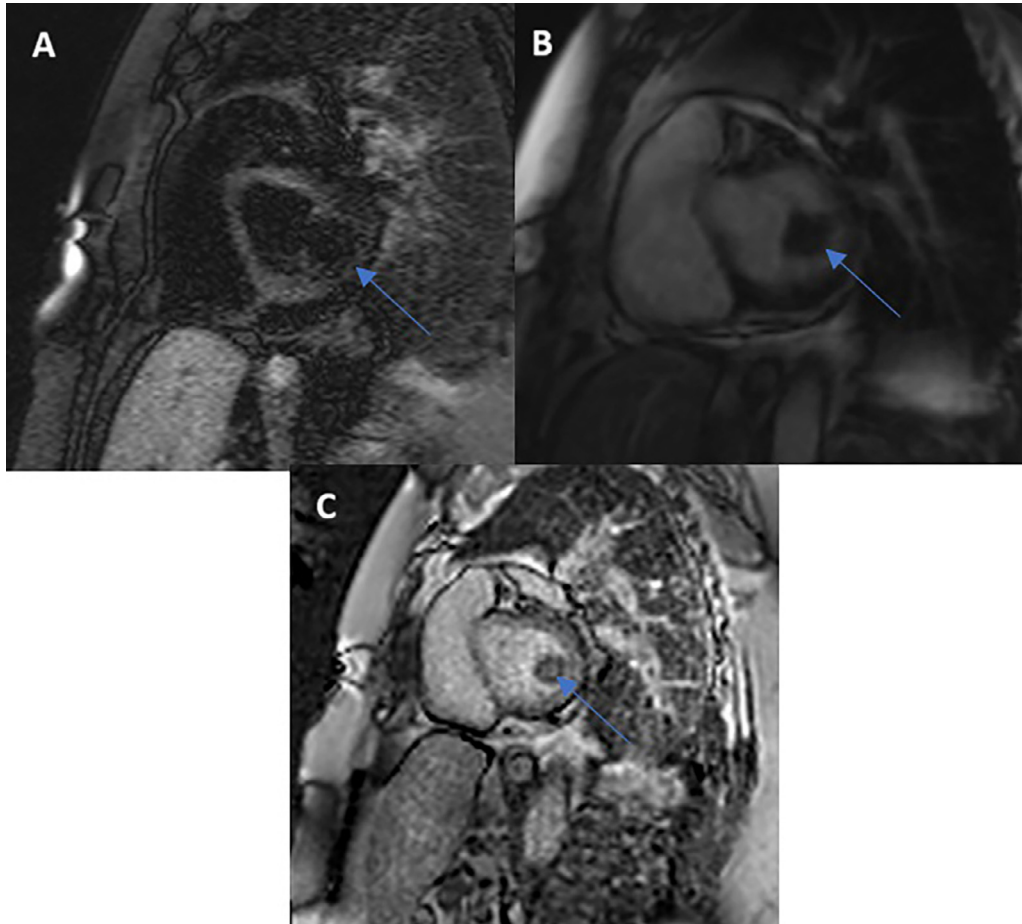


Fig. 4 – Cardiac magnetic resonance. Short axial sequence showing a hypointense mass (blue arrows) in the posterior region of the mitral annulus in both T2 (A) and cine (B) sequences with no late-phase gadolinium enhancement 20 minutes later (C).

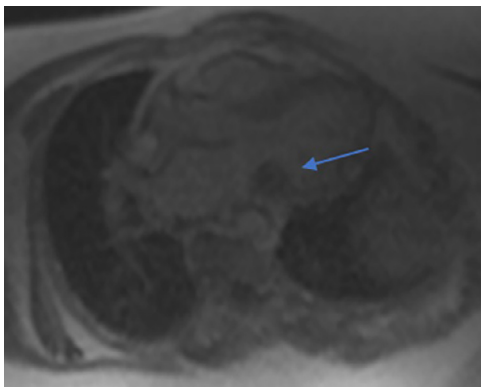


Fig. 5 – Cardiac magnetic resonance. Perfusion sequences depicting a hypointense mass in the posterior region of the mitral annulus (blue arrow).

The main differential diagnosis includes cardiac tumors, vegetation, thrombus or myxoma [4]. Cardiac tumors may appear like CCMA on TTE, emphasizing the role of multimodality imaging in confirming the diagnosis. CCMA has a typical

echocardiographic appearance characterized as large, round, attached, tumor-like calcifications, with well-defined edges, echogenic periphery, and a central anechoic area resembling liquefaction, and usually no posterior acoustic shadow artifacts behind it [6]. However, due to sometimes atypical characteristics, like our case where the mass was highly mobile, a tumor may be suspected instead.

Patients may be referred for transesophageal echocardiography for better evaluation of this mass, as it provides better visualization for posteriorly located structures. CCMA appears as a mass with a central anechoic area resembling liquefaction, with no flow in the central zone by color Doppler, surrounded by an echogenic structure [5].

Correlating the echocardiographic findings with imaging on computed tomography (CT) and CMR usually leads to a proper diagnosis without invasive tests. On CT scan, it appears as a well-defined oval or crescent-shaped hyperdense mass with peripheral calcifications, usually along the posterior mitral annulus, with a high number of Hounsfield units, and lacks contrast enhancement [7].

CMR is considered as the technique of choice in doubtful cardiac tumors cases. CCMA is seen as a hypointense lesion both in T1- and T2-weighted sequences [8]. Contrast en-

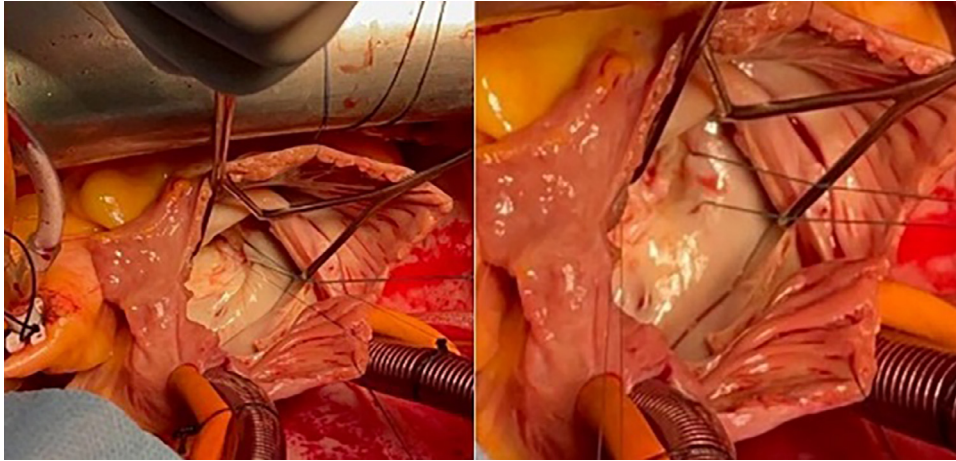


Fig. 6 – Intraoperative pictures. Depicting a caseous calcification of the mitral annulus cavity.

hancement on first pass sequences is not expected; However, it may show peripheral enhancement during delayed postcontrast sequences [4]. In our case, there was no delayed contrast enhancement, and the CMR did not show any calcification, therefore the possibility of a tumor (primarily a papillary fibroelastoma) couldn't be ruled out.

CCMA usually carries a benign prognosis. Most of the patients are asymptomatic requiring a careful follow-up [4]. Surgical indications include complications such as systemic embolization, severe valve dysfunction, and when a tumor cannot be ruled out, as in our case.

When surgery is performed, a calcific shell is usually found surrounding a central area filled with a toothpaste like material [9]. And histologic examination shows amorphous, acellular, basophilic material with chronic inflammatory reaction with macrophages.

Conclusion

CCMA is a rare and benign underdiagnosed variant of MAC. Differentiating it from other valvular masses can prove a real diagnostic challenge as highlighted in our observation. Although echocardiography is the mainstay imaging modality for diagnosis of CCMA, multimodality imaging techniques including cardiac CT and CMR can be used in doubtful situations to confirm the diagnosis and avoid unnecessary surgery.

Patient consent

Informed, written consent was obtained from the patient in accordance with COPE guidelines.

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