Unusual tumor-like calcification of the mitral annulus: diagnosis and tissue characterization by ultrasound, computed tomography and magnetic resonance imaging

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Calcification of the mitral annulus is a common echocardiographic finding in the elderly, particularly in females. Calcium deposits are generally located in the posterior mitral ring, sometimes extending to the whole mitral annulus and involving the mitral valve apparatus. The present report refers to 2 patients with a very atypical mass-like calcification of the mitral annulus resembling a cardiac tumor. A detailed evaluation of the mass was obtained at transthoracic and transesophageal echocardiography; the differential diagnosis with other intracardiac masses was aided by the use of computed tomography and magnetic resonance imaging. To our knowledge there has been no prior report of such a lesion evaluated at cardiac magnetic resonance imaging.

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Degenerative calcification of the mitral annulus is a common echocardiographic finding in the elderly, particularly in females[1]. The degree of calcification may range from focal deposits, usually in the posterior annulus, to extensive calcification involving the whole mitral apparatus and projecting into the adjacent left atrium or left ventricle. We report 2 cases of unusual, impressive mass-like mitral annular calcification, mimicking a cardiac tumor. A detailed evaluation of these lesions was obtained at transthoracic echocardiography (TTE), transesophageal echocardiography (TEE), 4-row multislice computed tomography (CT), and cardiac magnetic resonance imaging (MRI).

Description of cases

Case 1. A 64-year-old hypertensive woman was referred to our Institution because of mild effort dyspnea. Physical examination and 12-lead ECG were normal and her blood pressure was 140/80 mmHg. TTE showed a normal wall thickness and biventricular systolic function with evidence at Doppler of impaired left ventricular relaxation. Unexpectedly, a large (3.5 × 4 cm) round echodense structure, producing moderate shadowing artifacts and mimicking a tumor, was visualized in the posterior periannular region between the posterior mitral leaflet and the adjacent left ventricular and left atrial walls (Fig. 1). The mass had smooth borders, central areas of echolucency and its movements were synchronous with those of the annulus; the parasternal short-axis view (Fig. 1B) suggested at least partial involvement of the myocardial wall. At Doppler examination there were no signs of mitral regurgitation or left ventricular inflow obstruction. TEE (Fig. 1C) confirmed the findings of TTE showing that the posterior mitral leaflet was not included in the mass and that its excursions were normal. One year later the patient, who was clinically asymptomatic, was submitted to follow-up TTE showing no variation with respect to the previous picture. Further assessment with multislice CT and cardiac MRI was obtained. At CT examination a gross, dense homogeneous calcification was demonstrated in the posterior segment of the mitral annulus (Fig. 2). At this same site MRI study showed an area of signal absence on all sequences, without any perfusion after gadolinium administration, surrounded by a thin rim of high signal on both the pre-contrast T2-weighted and post-contrast late enhanced

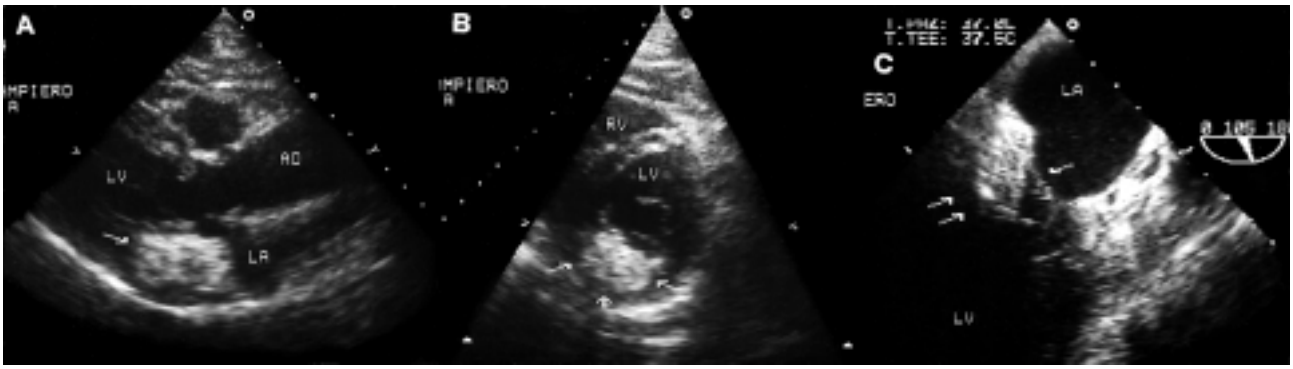

Figure 1. *Case 1. Transthoracic parasternal long-axis* (A) *and short-axis* (B) *views showing a large round echodense structure at the posterior left atrioventricular groove* (arrows). *The mass is characterized by areas of echolucency resembling liquefaction.* C: *transesophageal view at 105° (late systolic frame). The mass* (double arrow) *is located in the posterior annular region at the junction between the left atrium (LA) and the left ventricle (LV), inferior to the left posterior mitral leaflet* (single arrow) *which presents normal excursions. AO = aorta; RV = right ventricle.*

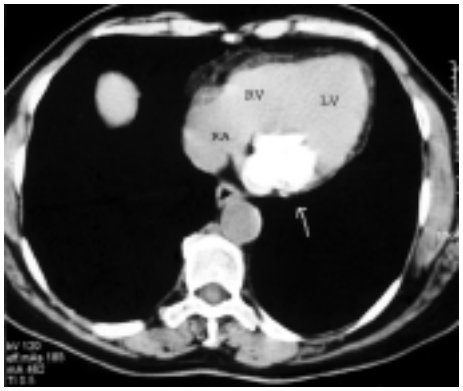

Figure 2. *Case 1. Unenhanced computed tomography. A large hyperdense calcified mass* (arrow) *may be seen at the posterior mitral annulus. LV = left ventricle; RA = right atrium; RV = right ventricle.*

images (Fig. 3), suggesting a thin edematous fibrous capsule enveloping gross calcifications.

Case 2. A 63-year-old woman with systemic hypertension, diabetes mellitus, obesity and chronic obstructive lung disease was referred because of worsening dyspnea. Physical examination revealed an apical grade 2 systolic murmur, a loud fourth heart sound and no pulmonary rales. ECG showed normal sinus rhythm with left atrial enlargement. TTE disclosed an hypertrophic left ventricle with normal systolic function and evidence of delayed relaxation; in the posterior annular region (Fig. 4A) a large (2 × 2 cm) round echogenic structure was found with the same characteristics seen in case 1 except for its smaller dimensions and for the presence of mild mitral regurgitation at Doppler investigation. TEE (Fig. 4B) demonstrated that the posterior mitral leaflet was stretched and arched over the mass, causing grade 1 mitral incompetence. Cardiac CT (Fig. 4C) showed a highly calcified area in the posterior aspect of the mitral annuls. Cardiac MRI (Fig. 4D), which was performed without contrast enhancement due to patient intolerance, confirmed the involvement of the mitral annulus and the calcified nature of the mass, as demonstrated by the lack of any signal on all sequences.

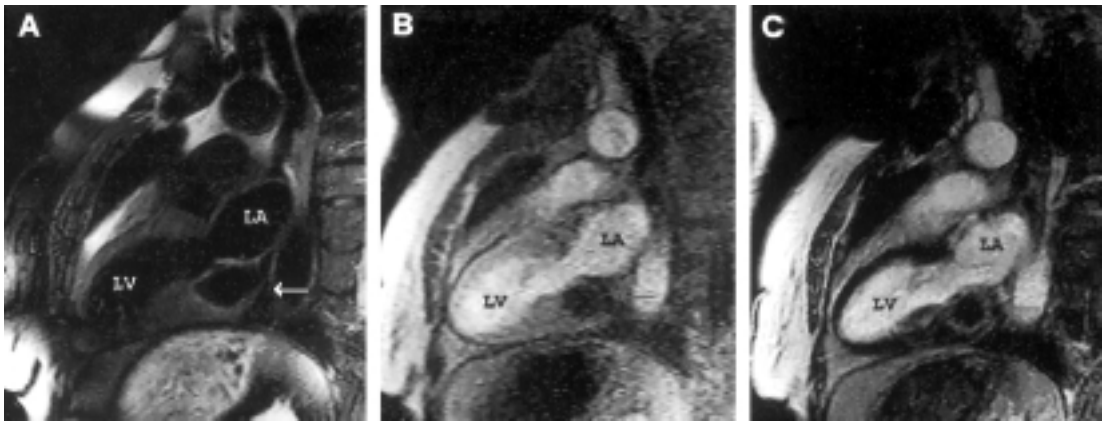

Figure 3. *Case 1. Cardiac magnetic resonance imaging.* A: *T2-weighted image, an area of absent signal due to calcification and surrounded by a thin hyperintense rim may be seen* (arrow). B: *first-pass acquisition frame, no perfusion of the mass.* C: *delayed contrast-enhanced acquisition, a hyperintense rim envelops the mass. LA = left atrium; LV = left ventricle.*

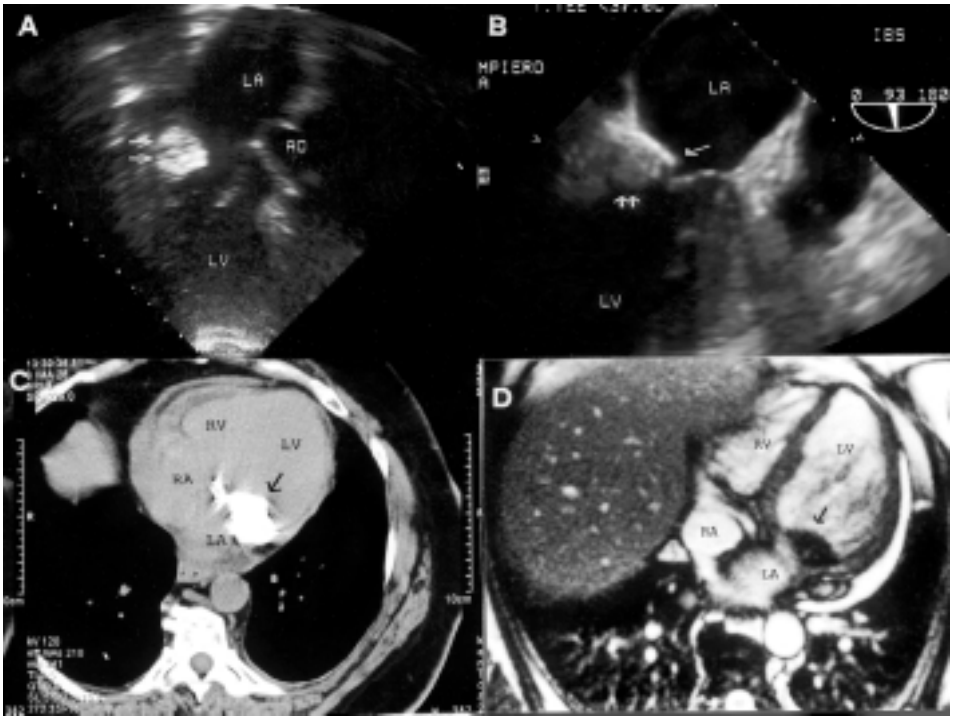

Figure 4. *Case 2.* A: *transthoracic 2-chamber view including the aorta (AO); a round echodense mass* (arrow) *may be seen at the posterior junction of the left ventricle (LV) and left atrium (LA).* B: *transesophageal 93° view; the basal and central segments of the posterior mitral leaflet appear attached to the mass* (double arrow) *and only the distal segment* (single arrow) *shows good mobility.* C: *unenhanced computed tomography; a gross calcification* (arrow) *may be seen in the posterior mitral annulus.* D: *unenhanced cardiac magnetic resonance imaging; owing to calcification, no signal may be seen in the mass* (arrow). *RA = right atrium; RV = right ventricle.*

Discussion

Calcification of the mitral annulus is a degenerative process involving the cardiac fibrous skeleton which occurs mainly in elderly individuals, particularly females[1]. The Framingham study reported, for patients over the age of 60, an incidence of 37% among females and 15% among males[2]. It may also occur in younger patients with advanced renal disease or other metabolic disorders that result in abnormal calcium metabolism[3]. Mitral annular calcifications are usually seen in the posterior or posterolateral atrioventricular groove, sometimes extending to the whole mitral annuls and involving the base of both mitral leaflets. Calcification of the papillary muscles and chordae tendineae may also be present, particularly in patients with end-stage renal disease. Depending on the location and extent of calcium deposits, various degrees of mitral incompetence or, less commonly, mitral stenosis may be present; rhythm disorders and systemic embolization have also been reported[4].

Unlike common mitral annular calcification, the findings observed in our 2 cases were very unusual because of the presence of a round tumor-like mass with well defined borders, high echogenicity and acoustic shadowing. Although this echocardiographic appearance and particularly the presence of shadowing artifacts suggested the presence of calcium deposits, consistent with the diagnosis of degenerative mitral annular calcification, an intramyocardial tumor could not be completely ruled out; in this regard, both CT and MRI confirmed the calcified nature of the mass, as demonstrated by the high CT density and MRI signal. Besides, in case 1, contrast-enhanced MRI revealed the presence of a thin edematous fibrous capsule enveloping and thus separating the mass from the adjacent myocardial wall, as also confirmed by the lack of perfusion after gadolinium bolus administration.

This unusual type of annular calcification, not adequately evaluated at echocardiography, has already been described, even though rarely, in the literature. The incidence at necroscopy reported by Pomerance[5] in 1970 for patients over 50 years of age was 2.7% of all autopsies with mitral annular calcification. Other than single case reports[6-8] only two series of *ante-mortem* diagnosis have been reported: Kronzon et al.[9] described 4 cases with intracardiac masses suggesting a prevalence of 0.055% (5 out of 9000 echocardiographic examinations). More recently, a prospective registry of patients with mitral annular masses was reported by Harpaz et al.[10]; in their series the prevalence was 0.63% of all mitral calcifications and 0.067% of all consecutive echocardiographic studies; these authors defined the lesion as "caseous calcification" of the mitral annulus and reported the operative findings of 3 patients referred to surgery due to mitral valve dysfunction or cerebral embolic events. Macroscopic examination of the sectioned masses highlighted the presence of a

toothpaste-like caseous material and microscopy revealed an amorphous acellular material with areas of multiple calcifications, necrosis and inflammatory infiltrate. In contrast to our echocardiographic findings, these authors did not report acoustic shadowing at TTE or TEE, probably due to a lower degree of calcific deposits. Other than tumors, the differential diagnosis of round echogenic structures adjacent to the left atrioventricular groove should include mitral annular abscess[11], infected mitral calcification[12], lipomatosis of the atrioventricular groove and enlarged lymph nodes[13]. We believe that the clinical presentation and echocardiographic picture (both at TTE and TEE, the latter for a better definition of the posterior mitral leaflet motion) should permit a correct diagnosis in the majority of cases. If the echocardiographic findings are not typical and the clinical data are uncertain, CT and MR imaging may provide additional information for the differential diagnosis[14,15]. Compared to conventional 4-row CT, MR permits a better tissue characterization and more clearly defines the relations of the mass with the surrounding structures and mitral valve apparatus; a similar diagnostic accuracy may probably be obtained with the last generation 16-row multislice CT with new reconstruction and post-processing programs, even though the radiation dose associated with CT imaging should be taken into account. From a clinical point of view these lesions, despite their impressive appearance, generally represent an incidental echocardiographic finding and carry a benign prognosis. Surgery should be reserved to patients with severe mitral valve dysfunction.

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