

# Caseous calcification of the mitral annulus presenting with symptomatic complete heart block



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## Introduction

Caseous calcification of the mitral annulus (CCMA) is a rare variant of mitral annular calcification (MAC). It is consequent to chronic degenerative process involving the fibrous annulus of the mitral valve.<sup>1,2</sup> The prevalence is highest among women and above 85 years of age. Most often it is an incidental finding on echocardiography.<sup>3</sup> MAC and vascular atherosclerosis are closely associated and share several risk factors such as advancing age, diabetes, and increased body mass index. Patients with MAC have demonstrated a higher prevalence of cardiac conduction disease.<sup>1</sup>

This case report illustrates a rare situation where CCMA was incidentally found in a relatively young male patient who had presented with complete heart block. It had infiltrated the basal septum and the His bundle. This case also proved to be a diagnostic challenge because CCMA was not readily confirmed on echocardiography. Further evaluation with cardiac magnetic resonance imaging (MRI) was required to arrive at the diagnosis.

## Case report

A 63-year-old man was referred to the Emergency Department with worsening chest discomfort, dizziness, and palpitations. He had a background medical history of type 2 diabetes, hypertension, and obesity. On examination he was euvolemic and his heart sounds were dual, with no murmurs or added sounds.

The electrocardiogram showed complete heart block with a ventricular rate of 36 beats per minute and a narrow QRS escape rhythm of 100 ms (Figure 1A). Serum troponin was not elevated. Routine blood chemistry was within normal limits. He was biochemically euthyroid.

**KEYWORDS** Cardiac magnetic resonance imaging; Caseous calcification of the mitral annulus; Complete heart block; His bundle pacing; Intrafascicular block; Mitral annular calcification  
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## KEY TEACHING POINTS

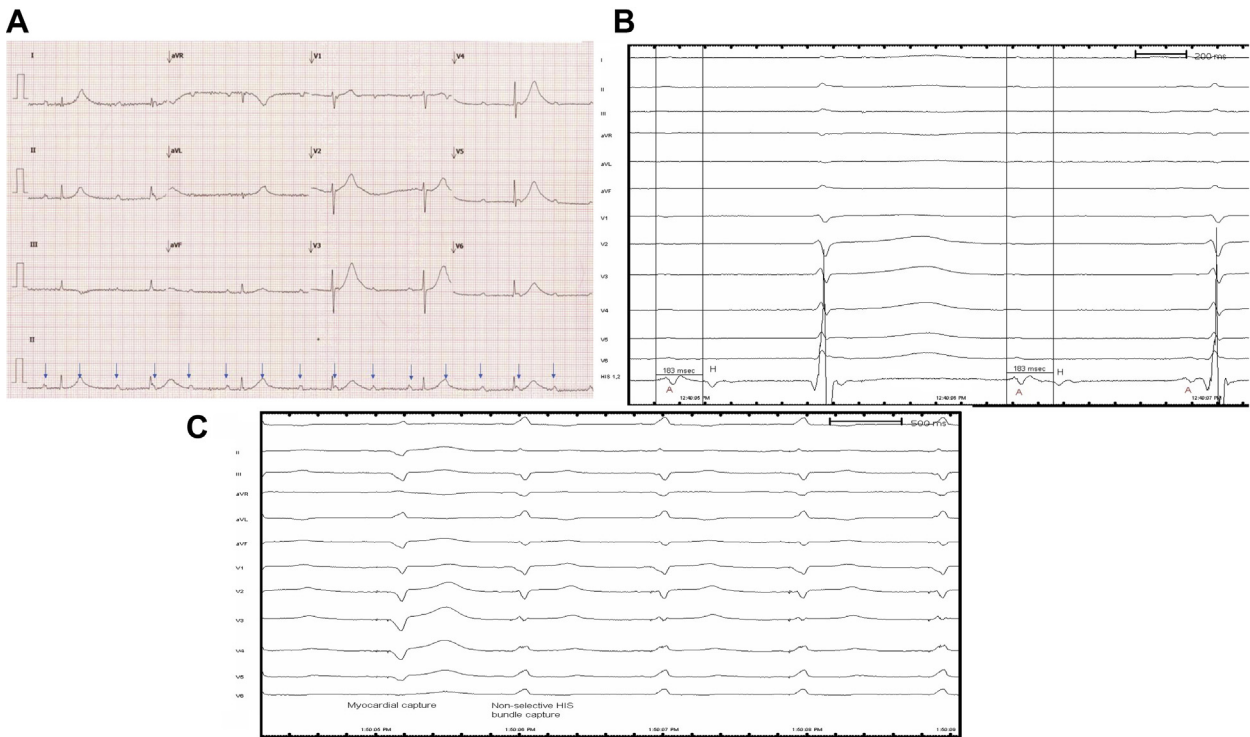
- Caseous calcification of the mitral annulus (CCMA) remains a largely underdiagnosed condition.
- It has a close association with cardiac conduction disease.
- It is predominantly observed in patients above the age of 85.
- The diagnosis of CCMA is typically made with transthoracic echocardiography.
- There is a role for cardiac magnetic resonance imaging in the diagnosis of complex cases.

A transthoracic echocardiogram revealed a normal ejection fraction of 55%–60%. An echogenic mass measuring  $3.1 \times 3$  cm in dimension was seen on the mitral annulus extending into the basal septum. It appeared as a bright echogenic lesion across the entire mitral annulus. There was mild mitral leaflet thickening with mild regurgitation and no stenosis (Figure 2A).

A transesophageal echocardiogram was done to delineate this further. This revealed a well-circumscribed circular intramyocardial mass in the basal posterior medial aspect of the left ventricle extending into the posterior mitral annulus. There was no mass effect or valvular impingement (Figure 2B). The differentials based on transesophageal echocardiography included lipomatous hypertrophy, noncaseous granuloma, and MAC. Central liquefaction was not appreciated on either of the modalities. Therefore, echocardiographic findings alone did not support the diagnosis of CCMA.

The relatively young age of presentation with conduction disease prompted us to evaluate further with computed tomography of the chest to exclude pulmonary sarcoidosis (Figure 2C). This did not demonstrate hilar or mediastinal lymphadenopathy. It did, however, reveal dense thick calcification of mitral annulus with basal septal extension that had an appearance suggestive of caseous calcification. A cardiac MRI was done to evaluate the mass further. This confirmed a hypointense lesion with septal extension. Low

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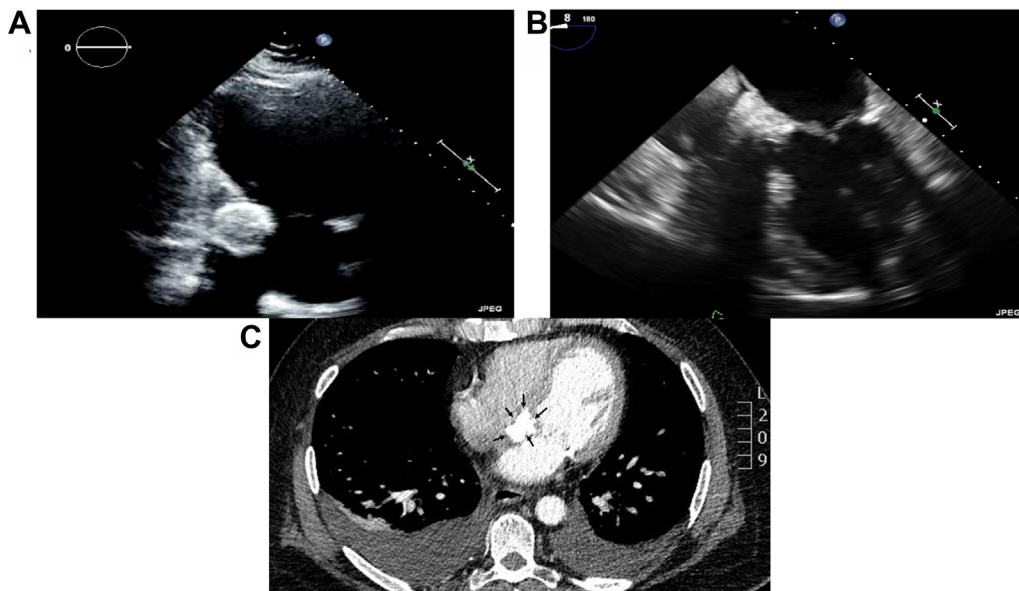


**Figure 1** **A:** Electrocardiogram (ECG) of the patient upon presentation to the hospital shows complete atrioventricular dissociation. The sinus rate is 100 beats/min (bpm) and ventricular rate of 36 bpm. **B:** Intracardiac electrogram done prior to pacemaker insertion, which shows complete heart block owing to an infrahisian block. Signals labeled as follows: A = atrium; H = His bundle potential. **C:** Twelve-lead ECG post pacemaker insertion. The first complex shows myocardial capture, and the second complex shows a nonselective His-bundle capture.

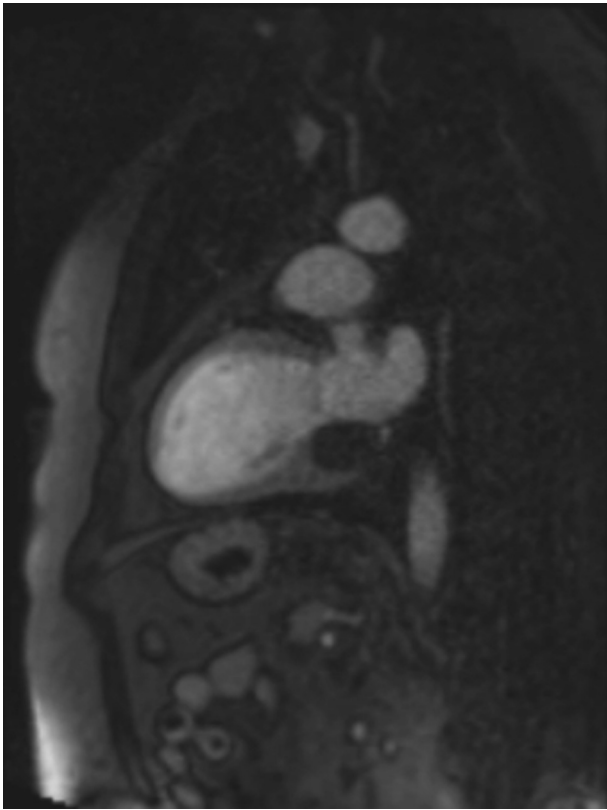
signal intensity was seen within the mass in all MRI sequences. There was a crescent of thin enhancement, posteriorly and inferiorly. The T2 weighted-Spectral Attenuated Inversion Recovery sequences demonstrated no myocardial

fat and confirmed the diagnosis of CCMA extending into the basal septum (Figure 3).

An electrophysiology study revealed evidence of infrahisian atrioventricular (AV) block with a distal hisian escape



**Figure 2** **A:** Transthoracic echocardiogram (apical 2-chamber view). Bright echogenic lesion measuring  $3.1 \times 3.0$  cm across the entire mitral annulus extending into the basal septum. **B:** Transesophageal echocardiogram (mid-esophageal 4-chamber long-axis view) showing well-circumscribed intramyocardial mass in the basal posterior medial aspect of the left ventricle measuring  $2.5 \times 2$  cm in diameter. It did not appear caseous and was echodense compared to the adjacent tissue. **C:** Computed tomography of the chest at the level of the heart. Dense thick caseous calcification of the mitral annulus with basal septal extension. Moderate bilateral pleural effusion noted.



**Figure 3** Two-chamber long-axis view demonstrating a hypointense lesion on the posterior and inferior portion of the mitral annulus with septal extension.

rhythm. A permanent pacemaker at the His bundle was implanted (electrocardiogram shown post pacemaker in Figure 1B). He continues to be pacing dependent and has since been asymptomatic.

## Discussion

The true prevalence of CCMA is currently unknown because of unfamiliarity with the diagnosis and consequent under-reporting.<sup>2</sup> A prospective analysis of patients referred for echocardiography found that a diagnosis of MAC was made in 10.6% of the study population. Approximately 0.64% of these had echocardiographic features consistent with CCMA. However, in a necropsy series, the prevalence was as high as 2.4%.<sup>4</sup>

CCMA is a rare variant of MAC.<sup>5</sup> MAC is the result of progressive calcium deposition along and beneath the annulus of the mitral valve. It is thought to be an active process similar to atherosclerosis and calcific aortic valve disease.<sup>4</sup> It begins with endothelial disruption at areas of increased mechanical stress at the junction between the mitral valve annulus and ventricular myocardium. Over time, areas of focal chronic inflammation of extracellular matrix results in remodeling and coalesces to form the dense, rigid, fibrotic band that is typically seen as a bright echodense region on echocardiography.<sup>5,6</sup> However, the pathogenesis of liquefactions and caseation in CCMA is not clearly understood.<sup>5</sup>

The most common presentation of CCMA is an incidentally discovered intracardiac mass on cardiac imaging. If present, the most common symptoms are palpitations and dyspnea.<sup>4</sup>

MAC has a close association with conduction system disease in the form of AV block, bundle branch block, and intraventricular conduction delay.<sup>1</sup> A study examining 104 patients with MAC matched them with 121 matched control subjects found that the conduction defects were present in 70% of patients with MAC as opposed to 34% of controls. Extension of calcific deposits into the region of the AV node and the bundle of His has been considered as the mechanism for conduction disease, as also described in this case.<sup>1</sup>

On echocardiography, the typical findings are that of a central echolucent zone and absent acoustic shadowing. CCMA is characteristically less echodense than MAC.<sup>5</sup> It appears as a large, round mass with smooth borders in the periannular region, with no acoustic shadowing artefact, and contains central areas of echolucencies consistent with liquefaction. Although transthoracic echocardiogram is considered the most reliable method to diagnose this condition, cardiac MRI is the modality of choice in complex cases.<sup>5</sup> In this case, echocardiography was useful in identifying the presence of the intracardiac mass and infiltration into the basal septum. However, the echodensity of the mass was difficult to gauge and its dimensions were overestimated.

The typical MRI features of CCMA, as noted in our case, have been described as a well-defined mass, with a peripheral rim of hypointensity with a hyperintense center. It is usually observed in T1-weighted imaging. On T2-weighted MRI sequences, CCMA appears as a mass devoid of a central signal but with a ring of high intensity in comparison with the surrounding myocardium.<sup>5</sup>

There is no consensus on the management of CCMA. Conservative management is preferred when the diagnosis is certain and there is no obstruction to atrial emptying.

The indications for surgical interventions include mitral valvular dysfunction (stenosis or regurgitation), embolic phenomenon, and excision when it is impossible to exclude a tumor.<sup>5</sup>

## Conclusion

Our case demonstrates a relatively young patient who had presented with complete heart block owing to CCMA, which had infiltrated the basal septum and the bundle of His. Although echocardiography is usually sufficient to diagnose the condition, further evaluation with a cardiac MRI was needed in this case to arrive at the diagnosis, suggesting the utility for MRI in diagnostically complex cases.

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