Case Report

Left Atrial Pseudotumor Caused By Caseous Calcification of the Mitral Annulus, Case Report with Review of Literature

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Caseous calcification of the mitral annulus (CCMA) is a rare form of periannular calcification, occurring in 0.06% of all echocardiographic studies and in 0.63% of all patients with mitral annular calcification. Here, we report a case of CCMA manifesting as a left atrial mass. The patient is a 78-year-old African American female with multiple co-morbidities, who presented with symptomatic coronary artery disease and chest pain. A pre-operative echocardiogram revealed a mass in the posterior wall of the left atrium, which did not appear to affect the mitral valve or protrude into the left atrial chamber. Intra-operatively, white toothpaste-like material was removed from the mass lesion. Gross examination revealed multiple fragments of grey-white friable caseous material, measuring 3 x 2.5 x 1 cm in aggregate. Bacterial and fungal cultures from this material were both negative. Microscopic examination showed this material to be amorphous, non-viable, basophilic, and acellular, with foci of calcification. These morphological features were diagnostic for CCMA. Review of the literature indicated that CCMA is a very rare benign lesion, presenting as a round, tumor-like mass with central echolucencies on echocardiography. We believe our case report will further raise the awareness of CCMA so that pathologists and radiologists can get familiar with this rare disease to avoid potential misdiagnosis.

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Key Words: *mitral valve annulus, caseous calcification, atrial pseudotumor, TTE (transthoracic echocardiogram), TEE (transesophageal echocardiogram)*

INTRODUCTION

Mitral annulus calcification (MAC) is defined as fibrosis and degenerative calcification of the mitral valve.[1] It is a common disorder seen more frequently in the older population, especially in osteoporotic women.[2] Patients on chronic dialysis as a result of end stage renal failure are particularly more affected.[3] Also involved are younger patients with abnormalities in calcium metabolism as well as those with severe mitral valve dysfunction.[4] In general, it is not an uncommon finding during routine evaluation of patients by echocardiographic studies.[5]

A study titled MESA (Multi-Ethnic Study of Atherosclerosis) concluded that MAC is independently associated with worsening severity of coronary artery atherosclerosis, which can be measured by the coronary artery calcium score (CAC). The study summarized that the occurrence of MAC is related to the burden of atherosclerosis rather than merely degenerative changes of the mitral valve.[1,6]

Calcified disease involving the mitral annulus can present as a spectrum depending on its stage of evolution. This spectrum, based on the point of presentation along this evolutionary timeline, includes mitral annulus calcification, calcified homogenous mass involving the mitral valve and liquefaction necrosis of the mass. Because liquefaction necrosis usually associates with reduction in mass dimension, the size of "mass" can be either stable or smaller during follow-up.[2] CCMA is therefore considered a dynamic entity, rather than a static one.[4]

Caseous calcification (in other words, liquefaction necrosis) is a rare variant, accounting for about 0.63% of all MAC cases[7] and typically involves the posterior annulus.[8] CCMA is composed of a mixture of cholesterol, fatty acids and calcium, imparting the mass a "toothpaste-like" consistency.

Grossly, the mass presents as soft periannular calcification and is often detected by transthoracic (TTE) or transesophageal (TEE) echocardiography, which shows a crescent or round shaped, echo-dense mass, often surrounding an echo-lucent central area of liquefaction.[9]

CASE REPORT

The patient is a 74-year-old African American female who was admitted for coronary artery bypass grafting. She has an extensive history of coronary artery disease, mitral valve

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replacement, morbid obesity, hyperlipidemia, hypertension, diabetes mellitus and non-oliguric renal failure. She did not have elevated blood calcium and phosphorus levels.

Preoperative transesophageal echocardiogram showed an ill defined mass in the wall of the left atrium and heavy calcification of the posterior valve leaflet with restricted motility. A planned coronary artery bypass grafting with exploration of left atrial mass was planned. Intraoperative, no mass was seen; however, on palpation, a mass was felt in the posterior wall of the left atrium. An incision made in this area resulted in the extrusion of white toothpaste like material. The planned coronary artery bypass was performed.

The postoperative course was complicated by encephalitis, cerebrovascular accident and aspiration pneumonia. Her clinical state consistently declined leading to her demise four months after the cardiac surgery was performed.

Grossly, we received multiple fragments of grey-white friable necrotic material, measuring 3 x 2.5 x 1 cm in aggregate. Bacterial, acid-fast bacilli and fungal culture from this material were all negative. Histopathologic examination of the mass revealed amorphous nonviable debris with foci of calcification (**Figure 1A** and **1B**). These morphological features were diagnostic of CCMA.

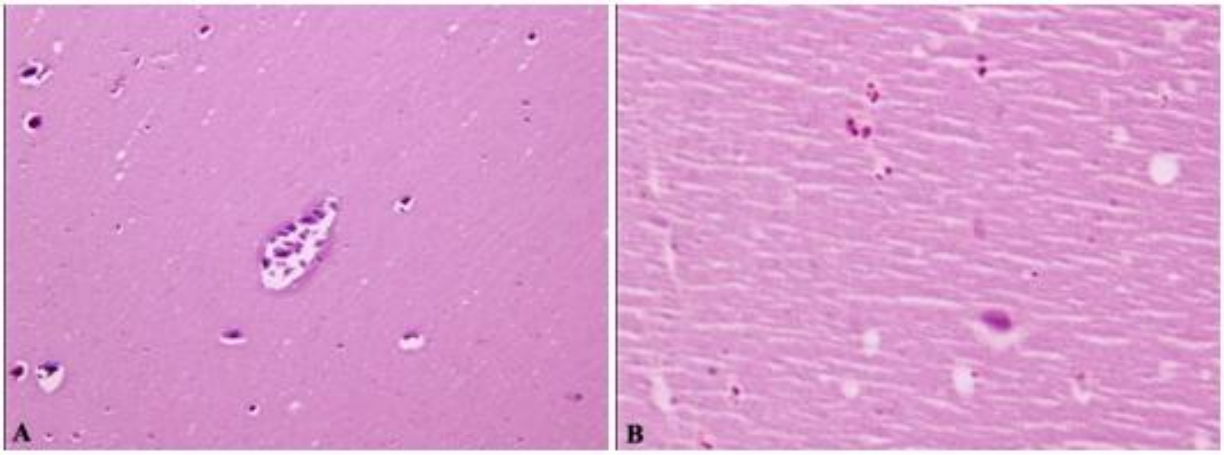

Figure 1. Morphological features if CCMA. **A.** Basophilic caseous material with interspersed areas of calcifications (hematoxylin-eosin stain: original magnification x100). **B.** Homogenous necrotic material with inflammatory infiltrate (hematoxylin-eosin stain: original magnification x400).

DISCUSSION

CCMA is a rare presentation of MAC and its pathogenetic mechanism is not known.[3] The prevalence of CCMA among all cases of MAC was 2.7%, as described in an autopsy series.[10] CCMA, an unusual variant of MAC, commonly presents as a large intracardiac mass with areas of echolucencies that resemble a tumor, often resulting in an exploratory cardiotomy.[11] Despite the benign, asymptomatic nature of this lesion, the clinical differential diagnoses should include calcified soft tissue sarcomas, calcified echinococcus cysts, calcified cardiac amorphous tumors and cardiac osteochondromas.[2] Other common mimickers include abscess or a thrombus. Misdiagnosis often results in unwarranted investigations and interventions. A useful tool in diagnosing this condition is a transesophageal echocardiogram (TEE). The findings on imaging are considered so distinctive that a pathological diagnosis is not habitually required.[7,12]

In the presence of typical radiological findings and histology coupled with negative bacteria, fungi and acid-fast cultures, the diagnosis is usually straightforward.[7] Histologically, CCMA has a core of eosinophilic amorphous acellular fluid, surrounded by a rim of lymphocytes and macrophages, with multiple areas of calcification and necrosis.[2,7,13,14]

When periannular abscess is coexistent with MAC, histological sections show amorphous basophilic calcific debris admixed with bacteria, which can be demonstrated with gram stain.[15] Cardiac tuberculoma can occur in all chambers of the heart, with the right atrium most commonly affected. The nodular form is characterized by caseous necrosis centrally. While Ziehl Neelson stain is most often negative, a definite diagnosis is made based on histological findings and positive cultures.[16] An intracardiac fungal mass is unusual and the degree of suspicion is often low.[17] Examples include cardiac zygomycosis[17] and cardiac aspergillosis,[18] which generally demonstrate the presence of characteristic hyphae in a necrotic background and a positive fungal culture.

CCMA usually presents with mitral regurgitation, mitral stenosis, and heart block. It can also interfere with the repair or replacement of the mitral valve. CCMA has been reported to embolize systemically as well.[19] Davidson, et al[10] also described the embolic potential of CCMA in a case of spontaneous cerebral embolization.

The absence of larger amounts of calcifications is one of the features used to distinguish mitral annular abscesses from

CCMA.[9] Furthermore, an abscess would be located at the mitral aortic fibrosa, usually along systolic flow in the cavity as seen with color Doppler.[2] Intracardiac tumors lack contrast enhancement and soft tissue densities, which are commonly seen in CCMA.[9] In addition, the different clinical presentation, typical location of the calcification, occasional extension to involve the mitral annulus, base of both mitral valve leaflets and/or papillary muscle and chordae tendinae are typically seen with CCMA.[3]

The prognosis of most CCMA is benign and management depends on the clinical situation.[7] CCMA can undergo spontaneous resolution.[3] Chahal, et al,[1,20] described a case in which echocardiographic examination (TEE and TTE) repeated after an year showed transformation of MAC to CCMA and nine months later, a repeat TTE and TEE showed a marked reduction of the mass with some residual MAC calcifications. In majority of asymptomatic cases that do not present with cardiac compromise or hemodynamic instability, close vigilance and medical follow up is recommended.[13]

Surgery is warranted in cases that present with co-existent mitral valve dysfunction from altered mitral annulus dynamics, destructive ulceration resulting in thrombus formation or endocarditis or uncertain diagnosis.[3,13] Prompt surgical intervention is also considered in cases presenting with embolic phenomenon.[10,13] The surgical procedure is however not free of complications which include the risk of atrio-ventricular perforation, vascular damage and embolization of necrotic calcified debris. In circumstances requiring mitral valve replacement, techniques such as "cavitron" ultrasonic surgical aspirator (CUSA) can be employed to enucleate the core, followed by reconstruction.[13] CUSA technique employs ultrasonic waves to disintegrate calcium and the resulting debris can be extracted without causing damage to adjacent structures.[21,22,23]

In conclusion, CCMA are entities that are rare and under-recognized variants of MAC, which need to be distinguished from common differentials such as intra-cardiac tumors, myocardial abscesses, thrombus[9] or cyst.[20]

CONFLICT OF INTEREST

None.

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