Massive calcification of giant left atrium in a rheumatic mitral stenosis patient: A case report

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ABSTRACT

Giant left atrium is a condition characterized by an extreme enlargement of the left atrium with a diameter more than 80 mm and it is usually associated with long standing rheumatic mitral valve disease. We present a case of giant left atrium with massive calcification in a female patient who had a history of rheumatic heart disease, severe mitral stenosis, permanent atrial fibrillation, and cerebral infarctions.

1. Background

Giant left atrium (GLA) is a condition characterized by an extreme enlargement of the left atrium with a diameter more than 80 mm [1]. Massive calcification of GLA in adults is extremely rare in the 21st century. Mitral valve disease (mitral regurgitation more than mitral stenosis) often leads to GLA [2]. Currently, there is no consensus regarding the management of GLA during mitral valve surgery. Most surgeons fix the mitral valve and do little to an oversized left atrium. Others only occlude the left atrial appendage [3]. We think that remodeling of the left atrium is part of the successful mitral valve surgery in the GLA. We report a case of giant atrium with massive calcification in the context of rheumatic mitral stenosis and severe tricuspid regurgitation in a middle-aged lady.

2. Case presentation

A 49-year-old woman presented to our emergency department with dyspnea, palpitations, and ankle edema. She had been diagnosed with rheumatic mitral stenosis for 13 years, and not compliant with injectable penicillin. 13 years ago, she presented with shortness of breath and was diagnosed with severe mitral stenosis and permanent atrial fibrillation. Because of personal reasons, no anticoagulant therapy was not taken by this lady. She presented with disturbance of consciousness and disorder of limb’s activity 3 years back.

Transthoracic echocardiogram showed a severe mitral stenosis (estimated mitral orifice size, 0.58 cm²) and moderate regurgitation, severe tricuspid regurgitation and a giant left atrium (96mm*135mm*98mm) with thrombi (Fig. 1A). The left ventricular size was 61 mm with an ejection fraction of 46%. The right ventricle was small with right ventricular dysfunction which was evident from a reduced tricuspid annular plane systolic excursion of 12 mm and a right ventricular ejection fraction of 32%. A computed tomography scan showed that the LA measured 20.0cm * 13.7 cm (coronal plane) with massive calcification and compressed the left ventricle, right ventricle and right atrium (Fig. 2 A&B). The lung function of the patient was severely impaired.

After a median sternotomy, cardiopulmonary bypass was established by standard aorta, superior and inferior vena cava cannulation. After the classical left atriotomy via Waterson groove, extensive calcification involved the entire atrium with the orifices of the mitral valve and pulmonary veins intact. The surface of the wall is irregular with adherent thrombus. The calcification was dissected from the atrial wall. The mitral valve was replaced with a mechanical valve (size 27; GK mechanical prosthesis); and the left atrial appendage was ligated; and the left atrium was plicated. The sites of plication include posterior mitral annulus (para-annular), lateral to right pulmonary veins and between right and left pulmonary veins. The posterior wall of left atrium between the right and left pulmonary veins was plicated in a semilunar fashion. It was plicated in a continuous running fashion. A superior plication was performed and in combination with the para-annular plication, then circumferentially around the left pulmonary veins...
reaching the mitral annulus. We then performed a tricuspid valvular annuloplasty with a MC3 ring (size 28; Edwards Life sciences). The pathology of the left atrium endocardium showed extensive myocardial collagenation with myxoid degeneration of the interstitial, little inflammatory cell infiltration and calcification.

The patient recovered very well and a post-operative chest CT scan showed a marked reduction in the cardiothoracic ratio (Fig. 3 A&B). The follow-up of the echo demonstrated the remarkably decreased left atrium with an anteroposterior diameter of 4 cm and well mechanical mitral valve function (Fig. 1B). The lung function of the patient was improved as well. The patient had an uncomplicated postoperative course. Twelve months after surgery, she was in NewYork Heart Association Class II.

3. Discussion

A review of the literature reveals that the majority of GLA cases are managed at the time of mitral valve surgery. Most authors agree that the main indication for its surgical management is the presence of compressive symptoms from neighboring organs [4]. We claim that by reducing the left atrial size, the pressure effect is reduced with a favorable effect on the postoperative course. A second indication in our opinion is the presence of thrombus and a history of thromboembolic events. Left atrial volume reduction can in theory prevent recurrent thrombosis by reducing intra-atrial stasis. Furthermore, a large atrial size increases thromboembolic risk and reduces the success rate of cardioversion [5].

In this case, the left atrial enlargement was due to the severe mitral stenosis and mitral regurgitation. This patient had a longstanding mitral stenosis associated with permanent atrial fibrillation. Both factors were responsible for the development of GLA and of thrombosis. In this case, the lung of the patient was severely oppressed and the atrium wall was calcified. So we did a LA reconstruction and plicated the dilated wall of the left atrium. Although valvular surgery is the fundamental operation, LA reduction may be a supplementary. LA reduction with mitral surgery seems to be effective for improving respiratory and circulatory function from the early postoperative period for patients with giant left atrium accompanying mitral lesion.

Giant atrium due to rheumatic etiology is still prevalent in developing countries. Some patients may remain asymptomatic despite with massive enlargement. The thromboembolism risk of symptomatic patient with atrial fibrillation is very high. Early recognition and appropriate prophylaxis of rheumatic fever could reduce the complications of rheumatic heart disease.

Fig. 1. A, Transthoracic echocardiogram showed a giant left atrium (96mm×135mm×98mm). B, The post-operative TTE showed the left atrium with an anteroposterior diameter of 4 cm.

Fig. 2. A, Axial computed tomography (CT) revealed a giant left atrium and calcification of the wall. B, Frontal chest radiograph of the patient with giant left atrium. The cardiothoracic ratio was almost 0.9.

Fig. 3. A, The post-operative CT scan of the chest showed a marked decrease of left atrium. B, Post-operative frontal chest radiograph of the patient showed a marked reduction in the cardiothoracic ratio.

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Competing interests

The authors declare no conflicts of interest.

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