Brief report

Caseous Calcification of the Mitral Annulus, a Rare Echocardiographic Finding

Nora García-Ibarrondo* and Roberto M. Lang

* Servicio de Cardiología, Hospital de Basurto, Bilbao, Vizcaya, Spain
Noninvasive Cardiac Imaging Lab, University of Chicago Medical Center, Chicago, Illinois, United States

Article history:
Received 3 November 2010
Accepted 22 December 2010
Available online 23 May 2011

Keywords:
Echocardiography
Mitral valve
Calcium

ABSTRACT

Caseous calcification of mitral annulus, also known as caseoma, is a rare echocardiographic finding. Because it often presents asymptptomatically, the diagnosis is usually incidental. The condition arises due to caseous degeneration in the internal material of the mitral annulus calcification. On echocardiography, the calcification is a round mass with a central echolucent area, typically located at the base of the posterior leaflet, and can be mistaken for cardiac tumors or abscesses. We describe 3 patients with the condition, one of whom required surgical removal of the mass to obtain a definitive diagnosis.

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INTRODUCTION

Caseous calcification of the mitral annulus (CCMA) is a rare variant of mitral annulus calcification, a chronic degenerative process that occurs at advanced ages, particularly in women, hypertensive individuals, and patients with chronic renal failure or calcium metabolism abnormalities. The condition usually arises in the basal area of the posterior mitral valve and can extend over the entire annulus.1,2

Caseous calcification of the mitral annulus represents a rare course of the process, in which caseous degeneration of the internal material occurs. It is composed of a calcified rounded covering and, in its interior, a “toothpaste-like” mixture of calcium, fatty acids, and cholesterol.1,2

METHODS

We present 3 patients with the condition, one of whom required surgical resection of the mass to obtain a definitive diagnosis.

RESULTS

Patient 1

A 66-year-old woman with a history of hypertension, hyperlipidemia, and coronary artery disease as well as percutaneous revascularization of the anterior descending artery was admitted for new angina symptoms upon minimal exertion. Coronary angiography confirmed severe, diffuse disease in the anterior descending artery and first diagonal branch that precluded percutaneous revascularization. The patient was scheduled for surgical revascularization.

Transsthoracic echocardiography (TTE) showed a rounded mass of 15 × 16 mm in the posterior mitral annulus that extended to the most basal area, the P1-P2 scallops, without causing valve stenosis and with only minimal mitral regurgitation (Figs. 1A and B).

Because a cardiac tumor was initially suspected and the patient was scheduled for revascularization surgery, a decision was made to remove the mass during the operation. During the procedure, an incision was made and a whitish pasty content was removed (Figs. 2C and D).

Patient 2

A 72-year-old woman with a prior history of hypertension, asthma, and deep vein thrombosis came to the hospital for fatigue and nonspecific dizziness in the previous 3 weeks.
At admission, paroxysmal complete atrioventricular block was diagnosed and a dual-chamber pacemaker was implanted during hospitalization.

The TTE showed a large calcified mass of $30 \times 30$ mm in the left atrium (Figs. 2A and B) that caused moderate valve stenosis with a mean pressure gradient of 6 mmHg. Cardiac magnetic resonance imaging showed that the lesion was located at the base of the posterior leaflet, extending medially toward the most apical portion of the interatrial septum and to the interventricular membranous septum. The lesion was hypointense with respect to the myocardium in the T1- and T2-weighted sequences and gradient echocardiography (Fig. 2C), and only presented late-phase
enhancement in the surrounding capsule. These findings, as well as
the presence of a calcified envelope in the cardiac computed
tomography scan (Fig. 2D), confirmed the suspicion of caseous
calcification of the mitral annulus.

The symptoms resolved once the pacemaker was implanted and
therefore a wait-and-see approach was taken.

Patient 3

A 71-year-old woman with a history of hypercholesterolemia and
complete hysterectomy for an ovarian tumor had experienced
2 episodes of syncope in the last few months, with no obvious
accompanying sweating or nausea. The electrocardiogram (ECG)
showed a new bifascicular block. No episodes of advanced
atrioventricular blocks or significant pauses were observed in
the 24-h Holter-ECG monitoring.

The study was completed with TTE, which revealed a rounded,
calcified mass of 15 × 14 mm at the base of the posterior mitral
leaflet, extending medially toward the interventricular septum but
without causing significant valve stenosis or regurgitation (Figs. 3A
and B).

DISCUSSION

Echocardiographic series report a low prevalence of CCMA,
between 0.06% and 0.07%, and up to 0.6% of patients with
calcification of the mitral annulus (CMA).1 The prevalence in
autopsies is much higher and is 2.7% of the population studied.

The condition is probably underdiagnosed because of the
asymptomatic course in most patients, although there have been
several reports of CCMA associated with mitral valvular
dysfunction, both stenosis4 and regurgitation, due to the structural
deforation it causes.

In our series, we observed a high frequency of conduction
abnormalities: one of our patients presented advanced atrioven-
tricular block, and another, the third patient, had a bifascicular
block with repetitive syncopes. Although this association has been
previously described in CMA, there is no evidence regarding a
specific association with CCMA, probably due to a low incidence.

The association between CMA and other diseases such as atrial
fibrillation, atherosclerosis, and coronary artery disease has been
widely studied. In the case of conduction system diseases, the
association is 26% to 70% according to the series.5,6

Because the mitral annulus is close to the region of the
atrioventricular node and the His-Purkinje system, any extension
of calcified deposits would affect how these electrical structures
function, leading to intraventricular conduction delays. This occurs
particularly in cases in which CMA extends to the anterior annulus
and more medial segments of the posterior annulus and mainly in
the form of complete left bundle branch blocks and bifascicular
blocks.7

Nevertheless, the frequency of this association must be
considered with caution because the occurrence of both phenom-
ena is related to age and the degenerative changes may take place
independently.

The initial diagnosis is made on echocardiography. On occasion,
the chest X-ray may reveal a calcified mass in the cardiac
silhouette. Transthoracic echocardiography will show a spherical
mass, with calcified borders and an echoluent area in its interior,
that is located near the annular region of the posterior mitral leaflet
and does not usually produce an acoustic shadow.1,2

The differential diagnosis should include cardiac abscesses and
tumors.8 In addition to the clinical course, the characteristic
calcified, well-defined borders and their typically posterior site
help differentiate them from abscesses, usually located in the
mitroaortic fibrous continuity. In the case of tumors, the absence of
intercholucency is observed.

Additional imaging studies may aid the final diagnosis.
Transesophageal echocardiography1 will provide more informa-
tion on the site and composition of the internal material although
the diagnosis will be greatly aided by computed tomography,4
given its capacity to detect the calcified envelope, and cardiac
magnetic resonance imaging.9 In cardiac magnetic resonance
imaging, the characteristics of the various sequences will help
distinguish the condition from other tumors, and the signal is
normally hypointense with respect to myocardium in the gradient
echocardiography and T1- and T2-weighted sequences, with little
to no contrast uptake at the periphery of the mass.

Due to the asymptomatic course, only monitoring of the
patient’s progress is recommended in most cases. Surgery should
be reserved for patients with significant valvular dysfunction or an
uncertain diagnosis.2,4,6 Self-limiting cases have been reported,1,10
which suggests a dynamic component of the lesion that can regress
to CMA during its course.

In conclusion, CCMA is a rare variant of CMA, but should be
considered in the differential diagnosis with other cardiac masses
to prevent unnecessary surgery.

This article indicates that, like CMA, CCMA may be associated
with infra-Hisian conduction abnormalities.

CONFLICTS OF INTEREST

None declared.

Figure 3. A: Transthoracic echocardiogram at long-axis parasternal view. B: 3-chamber apical view showing a typical image of caseous calcification of the mitral annulus with virtually no posterior acoustic shadow.
REFERENCES