Mitral annular calcification: Can CMR be useful in identifying caseous necrosis?

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Abstract: Mitral annular calcification (MAC) can resemble an intracardiac mass and it is defined as a chronic degeneration of the mitral annulus. Often reported is caseous mitral annulus calcification (CMAC), a periannular, extensive calcification resembling a tumor. We report the case of a 68-year-old woman who had been hospitalized for palpitations and dyspnea. The transthoracic and transesophageal echocardiography revealed a non-homogeneous, slightly mobile, round mass, attached to the ventricular side of posterior mitral leaflet, with central echo-lucent area and without acoustic shadowing. Therefore, a cardiac magnetic resonance (CMR) was performed; delayed enhancement sequences showed a non-enhanced central core surrounded by a hyperenhanced rim (fibrous cap). To confirm the diagnosis, a multidetector computed tomography (MDCT) was performed; the MDCT showed a hyperdense mass with a hypodense center and a calcified peripheral rim. The central content had heterogeneous fluid density without significant contrast enhancement. The MDCT findings were considered highly suggestive of CMAC. CMR may be useful for the identification and definition of pericardial and myocardial masses and CMAC.

Keywords: mitral annular calcification, caseous calcification, cardiac magnetic resonance, computed tomography

Introduction

Intracardiac masses are uncommon findings in the general population, with considerable problems of diagnosis. Mitral annular calcification (MAC) can resemble an intracardiac mass and it is defined as chronic degeneration of the mitral annulus, which mainly involves the posterior annulus. MAC is common in the elderly and may occur in patients with renal disease or abnormal calcium metabolism. Previous observational studies suggest that MAC might be associated with several other cardiovascular disorders, such as atherosclerosis, mitral regurgitation, stroke, atrioventricular conduction defects, and hypertrophic cardiomyopathy [1–4]. MAC may be accelerated by conditions that increase left ventricular pressure and by conditions that accelerate atherosclerosis [4, 5].

More frequently reported is caseous mitral annulus calcification (CMAC), a periannular, extensive calcification

resembling a tumor, which is composed of calcium, fatty acids, and cholesterol with a toothpaste-like texture. Differential diagnosis of round echogenic structures adjacent to the left atrioventricular groove should also include infected mitral calcification, lipomatosis of the atrioventricular groove, and enlarged lymph nodes [6].

CMAC is usually considered as a benign condition; however, it may cause mitral stenosis, mitral regurgitation, left ventricular outflow obstruction, or systemic embolization.

Case Report

A 68-year-old woman with arterial hypertension, hypertrophic cardiomyopathy, moderate mitral regurgitation, dyslipidemia, and carotid stenosis was hospitalized for palpitations and dyspnea. A couple of years ago, she had a lacunar infarct of the right lenticular nucleus.

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The transthoracic and transesophageal echocardiography revealed a non-homogeneous, slightly mobile, round mass, attached to the ventricular side of posterior mitral leaflet, with central echo-lucent area, and without acoustic shadowing (*Fig. 1a and 1b*).

A cardiac magnetic resonance (CMR) was therefore performed using 1.5T magnet (Philips Achieva, Eindhoven, The Netherlands) with a cardiac phased-array multi-coil to assess the cardiac mass. In T1-weighted (T1-W) sequences, the mass was isointense (*Fig. 1c*), and in

Fig. 1. (a) Transthoracic echocardiography in four-chamber view. (b) Transesophageal echocardiography in four-chamber view. The transthoracic and transesophageal echocardiography show an inhomogeneous round mass attached to ventricular side of posterior mitral leaflet, with central echo-lucent area and without acoustic shadowing. (c) T1-weighted sequences showing an isointense mass. (d) T2-weighted STIR short axis sequences with lack of signal. (e) Balanced steady-state free precession (bSSFP) in two-chamber sequences: the mass appears isointense with the normal myocardium. (f) bSSFP in four-chamber sequences: the mass appears isointense with the normal myocardium. (g) Delayed enhancement sequences showing a non-enhanced central core surrounded by a hyper-enhanced rim (fibrous cap). (h-j) Multidetector computed tomography: multiplanar reconstructions. The mass is hyperdense with hypodense center and a calcified peripheral rim

T2-weighted (T2-W) short tau inversion recovery (STIR) sequences, it lacks signal (Fig. 1d). However, the dark T2-W tissue signal was unusual for a cardiac mass [7]. In balanced steady-state free precession (bSSFP) images, the mass appeared isointense with the normal myocardium (Fig. 1e and 1f). Delayed enhancement sequences showed a non-enhanced central core surrounded by a hyperenhanced rim (fibrous cap) (Fig. 1g).

To confirm the diagnosis, a multidetector computed tomography (MDCT) was performed; the MDCT showed a hyperdense mass with a hypodense center and a calcified peripheral rim (Fig. 1b–1j). The central content had heterogeneous fluid density without significant contrast enhancement. The MDCT findings were considered highly suggestive of CMAC.

Discussion

The CMR features of CMAC are not yet well defined; in previous works, Monti et al. [8] reported two cases of CMAC and described *low signal* in both T1-W and T2-W sequences, both before and after contrast, associated with a slightly-darker-than-myocardium signal in SSFP sequences as the hallmarks. On the other hand, Di Bella et al. [9] found a hyperintense center and a hypointense rim in T1-FSE sequences, low signal in T2-STIR sequences, and strong peripheral enhancement 10 min after the contrast, administered using the contrast-enhanced inversion-recovery technique. However, Monti et al. [8] concluded that further cases need to be studied to prove the feasibility to diagnose CMAC by CMR without the need for further CT examination. According to Shriki et al., T1-W imaging can be a useful tool to differentiate MAC from CMAC. Precontrast T1-W sequences are helpful in demonstrating the distinction between MAC that shows low signal on T1 and CMAC that shows high signal on T1 [1, 10].

In our case, the mass was *isointense* in T1-W sequences with lack of signal in T2-W-STIR sequences. In bSSFP images, the mass appeared isointense with the normal myocardium. Delayed enhancement sequences showed a non-enhanced central core surrounded by a hyperenhanced rim.

Because of the observed discordance with previously reported cases, especially for the mass features in T1-W sequences, the patient underwent an MDCT that showed a hyperdense mass with hypodense center and a calcified peripheral rim.

On the basis of the above findings, a presumptive diagnosis of a centrally liquefied mass containing a high protein or hemorrhagic content was carried out.

The patient had also undergone surgery because of her history of stroke, presumably due to cardiac mass embolism. Mass histology showed a central liquefied

structure in free left ventricle wall attached to a thickened posterior mitral valve leaflet. The wall was fibrotic and thickened, containing areas of either calcification or inflammation, with a diagnosis of liquefaction necrosis of MAC.

Conclusions

CMR may be useful for the identification and definition of pericardial and myocardial masses. In case of CMAC, the interpretation of CMR images could be still challenging; however, the T1-W imaging, as our case demonstrated, can be a helpful tool to differentiate MAC from CMAC. Further studies are required for the best definition of CMAC features at CMR.

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