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Cerebral Infarction Due to Calcified Amorphous Tumor: A Rare Case Report

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ABSTRACT

A Calcified Amorphous Tumor (CAT) consists of calcified nodules embedded within an amorphous fibrous material and represents an uncommon non-neoplastic intracavitary cardiac mass. In this article, we present the case of a 38-year-old Iranian gentleman who experienced a sudden loss of consciousness and right hemiparesis. Brain imaging revealed acute infarction in the left basal ganglia. Echocardiography identified a large heterogeneous echodensity in the posterior AV groove, with central echolucency extending to the base of the posterior left ventricular (LV) wall and the basal posterior mitral valve leaflet (PMVL), findings consistent with CAT.

Cardiac Magnetic Resonance Imaging (CMR) demonstrated an intramural calcified mass with a necrotic core located in the lateral annulus of the mitral valve, extending into the left atrium and ventricle. The mass was deemed responsible for the cerebral infarction, which was classified as cardioembolic. This case report highlights Calcified Amorphous Tumor (CAT) as a potential embolic source, underscoring the importance of early recognition through multimodality imaging. Comprehensive management, which may include regular follow-ups or surgical intervention, is crucial for patients diagnosed with CAT.

Introduction

The calcified amorphous tumor (CAT) of the heart is an uncommon non-neoplastic intracavitary cardiac mass. It consists of calcified nodules embedded within an amorphous fibrous material and can cause symptoms of embolization or obstruction due to calcified fragments [1]. CAT is estimated to account for 2.48% of primary cardiac neoplasms [2]. The echocardiographic prevalence of CAT is reported as 0.64% among patients with Mitral Annulus Calcification (MAC) and 0.068% in the general population [3]. Autopsy findings indicate a higher prevalence (2.7%), suggesting that this condition may be underrecognized [4]. CAT is more frequently observed in older individuals, females, and those with risk factors such as hypertension, chronic kidney disease (CKD), or altered calcium-phosphate metabolism.

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In our literature review, we identified that cardiac CATs rarely result in cardioembolic strokes. Here, we present the case of a young patient with a cerebral infarction caused by a cardiac CAT.

Case Presentation

A 38-year-old Iranian gentleman was referred to our hospital after experiencing a sudden loss of consciousness and right hemiparesis. He had no significant past medical or family history and no prior episodes of chest trauma, tuberculosis, or unexplained high fever. Upon presentation, his vital signs were stable, with a blood pressure of 135/85 mmHg, a pulse rate of 58 beats per minute, and a body temperature of 36.8 °C.

Laboratory analysis showed that serum parathyroid hormone, calcium, creatinine, and glucose levels were within normal limits. Additional findings included sodium at 136 mEq/L, potassium at 4.6 mEq/L, blood urea nitrogen at 16 mg/dL, serum creatinine at 0.9 mg/dL, magnesium at 1.8 mg/dL, calcium at 8.9 mg/dL, phosphorus at 3.9 mg/dL, aspartate aminotransferase at 23 U/L, alanine aminotransferase at 17 U/L, and total bilirubin at 4.2 mg/dL. His complete blood count (CBC) revealed a hemoglobin level of 14.3 g/dL, a white blood cell count of 5100/µL, and a platelet count of 156,000/µL. C-reactive protein was measured at 16 mg/L. The intact parathyroid hormone level was 143 pg/mL, and the parathyroid hormone-related peptide level was less than 1.0 pmol/L. The tuberculin skin test was negative.

An electrocardiogram (ECG) revealed nonspecific ST-T changes (Figure 1).

Upon arrival at the hospital, the patient exhibited right hemiparesis and motor aphasia. Imaging studies, including brain computed tomography (CT) and magnetic resonance imaging (MRI), revealed acute infarctions in the head of the left caudate nucleus, basal ganglia, and the splenium of the corpus callosum (Figure 2).

A plain chest radiograph showed diffuse linear pericardial thickening and a calcification ring encircling the ventricle (Figure 3).

Based on these findings, an echocardiogram was performed. The initial echocardiography identified a large heterogeneous echodensity (25 × 17 mm) in the posterior atrioventricular (AV) groove with central echolucency, extending to the base of the posterior left ventricular (LV) wall and the basal posterior mitral valve leaflet (PMVL), findings consistent with a calcified amorphous tumor (CAT). A small mobile echodense mass (8.9 mm) attached to the basal posterior LV wall was also detected (not shown).

Additionally, the echocardiogram revealed a normal LV size with mild systolic dysfunction (LVEF = 50%), inferolateral and posterior wall hypokinesia, mild to moderate mitral regurgitation, and mitral annular calcification (MAC) (Figure 4).

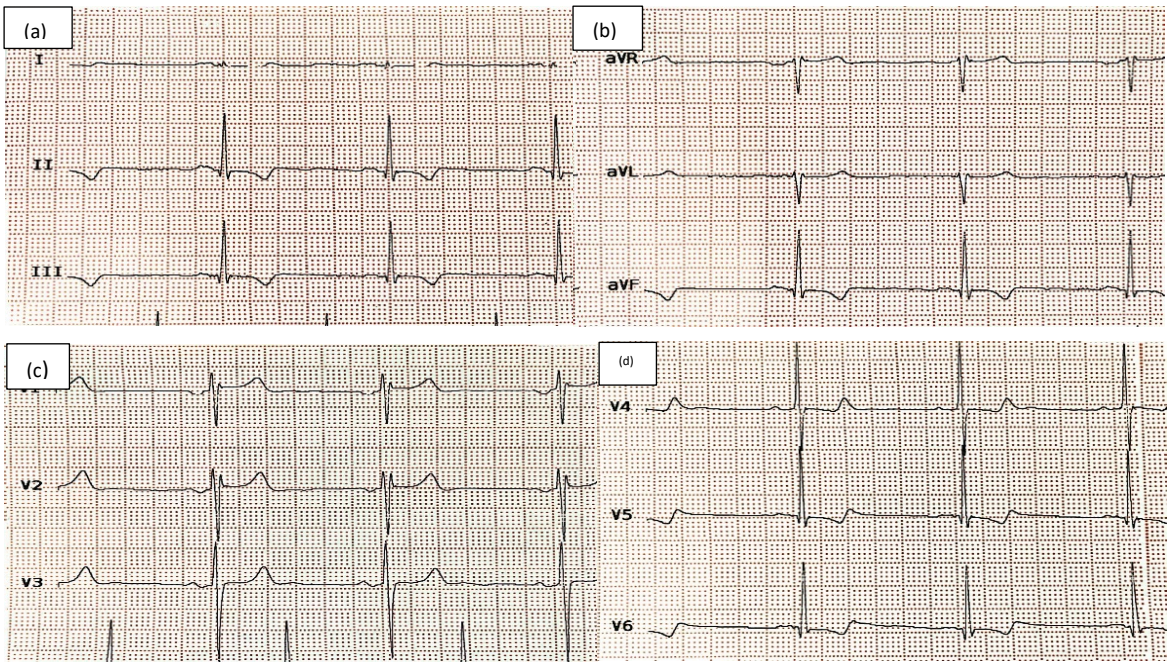

Fig. 1. 12 lead ECG shows left atrial abnormality and T wave inversion in inferior and V3-V4

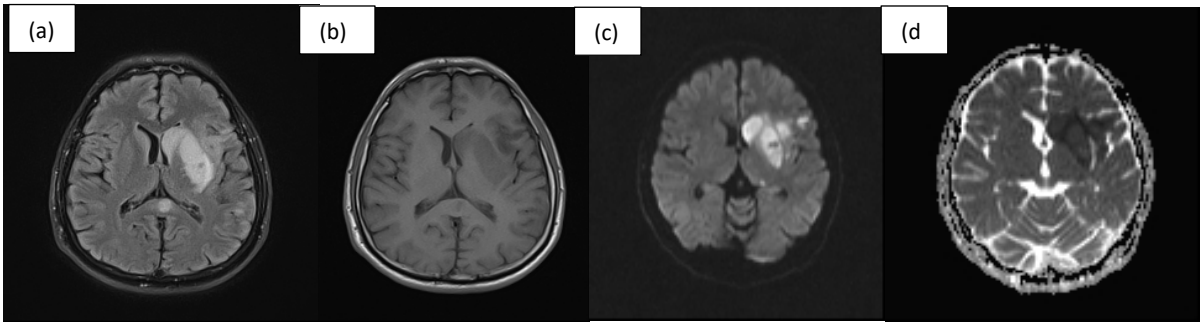

Fig. 2. Axial view of brain MRI a)Flair sequence showing high signal intensity in the in the head of left caudate nucleus, basal ganglia and the splenium of corpus callosum. b)T1 weighted sequence showing hypointense signal in the head of left caudate nucleus, basal ganglia and the splenium of corpus callosum. c&d)diffusion weighted imaging (DWI) & apparent diffusion coefficient (ADC) showing restricted area in left basal ganglia and splenium of corpus callosum.

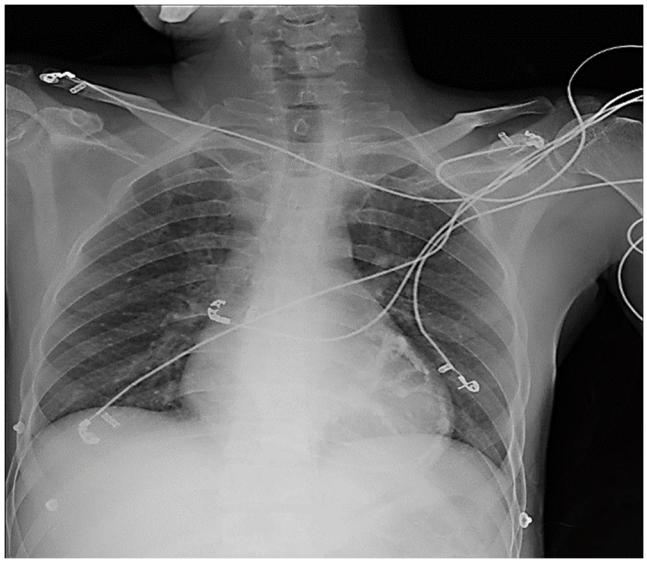

Fig. 3. Anteroposterior Chest X-ray showing linear calcification encircling the ventricle

A cardiovascular magnetic resonance (CMR) scan was requested for further investigation. The patient underwent cardiovascular MR with a mass protocol, which revealed extensive calcification of the pericardium near the annulus and the base of the heart, as well as around the right ventricle (RV). Notably, there was depression of the RV-free wall without evidence of constriction.

Additionally, an intramural calcified mass with a necrotic core, measuring 32 × 26 mm, was identified in the lateral annulus of the mitral valve, extending into both the left atrium and left ventricle (Figure 5). The remarkable aspect of this case was the penetration of the affected tissue into the myocardium and even into the left ventricular cavity, with a tiny mobile particle attached to the mass.

To further assess pericardial involvement and visualize the coronary arteries, coronary CT angiography was performed. The scan revealed diffuse calcification within the pericardium, extending into the atrioventricular groove, with more pronounced involvement in the posterior LV due to the CAT (Figure 6). Notably, there was no evidence of coronary artery stenosis.

Based on the clinical course and imaging findings, the diagnosis of CAT was confirmed. The mass was determined to be related to the cerebral infarction, now classified as cardioembolic. Given the severity and extent of pericardial and myocardial involvement, surgical excision was deemed unsuitable by the surgical team. The patient was treated medically with anticoagulant agents. Neurological symptoms improved rapidly, and he was discharged with a prescription for anticoagulant therapy. At a six-month follow-up, the patient remained well, with no evidence of recurrent cerebral infarction.

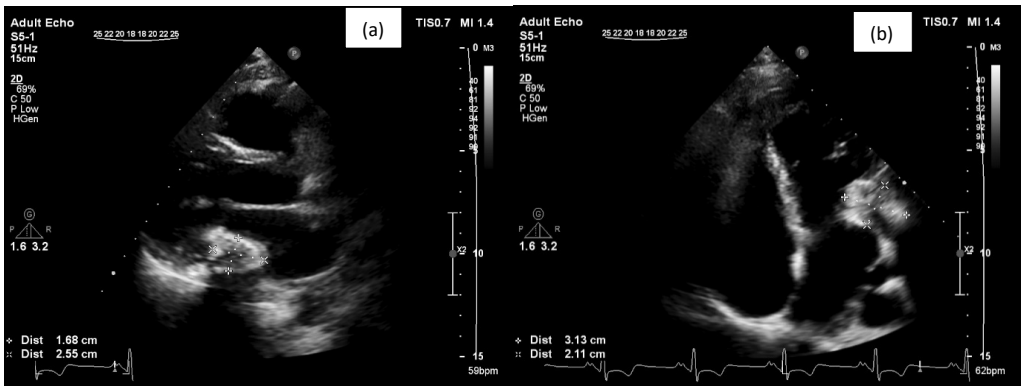

Fig. 4. Transthorasic echocardiography in a)parasternal & b)apical 4chamber view shows a heterogenous echodensity in posterior AV groove with central echolucency.

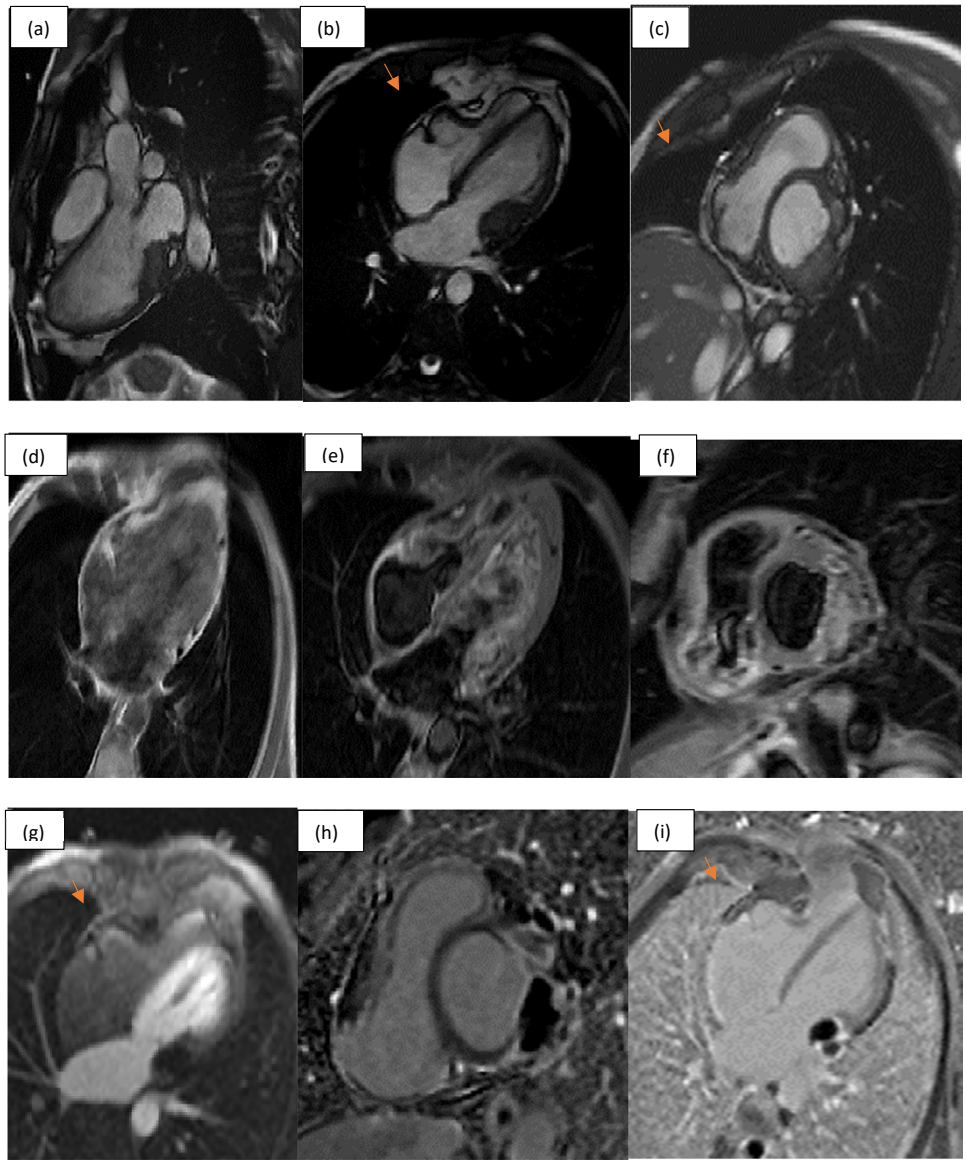

Fig. 5. a,b,c)Balanced steady-state free precession(bSSFP) in 2ch,4ch &short axis sequences showing an isointense mass. d)T1-weighted axial sequence showing isointense mass. e&f)T2-weighted STIR short axis sequence with heterogenous iso to high intense. g)1st pass perfusion sequence with trace perfusion. h & i)Delayed enhancement sequence with no enhancement. Also, extensive pericardial calcification with greater intensity in the anterior RV is evident along with compressive effect(orange arrow)

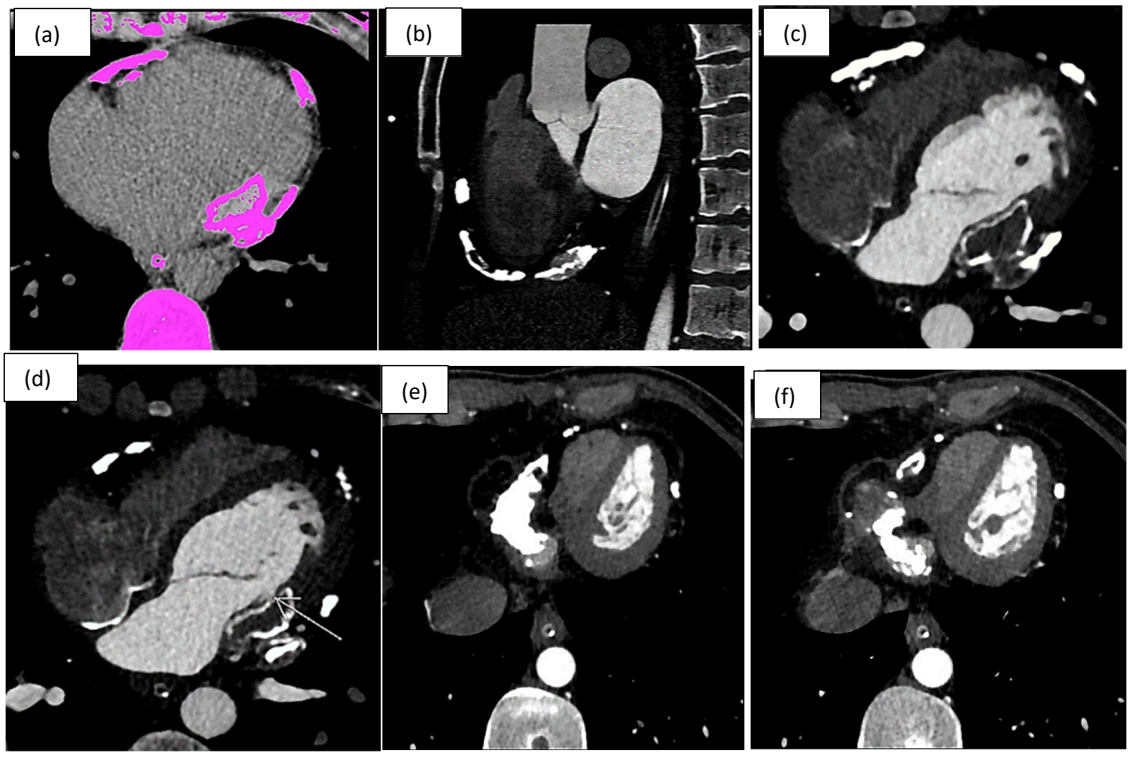

Fig. 6. Multidetector computed tomography.a)Non contrast image showing calcified pericardium and mass with myocardial penetration.b-f)Multiplanar reconstruction.The mass is hyperdense at peripher with hypodense core with no enhancement.

Discussion

The most important differential diagnoses of CAT include cardiac myxoma, caseous calcification of the mitral annulus (CCMA), and calcified thrombus.

Cardiac myxoma is the most common primary cardiac tumor in adults, accounting for approximately half of all primary benign cardiac tumors. Myxomas are typically located in the left atrium and attached to the interatrial septum by a peduncle; however, they can occasionally be found in the ventricles [3]. Dystrophic calcifications, caused by repeated episodes of hemorrhage, can sometimes be observed in myxomas, with the calcific components appearing hypointense on MRI scans. A key imaging feature distinguishing myxomas from thrombi and CATs is their enhancement following the administration of gadolinium-based contrast agents during MRI scans [5, 6].

CCMA is a rare condition characterized by central degenerative softening of calcifications in the mitral annulus [5]. CCMA is usually focal and located adjacent to the posterior mitral leaflet. Its appearance on MRI scans varies but is generally hypointense on T1- and T2-weighted images. On CT imaging, CCMA often appears as a well-defined, peripherally calcified mass near the mitral valve with variable attenuation. CCMA may exhibit subtle peripheral enhancement on late gadolinium-enhanced MRI scans, likely due to a thin fibrous cap. The diagnosis of MAC is confirmed by identifying a densely calcified lesion in the posterior mitral annular ring [5]. Notably, the primary difference between CCTA and CCMA lies in the lesions' specific locations.

A calcified thrombus should be considered in the differential diagnosis of CAT. Ventricular thrombus is typically associated with myocardial scars or severe left ventricular dysfunction. The left ventricular thrombus often exhibits a more linear shape and can be found along the endocardial surface at the mid-basal level of the left ventricle or mid-ventricle, even in the absence of a left ventricular aneurysm [7].

The MRI signal characteristics of a thrombus vary depending on its age. Chronic organized left ventricular thrombi generally appear as low-signal-intensity intracavitary masses, with or without calcifications, adjacent to a thinned and/or dysfunctional left ventricular wall [5]. Thrombus calcifications are usually limited to small foci, while diffuse calcifications are uncommon. Contrast administration during MRI scans can help differentiate thrombus from cardiac tumors, except for CAT. While contrast enhancement

is not expected in either thrombus or CAT, myocardial infiltration is a distinguishing feature seen in CAT [8]. Furthermore, the presence of atrial fibrillation or left ventricular dysfunction strongly suggests the diagnosis of a thrombus [5, 9].

Calcified Amorphous Tumor (CAT) is a non-cancerous cardiac tumor first described by Reynolds et al. in 1997 [1]. The clinical differential diagnosis of cardiac CAT includes both benign and malignant cardiac tumors, predominantly myxoma, as well as non-neoplastic processes such as thrombosis, embolism, and vegetation [1]. Echocardiography is instrumental in identifying the morphology, location, and echogenicity of intracardiac masses. Cardiac CATs are pedunculated and calcified, and they can be located in any of the four chambers, though they are most commonly found in the left ventricle [10]. In our case, the CAT was located in the atrioventricular (AV) groove and extended to the left ventricular wall.

Historically, the diagnosis of cardiac tumors relied on histological specimens obtained during post-surgical procedures or postmortem examinations. However, advancements in cardiovascular imaging—such as cardiac magnetic resonance imaging (CMR) and cardiac computed tomography (CT)—have greatly improved the characterization of masses and tissues. Used together, these modalities provide superior tissue characterization [11].

A CT scan demonstrating a partially or diffusely calcified mass is critical for the initial diagnosis of CAT [8]. In our case, the mass appeared hyperdense at the periphery with a hypodense core and showed no enhancement on the CT scan. CMR, with its higher contrast resolution, offers greater efficacy in characterizing these masses. Previous studies suggest that CATs may exhibit high, intermediate, or low signal intensities on both T1- and T2-weighted images [12].

The CAT in our case exhibited a heterogeneous appearance, with isointense signal intensity on T1-weighted images and iso to high signal intensity on T2-weighted images. Additionally, Balanced Steady-State Free Precession (bSSFP) sequences in 2-chamber, 4-chamber, and short-axis views demonstrated an isointense mass, while the delayed enhancement sequence showed no enhancement. This finding is consistent with observations in a few previous studies [13]. However, most CAT cases in prior studies displayed low signal intensity on both T1- and T2-weighted spin-echo sequences.

Yilmaz et al. retrospectively examined CAT configurations and shapes on MRI and CT scans in 12 patients, noting partial calcifications with hypodense or diffusely calcified masses on CT. On cardiac MRI, CATs exhibited a homogeneous appearance with low signal intensity on T1- and T2-weighted spin-echo sequences and showed no contrast enhancement after gadolinium injection in both early and delayed sequences [2]. Tian et al. presented a case in which CMR revealed a heterogeneous appearance with low signal intensity on both T1- and T2-weighted images. They suggested PET scans as a valuable tool for the early detection of cardiac tumors or metastases [14].

No specific features for CAT have been established on CMR, underscoring the importance of multimodality imaging for identifying cardiac CAT, determining treatment plans, and performing serial follow-ups post-treatment.

Data on CAT's pathophysiology, treatment, and prognosis remain limited. The majority of CAT cases are associated with valvular heart disease (31%), end-stage renal disease (21%), mitral annular calcification (MAC) (14%), or diabetes mellitus (14%) [15]. However, none of these conditions were present in our patient. Symptoms in patients with cardiac CAT include dyspnea, syncope, and embolism-related manifestations [2]. Reports have indicated that CAT can lead to cerebral infarction [4, 16]. The case we described involved a cardiogenic cerebral embolism caused by CAT in a young patient. The absence of alternative causes supports the role of CAT in this ischemic episode.

Yamato Nishiguchi et al. reviewed all previously reported cases of CAT-induced cerebral infarction up to July 31, 2020, identifying 17 cases [17]. They reported their own case in 2021 involving a 63-year-old female who presented with a visual field defect and achromatopsia, accompanied by brain infarction in the territories of the bilateral PCA and SCA due to CAT. She underwent surgical treatment and was successfully healed.

More recently, Yoshifumi Ogasawara et al. described a case of caseous calcification of the mitral annulus mimicking CAT, which led to multiple cerebral infarctions in a 73-year-old woman in Japan. She presented with dysarthria, and a head MRI revealed multiple acute cerebral infarctions in the bilateral cerebral and cerebellar hemispheres. Surgical resection of the mass and mitral valve replacement were performed [18].

Julie Lorraine Rosenthal et al. reported a 64-year-old male patient with acute vision loss. Echocardiography revealed a mobile echo density on the posterior

papillary muscle and echogenic myocardium. CT and MRI imaging confirmed that the echo density was associated with myocardial calcium. Post-surgical pathology supported the diagnosis of CAT [19].

To our knowledge, our case report about cerebral infarction due to CAT represents the 19th known case as of November 2024, and it is only the 4th case where medical treatment was chosen instead of surgery. Notably, most previously reported cases involved middle-aged or elderly patients with multiple underlying conditions that predisposed them to a hypercoagulable state. However, in our case, no prior medical history was identified

CAT is a potential source of embolism, necessitating careful monitoring. Surgical resection [4] or long-term anticoagulant therapy should be considered essential for management. Most previous studies suggest that surgical excision is mandatory for both diagnosis and treatment, with postoperative recurrence of CAT being rarely reported [20]. Benedetta Formelli et al. [21] managed a patient with cerebral infarction due to CAT conservatively, documenting no significant progression of the lesion over four years. This supports the feasibility of medical treatment, including anticoagulant therapy and close monitoring, as a valid option in cases where surgery poses high risks.

In our case, surgical excision was not feasible due to the severity and extent of pericardial involvement and the absence of risk factors for thromboembolism. The patient responded well to anticoagulant therapy, showing no recurrence during the six months following discharge.

What makes this case unique is the combination of extensive pericardial calcification and CAT, presenting in a young patient with no prior medical history or abnormal laboratory test results. Therefore, when surgical excision is deemed too risky or invasive, anticoagulation and long-term follow-up may serve as effective therapeutic alternatives. We also emphasize the importance of multimodality imaging for detecting cardiac CAT, aiding in treatment planning, and conducting serial follow-ups after treatment.

Conclusion

CAT is a potential source of embolism, requiring careful monitoring. Surgical resection or long-term anticoagulant therapy should be considered as necessary management options. Since no specific features for CAT have been established on CMR, the findings of this case, along with the literature review, emphasize the critical role of multimodality imaging in detecting cardiac CAT, formulating treatment strategies, and facilitating serial follow-ups for patients after treatment.

Acknowledgments

The author has nothing to report.

Ethical Considerations

Ethics Statement

This case report was conducted in accordance with the principles outlined in the Declaration of Helsinki.

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Conflict of Interests

The authors declare no known competing financial interests or personal relationships that could have influenced the work reported in this paper

Patient Consent

Written informed consent for the publication of this case report was obtained from the patient. The consent document will remain on file for our records.

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