Diagnostic Imaging

Massive mitral annular calcification mimicking intracardiac mass: Multimodality approach to diagnosis

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ABSTRACT

The correct differential diagnosis of cardiac masses can be challenging and often carries important clinical implications. We present the case of a 78-year-old man with a cardiac mass of unclear etiology diagnosed on echocardiography. Using a multimodality approach with cardiac magnetic resonance and computed tomography, it was possible to define the real nature of the mass as composed of 2 voluminous calcifications of the mitral annulus.

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Case report

We present the case of a 78-year-old man who was referred to our cardiologic outpatient clinic. The man’s medical history included hypertension, dyslipidemia, and chronic obstructive pulmonary disease. In December 2015 a transthoracic echocardiography identified the presence of a voluminous oval hyperechoic mass, located at the level of the posterior cusp of the mitral valve. The characteristics of the mass suggested a calcified nature, but other etiologies (as an atypical form of myxoma) could not be excluded. A chest radiograph provided no evidence of calcification. A transesophageal echocardiography was therefore recommended but not performed. One year later, a second transthoracic echocardiography confirmed the presence of an inhomogeneous and irregular mass, located at the level of the posterolateral portion of the mitral annulus, which was essentially unchanged. The motion of the valve leaflets was not significantly affected, and mild mitral regurgitation, along with atrial dilatation, was pointed out (Fig. 1).

A cardiac magnetic resonance (CMR) was therefore requested to complete the differential diagnosis and to characterize the mass. At the level of the lateral segment of the mitral annulus, 2 contiguous masses (maximum diameter of 17 and 12 mm, respectively) were observed, which appeared hypointense on T1-weighted, T2-weighted, and short tau inversion recovery sequences. No contrast uptake or late enhancement was observed after the administration of gadolinium. These characteristics were highly suggestive of degenerative mitral annular calcification (MAC). To confirm the presence of the calcifications, a multimodality approach was implemented and a computed tomography (CT) was performed, clearly demonstrating the presence of 2 contiguous calcified masses (Fig. 2).
Discussion

The majority of cardiac masses are detected incidentally during routine echocardiographic studies for other indications. In many cases, echocardiography reveals characteristic anatomic and functional features that already enable differential diagnosis of a cardiac mass.

When diagnosis is doubtful, a multimodality imaging approach is generally necessary for precise tissue characterization and risk stratification and for choosing the best therapeutic option. Cardiac CT is a commonly used second-line diagnostic modality to assess cardiac masses [1,2]. In addition, incidental findings of cardiac masses are becoming more common as CT is increasingly used to evaluate coronary artery disease. Several technological advances in CT, including submillimeter detector arrays, increased rows of detectors, half-scan postprocessing algorithms, and electrocardiographic gating, have resulted in improved imaging of cardiac structures, including cardiac masses. CT can also offer information regarding vascularity by means of contrast enhancement, presence of calcifications (unlike magnetic resonance imaging [MRI]), and presence of fat. Limitations of CT include exposure to ionizing radiation, lower temporal resolution compared with echocardiography or MRI, and lower soft-tissue contrast resolution compared with MRI. MRI characteristics can be used to predict the likely malignancy of a cardiac mass. Compared with CT, MRI offers higher temporal resolution and better tissue characterization, and MRI does not expose patients to ionizing radiation. However, although access to cardiac MRI is increasing, the procedure remains less available than echocardiography or CT.

Cardiac masses include a variety of conditions for which the treatment can be completely different, ranging from normal anatomic variants (embryonic remnants like the Chari network or the Eustachian valve, false tendons, the moderator band, etc), pseudotumors (thrombus, pericardial cysts, and caseous calcifications of mitral valve), benign lesions (myxoma, lipoma, papillary fibroelastoma, fibroma, and hemangioma), and malignant tumors (cardiac metastases and sarcomas).

MAC is characterized by a chronic degenerative process of the fibrous support structure of the mitral valve, which mainly occurs at the level of the posterior subvalvular angle. The re-
ported prevalence is between 8% and 15% [3,4]. The incidence of MAC increases with age, female sex, renal failure, hypertension, and hyperparathyroidism, and it seems to share the same risk factors as atherosclerotic disease. Rarely are these calcifications so widespread that they assume a real “tumor-like” appearance [5]. The pathophysiology is unclear but could be related to changes in calcium-phosphorous metabolism. Microscopy shows nonspecific inflammatory changes and calcium deposits [6].

MAC is generally an asymptomatic condition, but in severe cases, it can have clinical implications and it has been found to increase the incidence of mitral valve disease and arrhythmias [4,7]. It has also been associated with an increased rate of mortality and cardiovascular disease [8].

On echocardiography, MAC is usually visualized as an echodense (hyperechoic) irregular structure involving the mitral annulus with associated acoustic shadowing [4]. On CMR, MAC displays a loss of signal on both T1- and T2-weighted images, and it shows no contrast uptake during first pass or late gadolinium enhancement (LGE) sequences (Fig. 3) [1]. CT can offer information regarding the presence of calcifications, as in our case report, using the multimodality approach, CT permits the confirmation of the calcified nature of the lesions, an element that, added to the MRI signal characteristics and the complete lack of contrast medium capitation, allowed the diagnosis of MAC (Fig. 4).

The presence of the calcifications can nevertheless be present also in other conditions:

- Caseous calcification of the mitral annulus is a rare form of degenerative MAC (0.06-0.07 of the general population) and typically involves the posterior mitral annulus and the antroventricular groove [9]. The unusual characteristics can lead to misdiagnosis as a cardiac tumor, thrombus, or abscess. MRI can be very useful for tissue characterization: in the early phase, the associated mass is hyperintense at both T1- and T2-weighted imaging (high fluid content and liquefactive necrosis). Associated calcifications within and

**Fig. 3** – T1-weighted scans along the short axis with contrast medium: hypoenhancement in the first pass (A) and in the late phase (B) after contrast medium (masses depicted by arrows).

**Fig. 4** – Computed tomography scans: paracoronal plane (A) and parasagittal plane (B): the calcified nature of the lesions is clearly shown (arrows).
around the mass appear hypointense at both T1- and T2-weighted imaging but are better characterized with CT. LGE imaging typically shows a peripheral rim of hyperenhancement (fibrous cap around the avascular core of necrotic caseous material) [1].

- Myxomas are the most common type of primary cardiac tumor (25%-50%) and usually occur in the fourth to the seventh decade of life. Atrial myxoma usually has quite distinctive features, which include the presence of a single pedunculated mass arising from the fossa ovalis and protruding into the left atrium; it can often have an irregular shape with nonhomogeneous echogenicity, and sometimes areas of calcifications [10]. These atypical characteristics can sometimes be a potential limitation for definite echocardiographic differential diagnosis and prompt the need for a multimodality approach with CMR or CT. Myxomas appear mostly isointense on T1-weighted images and hyperintense on T2-weighted images. Myxomas may contain cysts, necrosis, fibrosis, hemorrhage, and calcification, which lead to a typically heterogeneous appearance at contrast enhancement. Many myxomas have a layer of surface thrombus, with low signal intensity on LGE images. CMR cine imaging is very useful in the workup of myxomas as they are highly mobile, occasionally prolapsing through the mitral valve and causing obstruction. With steady-state free precession cine techniques, myxomas appear hyperintense relative to the myocardium but hypointense relative to the blood pool.

- Fibromas—cardiac fibromas are the second most common congenital tumor and typically present in pediatric or young adult life; these typical characteristics differentiate the fibroma from our case report. They are usually solitary tumors (unlike rhabdomyomas) and are most often located intramurally in the ventricles involving the interventricular septum. With MRI, fibromas are isointense relative to normal myocardium on T1-weighted images and are characteristically hypointense on T2-weighted images (unlike other masses). They are generally homogeneous unless there is central calcification, which may be seen as patchy central hypointensity. With gadolinium-based contrast agent administration, fibromas generally show no contrast enhancement during perfusion imaging because of their avascularity. However, 7-10 minutes later, they classically show intense hyperenhancement on LGE images.

Although these cardiac conditions are rare, in some cases, it is useful and necessary to employ a multimodality approach with echocardiography, CMR, and CT to accurately differentiate the various masses.

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