

Radiological Case Report / Caso Clínico

Caseous Calcification of the Mitral Valve Annulus: A Case Report

*Calcificação Caseosa do Anel da Válvula Mitral: A Propósito de um Caso Clínico*João Rebelo¹, André Carvalho¹, Inês Portugal¹, Rui Cunha^{1,2}¹Serviço de Radiologia do Centro Hospitalar de São João, Porto, Portugal²Faculdade de Medicina da Universidade do Porto, Portugal

Address

João Ferreira Rebelo
 Serviço de Radiologia
 Centro Hospitalar de São João
 Alameda Prof. Hernâni Monteiro
 4200-319 Porto, Portugal
 e-mail: rebelo.jmr@gmail.com

Abstract

Annular calcification of the mitral valve annulus results from chronic degeneration of its fibrous valve annulus that may present as caseous calcification. This represents a rare and idiopathic variant whose lack of knowledge can lead to misdiagnosis.

The authors present a case report of caseous calcification of the mitral valve annulus, including its main imaging findings and differential diagnoses.

Keywords

Mitral valve; Ectopic calcification; Computed tomography; Magnetic Resonance.

Resumo

A calcificação caseosa do anel da válvula mitral resulta da degenerescência crónica do seu anel valvular fibroso que se apresenta sob a forma de calcificação caseosa. Esta representa uma variante rara e idiopática, cujo desconhecimento pode levar a diagnósticos equívocos.

Os autores apresentam um caso clínico de calcificação caseosa do ânulo da válvula mitral, incluindo os seus principais achados imagiológicos e diagnósticos diferenciais.

Palavras-chave

Válvula mitral; Calcificação ectópica; Tomografia computadorizada; Ressonância magnética.

Clinical History

A 72-year-old female patient was admitted to the Emergency Room (ER) after cardiorespiratory arrest due to ventricular fibrillation that was quickly assisted and reversed after four cycles of advanced life support. In the emergency room, an electrocardiogram (ECG) was performed, which demonstrated atrial fibrillation and ST-segment elevation in the lower leads and in V5-V6. The patient underwent emergent catheterization, where moderate to severe left ventricular dysfunction was observed, with marked hypokinesis of the apical segments and moderate of the remaining segments, with the exception of the basal joints which were hypercontractile. During the examination, the presence of a calcified image in close relationship with the lower basal myocardium was also identified.

After the procedure, which took place without complications, the patient was admitted to the Intensive Care Unit, with the diagnosis of acute inferolateral myocardial infarction and a calcified juxtacardiac mass of unknown origin. During hospitalization, a transthoracic echocardiogram (TTE) was performed, which confirmed the presence of a calcified mass, involving the posterior mitral annulus and the lower, posterior and lateral basal segments of the left ventricle (Fig. 1), raising the suspicion of caseous calcification of the mitral valve annulus.

Additional assessment by thoracic Computed Tomography (CT) (without ECG synchronization) documented a grossly calcified lesion in the topography of the mitral valve, measuring about 6x5x5 cm in major axes (transverse x anteroposterior x longitudinal), with no apparent involvement of its leaflets, projecting itself to the left ventricular cavity (Fig. 2). These findings were in

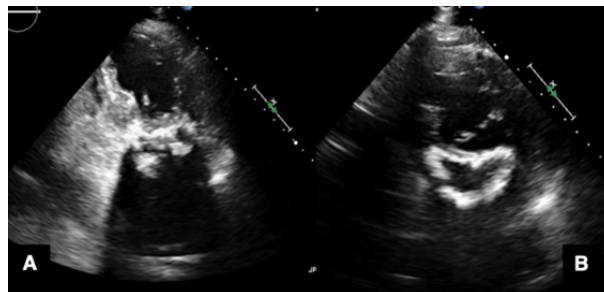


Fig. 1 – Transthoracic echocardiogram. In the topography of the mitral valve, a grossly calcified structure is observed, conditioning intense acoustic shadow (A), with a central area markedly hypoechogenic, in probable relationship with changes of liquefaction (B). Apparent extension to the left ventricular cavity.

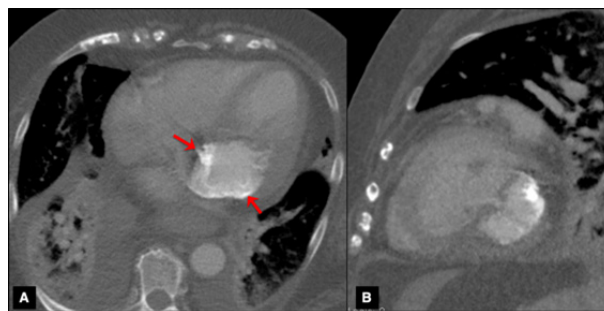


Fig. 2 – Computed Tomography (without ECG synchronization) with intravenous contrast, axial (A) and sagittal (B) images. A slightly hyperdense mass centered on the mitral valve annulus can be observed, showing a grossly calcified peripheral border (arrow). Bilateral pleural effusion and consolidation of the pulmonary parenchyma of the right lower lobe are also seen.

agreement with the echocardiographic changes previously described. The presence of calcifications to the extent described indicated a probably chronic etiology, being this presentation compatible with the diagnosis of caseous calcification of the mitral valve annulus.

Additionally, a Cardiac Magnetic Resonance (CMR) was performed, which confirmed the presence of a “mass” centered on the lower and lateral slope of the mitral annulus, with infiltration of the lower basal segment of the left ventricle, conditioning the reduction of its wall thickness. The mass showed intermediate signal in the T2-weighted sequences and hypointense with signal void at the periphery in the gradient echo sequences. Anomalous contrast uptake was not identified, however, there was a slight peripheral late enhancement in the PSIR (Phase-Sensitive Inversion Recovery) sequences, an aspect suggesting the presence of fibrosis (Fig. 3).

In retrospect, it was confirmed that this lesion was already evident in previous chest radiographs, with relative imaging stability over time (Fig. 4).

The case was presented and discussed in a multidisciplinary meeting, in order to assess the need for surgical excision of the mass. Given the extension of the calcifications and because it was an asymptomatic lesion, it was decided not to proceed with valve surgery.

Currently, the patient is asymptomatic, with no complaints of dyspnea or chest pain, stable and with a NYHA (New York Heart Association) class II heart failure.

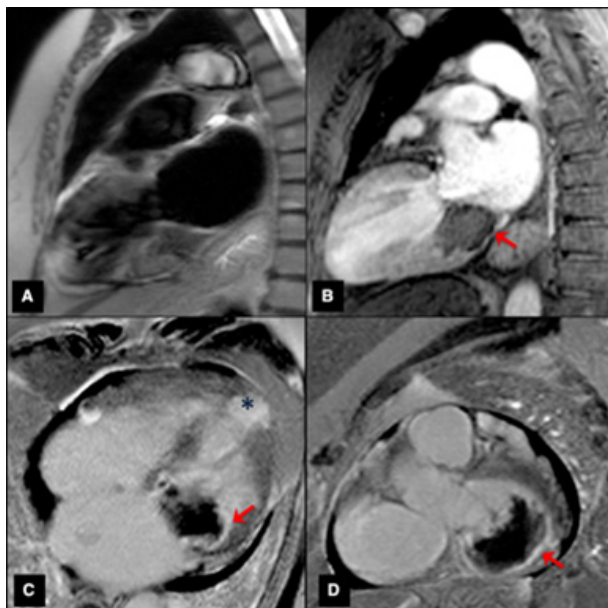


Fig. 3 – Cardiac Magnetic Resonance with gadolinium administration. (A) T2 HASTE (Half Fourier Acquisition Single Shot Turbo Spin Echo) sequence in vertical long-axis (B). Cine-SSFP sequence in vertical long-axis and late enhancement images after gadolinium (PSIR) in horizontal long-axis (C) and short axis (D). A hypointense mass of expansive characteristics is documented, centered on the mitral valve annulus, with a void peripheral signal reflecting the presence of calcification (arrow B) that becomes much more evident in late enhancement images (arrows C and D). Apical transmural apical enhancement is also present, meaning myocardial infarction (star C). Small volume pericardial effusion.

Discussion

Most cardiac masses are detected incidentally during routine tests performed for other indications.^{1,2,3} Caseous calcification of the mitral valve annulus is a chronic degenerative process that usually involves the posterior

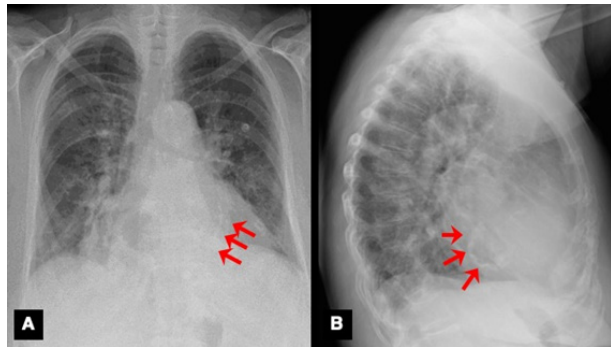


Fig. 4 – Chest radiography, face (A) and profile (B). As indicated by the arrows, more evidently in the profile view, a grossly calcified “mass” is observed in the expected topography of the mitral valve. Differential diagnoses could include rheumatic valve disease, caseous calcification of the mitral valve or even tumor pathology with calcifications on it.

ring.^{1,2,4,5} The terminology is peculiar, since the term caseous is generally associated with a type of necrosis found in the context of tuberculosis.² It is more prevalent in elderly women and in patients with chronic kidney disease, as in the case described. Patients on hemodialysis or with metabolic disorders that induce changes in normal calcium homeostasis appear to induce an increased risk.^{1,2} Its pathogenesis is not yet fully understood, but due to its association with atherosclerotic pathology, it presents risk factors similar to other cardiovascular diseases.^{1,2,3} Most patients are asymptomatic, and although it is predominantly a benign process, in some cases it can condition mitral valve dysfunction, due to chronic mitral regurgitation or in rarer cases due to mitral stenosis.^{2,3,6} In the present case, the presence of associated severe mitral regurgitation was a feature.

The mitral annulus has a very particular location, being close to the atrioventricular node and the conduction system, consequently pathology in this topography can give rise to bradyarrhythmia or other changes in the normal rhythm. Rare cases of systemic embolization have also been described in the literature.^{2,3} During patient follow-up, no rhythm abnormalities or embolic phenomena were detected.

Transthoracic echocardiography (TTE) is considered the most reliable method for such diagnosis. Usually, it translates into a round mass of large dimensions and well-defined limits, presenting peripheral hyperechogenicity and central hypoechoic areas corresponding to areas of liquefaction.^{3,5,7} The reflexivity of the calcifications is lower compared to typical calcifications, generally without an associated posterior acoustic shadow effect.^{1,3,5} Due to its low incidence and its expansive presentation, as a pseudo-mass, the caseous calcification of the mitral valve annulus can be wrongly interpreted as a neoplastic process. The presence of some findings favors the diagnosis of benignity, namely its typical location, with possible extension to the entire mitral annulus, the presence of well-defined limits and the existence of a central hypoechoic area.^{2,3} In doubtful situations, an additional imaging evaluation is recommended for adequate characterization of the lesion, risk stratification and eventual surgical planning, namely through CT and MRI.

Cardiac CT is used as a second-line diagnostic modality in the assessment of cardiac masses. Recent technological advances, namely the introduction of a greater number of detectors and the possibility of making acquisitions with electrocardiographic synchronization, have resulted

in a better spatial resolution of cardiac structures with this imaging technique. CT also provides some additional information, like the presence of calcifications (being better than MRI in this evaluation), fat or vascularization.^{2,3,6}

CMR can be used to predict the malignancy of a cardiac mass. In comparison with computed tomography, magnetic resonance imaging offers greater temporal resolution and better tissue characterization, without exposure to ionizing radiation. However, accessibility to this imaging technique remains limited when compared to other techniques.^{2,3,6,8}

Caseous calcification of the mitral valve annulus is translated into Cardiac CT by a well-defined hyperdense mass, oval or crescent-shaped, with peripheral calcifications and generally along the posterior mitral annulus, with no evidence of contrast uptake.^{1,2,5} Central hypodensity is believed to be secondary to the liquefied material that fills the center of the lesion.^{2,3,5,6}

In the presented case, ECG synchronization or images without contrast were not used, due to technical impossibility, aspects that could affect the assessment of cardiac structures. However, given the dimensions, the location and the high density demonstrated by the mass, compatible with calcium content, the most likely diagnosis of caseous calcification of the mitral valve annulus remained.

In CMR, it presents itself as a well-defined mass, which at T1 shows central hypersignal and peripheral hyposignal. In T2-weighted sequences, exhibit central hyposignal, but with a hypersignal annulus relative to the adjacent myocardium. It does not show uptake of paramagnetic contrast, although sometimes there is late enhancement indicating associated fibrosis.^{1,2,3,5,6,8}

Findings of the present case, although not pathognomic, are very suggestive of caseous calcification of the mitral valve. Thus, given the low probability of other etiologies, namely malignant ones, and given the clinical stability of the patient, it was decided to choose surveillance in consultation instead of invasive procedures such as biopsy or surgical excision. The good general condition of the

subject, without significant repercussions, is adequate for the proposed diagnosis.

Because it is an uncommon pathology and unfamiliar to radiologists not dedicated to the area of cardiac radiology, it can be potentially misinterpreted. The main differential diagnoses to consider include other intracardiac masses, such as tumors (most commonly myxoma), abscesses or vegetations.^{2,3,6}

Myxoma is a common intracardiac lesion, with distinct characteristics, presenting as a single pedunculated mass, originating in the oval fossa, and projecting to the left auricle.^{2,3}

In atypical cases, it may demonstrate irregular morphology and heterogeneous texture, leading to the need of resorting to other techniques as a complement to the echocardiography. In CMR, it exhibits isosignal in T1 and hyposignal in T2. The use of cardiac gating is useful in the study of myxomas, due to their high mobility.^{2,3}

Other unusual tumors to consider include mitral valve hemangioma and malignant lesions such as leiomyosarcoma.

In the ultrasound study, hemangiomas have hypoechogenic areas that represent tumor vascularization and that extend peripherally, with no calcifications identified in their dependence. On the other hand, leiomyosarcoma is more heterogeneous, with areas of necrosis and calcifications.^{2,3}

Infectious pathology should always be included in the list of differential diagnoses, and intracardiac abscesses appear as collections within the myocardium / annular region, with homogeneous echogenicity on the echocardiogram. These do not show calcifications; however, they can demonstrate peripheral flow at Doppler study.^{2,3}

In summary, caseous calcification of the mitral valve annulus is a rare and benign pathology, associated with a good prognosis and, in most cases, with no need for targeted therapy. Although it has well-established imaging diagnostic criteria, it is still unknown to many cardiologists and radiologists, which can lead to possible incorrect diagnostic interpretations and unnecessary surgical procedures.

Received / Recebido 24/11/2019

Acceptance / Aceite 26/03/2020

Ethical disclosures / Divulgações Éticas

Conflicts of interest: The authors have no conflicts of interest to declare.

Conflitos de interesse: Os autores declaram não possuir conflitos de interesse.

Financing Support: This work has not received any contribution, grant or scholarship.

Suporte financeiro: O presente trabalho não foi suportado por nenhum subsídio ou bolsa.

Confidentiality of data: The authors declare that they have followed the protocols of their work center on the publication of data from patients.

Confidencialidade dos dados: Os autores declaram ter seguido os protocolos do seu centro de trabalho acerca da publicação dos dados de doentes.

Protection of human and animal subjects: The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).

Proteção de pessoas e animais: Os autores declaram que os procedimentos seguidos estavam de acordo com os regulamentos estabelecidos pelos responsáveis da Comissão de Investigação Clínica e Ética e de acordo com a Declaração de Helsínquia da Associação Médica Mundial.

References

1. Mallat N, Limeme, M, Zaghoulani H, Mootemri F, Majdoub S, Amara H. Caseous calcification of the mitral annulus on MDCT: a rare intracardiac mass. *Acta Radiologica Short Reports*. 2013;2.
2. Elgendy, I. Y. and Conti, C. R. Caseous calcification of the mitral annulus: a review. *Clin Cardiol*. 2013;36:E27-E31.
3. Caruso F, Ventura L, Viliari D, Della Sala SW. Massive mitral annular calcification mimicking intracardiac mass: multimodality approach to diagnosis. *Radiol Case Rep*. 2018;13:376-9.
4. Hari Y, Hiraoka A, Yositaka, H. et al. *Int J Cardiovasc Imaging*. 2018;34:1471.
5. Assunção FA, Oliveira DCL, Santos AAD, Nacif, MS. Caseous calcification of the mitral annulus: computed tomography features. *Radiologia Brasileira*. 2016;49:273-4.
6. Ribeiro S, Salgado A, Salomé N, Bettencourt N, et al. Caseous calcification of the mitral annulus: a multi-modality imaging perspective. *Rev Port Cardiol*. 2012;31:313-6.
7. Lopez S, Clavaud M, Elbeze JP, Lena, D. Isolated caseous calcification of the mitral valve. *J Card Surg*. 2018;33:142-3.
8. Ghazawi FM, Vinco G, Walker M. Caseous calcification of the mitral annulus: a rare entity confirmed by cardiovascular magnetic resonance imaging. *Int J Cardiovasc Imaging*. 2018;34:25-6.