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.²⁻³ It usually occurs in women older than 70, being an important differential diagnosis of cardiac tumors, thrombi, cysts or abscesses. 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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Discussion

CDMA is a chronic degenerative process mainly involving the posterior annulus.¹ Calcification of the mitral annulus is observed in the autopsy of 3% to 8% of the general population,¹ 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 atheromatosis 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...
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 first.

Transthoracic echocardiography, in most cases, is sufficient for the diagnosis of CDMA. When in doubt, the investigation can be complemented with transesophageal echocardiography or MRI. 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. Sometimes, the discovery of intracardiac masses is an accidental echocardiographic finding, but may arise during follow-up of patients with heart failure or thromboembolic phenomena. The differential diagnosis of masses visible in the cardiac cavities includes thrombi, abscesses, cysts, vegetation and tumors. 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 interventricular mitroaortic fibrous portion. The tumors have no central echolucency as observed in the case of CDMA.

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 (postcontrast T1). 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 surgery. Surgery should be reserved for symptomatic cases of severe valvular dysfunction or patients with cerebral embolism related to calcified lesion.

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. Histological test reveals a periannular calcification predominantly with acellular substances, negative and free of cancer or inflammatory cells.

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 questionable.

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.

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There are no relevant potential conflicts of interest.

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**References**


