

Utility of cardiac magnetic resonance imaging for pre-operatively evaluating the degree of caseous calcification of the mitral annulus infiltration within the left ventricular myocardium: a case report

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Background

Depending on the degree of infiltration of dry mitral annulus calcification (MAC) into the left ventricular myocardium, calcification removal may cause thinning of the left ventricular wall and increase the risk of post-operative left ventricular rupture. Therefore, the degree of pre-operative infiltration into the left ventricular myocardium should be assessed as accurately as possible. We report a case of caseous calcification of the mitral annulus (CCMA), a subtype of MAC, in an 84-year-old woman; cardiac magnetic resonance imaging (MRI) was useful in assessing the degree of left ventricular infiltration of calcification.

Case summary

The patient was referred to our hospital for surgery due to respiratory distress and severe mitral insufficiency. Echocardiography revealed a mass under the posterior mitral annulus adjacent to the mitral valve ring, suspected to be a CCMA. Cardiac MRI was performed to assess the risk of left ventricular rupture. Pre-operatively, no valve ring or left ventricular myocardium infiltration was confirmed. Surgical caseous calcification removal and artificial valve implantation within the preserved valve annulus were then performed. The patient had an uneventful post-operative course and was discharged without complications on the 21st post-operative day.

Discussion

In this CCMA case, a pre-operative cardiac MRI was useful for evaluating the degree of the valve annulus and left ventricular myocardial involvement and estimating the risk of left ventricular rupture.

Keywords

Caseous calcification of mitral annulus • Cardiac magnetic resonance imaging • Pre-operative risk assessment • Case report

ESC Curriculum

2.1 Imaging modalities • 2.3 Cardiac magnetic resonance • 2.2 Echocardiography • 4.3 Mitral regurgitation • 7.5 Cardiac surgery

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Learning points

- Although CCMA is a benign disease, 19.2% of patients will have an embolic stroke, and surgery should be considered.
- Assessment of the degree of CCMA infiltration into the left ventricular myocardium is important to assess the risk of post-operative left ventricular rupture.
- Cardiac MRI is useful in assessing the internal nature of mitral annulus calcification and aids in diagnosis.

Introduction

Caseous calcification of the mitral annulus (CCMA) is a rare subtype of mitral annulus calcification (MAC). Its prevalence is 0.06% in patients undergoing transthoracic echocardiography.^{1,2} CCMA is a benign disease; however, 19.2% of patients with CCMA will experience an embolic stroke. Therefore, surgery to correct CCMA is often considered.³ It is crucial to evaluate the extent of cardiac involvement of the CCMA pre-operatively. This is because calcification invasion within the valve ring or left ventricular myocardium can occur, causing intra- or post-operative left ventricular rupture. Here, we report a case where we could evaluate CCMA involvement pre-operatively using cardiac magnetic resonance imaging (MRI), which facilitated pre-operative risk assessment.

Timeline

Timeline	Event
August 2012	A local doctor began monitoring the patient for mitral regurgitation, tricuspid regurgitation, and atrial fibrillation.
February 2022	The patient had respiratory distress on exertion (NYHA 3) and was referred to our hospital for surgical treatment.
16 March 2022	The patient underwent cardiac surgery.
18 March 2022	Extubation was performed (POD2).
6 April 2022	The patient was discharged uneventfully from the hospital (POD21).

Case presentation

An 84-year-old woman started follow-up consultations for atrial fibrillation in 2012. During follow-up, she developed New York Heart Association functional classification II–III symptoms of dyspnoea on exertion. She was referred to our hospital for surgical correction of worsening mitral and tricuspid regurgitation. Pre-operative echocardiography revealed a 22.4 mm × 23.7 mm posterior mitral valve mass (Figure 1). Internal echoes of the mass were heterogeneous, with high echoes at the margins and low echoes within the interior mitral valve. The lesion was clearly static, and CCMA was suspected. Mitral regurgitation was moderate–severe, with an effective regurgitant orifice area of 0.37 cm² and mitral regurgitant volume of 51 mL.

The risk of a left ventricular rupture was difficult to assess because computed tomography (CT) failed to reveal if the posterior mitral valve leaflet calcification had infiltrated the valve ring and left ventricular myocardium (Figure 2). Consequently, an MRI was performed, which revealed that the valve ring was in contact with valve leaflets and that the valve leaflet structure was intact (Figure 3). Therefore, we concluded that mass removal was not likely to increase the risk of left ventricular rupture and proceeded with the surgery.

Circulation, through an artificial heart–lung machine, was established via median sternotomy, and the mitral valve was approached through a right-sided left atrial incision under cardiac arrest. The mass was located under the posterior valve ring (Figure 4A). Paste-like caseous calcification was found in the incision (Figure 4B), and the contents were then removed. The mitral valve was excised as the annulus structure was maintained. Then, a 2-0 ETHIBOND EXCEL[®] Polyester Suture (Ethicon US, LLC, Somerville, NJ, USA) with a pledget was threaded over the annulus with an everting mattress. Afterwards, an artificial bio-prosthetic valve (Epic 29 mm, Abbott, Santa Clara, CA, USA) was attached to the mitral annular ring. Tricuspid valve plication, a maze procedure, and left ventricular closure were also performed.

On post-operative pathological examination, the caseous calcification (Figure 4C) was accompanied by fine calcification. A few cellular components, such as inflammatory cells, were observed (Figure 5). These findings were consistent with CCMA, with no evidence of malignancy. Echocardiography revealed no mitral regurgitation. The patient was discharged on post-surgical day 21 and continued follow-up at the outpatient clinic. Echocardiography performed in the outpatient clinic after discharge showed no abnormalities in the function of the mitral valve prosthesis. Dyspnoea on exertion also improved. There were no surgical complications such as bleeding, infection, respiratory failure, heart failure, or cerebral nerve complications.

Discussion

The risk of MAC and CCMA increases in patients with end-stage renal failure, especially those on dialysis.^{4–6} Pathological findings tend to show various calcifications and necrotic areas surrounding an internal cell-free eosinophilic substance.^{1,2} The present case had such findings; therefore, infection and malignancy could be ruled out.

CCMA often involves the formation of masses on the posterior mitral annulus and, less commonly, on the anterior mitral annulus. It has been suggested that calcification may affect the conduction system, causing bradycardia and an atrioventricular block.^{1,2} CCMA diagnosis can be achieved using echocardiography, CT, and MRI. Echocardiography reveals that most masses are intracardiac, round, internally hyperechoic, and well-defined with a low-signal centre and no signal on colour Doppler.¹ Similarly, CT shows a round, smooth mass with calcification around the periphery and a low- or high-density centre.⁷

Conditions that must be ruled out in CCMA include infectious endocarditis, abscesses, tumours, and intracardiac thrombi. This can be challenging. Mucinous species, accounting for 75% of cardiac tumours, are well-demarcated, round, and have an iso-signal or low-signal interior on echo, findings similar to those of CCMA.⁸ MRI may be useful for

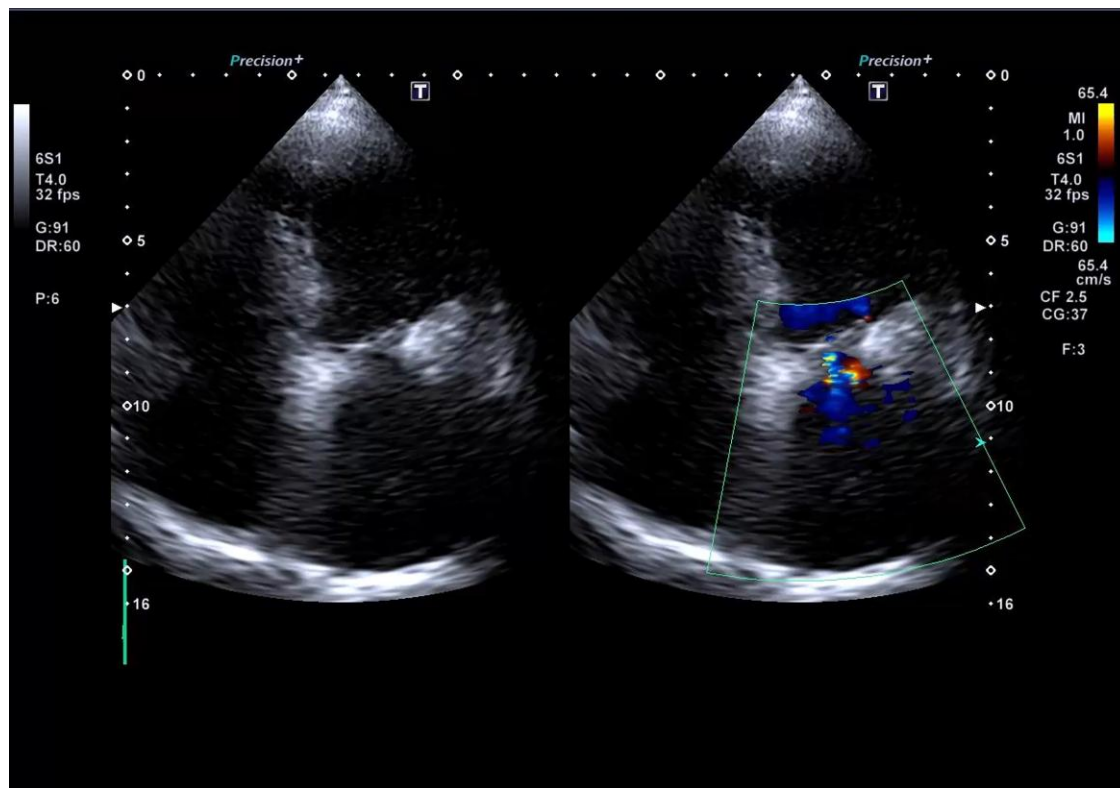


Figure 1 Echocardiogram revealing a large, 22.4 mm x 23.7 mm mass-like lesion in the posterior region of the mitral valve (white arrow ①). Severe mitral regurgitation is present (white arrow ②).

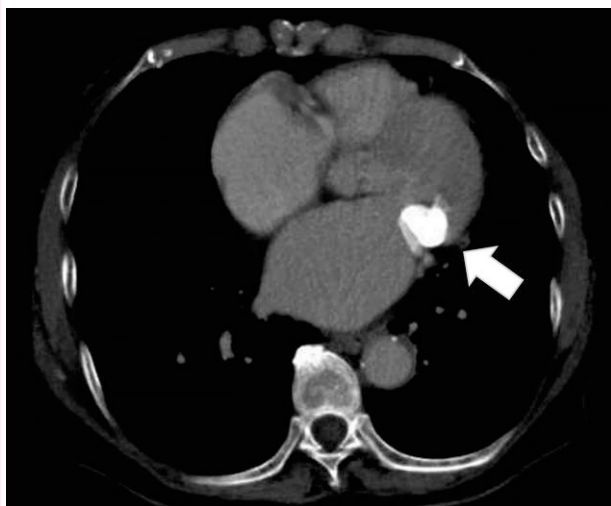


Figure 2 Contrast-enhanced computed tomography findings. Imaging reveals calcification of the mitral valve annulus (white arrow). However, assessment of infiltration within the annulus and left ventricular myocardium was difficult.

differentiating CCMA from other types of cardiac tumours because low iso-intensity at T1 and high intensity at T2 can be shown.⁷ Cardiac MRI is useful for evaluating the degree of invasion because it can be employed to identify the mitral valve ring and left ventricular myocardium boundary. In this case, intra-operative contents included dry, necrotic material, a finding consistent with CCMA.

No clear indications for CCMA surgery have been reported. However, 19.2% of patients with CCMA experience embolic cerebral infarction, a significantly higher risk than that of patients with MAC (11.8%).³ The indication for surgery in our patient was severe mitral regurgitation caused by a necrotic substance that compressed the posterior leaflet of the mitral valve, resulting in desynchrony of the anterior and posterior leaflets and respiratory distress symptoms. The mass was located under the valve ring; therefore, an artificial valve was attached to the valve ring. In such cases, the possibility of performing an incision and internal drainage should be considered, with a closure of the incision, and valvuloplasty or valve replacement performed, if necessary.

CCMA is a rare subtype of MAC. Although benign, it can cause an embolism, and drainage and closure of the endocardium should be considered in high-risk cases, such as those with residual enlargement. Pre-operative evaluation of the degree of infiltration of the valve ring and left ventricular myocardium can reduce the risk of intra-operative or post-operative left ventricular rupture. This case demonstrates that cardiac MRI is useful for assessing the risk of rupture.

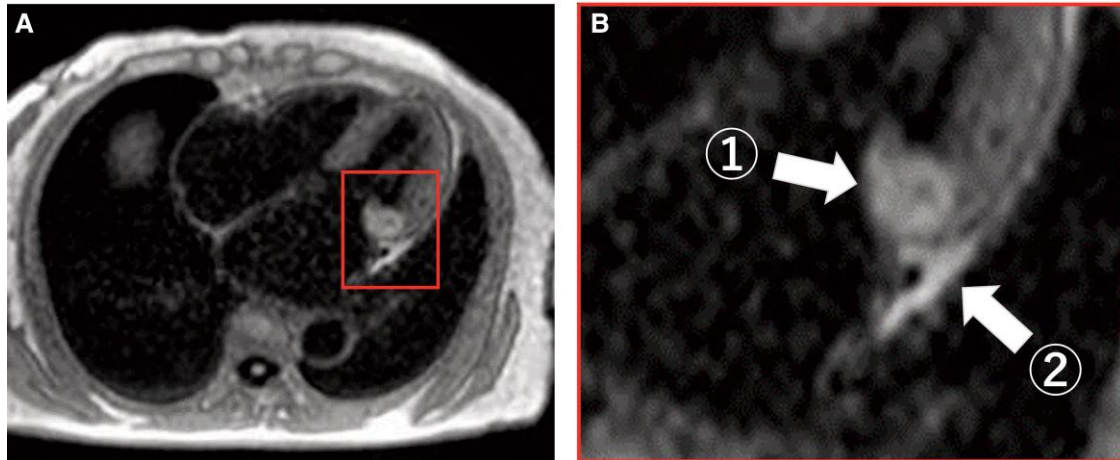


Figure 3 Cardiac magnetic resonance image shows the mass (white arrow ①) in contact with the valve ring (white arrow ②) without infiltration or invasion of the left ventricle.

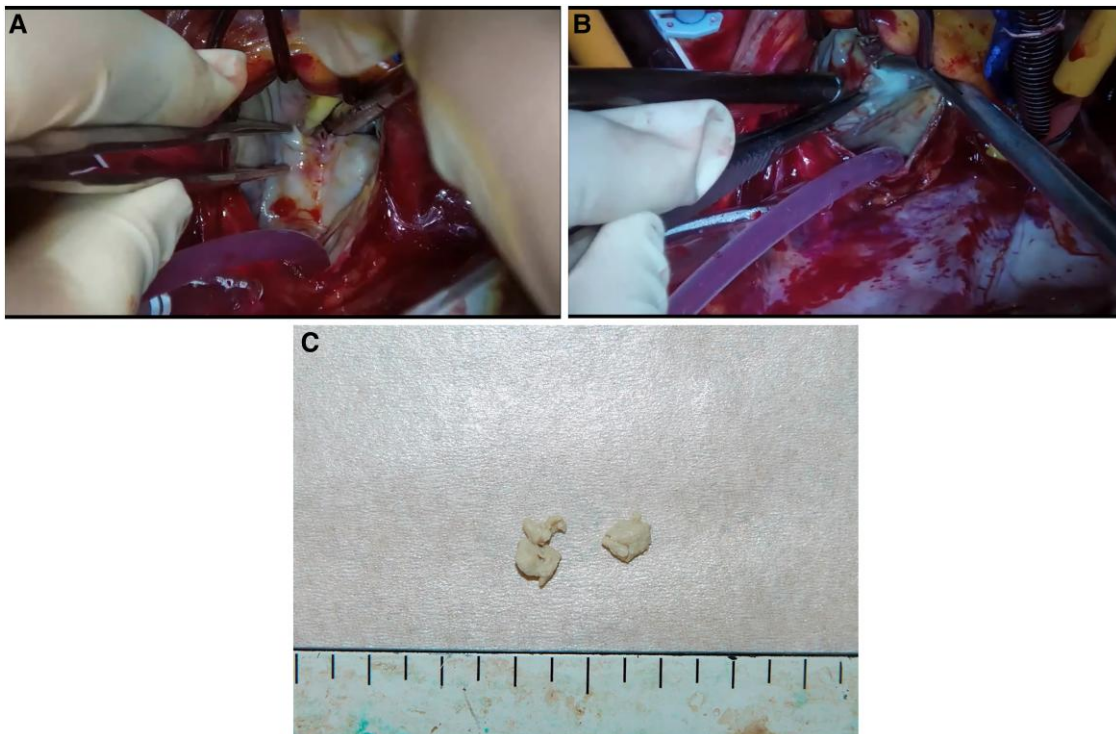


Figure 4 Intra-operative images. (A) A movie of the mass lesion in the annulus of the mitral valve's posterior. (B) A movie revealing caseous calcification within the mass. (C) A photograph of the caseous calcification after surgical removal.

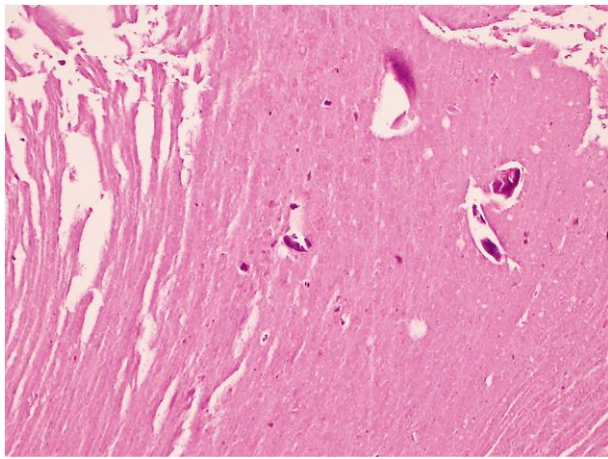


Figure 5 An image of the post-operative pathological examination findings revealing calcification and a few cellular components, such as inflammatory cells ($\times 100$ magnification).

Lead author biography



Takanori Hishikawa is a cardiac surgeon at Nagoya Tokushukai General Hospital. He graduated from the Department of Medicine of Nagoya City University in 2018 and worked as a resident at Tokyonishi Tokushukai Hospital till March 2020. Presently, he is affiliated with Nagoya Tokushukai General hospital as a clinical fellow, a position he has held since April 2020.

Supplementary material

Supplementary material is available at *European Heart Journal – Case Reports* online.

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Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as [Supplementary data](#).

Consent: Informed consent was obtained from the patient included in the study in accordance with COPE guidelines. The ethics committee approved this study.

Conflict of interest: None declared.

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Data availability

Data sharing is not applicable to this article as no new data were created or analysed in this study.

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