

A Rare Beast: Cardiac Calcified Amorphous Tumor



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INTRODUCTION

Calcified amorphous tumor (CAT) of the heart is a rare non-neoplastic intracardiac mass first described in 1997 by Reynolds *et al.*¹ Although case reports of cardiac CAT have been published since, early attempts at identifying characteristic features of this condition and distinguishing it from other cardiac masses relied on limited information.^{2,3} More data on cardiac CAT are required to develop evidence-based approaches to its diagnosis, treatment, appropriate follow-up, and possibly prevention strategies.

CASE PRESENTATION

A 54-year-old man with a history of hypertension and coarctation of the aorta (repaired at age 7) presented reporting 1 week of shortness of breath at rest, particularly when lying flat, and palpitations. He denied any fevers, chills, or chest pain. Physical examination revealed clear lungs; normal heart sounds; no murmurs, rubs, or gallops; no jugular venous distension; and no peripheral edema. Laboratory analysis was significant for a troponin peak of 0.3 ng/mL. Electrocardiography revealed sinus rhythm with prolonged PR interval, intraventricular conduction delay, and early repolarization pattern. Chest radiography showed no focal infiltrate, pleural effusion, or pneumothorax. Computed tomography of the chest showed a reticulonodular infiltrate in the right upper lobe. Transthoracic echocardiography revealed mild concentric left ventricular hypertrophy, normal left ventricular ejection fraction (55%–60%), no regional wall motion abnormalities, and no significant valvular abnormalities. There was, however, a prominent calcified mobile echo density (1.2 × 0.5 cm) within the left ventricular cavity adjacent to the anterolateral papillary muscle. The differential diagnosis at that point included neoplasm, thrombus, or vegetation. Subsequent cardiac magnetic resonance imaging confirmed a 1.1 × 0.9-cm mass adjacent to the anterolateral papillary muscle. However, because of cardiac arrhythmia and the patient's difficulty complying with breath-hold instructions, the magnetic resonance images were suboptimal, and further imaging was needed. Transesophageal echocardiography revealed a broad-based, calcified mass arising from the myocardium adjacent

to the anterolateral papillary muscle and a large mobile component with independent motion (Figures 1–3, Videos 1–3). Given the size, location, and embolic potential of the mass, surgical resection was advised. The patient underwent successful resection of the mass, with a total bypass time of 36 min, and recovered uneventfully. Gross pathology showed a 1.5 × 0.5 × 0.3-cm mass composed of soft tan-white membranous tissue and friable debris (Figure 4). Microscopic pathology revealed a calcific nodule with eosinophilic amorphous fibrinous material consistent with cardiac CAT (Figure 5). There was no evidence of a neoplastic process or sarcoidosis.

The patient underwent repeat transthoracic echocardiography 8 months after surgery, which showed no recurrence and no systolic or valvular abnormalities. There is a plan for follow-up of this patient with his cardiologist every 6–12 months to closely monitor signs and symptoms. Follow-up imaging will also be coordinated to assess for recurrence.

DISCUSSION

Cardiac CAT is a non-neoplastic cardiac mass of unknown etiology.¹ The pathogenesis of CAT remains poorly understood. Various etiologies have been proposed, including organized calcific mural thrombus,^{1,3} hypercoagulability,^{1,3,4} abnormalities of calcium and phosphate metabolism,^{1,4,5} and chronic inflammation.⁶ In some cases of cardiac CAT, underlying hypercoagulability was attributed to atrial fibrillation, trauma, antiphospholipid syndrome, malignancy, and genetic diseases.^{1,4,5} Abnormalities of calcium and phosphate metabolism have been linked to patients on hemodialysis⁷ and patients with abnormal regulation of parathyroid hormone and vitamin D.⁴ It has been suggested that end-stage renal disease may play an important role in the pathogenesis of mitral annular calcification–associated CAT formation, which may be distinct from CAT occurring at other sites in the heart.⁴ We could not identify any obvious etiologic factors in our patient to support any of these hypotheses. Histologically, cardiac CAT is characterized by calcified amorphous debris in the background of fibrinous material. Limited data suggest female predominance (approximately 68%) and a wide age range (16–82 years), with a mean age of 55 years.^{2,8} In addition, cardiac CAT has been associated with hypertension (16%), valvular heart disease (31%), end-stage renal disease (25%), diabetes (23%), and coronary artery disease (18%).^{3,8}

Patients with cardiac CAT frequently present with dyspnea, syncope, and/or ventricular arrhythmias.⁹ On echocardiography, cardiac CAT usually appears as a calcified intracardiac mass with or without mobile components. Although cardiac computed tomography and magnetic resonance imaging are slowly gaining more favor, current cardiac imaging techniques do not reliably differentiate cardiac CAT from other masses.¹⁰ Cardiac CAT has been found in various intracardiac locations, including the mitral valve (32%), right atrium (14%), right ventricle (16%), tricuspid valve

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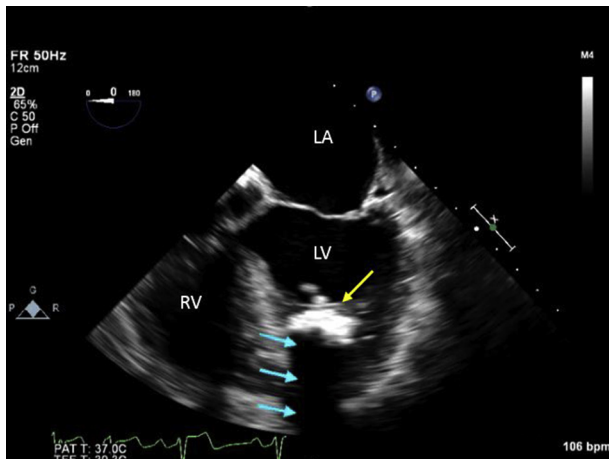


Figure 1 Transesophageal echocardiography. Midesophageal five-chamber view showing large irregularly shaped calcified left ventricular mass (yellow arrow). Note the dense acoustic shadow cast by the mass consistent with dense calcification (blue arrows). See [Video 1](#). LA, Left atrium; LV, left ventricle; RV, right ventricle.

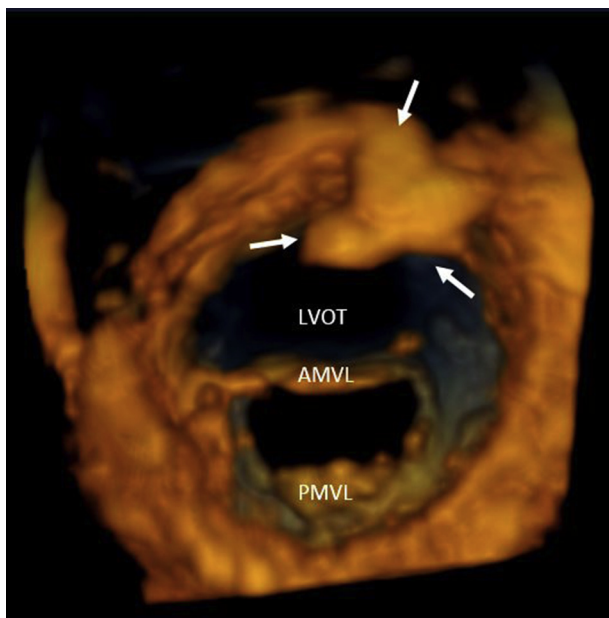


Figure 2 Three-dimensional transesophageal echocardiography. Left ventricular perspective. The mass (white arrows) is seen broadly attached to the midanterior segment of the left ventricular myocardium. See [Video 2](#). AMVL, Anterior mitral valve leaflet; LVOT, left ventricular outflow tract; PMVL, posterior mitral valve leaflet.

(5%), left atrium (11%), and left ventricle (22%).^{1,3,8} The cardiac CAT in our case is unique because of its location in the left ventricle, as most have been found on the mitral valve and annulus. The proximity of the mass to the mitral subvalvular apparatus is an important consideration in surgical planning in order to avoid injury to papillary muscles and chordae. In our case, although the mass was close to the anterolateral papillary muscle, there was

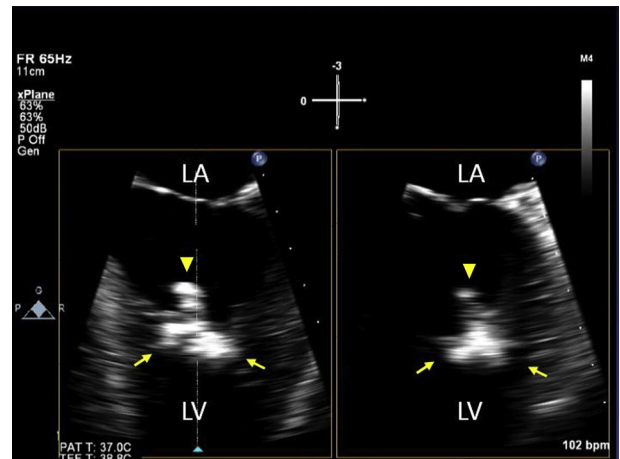


Figure 3 Transesophageal echocardiography. Midesophageal four-chamber view in the biplane mode showing spatial orientation of the mass (arrowheads, mobile component; arrows, fixed component). See [Video 3](#). LA, Left atrium; LV, left ventricle.

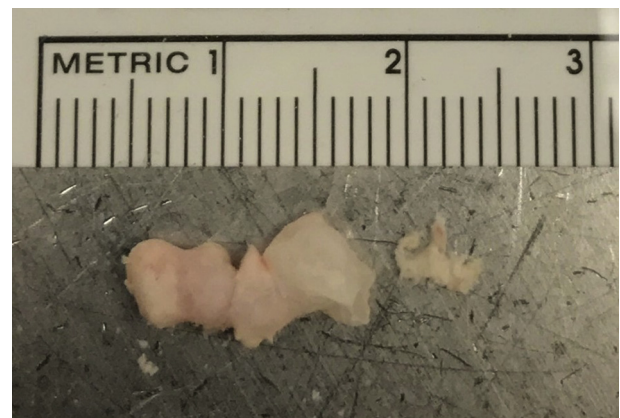


Figure 4 Gross anatomic pathology of the excised left ventricular mass showing the tan-white mass with soft friable debris.

no tumor invasion into the muscle itself, and no injury to the subvalvular apparatus occurred during surgical resection. The embolic potential of these lesions has been reported as high as 31%, which seems to justify surgical resection performed in most cases.^{1,11,12} In a recent case report, cardiac CAT was associated with multiple small cerebral and cerebellar infarctions.⁸

Although most published cases of CAT to date were treated surgically, conservative management may be preferred in some cases, such as asymptomatic elderly patients.¹³ In addition, caution should be used in cases with extensive invasion of the myocardium by the tumor, as wide surgical resection could result in fatal complications.¹⁴ Other imaging modalities (computed tomography, magnetic resonance imaging) may have a role in noninvasive diagnosis of cardiac CAT and thus may help avoid surgery in some patients.¹⁰ More investigation is needed to determine which patients will benefit more from conservative management rather than surgery.

It is not yet clear how rapidly these masses grow. In one reported case, a cardiac CAT measuring 1.0 × 0.7 cm grew to 2 × 3 cm in <1 year.⁵ Our patient did not have any prior echocardiographic studies to help assess the rate of progression of the cardiac CAT.

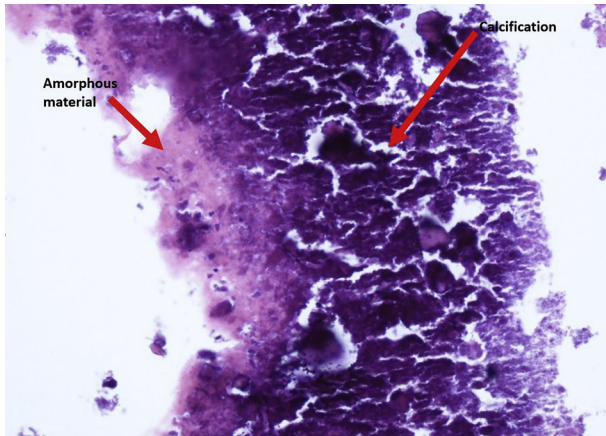


Figure 5 Microscopic pathology showing nodules of calcification and eosinophilic amorphous material in the background consistent with CAT.

Data on recurrence are scant, but at least one study found recurrence 2 years after surgical resection, thus raising the possibility that surveillance echocardiography may have a role in long-term follow-up of these patients.⁵

CONCLUSION

Cardiac CAT is a rare non-neoplastic intracardiac mass usually diagnosed by echocardiography and usually managed with surgical resection because of risk for embolism. Overall prognosis appears to be favorable, on the basis of limited available information. More data are required to determine the best diagnostic and management strategies, including selecting the optimal diagnostic imaging techniques, clarifying the role of systemic anticoagulation, and determining the frequency of postsurgical surveillance for recurrence.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at <https://doi.org/10.1016/j.case.2018.04.003>.

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