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Giant Left Atrium in Rheumatic Mitral Disease: Beyond a Simple Adaptive Mechanism: 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 in a 47-year-old female patient who had a history of rheumatic heart disease, severe mitral disease, permanent atrial fibrillation and causing the compression of adjacent intra-thoracic structures. The patient underwent a successful mitral valve replacement with reduction of the enlarged sized left atrium with good outcomes.

KEYWORDS: Giant Left Atrium, Rheumatic Mitral Disease, Atrial Fibrillation, Compression, Plication.

INTRODUCTION

Giant left atrium (GLA) is a rare condition, with a reported incidence of 0.3%, characterized by an extreme enlargement of the left atrium (LA) with a diameter more than 80 mm [1]. Following mainly long standing rheumatic mitral valve regurgitation or mixed mitral disease with predominant regurgitation [2]. It can be associated with atrial fibrillation, thromboembolic, hemodynamic and respiratory complications. [2, 3] Delaying the mitral valve replacement can lead to fatal outcomes; optimal timing is crucial for reducing mortality rates. We report the case of a giant left atrium in the context of a neglected rheumatic mitral mixed disease with tricuspid regurgitation in a middle-aged lady.

CASE REPORT

A 47-year-old woman was admitted to our institution with progressive worsening dyspnea, repeated coughing and recurrent paroxysmal nocturnal dyspnea with palpitations. She has been followed for rheumatic mitral stenosis for 13 years; Mitral valve replacement was indicated 10 years ago but the patient refused surgery when it was suggested at that time. Since then, she had been receiving medical treatment. Apart from dyspnea, there were complaints of voice hoarseness, some episodes of dysphagia with no other gastrointestinal symptoms.

The physical examination revealed a blood pressure at 100/65 mmHg, the heart rate was of 98B/min with a rhythm of atrial fibrillation, her neck veins were distended, no crepitations were found. Cardiac auscultation revealed a soft S1 along with a pansystolic murmur at apex radiating to axilla. A long mid diastolic rumbling murmur across the mitral area was also heard. Examination of other organ systems was normal. Chest X-Ray showed increased cardiothoracic ratio with splaying of the carina and elevation of left main bronchus suggestive of left atrial enlargement. At the time of admission, the patient was on treatment including diuretics, digoxin and warfarin with an INR of 2,9.

Transthoracic echocardiography (figure 1) showed a hugely dilated left atrium, with an anteroposterior diameter of 15 cm in the parasternal long axis view. The estimated left atrial volume was 1375 ml with an area surface estimated at 154 cm^2. There was severe mitral stenosis (MVA = 0.5 cm^2 and a mean diastolic pressure gradient of 15 mmHg at rest) associated with severe mitral regurgitation (PISA = 17 mm, ORE = 2.59 cm^2 and regurgitating volume of 409 mL) as shown in figure 2. The left ventricle wasn't dilated with a preserved systolic ejection fraction of 68 %. Tricuspid valve was moderately incompetent with an estimated pulmonary arterial pressure of 55 mmHg. There was trivial aortic incompetence.

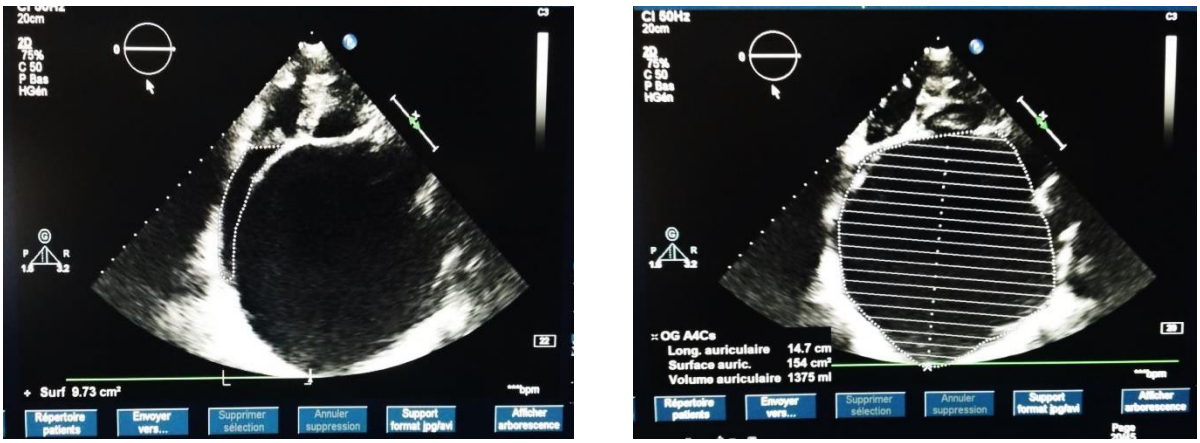

Figure 1: Transthoracic echocardiography views showing in four chambers views the huge left atrium, with a small left ventricle and normal sized right cavities.

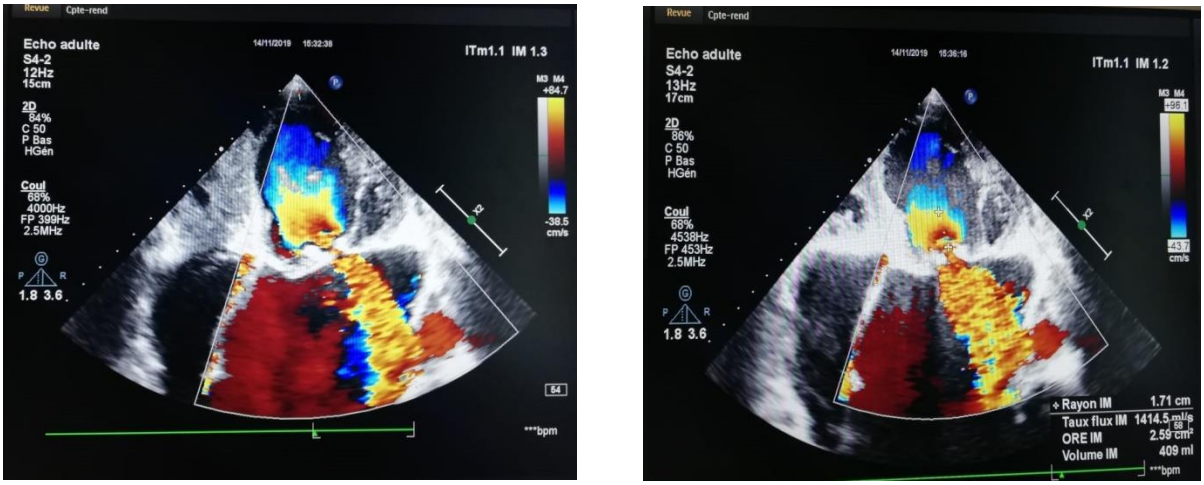

Figure 2: Four chambers echocardiographic views showing the severity of mitral regurgitation; note the direction of the eccentric intra-atrial jet.

Mitral valve was replaced with St. Jude Medical valve and the tricuspid valve was repaired using De Vega's tricuspid valve annuloplasty with reduction of the enlarged sized left atrium. Postoperatively, the patient remained hemodynamically stable.

One week after surgery, the echocardiographic control showed a well-functioning prosthetic valve, a reduced-sized left atrium with an estimated size of 10 × 11 cm; chest X-ray showed a reduced cardiothoracic ratio with clear lung fields. The evolution was favorable with uneventful postoperative course; she was discharged after 12 days with a regular follow-up.

DISCUSSION

Rheumatic heart disease continues to be a common health problem in the developing world, causing morbidity and mortality among both children and adults. [4]

Long standing mitral valve disease is associated with enlargement of the LA. Though, the mechanism of formation of GLA is not fully understood.

Patients with chronic mitral valve disease are not always associated with GLA. Only 19% may develop such a condition [5]. Previous studies showed that chronic pressure and increased volume in the LA is not the only cause of GLA, but weakening of the left atrial wall by rheumatic pancarditis causing chronic inflammation and fibrosis is also implicated [2, 6].

Extreme enlargement of the left atrium more than 65 mm is known as giant left atrium. [7] Hurst [2] defined a giant left atrium as "one that touches the right lateral side of the chest wall" on chest-X ray (CXR) and that "the condition is almost always caused by rheumatic mitral valve disease".

Piccoli *et al* [8] defined the GLA as a cardio-thoracic ratio on CXR of >0.7 combined with a left atrial anterior-posterior diameter of >8 cm on transthoracic echocardiography.

Other investigators [7] have decreased the LA size for diagnosis to 6.5 cm in the parasternal long axis view.

Patients with GLA are usually presented with symptoms related to mitral valve disease. Specific symptoms may occur due to compression of the esophagus and airway by the enlarged posterior wall of the LA causing dysphagia and respiratory dysfunction which was the case of our patient with her complaints of voice hoarseness and some intermittent episodes of dysphagia.

Cardiac output may be reduced by obstruction of inferior vena cava (IVC) due to displacement of atrial septum and compression of the postero-lateral wall of the left ventricle by the enlarged LA [9]. Minagoe *et al* [10] studied the IVC flow in such cases and found a significant reduction in IVC orifice with significant increase in flow velocities, explained by the inter-atrial septal bulge occurring in this condition. This explains the right-sided manifestations that can occur even without significant tricuspid regurgitation. [11]. In rare situations, patients are completely asymptomatic [8].

Enlargement of the LA is associated with development of atrial fibrillation (AF), which in return can lead to further enlargement of LA. Such an observation was seen even among patients who had AF with absence of cardiac pathology [12].

Various complications were described with this enlargement, including thrombus formation, thromboembolic events, hemodynamic derangements and sudden death [8, 9].

Our patient had a longstanding mitral stenosis and severe regurgitation associated with chronic AF. Those factors were responsible for the development of GLA. There was no echographic evidence of intra-cardiac thrombosis and no history of thromboembolic events with a well-controlled INR.

Although rheumatic valvular heart disease is widespread, GLA is rare and only seen in 4-5 % of patients. Such a low incidence is probably related to early development of pulmonary hypertension and its subsequent effect on reducing right ventricular output, so both pressure and volume load in left atrium are reduced; [13] due to these hemodynamic significant effects, optimal timing for surgery is crucial.

The aim of surgery in GLA is to correct the mitral valve abnormalities, to treat compression manifestations, to prevent thromboembolism and to revert if possible, atrial fibrillation to normal sinus rhythm. [9]

Two strategies had been applied with some controversies regarding the management of GLA at the same operating time. The first is to perform mitral valve surgery with left atrial volume reduction while the second is to perform mitral valve surgery alone. [9]

It has been shown through previous studies that plication at the time of mitral valve replacement (MVR) is essential to reduce LA size to eliminate its compressing symptoms and thromboembolic complications [9] by reducing its volume and thus reducing intra-atrial stasis. They claimed that the pressure effect is reduced with favorable effect on the postoperative course [14]. The standard is to resect the LA until its diameter decreases to 5 cm.

While some surgeons believe that successful MVR alone will result in the eventual reduction of LA size as the volume and mean atrial pressure decline. [9] They believed that prolongation of the cardiopulmonary bypass time in these patients does not warrant LA plication except when the atrium compresses the lung as to produce atelectasis.

A group of surgeons in 2001 [15] compared both surgical strategies and showed that the reduction in left atrial size was comparable in both groups.

Tonguc and colleagues [14] compared patients undergoing MVR with and without plication, found no significant difference in hemodynamic improvement and reduction of LA diameter especially when the diameter was below 80 mm. It is admissible that the size of LA will not change significantly after MVR due to rheumatic irreversible damage of LA muscle by fibrosis but hemodynamics will improve after MVR with or without gesture on the LA. Plication is to be considered especially in the presence of compressive symptoms from neighboring organs [16].

The restoration of sinus rhythm in GLA was long debated. GLA has been recognized as refractory for regular cardioversion procedures either medically or electrically. [17] This is possibly due to fibrotic and calcified degeneration of the LA myocardium. Maze procedure has been used to treat longstanding persistent AF by dividing the macro-reentry circuits. Normal sinus rhythm had been reported to range from 67% to 98%. [17].

Yuda *et al* [17] studied the efficacy of the Maze procedure in restoring the atrial contraction in patients with GLA compared to those without GLA. After one year follow-up, normal sinus rhythm was obtained in 53% of the GLA in comparison to more than 75% in non-giant left atrium. Atrial contraction was regained in 20% of patients with GLA in comparison to more than 50% in non-giant left atrium.

Wang et al in a recent study [18] used Maze procedure concomitant with MVR with or without tricuspid valve surgery and bi-atrial reduction surgery. The results of this study confirmed the previous one. The bigger the LA, the lower is the rate of restoring the sinus rhythm.

Our patient was suffering from longstanding rheumatic mitral valve disease with GLA and manifesting compressive symptoms on respiratory and digestive tracts with hoarseness. She had MVR with De Vega's tricuspid annuloplasty and left atrial plication at same operating time, without Maze procedure with great improvement of symptoms in the early postoperative period. Atrial fibrillation rhythm remained and was medically controlled with oral anticoagulation to prevent thromboembolism.

CONCLUSION

Giant atrium due to rheumatic etiology is still prevalent in developing countries. Appropriate prophylaxis of rheumatic heart disease and early recognition could reduce the consequent complications. Some patients may not develop symptoms in spite of massive enlargement. The thromboembolism risk of symptomatic patients with atrial fibrillation is very high.

LA debulking with mitral valve surgery seems to be effective in the presence of compressive symptoms with a significant improvement on respiratory and circulatory functions from

the early postoperative period and may also prevent thrombosis for those patients.

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