Case Report

Caseous calcification of the mitral annulus mimicking benign cardiac tumour of the mitral valve

Huanhuan Gao, Lei Yao, Yan Cheng, Chao Wu, Xiaoli Mei, Yun Mou, Lijun Jiang, Zhelan Zheng

Abstract

Caseous calcification of the mitral annulus (CCMA) is a rare subtype of mitral annular calcification. It usually appears as a large, round, mass-like calcification with an echolucent core, which may be misdiagnosed as an intracardiac mass, cyst, thrombus or abscess of the mitral valve.

We present a case report of CCMA that was misdiagnosed by echocardiography as a benign tumour due to its atypical imaging. The mitral valve mass was resected and it was pathologically confirmed to be a myxoid change with calcification.

Echocardiography is the preferred initial diagnostic tool. Myocardial contrast echocardiography (MCE) is used to evaluate the vascularity of intracardiac masses or mass-like lesions, but neither echocardiography nor MCE is reliable for identifying atypical lesions. Cardiac computed tomography is helpful in establishing a diagnosis by showing dense calcifications, while cardiac magnetic resonance imaging is used primarily as a credible tool. We therefore recommend that a diagnosis should be made based on various imaging modalities, if necessary, and operators should be skilled to avoid misdiagnosis.

Keywords: caseous calcification, mitral annulus, masses, case report

Submitted 14/1/21, accepted 27/2/21
Cardiovasc J Afr 2021; 32: online publication www.cvja.co.za

DOI: 10.5830/CVJA-2021-007

Echocardiography and Vascular Ultrasound Center, the First Affiliated Hospital, College of Medicine, Zhejiang University, Hangzhou, China
Huanhuan Gao, MD Lei Yao, MD Yan Cheng, MD Chao Wu, MD Yun Mou, MD Zhelan Zheng, MD, 1186034@zju.edu.cn

Department of Ultrasound, the People’s Hospital of Deqing County, Zhejiang, China
Xiaoli Mei, MD

Department of Thoracic and Cardiovascular Surgery, the First Affiliated Hospital, College of Medicine, Zhejiang University, Hangzhou, China
Lijun Jiang, MD

Mitrval annular calcification is a common echocardiographic finding in our daily work, and is considered chronic degeneration of the mitral valve fibrous ring, involving mainly the posterior leaflet of the annulus. Caseous calcification of the mitral annulus (CCMA) is a rare variant of mitral annular calcification, which tends to occur in the elderly, with female, hypertensive individuals and patients with chronic renal failure or calcium metabolism abnormalities being the most vulnerable.1

This disorder usually appears as a large, round, mass-like hyperechoic lesion that may cause diagnostic mistakes. We present a case of CCMA that was misdiagnosed by echocardiography as a benign tumour located in the base of the posterior leaflet of the mitral valve, accompanied by secondary mitral stenosis.

The study was approved by the Institutional Review Board at the First Affiliated Hospital, College of Medicine, Zhejiang University. The procedures were conducted according to the principles of the Helsinki Declaration. Written informed consent was obtained from the patient for publication of this case report and the accompanying images.

Case report

A 64-year-old Chinese female with a suspicious-looking tumour at the posterior leaflet of the mitral valve was referred to our hospital for further diagnosis. She had no chest pain or dyspnoea. The physical examination, electrocardiogram and chest X-ray were unremarkable. Her vital signs were normal with a heart rate of 94 beats/minute, a respiratory rate of 20 breaths/minute, a body temperature of 37°C and a blood pressure of 134/80 mmHg. No cardiac murmur or thrill was present. Initial laboratory tests, including serum creatinine (51 μmol/l), sodium (136 mmol/l), chlorinum (97 mmol/l), potassium (4.89 mmol/l), calcium (2.04 mmol/l) and magnesium (0.98 mmol/l) were all in the normal range.

Four years earlier, the patient underwent a routine echocardiography examination and a hyperechoic lesion in the posterior leaflet of the mitral valve was discovered (Fig. 1A). It was 15 × 9 × 8 mm with no significant mitral regurgitation or stenosis. On presentation, echocardiography revealed that the hyperechoic lesion had increased to 22 × 20 × 16 mm and was attached to the posterior leaflet of the mitral valve (Fig. 1B). It caused moderate mitral stenosis.

Myocardial contrast echocardiography (MCE) was further performed to evaluate the vascularity of the mass and it was found that it was a ring-enhancement mass with distinct borders (Fig. 2A). Quantitative analysis revealed that when compared to ventricle myocardium, the intensity of the mass was higher,
which meant that the lesion had a greater microvascular blood volume (Fig. 2B). Based on its location, appearance and microperfusion, the diagnosis of a benign cardiac tumour (most probably papillary fibroelastoma) was made.

Surgery was performed and it showed that a solid, mixed cystic mass, originating from the posterior leaflet of the mitral valve, approximately 20 mm in length, protruded into the left atrium. The mitral valve mass was resected, while further intraoperative transoesophageal echocardiography revealed severe mitral regurgitation. An Edward bioprosthetic valve (25#) was imbedded. The patient’s recovery was uneventful.

A postoperative pathological examination revealed that the mass was a mitral valve myxoid change with calcification (Fig. 3).

Discussion
CCMA, also called liquefaction necrosis, is a rare type of mitral annular calcification that describes chronic degenerative changes of the cardiac fibrous skeleton, and mainly involves the area between the crest of the posterior left ventricular muscle and the posterior mitral annulus. The elderly female population is the most vulnerable although no clinical significance has been found at present.

CCMA comprises a calcified rim and surrounding caseous material that is composed of calcium, fatty acids and cholesterol, with a toothpaste-like texture. Under the microscope, the CCMA manifested as an amorphous, acellular, basophilic and calcific structure, with a chronic inflammatory response with macrophages as the most numerous cell type. CCMA usually...
behaves as a benign and asymptomatic lesion and is easily confused with other intracardiac masses, cysts, thrombus or abscess.

Under this condition, an association between the CCMA and a medical history of hypertension, chronic renal failure or haemodialysis, and abnormal calcium metabolism should be checked. No related medical history was found in our patient, which was one of the reasons for the misdiagnosis.

Another reason that led to misdiagnosis may have been the relatively small size and atypical imaging of the mass. CCMA is usually large, round, calcified and enveloped in a echolucent core, typically located at the base of the posterior leaflet, and can be misdiagnosed as a cardiac tumour or abscess on echocardiography. The posterior leaflet becomes thickened, stretched and arched over the mass.

Secondary to these anatomical changes, mitral valve dysfunction (either stenosis or regurgitation) can be detected. Since the mass was mimicking a benign cardiac tumour that had increased in size and could not be differentiated from the degenerative valves and with significant valvular dysfunction, surgery was performed on the patient.

MCE may provide much more information about the location, border and perfusion of the CCMA, however no report has been published on the details. According to its pathological findings, the authors concluded that a ring-enhancement mass with a no-perfusion core should be detected by MCE. However, the present case did not seem to fit the characteristics.

The CCMA had a small volume and edge with abundant neovascularisation, which was misleading since the mass was vascular. Another potential contributing factor was the ‘bleeding’ effect, which means the blood of the surrounding left ventricular cavity was moving into the regions of interest as the mass was rapidly oscillating in and out of the imaging plane. We therefore suggest that when MCE is used to evaluate the vascularity of an intracardiac mass, operators need to be aware of these potential pitfalls.

Cardiac computed tomography (CT) and magnetic resonance imaging (MRI) also can be helpful in confirming or establishing diagnosis. Non-contrast CT showed a large calcified mass at the base of the posterior mitral annulus, extending to the adjacent mitral valve and myocardium. On contrast-enhanced CT, the central part appeared less hypodense due to the caseous toothpaste-like material contained within the denser calcified peripheral rim. MRI has shown low signal on both T1- and T2-weighted images due to calcification but was inferior in showing the calcification directly.

Surgical intervention is not only the definitive treatment to remove the potential obstacle of obstruction or embolisation the mass brings about, but also a way to accurate diagnosis and therapy. Therefore our patient had the mass resected and the valve replaced. She is now in a good general condition and undergoing out-patient care with follow up and further management.

Conclusion

CCMA is an exceedingly rare valvular lesion with an excellent outcome following complete surgical removal. Histopathological findings of an amorphous, acellular, basophilic and calcific structure, with a chronic inflammatory response, is the gold standard for diagnosis. Although it was difficult to differentiate from other cardiac masses via echocardiography, diagnosis could be made based on combined imaging modalities. This incidental lesion may be encountered in clinical practice now and then, and cardiac imaging interpretation experts should be familiar with it in order to avoid misdiagnosis.

This study was supported by grants from the National Natural Science Foundation, People’s Republic of China (No. 81371571); Zhejiang Provincial Natural Science Foundation, People’s Republic of China (No. LY13H180008); Scientific Research Fund of Zhejiang Provincial Education Department (No. Y200907825); Health and Family Planning Commission of Zhejiang Province (2016KYA078); and the platform key programme of Health and Family Planning Commission of Zhejiang Province (grant 2015ZDA013).

References


