

Caseous mitral annulus calcification mimicking a cardiac tumor

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September 11, 2023

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Data availability statement

The data of this case are available from the corresponding author upon reasonable request.

Funding information

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Ethical approval

Written informed consent has been obtained from the patient.

Consent

The authors have confirmed during submission that patient consent has been signed and collected in accordance with the journal's patient consent policy.

Manuscript:

Key clinical message:

This report highlights the contribution of cardiac magnetic resonance and cardiac computed tomography to the diagnosis of caseous mitral annulus calcification which can be misdiagnosed as a cardiac tumor on echocardiography as in the reported case.

Key words: mitral annulus; calcification; tumor; echocardiography; CT; CMR

Introduction:

Caseous mitral annulus calcification (CMAC) is an extremely rare condition with controversial etiopathogeny and few clinical symptoms occurring mainly in old women (1). Its diagnosis on echocardiography may be confusing on echocardiography. Computed tomography (CT) is the key diagnostic imaging technique. We here report the case of a 54-year-old women diagnosed with a mitral tumor on echocardiography. The diagnosis was doubtful on cardiac magnetic resonance (CMR). CT concluded to CMAC.

Case History:

A 54-year-old woman, treated for invasive ductal carcinoma of the left breast by mastectomy, chemotherapy and radiotherapy 8 years ago, presented at the cardiology department for a systematic echocardiography before cataract surgery. Echocardiography showed a soft tissue mass with central echolucencies of the posterior mitral annulus blocking the posterior leaflet of the valve with mitral regurgitation. There were neither acoustic shadowing nor blood flow on color Doppler (figure 1). The diagnosis of a cardiac tumor, particularly a metastasis, was suggested. The patient underwent a cardiac magnetic resonance (CMR) within few days (figure 2). Multi-planar cine images showed a low signal mass of the posterior mitral annulus blocking the posterior leaflet with mitral regurgitation, extending to the myocardium. The mass was in high signal on T1 black blood weighted images and was confused with the blood on STIR weighted images. Late gadolinium enhancement (LGE) sequences showed peripheral intense enhancement. A cardiac CT with prospective gating and no contrast material administration showed that the mass was a huge calcification of the posterior mitral annulus with lower density than the other calcifications of the mitral and the aortic annuli. Review of the radiological data of the patient at the work up of her breast cancer showed tiny calcifications of the mitral annulus (figure 3). Cardiac tumor was eliminated and the diagnosis of CMAC was retained.

Discussion:

Degenerative mitral annulus calcification (MAC) is identified in 10.6% of the population. Among them, only 0.6-0.7% present CMAC on echocardiography. Their prevalence in autopsy series is higher (about 2.7% of MAC cases), suggesting underdiagnoses on imaging (1). It is frequently observed in elderly women with hypertension and dyslipidemia as well as in patients with kidney failure, in whom, it is linked to calcium – phosphate metabolism deregulation (1, 2). However, caseous necrosis of MAC is not clearly explained (3). The patient in our case is young. We believe that the size of the calcification in her can be explained by the radiotherapy she received that accelerated the process of mitral valve degeneration. CMAC is often an asymptomatic condition (as in the case we report) but patients can present with conduction abnormalities or with embolization of caseous material (3). CMAC is a dynamic process, can grow in size and infiltrate the adjacent myocardium (4), as reported in our case. The center of CMAC contains liquefied calcifications, cholesterol and fatty acids. Microscopic examination shows a central amorphous acellular eosinophilic material with macrophages and lymphocytes as well as peripheral fibrocalcic envelope (1, 4, 5). These features explain the radiological findings. In fact, on ultrasonography, CMAC has a central echolucency with a hyperechoic

rim, a calcified envelop, no blood flow and no acoustic shadowing (3, 4). CT confirms calcifications. The center is less hyperdense than the periphery (4). CMR offers a better assessment of the mass that has a low signal on T1W and T2W images. First pass perfusion shows no enhancement whereas late gadolinium enhancement shows a peripheral enhancement (4). Because of its scarcity, and variable appearance on ultrasound examination, CMAC is frequently confused with infective myocarditis vegetation, myocardial abscess, thrombus and cardiac tumor like in our case (1, 3). Cardiac tumors include benign and malignant primitive neoplasms as well as metastases. Primitive cardiac tumors are rare with an incidence varying between 0.002 and 0.3% (2). Benign tumors include myxomas, rhabdomyomas, papillary fibroelastoma, fibromas, hemangiomas, lipomas, and leiomyomas (6). Myxoma is the most frequent benign type in adults that often arises from the interatrial septum in the region of the fossa ovalis and develops in the left atrium. It is generally a mobile hypoechoic mass on ultrasonography, hypodense on CT. Calcifications are observed in 14% of cases. It has a characteristic signal on MRI. In fact, It is hypointense on T1W images, hyperintense on T2W images, but may be heterogeneous, depending on its content. Enhancement is generally patchy (1, 2, 6). Fibroelastoma is rare and is located downstream the valve. It shows low signal on T2W images because of its fibrous content and is isointense on T1W images (6). Hemangioma is a vascular tumor that can be found in any chamber. It has typically a heterogeneous high signal on T1W and T2 W images with intense enhancement (5). Rhabdomyoma and fibroma are common in pediatric population (6). Primitive malignant cardiac tumors are extremely rare and are sarcomas in most cases. Malignancy is suggested by rapid growth, invasion of adjacent structures, necrosis, hemorrhage, feeding vessels and involvement of more than one chamber (6). Angiosarcoma is the most frequent type in adults and typically arises in the right atrium. It has a poor prognosis with a frequent metastatic spread (5, 6, 7). Metastases are much more frequent and are found in 10-12% of patients with a primitive cancer in post-mortem series (5). Primitive cancers spreading to the heart include lung cancers, breast cancers, lymphomas and melanomas (5). As the patient of the case we report had a history of a treated breast cancer, metastasis was the first suggested diagnosis. Metastases show low signal on T1W and high signal on T2W images with heterogeneous enhancement (5).

Conclusion:

In conclusion, cardiologists as well as radiologists should be aware of the imaging characteristics of CMAC despite its rareness in order to distinguish it correctly from cardiac tumors.

Acknowledgements

A special acknowledgment to all contributors.

Conflict of interest

None.

Authors' contribution:

Henda Nèji and Emna Bennour: Writing – original draft

Mariem Affes: Conceptualization

Salma Kchaou and Ines Baccouch: Investigation

Saoussen Hantous-Zannad and Ikram Kammoun: Supervision and validation

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Figures legend:

Figure 1: Echocardiography showing an echoic mass of the posterior annulus with a central echolucency (arrow in A) and no color Doppler flow (arrow in B).

Figure 2: CMR showing a low signal mass of the posterior annulus (yellow arrow) with mitral regurgitation (arrowhead) on cine image (A). The mass has a central low signal with a peripheral high signal rim on STIR W images (red arrow, B) and a high signal on black blood T1W images (blue arrow, C). It shows a peripheral enhancement on late gadolinium enhancement sequences (white arrows, D – E – F).

Figure 3: cardiac CT without contrast material showing a fully calcified mass of the posterior annulus (arrows, A – B). CT in the work up of the breast cancer 8 years ago, showing tiny calcification of the posterior annulus (arrow, C)

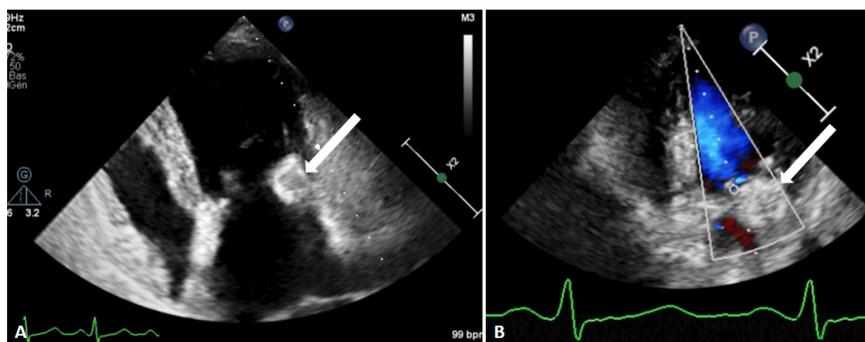


Figure 1

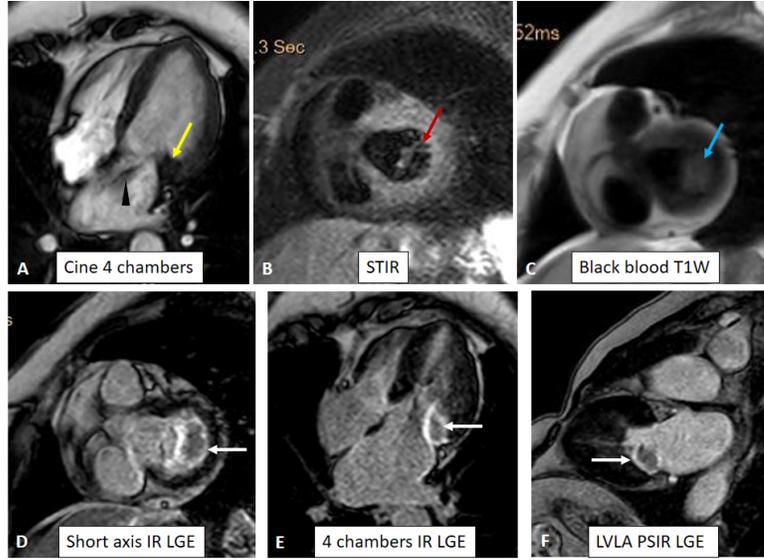


Figure 2

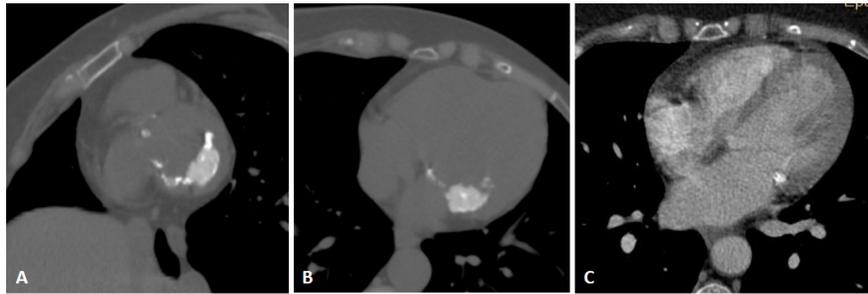


Figure 3