Mitral Annular Mass

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Case Presentation
A 74-year-old man presented with a multifocal acute cerebrovascular accident. Laboratory data revealed normal renal function and serum calcium level. Chest radiograph demonstrated an enlarged cardiac silhouette, but was otherwise unremarkable for age. A cardiac source for the emboli was suspected, and an echocardiogram was requested. Echocardiogram showed an intracardiac mass (Figure 1A), which was further investigated with computed tomography (CT), (Figure 1B, 1C) and MRI (Figure 1D).

FIGURE 1. Two-dimensional echocardiogram apical four-chamber view (A) shows a lobulated echogenic 3 x 2-cm mass in the region of the posterior mitral valve leaflet (arrow) that was mobile. Unenhanced CT four-chamber view image (B) shows a rim-calcified mass (arrow) corresponding to the echocardiogram abnormality with adjacent sheet-like calcifications (arrow head) that are likely in the mitral valve leaflet. EKG-gated enhanced CT maximum intensity projection (MIP) image in short axis projection though the mitral valve (C) reveals a centrally hypodense, rim-calcified mass (white arrow) in the atrioventricular groove with close relationship to the posterior mitral valve leaflet. Calcifications of the mass are contiguous with the densely calcified inferior mitral annulus (arrowhead). The anterior mitral valve leaflet (black arrow) exhibits degenerative calcifications. EKG-gated cardiac MRI fast imaging employing steady-state acquisition (FIESTA) short axis image (D) through the same level as image C shows that the central content of the mass (arrows) has high-signal intensity surrounded by low-signal rim.
Key imaging finding(s)
Lobulated mass contiguous with the mitral annulus and posterior leaflet (CT and MRI).

Differential Diagnoses
Caseous calcification of the mitral annulus
Myxoma
Mitral para-annular blood cyst
Mitral annular abscess
Infective endocarditis

Discussion
The differential diagnosis for a mass closely related to the mitral annulus differs from the nonspecific cardiac mass differential. The most common lesions associated with the mitral annulus include caseous calcification of the mitral annulus, myxoma, para-annular cyst, annular abscess, and infective endocarditis.

Caseous Calcification of the Mitral Annulus
Caseous calcification of the mitral annulus (CCMA) accounts for < 1% of mitral annular calcification (MAC) cases. It most commonly occurs in elderly women; however, it may also affect younger patients with advanced renal disease or other metabolic disorders that result in abnormal calcium metabolism. A calcified rim surrounds caseous or toothpaste-like material composed of calcium, fatty acids, and cholesterol, typically located on the posterior mitral annulus. On transthoracic echocardiogram (TTE), CCMA usually appears as a large,round,echogenic mass with smooth borders and central echolucency.1 On cardiac MRI, the mass most commonly has low-signal intensity on T1- and T2-weighted sequences. On CT, it typically appears as a round, smooth mass with central hyperdensity and peripheral dense calcifications. Due to the variable appearance of caseous calcifications as a result of liquefaction necrosis on imaging, the central component of the mass may less frequently appear hypodense on CT and hyperintense on MR sequences, as is seen in the illustrative case. Nevertheless, the densely calcified rim consistently appears hyperdense on CT and hypointense on MR. The lack of soft tissue attenuation and enhancement is a clue to differentiate CCMA from a tumor. Because misdiagnoses and unnecessary surgical treatment may occur, a combination of multiple imaging modalities is often necessary to establish or suggest the diagnosis.3 Microscopic examination shows an amorphous, acellular, basophilic, and calcified structure, with chronic inflammatory macrophages.1,2
CCMA is usually a benign and asymptomatic lesion with no need for therapeutic intervention. Rarely, if mitral dysfunction or conduction disturbance occurs, mass excision and valve repair or replacement is the preferred treatment.2 Cases of systemic embolization by CCMA have been reported, which likely explains the symptoms in our patient, and prompt intervention.3

Myxoma
Cardiac myxoma represents the most common primary cardiac tumor in adults. It is typically a polypoid, intracavitary left atrial mass that arises from the interatrial septum, but it may originate in any cardiac chamber.4,5 Most affected patients present with at least one feature of a classically described triad that includes cardiac obstructive symptoms, constitutional symptoms, and embolic events.
Echocardiogram features include an echogenic round, lobular, mobile mass, which seldom originates from the valves. Common nonspecific radiographic features include cardiomegaly and intracardiac calcification. Myxomas may cause elevated left atrial pressures with associated left atrial enlargement, vascular redistribution, a prominent pulmonary trunk, pulmonary edema, pleural effusions, and rarely an enlarged left atrial appendage.4
On CT, myxomas are typically spherical or ovoid with a lobular or smooth contour and an attenuation less than or equal to that of the myocardium on postcontrast studies. They may be heterogeneous or homogeneous, sometimes containing coarse or punctate calcification. If located in the right atrium, the tumor may extend into the superior or inferior vena cavae and may embolize, leading to pulmonary infarcts. If situated in the left heart, the tumor may embolize to the systemic circulation, resulting in end organ infarction.6 On MRI, myxomas demonstrate variable heterogeneous signal intensity with most lesions being isointense and the remaining lesions being hyper- or hypointense compared to myocardium on T1-weighted images. There may be heterogeneous or homogeneous enhancement.4

Mitral Para-annular Blood Cyst
Congenital blood cysts of the heart valves most often involving the tricuspid and mitral valves of fetuses and infants are generally considered of no clinical significance. However, lesions may persist and enlarge to form giant cysts of the heart valves. Even when large, blood cysts are usually asymptomatic in adults and are often discovered incidentally during routine echocardiographic evaluation. In rare cases, mitral regurgitation can occur due to the para-annular location of the cyst. Cysts may be a potential source of cerebrovascular embolism. There is not necessarily a correlation between the size of the cyst and hemodynamic consequences, and there is no consensus regarding the management of blood cysts. Some authors suggest that asymptomatic cysts, because of their benign character, can be monitored with echocardiography, and that resection should be reserved for cysts that interfere with normal cardiac function.6
On echocardiogram, blood cysts appear as hypoechoic cystic lesions, which may contain internal echoes, often with smooth outer surfaces. On CT, they appear as hypodense cystic masses with or without wall calcification. On MRI, blood cysts demonstrate low to intermediate signal intensity on T1-weighted images and high signal intensity on T2-weighted images without enhancement.
**Mitral Annular Abscess**

Mitral annular abscesses are most often seen in patients who have had endocarditis or bacteremia. The patients typically present with fever, pain, fatigue and shortness of breath. The lesions usually appear along the mitral-aortic fibrosa. They may closely resemble CCMA on imaging, but lack calcifications, typically have a homogeneous core, and may show systolic blood flow by color Doppler. On echocardiogram, myocardial abscesses appear as hypoechoic masses. Hypokinesis of the surrounding myocardium can be present, along with pericardial effusion. CT or MRI can be used to accurately define the anatomical location, as needed for surgical intervention. Abscesses appear on CT scan as loculated lesions with fluid density and peripheral contrast enhancement with surrounding myocardial wall thickening. MRI is more sensitive for abscess detection than other imaging modalities due to its superior temporal and spatial resolution; in addition, its use is advantageous for morphological evaluation, since it provides surgeons with more accurate anatomical information.8

**Infective Endocarditis**

Infective endocarditis is a rare but potentially life-threatening disease associated with intravenous drug use, prosthetic valve replacement, congenital heart disease, human immunodeficiency virus, and poor dentition. When involving the mitral valve, infectious vegetations typically form on the left atrial side of the mitral valve.9 Transeophageal echocardiogram is the most reliable means of identifying lesions as mobile echogenic masses implanted in a valve, mural endocardium in the trajectory of a regurgitant jet, or in prosthetic material with no alternative anatomical explanation.10 On CT, mitral vegetations > 10 mm in diameter are almost always detected. They appear as a hypoattenuating, mobile mass adhering to the valve leaflet.9 On MRI, large vegetations typically appear as low signal masses attached to a valve leaflet and are almost always associated with mitral regurgitation. MRI is also useful to detect embolization of large vegetations.10

**Additional diagnostic considerations**

While metastases represent the most common cardiac neoplasms, they were not included in this differential based on the imaging appearance of the mass in the illustrative case, as well as the clear association with the mitral annulus.

**Diagnosis**

Caseous calcification of the mitral annulus (CCMA)

**Summary**

The differential diagnosis for a mitral annular mass includes a variety of entities, to include CCMA, myxoma, peri-annular blood cyst, abscess, and infective endocarditis. The imaging appearance of the cardiac lesion presented in this case favors CCMA based on the lesion morphology, location, and presence of rim calcifications. CCMA may be distinguished from myxoma based on the fact that myxoma is usually mobile, pedunculated, and attached to the myocardial wall. Unlike CCMA, myxoma lacks peripheral calcifications, rarely involves the valve, and is highly vascular. Blood cysts appear as hypoechoic cystic lesions on ultrasound with smooth walls and hypodense cystic masses on CT with or without wall calcification; when wall calcifications are present, the lesions can mimic CCMA. Mitral annular abscess would not be expected to show extensive peripheral calcifications on CT. Lastly, in cases of infective endocarditis, vegetations typically affect the valve leaflet, rather than mitral annulus.

**References**