Caseous Calcification of the Mitral Annulus

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Introduction

Calcification of the Mitral Annulus (CMA) is a chronic and degenerative process that is easily recognized by transthoracic echocardiography (TTE). It is a common finding in the elderly, but it may also occur in young patients with advanced kidney disease and other disorders related to abnormal calcium metabolism, or in those with severe mitral valve prolapse[1-3]. Caseous Calcification of the Mitral Annulus (CCMA) is a lesser known entity of the CMA, defined as a rounded mass with hypoechoic center composed of a mixture of fatty acids, cholesterol and calcium[3-4]. This variant may be mistakenly confused with other diagnoses such as myocardial abscess, tumors and thrombi[5-7]. Below,we present the morphological findings on echocardiography and the clinical profile of nine patients diagnosed with caseous calcification of the mitral annulus between August 2009 and February 2011 in our institution.

Case Report

Between 2009 and 2011, nine patients were diagnosed with CCMA on TTE at the Echocardiography Service of Hospital Pró Cardíaco. No patient underwent Transesophageal Echocardiography (TEE). The age of those patients was 72 to 97 years, including eight women, six hypertensive patients and four diabetic patients. Among the patients studied, there was no history of chronic kidney disease. Among these patients, two underwent outpatient examination. The other seven were admitted at the emergency department for various reasons, such as: stable angina, atypical chest pain, fever, and dyspnea. Four had a history of coronary artery disease. In none of the patients blood cultures were performed. One patient with stable angina underwent coronary angiography and was diagnosed with multivessel coronary disease and underwent coronary artery bypass grafting. None of the nine patients underwent mitral valve replacement.

In all patients, the echocardiographic findings consistent with CCMA were related to the posterior annulus and most of them caused no restriction to the opening of the cusps. However, in three patients there was reduction of the posterior mitral cusp mobility and in one patient the maximum diastolic gradient was found to be 18 mmHg and the mean gradient was 9 mmHg, with mitral valve area of 1.3 cm^2 calculated by PHT. In another patient, the maximum gradient found was 8.14 mm Hg and the mean gradient was 2.27 mmHg and mitral valve area of 2.08 cm^2 estimated by PHT. In the third patient, the decreased mobility of the posterior cusp showed no significant change in the transmitral flow. In these three patients, restriction on the movement of the posterior cusp was due to the mechanical restriction on its opening, rather than by secondary problems of cusp anatomy.

Discussion

The true prevalence of CCMA is not known. In 1970, through a study of necropsy series, Pomerance reported the incidence of this variant in approximately 3% of 258 patients older than 50, diagnosed with CMA[1]. Harpaz et al.[8] used TTE to demonstrate this variant in 0.63% of 3,007 patients with CMA. Considering the incidence on 28,384 patients (with or without CMA), the occurrence of CCMA was 0.067%. Kronzon et al.[4] also found that entity in 0.055% of nine thousand TTE performed by the laboratory until the date of their study. In this study, nine cases of CCMA were found between August 2009 and February 2011. As the occurrence of CCMA is rare, the way to calculate the incidence may greatly affect the results. Considering the period between the first and the last case of the study (19 months), the calculated incidence is 0.084%. However, if we consider all examinations (20,271 TTE) from 2009 to 2011 (36 months), the calculated incidence is 0.044%. Note that, regardless of the calculation method, the values found are very close to the values reported in the literature. The cases reported are also consistent with the literature in terms of age and gender. The occurrence is more common in the elderly and women and usually presents a benign evolution.

The differential diagnosis of rounded and hypoechoic structures adjacent to the left atrioventricular annulus includes lipomatous infiltration of the atrioventricular annulus, infected mitral calcification, dilated coronary sinus, large aneurysm of the circumflex coronary artery, bulky lymph nodes, tumors, abscesses and thrombi[3,9,10]. CMA with prominent coronary sinus may also mimic CCMA. In fact, the dilated coronary sinus can be seen in the left atrioventricular annulus as a rounded structure with a hypoechoic central area. Its location within the pericardium is crucial to differentiate it from other vessels. An echo-free rounded structure above the posterior mitral annulus and close to the left atrial posterior wall is the finding that best characterizes persistent left superior vena cava draining into the coronary sinus.

Keywords

Calcinosis; Heart; Echocardiography; Mitral Valve; Calcification; Heart Valve Diseases.

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As described by Deluca et al.[3], an additional transthoracic image using saline solution injected into a peripheral vein of the left arm confirms the presence of dilated coronary sinus with persistent left superior vena cava. The descending aorta appears in the long axis parasternal view as an echo-free rounded structure immediately behind the mitral annulus and left atrium.

In contrast to the dilated coronary sinus, the aorta is seen outside the pericardium. A large aneurysm of the circumflex coronary artery with thrombus formation seen on parasternal long axis view on TTE or two-chamber view on transesophageal echocardiography (TEE) may also mimic CCMA. An abscess in the posterior mitral annulus can be confused with CCMA and its distinction depends on the clinical presentation, absence of a large amount of calcification, and systolic flow is often seen on color Doppler in the abscess cavity.

Large lymph nodes in the mediastinum may also be rare causes of rounded structures adjacent to the left atrioventricular annulus and can be found in patients with lymphoproliferative disease. The sonographic image of the lymph node tissue is hypoechoic with undefined edges. The lymph nodes are seen as multiple rounded structures of low density in the left atrioventricular annulus behind the left atrial posterior wall.

Other ecoluscent or solid structures can be considered, such as cysts, benign or malignant tumors, with compression or infiltration of normal anatomic structures[3]. Therefore, it is important to consider other imaging methods such as nuclear magnetic resonance and computer tomography scans to complement TTE in the diagnosis of CCMA.

The CCMA can be defined by echocardiography as a large, rounded mass with a hyperechoic peripheral annulus with defined edges, located in the periannular region without causing acoustic shadowing artifact, containing a hypoechoic central area composed of liquefied material[8]. The diagnostic is generally obtained by TTE. However, some studies consider that TEE is important to better evaluate the mass and its location[11].

When there is perforation of the CCMA, a large amount of milky material, resembling toothpaste, drain off the mass. The existence of large and dense calcified deposits in the left atrioventricular annulus with significant artifact of acoustic window, allows ruling out the diagnosis of CCMA.

Situations of CCMA are usually found randomly, since there are no reports of related symptoms[9]. Since it is an abnormality with benign evolution, most cases require clinical follow-up only[12]. Surgery is rarely recommended, but it should be considered in the case of concomitant valvular dysfunction[13].

According to Harpaz et al.[8], at five years follow-up, 43% of patients diagnosed with CCMA revealed different TTE images at the initial examination, making this entity a mutable condition. There are also literature reports of spontaneous CCMA resolution in renal patient after reduction of calcium concentration in hemodialysis. The authors assume that acute changes in serum calcium level should have contributed to the dissolution of calcium mass[14]. Novaro et al.[15] described the case of a 78-year-old woman with CMA; three months later, she presented lethargy and increased serum calcium concentration and reduced parathyroid hormone values, and the new TTE, in this context, revealed the caseous transformation of calcification. Thus, we should think of CCMA as a dynamic, not static, entity.

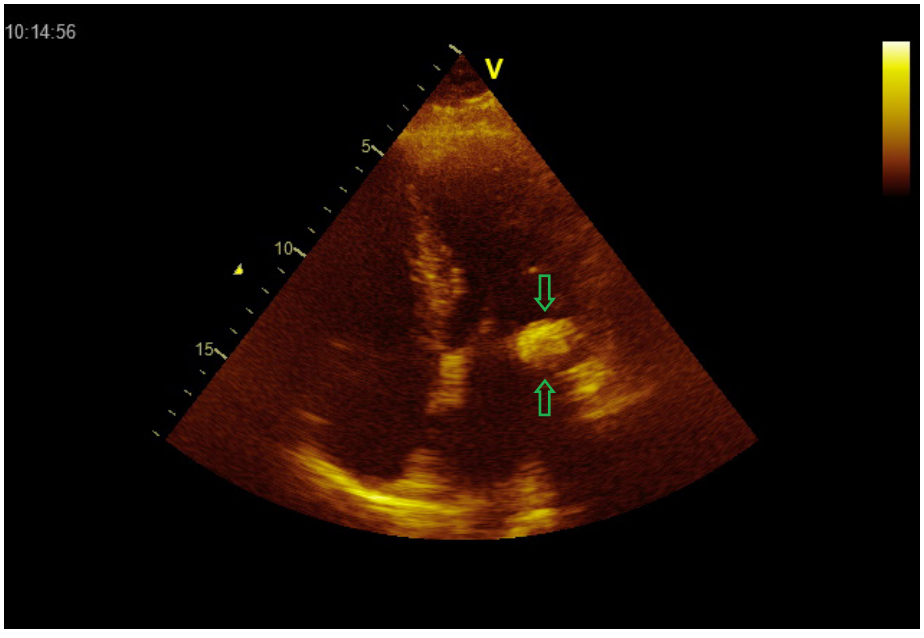

Figure 1 – *Apical 4-chamber view showing caseous calcification in the posterior annulus of the mitral valve.*

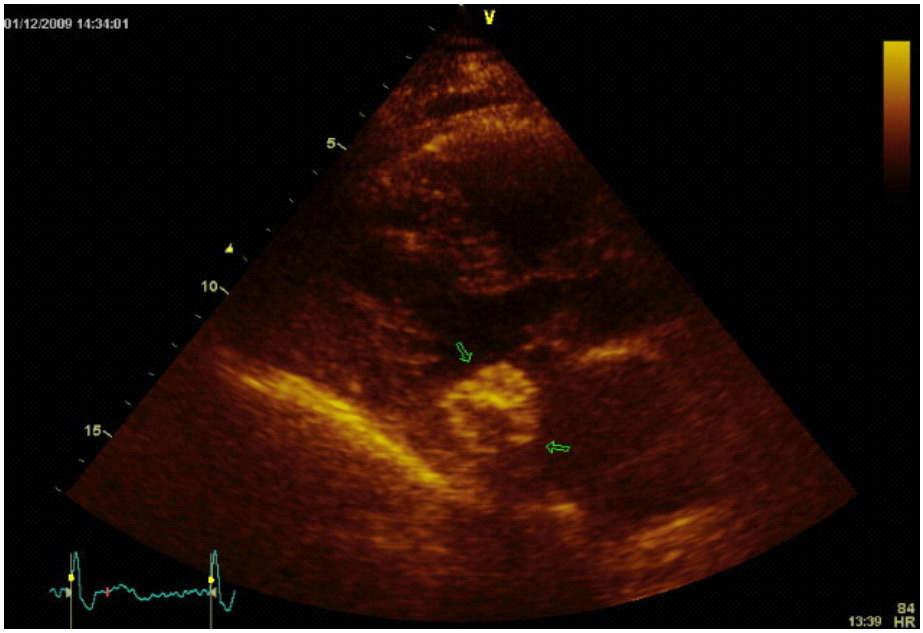

Figure 2 – *Longitudinal parasternal view showing caseous calcification of the mitral annulus.*

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