Rare disease

Cardiac tumour in a patient with rheumatic heart disease

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Summary

A 45-year-old female known to suffer from rheumatic heart disease (RHD), presented with breathlessness of 1 year duration. Two-dimensional echocardiography revealed significant mitral and aortic valve disease mandating double valve replacement. In addition, an unusual finding in the form of a well-defined, densely calcified intramyocardial left ventricular mass was noted on echocardiography. The nature and extent of the mass was assessed by additional imaging modalities. Patient underwent excision of the mass followed by double valve replacement. Histopathology was consistent with cardiac calcific amorphous tumour (CAT). An unusual occurrence of CAT in a patient with RHD is presented herein. This rare tumour has not been previously described in patients with RHD.

BACKGROUND

Cardiac tumours are uncommon in patients with rheumatic heart disease (RHD). Thrombus and vegetation are the usual masses encountered in this subset. In their absence, rare differentials should be considered. Cardiac calcific amorphous tumour (CAT) is one such rare condition. It was recognised as a distinct pathological entity in 1997 and thereafter, only a few cases have been reported in literature. More significant, this has not been described in the subset of patients with RHD.

CASE PRESENTATION

A 45-year-old female with RHD, presented with New York Heart Association class-III breathlessness of 1 year duration. There were no symptoms of angina, syncope, palpitation, extremity oedema or fever. There was no history of tuberculosis, diabetes mellitus or chronic kidney disease. On examination, her heart rate was 76 beats per minute and blood pressure was 110/70 mm Hg. Auscultation of the precordium revealed a long middiastolic murmur at apex and an ejection systolic murmur in right second intercostal space. Examination of other systems was normal.

INVESTIGATIONS

Laboratory studies revealed a normal complete haemogram with erythrocyte sedimentation rate of 20 mm/h and C reactive protein of 1 mg/dl. Blood sugar, renal and liver function tests were within normal limits. The serum calcium level was 9.1 mg/dl. Antinuclear antibodies and rheumatoid factor was negative. Chest x-ray in posteroanterior and lateral view (figure 1A,B) revealed a homogenous opacity. Unlike valvular calcification, which is non-homogenous and confined to specific anatomic location, here a uniform calcified mass was noted to the left side of spine, above the dome of diaphragm (→). Two-dimensional echocardiography confirmed the clinical suspicion of significant mitral and aortic valve disease, warranting surgery.

A well-defined, densely calcified intramyocardial mass was noted at the level of mitral valve. (figure 2A–C) (videos 1 and 2). On three-dimensional echocardiography, the mass was seen to arise from posterior wall of left ventricle (LV), adjacent to medial commissure (figure 2D,E). The calcific nature and extent of the mass was confirmed by fluoroscopy (figure 3A) and CT imaging (figure 3B,C). The coronary angiogram was normal.

Video 1 Transthoracic two-dimensional echocardiographic parasternal long axis view. The mitral valve leaflets are thickened with restricted mobility. A well-defined densely calcified mass was noted posterior to posterior mitral leaflet.

Video 2 Transthoracic two-dimensional echocardiographic short axis views at the level of mitral valve. The stenotic mitral valve with fused commissures can be seen. The intramyocardial location of the mass could be appreciated.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis of calcified LV mass includes non-neoplastic conditions like thrombus, hydatid cyst, tuberculoma and cardiac CAT. Neoplasms like myxoma, fibroma and teratoma can present with a similar picture.

TREATMENT

Patient underwent excision of the mass (figure 4A) followed by mitral and aortic valve replacement. The mass had a yellowish-grey appearance (figure 4B) and on microscopy revealed variably sized areas of calcification with surrounding eosinophilic amorphous material, collagen and chronic inflammatory cells (figure 4C). These findings were consistent with cardiac CAT.

OUTCOME AND FOLLOW-UP

The in-hospital course was uneventful. Postoperative echocardiogram showed no evidence of residual tumour and normally functioning prosthetic valves. She is asymptomatic at 4 months of follow-up.

Figure 1 X-ray in rheumatic heart disease. Chest x-ray in posteroanterior view (A) showed cardiomegaly of LV contour, LA enlargement and pulmonary venous hypertension. A uniform calcified mass was noted to the left side of spine above the dome of diaphragm (→). In the lateral view (B), this was seen at the junction of posterior and inferior surface of cardiac silhouette (→).

Figure 2 Two-dimensional and three-dimensional echocardiographic assessment of cardiac mass. A well-defined densely calcified mass noted posterior to posterior mitral leaflet in the parasternal long axis view (A) (video 1). In the short axis view (B) (video 2) at the level of mitral valve, the intramyocardial location of the mass could be appreciated. It is mainly confined to basal posterior segment without involvement of the leaflet. The above findings were also seen in apical four-chamber view with posterior angulation (C). Three-dimensional echocardiography with enface view of mitral valve through left atrium demonstrating the extent of the mass (D,E) and protrusion into left atrium.

Figure 3 Fluoroscopic and CT images. Coronary angiogram in right anterior oblique caudal view showing the mass in relation to left circumflex coronary artery. The absence of vascularity can also be noted (A). On CT imaging, the bone-like ossifying nature of the tumour is visualised (→) (B). The site and extent of the mass is confirmed in the sagittal section (→) (C). The mass was confined to the LV myocardium without infiltration of adjacent structures.

Figure 4 Gross anatomy and histopathology of cardiac mass. The LV mass had an 'egg-shaped' contour when seen from LA (→) side intraoperatively (A). The excised mass measured $4 \times 3.5 \times 2$ cm with pultaceous material extruding from the cut surface (→) (B). On microscopy, there are areas of extensive calcification with eosinophilic amorphous material. There was no evidence of tumour pathology (C).

DISCUSSION

Cardiac tumours are uncommon in patients with RHD. Thrombus and vegetation are the usual masses encountered in this subset. Clinical history and imaging forms the corner stone of evaluation. In the absence of evidence for thrombus or vegetation, rare differentials should be considered. Cardiac CAT is one such rare condition which has not been described in patients with RHD. It is a non-neoplastic, endocardially based, intracavitary mass with no specific predilection for any of the cardiac chambers.^{1 2} It may be an isolated finding or may be seen in association with conditions like diabetes mellitus, chronic renal failure, coronary artery disease and vasculitis.¹ Clinically, the tumour may be asymptomatic or manifest with features of tumour embolisation.³ Clinical and radiological differentiation of this lesion from other cardiac masses is not possible. Multimodality imaging may give a clue to possible alternative diagnosis. Cardiac CAT can be confirmed only by histopathological examination. It is possible that cardiac CAT is an extreme manifestation of degenerative, calcific process of RHD which extends to the posterobasal myocardium. Alternatively, it could represent organised and calcified mural thrombus. Long-term prognosis is excellent following complete removal of the mass. However, recurrences are reported³ and long-term surveillance may be justified if there is evidence of incomplete excision.

Learning points

- ▶ Cardiac CAT should be considered in the differential diagnosis of calcified intracardiac tumours.
- ▶ Although described in various settings, it has not been previously reported in patients with RHD.
- ▶ Despite multimodality imaging techniques, histopathology remains the gold standard for confirmation of diagnosis.
- ▶ Complete excision is associated with favourable outcome.

Competing interests None.

Patient consent Obtained.

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