

Caseous Degeneration of the Mitral Annulus Associated with Severe Mitral Regurgitation

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Introduction

The caseous degeneration of the mitral annulus (CDMA) is a typically benign entity that is rarely seen in images. It accounts for about 0.5% to 1% of mitral annulus calcifications and its etiology is not well understood^{1,2}. It usually occurs in women older than 70, being an important differential diagnosis of cardiac tumors, thrombi, cysts or abscesses¹. In this paper, we report a case of CDMA associated with severe mitral regurgitation requiring surgical intervention.

Case Report

Female patient, 73 years old, hypertensive, was admitted in the emergency room with intense dyspnea associated with retrosternal discomfort. After the cardiologist's assessment, transthoracic echocardiographic investigation was conducted. Transthoracic echocardiogram showed severe dilation of the left atrium (62 mm) and mild eccentric myocardial hypertrophy. Left ventricular systolic function was preserved with no abnormalities in segmental myocardial contractility. The aortic and tricuspid valves presented thickening and discrete deposit of calcium with a slight degree of failure. The mitral valve was thickened with calcification of the posterior cusp and annulus. Doppler test revealed severe mitral insufficiency. Maximum systolic pulmonary artery pressure was estimated at 59 mmHg (calculated by tricuspid regurgitation and right atrial pressure). The ascending aorta and the pericardium showed normal echocardiographic aspects.

The transthoracic study revealed a hyperechoic mass (29 x 26 mm), non-mobile, with a slightly irregular surface, with a hypoechoic area inside, compromising the basis of the posterior cusp and the mitral annulus (posterior and lateral left area) suggestive of CDMA (Figure 1).

Keywords

Heart Valve Diseases/physiopathology; Mitral Valve/physiopathology; Mitral Valve Insufficiency/physiopathology; Echocardiography; Heart Atria/abnormalities.

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Manuscript received May 22, 2015; revised manuscript June 1, 2015; accepted June 10, 2015.

DOI: 10.5935/2318-8219.20150033

Magnetic resonance imaging (MRI) of the heart was later performed to complement the diagnostic investigation. MRI revealed enlarged left atrium, severe mitral regurgitation, ejection fraction of 68% and hypointense intracardiac mass located in the mitral valve plane affecting the annulus and the posterior cusp (Figure 2). Coronary angiography showed coronary arteries free of significant lesions and ventriculography was not performed.

Considering the severe valve impairment, the patient underwent surgery (Euroscore = 10.4%) to replace the mitral valve for biological prosthesis n° 29 Braile. The caseous "toothpaste-like" material was present in the mitral annulus and stretched from the annulus to the free wall of the left ventricle (Figure 3).

After surgical cleaning of all the caseous material, considerable weakening of the mitral annulus and thinning of the left ventricular wall was found, being necessary to strengthen it with stitches anchored in bovine pericardium strips to attach the prosthesis safely. Cardiopulmonary bypass was performed conventionally with antegrade myocardial protection and systemic hypothermia of 30 °C with total time of 140 minutes.

Postoperative echocardiography revealed moderate impairment in the overall left ventricular systolic function, normal functioning prosthesis and residual thickening with calcification at the base of the posterior cusp and annulus. The patient required circulatory support with intra-aortic balloon and temporary transvenous pacemaker due to complete atrioventricular block. The patient later presented episodes of atrial fibrillation, hemodynamic instability and death after 48 hours of surgery. Histopathological examination revealed mitral valve with fibrosis and calcification, as described in the literature.

Discussion

CDMA is a chronic degenerative process mainly involving the posterior annulus³. Calcification of the mitral annulus is observed in the autopsy of 3% to 8% of the general population^{1,3}, but it is rarely seen in images³. The largest series in the literature analyzed 18 cases⁴. The echocardiographic prevalence is of 0.6% in patients with mitral annular calcification and 0.06%-0.07% in patients of all ages⁵.

CDMA is considered a form of expression of atherosclerotic disease associated with hypertension, coronary artery disease and aortic atheromatosis¹ with the same risk factors identified for cardiovascular disease⁶.

Generally, the presence of CDMA is not accompanied by symptoms. However, most often, the symptoms are due

Case Report

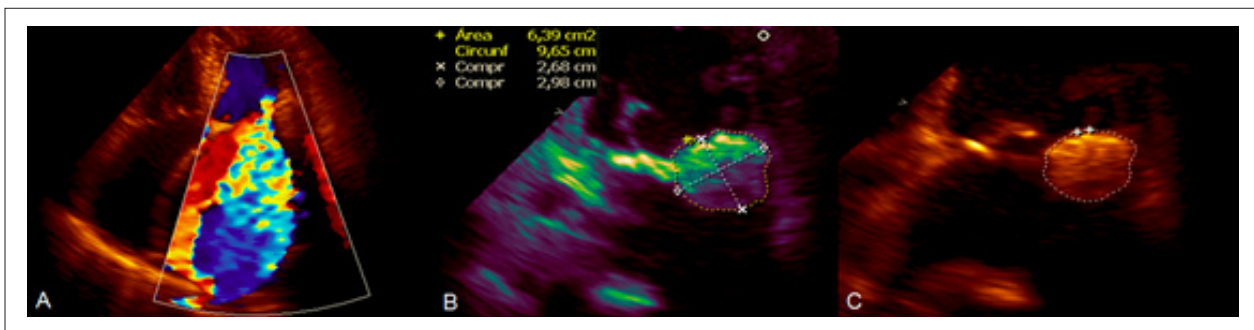


Figure 1 – Transthoracic echocardiography: A: severe mitral insufficiency; B: Hyperechoic mass measurements; C: Hyperechoic mass, non-mobile, of slightly irregular surface with a hypoechoic area inside, compromising the basis of the posterior cusp and mitral annulus.



Figure 2 – Magnetic resonance imaging of the heart: A: 4 chambers in Cine-MRI — severe mitral regurgitation (arrow) and hypointense mass (*); B: 3 chambers in Cine-MRI — hypointense mass in the posterior annulus (arrow); C: delayed enhancement sequence with contrast at the edge of the mass (arrow).

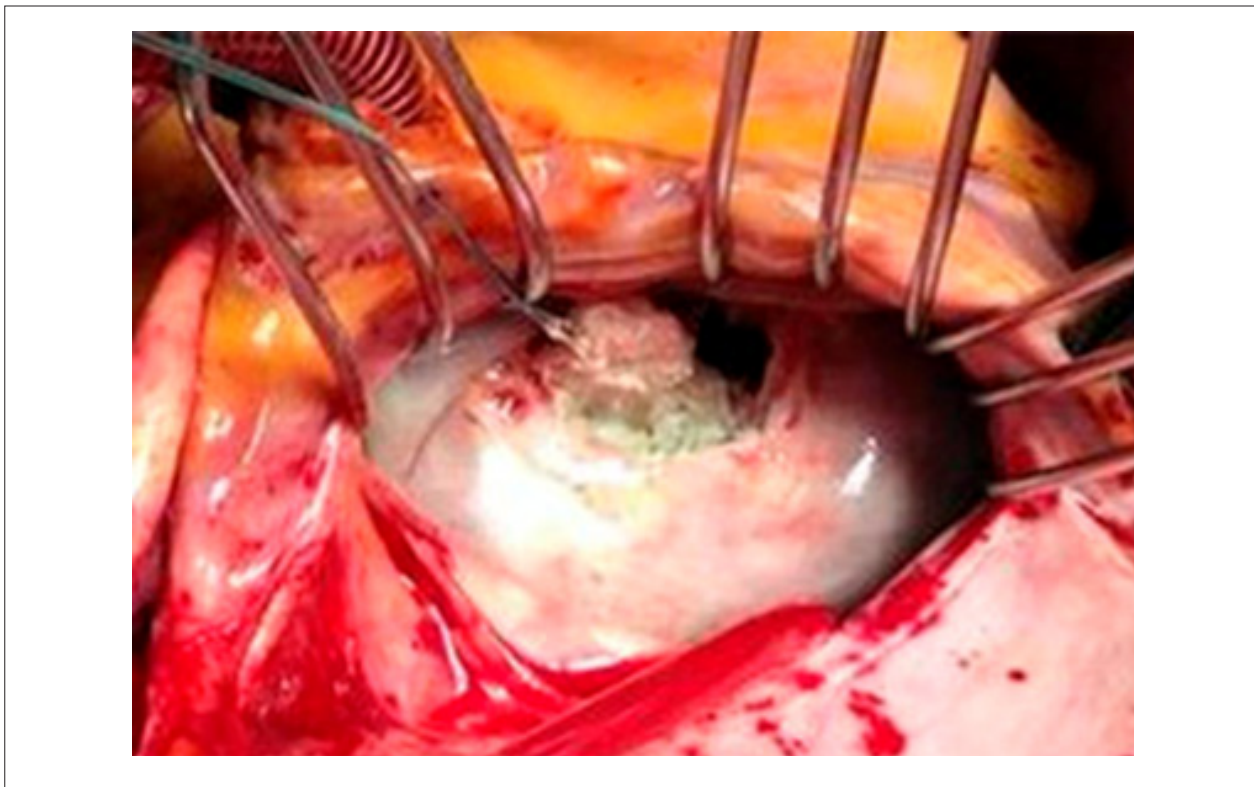


Figure 3 – “Toothpaste-like” intraoperative image characterizing CDMA, extending from the mitral annulus to the free wall of the left ventricle.

to valve impairment, such as dyspnea secondary to mitral insufficiency or mitral stenosis. Embolic phenomena are rare, but can also occur first¹.

Transthoracic echocardiography, in most cases, is sufficient for the diagnosis of CDMA. When in doubt, the investigation can be complemented with transesophageal echocardiography^{1,3,6} or MRI^{7,8}. The typical echocardiographic image is a large hyperechoic rounded semilunar-shaped mass of heterogeneous aspect with an echolucent area inside, usually located in the posterior annulus of the mitral valve consistent with the findings in this paper^{1,3,6}. Sometimes, the discovery of intracardiac masses is an accidental echocardiographic finding, but may arise during follow-up of patients with heart failure or thromboembolic phenomena⁶. The differential diagnosis of masses visible in the cardiac cavities includes thrombi, abscesses, cysts, vegetation and tumors^{1,3,6}. The distinction with abscesses is made by the clinical picture with benign evolution and typical location in the posterior annulus, since the abscesses are usually located in the intervalvular mitroaortic fibrous portion. The tumors have no central echolucency as observed in the case of CDMA¹.

MRI can assist in the differential diagnosis of intracardiac masses and in the research of involvement of surrounding structures. The characteristics that suggest the CDMA to the method are: hypointense (dark) images in the dark blood T1 and T2-weighted sequences with fat suppression (compatible with calcification); hypointense mass in relation to the myocardium in Cine-MRI sequences; absence of mass perfusion and peripheral contrast enhancement with central core without contrast in late enhancement sequences (post-contrast T1)^{7,8}. All these characteristics were present in the patient's test.

It is important to suspect of this disease and perform the correct differential diagnosis, since CDMA is a benign condition, which does not imply indication for surgery⁶. Surgery should be reserved for symptomatic cases of severe valvular dysfunction or patients with cerebral embolism related to calcified lesion^{1,3,6}.

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When surgery is performed, it identifies a calcified lesion, usually around a central region filled with a "toothpaste-like" material made up of calcium, fatty acids and cholesterol^{3,6}. Histological test reveals a periannular calcification predominantly with acellular substances, negative and free of cancer or inflammatory cells⁹.

Complications of calcification of the mitral annulus, such as secondary infections, arrhythmias, failure or significant mitral stenosis and stroke have been rarely reported. While CDMA may be present in 25% of individuals who have had a stroke, the direct relationship of CDMA with embolic phenomena is questionable⁶.

CDMA is benign per se with good long-term prognosis. Therefore, it is imperative to recognize this disease on imaging studies to deliver an accurate differential diagnosis, so that surgical indication is accurate and only intended for significantly affected patients as described in this paper.

Authors' contributions

Research creation and design: Bohatch Jr MS, Dietrich A; Data acquisition: Bohatch Jr MS, Dietrich A, Fiamoncini A, Azevedo GSA, Varella EL; Data analysis and interpretation: Bohatch Jr MS, Dietrich A, Azevedo GSA, Di Giovanni FJ; Manuscript drafting: Bohatch Jr MS, Di Giovanni FJ; Critical revision of the manuscript for important intellectual content: Bohatch Jr MS, Dietrich A, Fiamoncini A, Azevedo GSA, Varella EL, Di Giovanni FJ.

Potential Conflicts of Interest

There are no relevant potential conflicts of interest.

Sources of Funding

This study had no external funding sources.

Academic Association

This study is not associated with any graduate program.

Case Report
