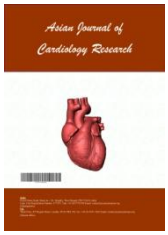

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Giant Left Atruim in Rheumatic Mitral Valve Disease, What Prognosis?

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Rheumatic mitral stenosis (MS) remains the most common type of valvular heart disease worldwide. While the prevalence of mitral valve rheumatism is decreasing in industrialized countries, it continues to be endemic in most countries in Asia, Latin America, the Middle East and Africa, including Morocco, which delimits the belt of shame.

Giant left atrium (ectasia of the left atrium) is due to long-term evolution of rheumatic mitral valve disease, approximately 10 years after diagnosis of MS can be really challenging when it comes to therapy decision. Transthoracic echocardiography is first tool used in heart valve evaluation and evaluating heart remodeling during advanced stage valve diseases. The prognosis of patients diagnose lately for rheumatic mitral stenosis with giant left atrium (ectasia) depends largely on some factors especially reversible pulmonary hypertension, possible left atrium reduction (LAR) associated with other comorbidities.

We hereby report cases of 3 female adult patients lately diagnosed for rhematic mitral stenosis with giant (ectasia) left atrium (GLA) and severe pulmonary hypertension who in spite of surgical valve replacement shortly died after surgery because of persistent pulmonary hypertension. What determines their poor prognosis?

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Keywords: Giant left atrium (ectasia); mitral stenosis; pulmonary hypertension; LAR.

1. INTRODUCTION

Acute rheumatic fever (RF) is an autoimmune inflammatory process that develops as a sequela of group A β-hemolytic streptococcal (GAS) tonsillopharyngitis. It may lead to rheumatic heart disease (RHD) and it constitutes the major etiology of mitral stenosis (MS) [1]. While the prevalence of mitral valve rheumatism is decreasing in industrialized countries, it continues to be endemic in most countries in Asia, Latin America, the Middle East and Africa, including Morocco, which delimits the belt of shame. Statistics indicate that 15.6 million children and young adults are affected by the rheumatic disease and that 233,000 patients die from it each year in these countries [2].

The mitral valve is affected in 65–70% of patients and rheumatic fever is the first cause of mitral stenosis in developing countries [3]. One of the problems actually it's the late diagnosis of rheumatic mitral stenosis and late surgical therapy which increase poor prognosis in affected patients in case of ectasia of the left atrium and irreversible pulmonary hypertension. Trans thoracic echocardiography is the key tool to screen heart valves in rheumatic fever patients especially after 5-10 years of diagnosis (4). Early detection of valvular heart rheumatic disease is of better prognosis than late diagnosis.

We hereby report cases of 3 patients lately diagnosed for rheumatic mitral stenosis with giant (ectasia) left atrium and severe pulmonary hypertension of which, one the patient, in spite of surgical valve replacement shortly died because of persistent pulmonary hypertension due to lack of left atrial remodeling and left ventricular adaptation. The interest of this clinical cases is evaluating the risk of mortality secondary to thrombo-embolic events and mortality in operated or non-operated subjects.

2. PATIENTS AND METHOD

Case 1

The first patient was a young female aged 48 years with a history of non-surgical rheumatic mitral valve disease evolving over 20 years under medical treatment based on oral anticoagulation only, acenocoumarol (sintrom) tablets daily without regular fellow-up. She had no other underlying personal or family history of heart disease. She consulted at the emergency department of cardiology at the University teaching Hospital, Ibn Rochd of Casablanca for worsen dyspnea and altered general health state. Clinical examination findings were, stage IV NYHA dyspnea, edema of both lower limbs rising to the knees evoking heart failure with no acute articular symptom. Pulmonary auscultation found bilateral midfield crackles whilst heart assessment found an irregular heartbeat, diastolic rolling at the mitral focus associated with burst of B2 sound at the pulmonary valve focus. The ambient air oxygen saturation of the patient at admission was 96% and blood pressure of 130/75mmHg. The routine ECG showed accelerated atrial fibrillation with a heart frequency of 125beats per minute(bpm) Fig. 1.

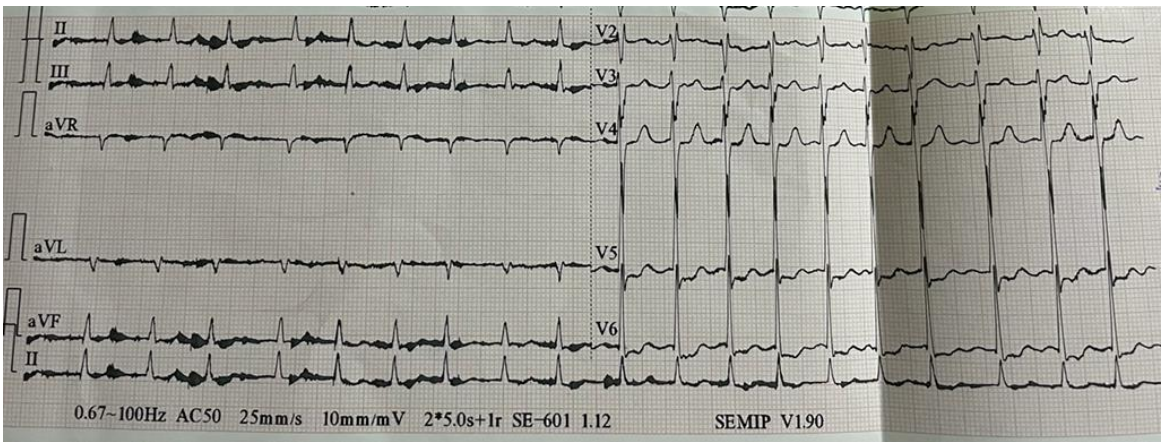


Fig. 1. Electrocardiogram of patient Case 1: Accelerated atrial fibrillation (AF) at a frequency of 125bpm

Initial biological analyses of the patient found a high white blood count at 12000/µL with predominant neutrophil with a normal INR ratio of 2.5 whilst the rest of the biological analyses was without abnormality.

Transthoracic echocardiography confirmed a severe mitral stenosis with mitral surface area of 0.6cm^2**,** and transvalvular gradient of more than 8mmHg associated with a mild mitral regurgitation. The mitral valves where heavily calcified at their free leaflet sides with commissural fusion. The ventricular ejection fraction was estimated by Simpson bipolar plane to 55%, ectasia of the left atrium with the highest volume and atrial surface area found in the patient was 980ml/m^2 indexed volume and 129.4cm^2 left atrial (LA) surface area (Fig. 2), presence of spontaneous contrast in the left atrium**,** associated with severe tricuspid regurgitation estimated with a systolic pulmonary pressure at 80mmHg.

The patient was put under medical therapy based on restriction hydric intake with enzyme converter inhibitor (coversyl 5mg daily), mineralocorticoid (Aldactone 50mg ½ tablet a day), high dose loop diuretic (lasilix intravenously) for decongestion, and betablocker (Cardensiel; initial dose of 1.25mg daily titrated) for rhythm control. Three days after heart decongestion therapy with diuretics, the patient was programmed for surgery. Right heart catheterization was

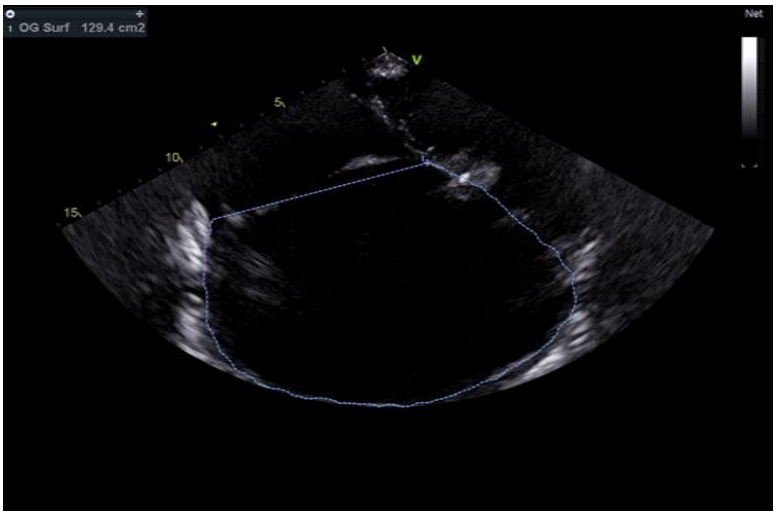


Fig. 2. Transthoracic echocardiography (TTE): 4 chambers apical view: showing a giant left atrium (ectasia) occupying the whole monitor screen with a surface area of 129.4cm^2

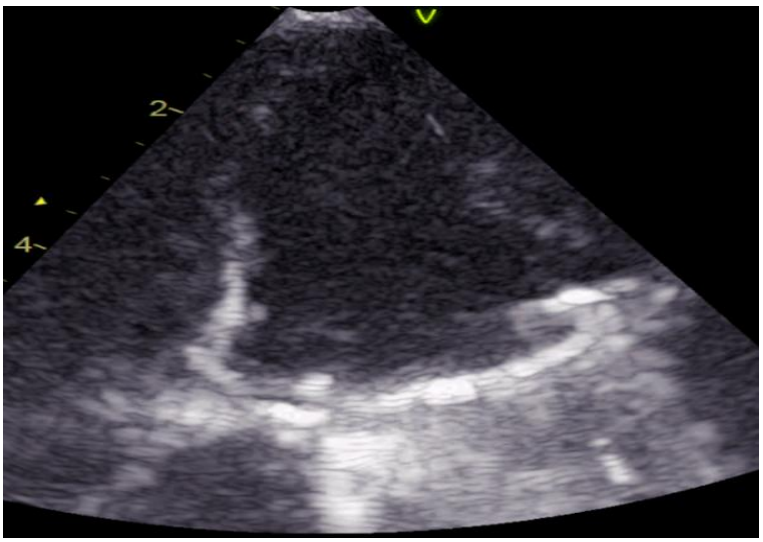


Fig. 3. TTE: 4 chambers apical (operated patient) view zoomed at the mechanical mitral prosthetic valve replacement with shadows of rings in a dilated atrium

not performed in the patient for other reasons, a surgical replacement of the diseased valve with prosthetic mechanical valve **(**Fig. 3) plus tricuspid plasty without left atrium reduction (LAR) was performed (especially left atrium plication) Fig. 3. Patient died 24hrs after surgery because of persistent pulmonary hypertension associated.

Case 2: The second patient was the youngest amongst the three female patients aged 42 years with a history of non-surgical rheumatic mitral valve disease evolving over 17 years under medical treatment based on oral anticoagulation only, acenocoumarol (sintrom) tablets daily without regular fellow-up. She had no other underlying personal or family history of heart disease. She consulted at our out-patient department of cardiology at the University teaching Hospital, Ibn Rochd of Casablanca for specialized medical follow-up referred by another hospital in the sub-region of Casablanca. Clinical examination findings showed patient in a fairly good general health state. Her blood pressure (Bp) was normal whilst cardiac assessment concluded an irregular heartbeat and diastolic rolling at the mitral focus. There were no signs of congestive heart failure and the ECG findings showed an atrial fibrillation at a frequency of 80bpm. The biological analyses were normal with an INR ratio within the therapeutic objective (3.14). TTE showed a heavily calcified mitral valves predominantly at their free leaflet sides with commissural fusion associated with severe mitral stenosis with a mitral surface area of 0.9cm^2 without no mitral regurgitation and transvalvular gradient of 12mmHg. The ventricular ejection fraction was good at 60% associated with a moderate tricuspid regurgitation with an estimating resting systolic pulmonary pressure of 35mmHg. Ectasia of the left atrium (LA) with a surface area volume of 390ml/m^2 indexed volume and 80cm^2 LA surface area and presence of spontaneous contrast (Fig. 4). The patient's treatment was adjusted by adding an enzyme converter inhibitor (coversyl 1.25mg daily) to a betablocker (Cardensiel; initial dose of 1.25mg daily titrated) for rhythm control. Patient was proposed for surgery which she refused in-spite all advices given.

Case 3: The third patient was a the eldest amongst the three female patients aged 58 years with a history of non-surgical rheumatic mitral valve disease evolving over 15 years under medical treatment based on oral anticoagulation, acenocoumarol (sintrom) tablets daily, mineralocorticoids and diuretics with recent regular fellow-up. She presented the same signs and symptoms presented by the first patient (case1) upon her arrival at the emergency department. The clinical examination found a patient in a very altered general health state, orthopnea, bilateral diffuse edema of the lower limbs rising over the knees with no acute articular symptoms. Heart and lung assessment concluded a bilateral crackle rising over lung's midfield, tachycardia, diastolic rolling at the mitral focus associated with abnormal pulmonary bruit at pulmonary focus. The ambient air oxygen saturation of the patient at admission was 95% and blood pressure of 150/90mmHg and routine ECG performed showed an accelerated atrial fibrillation with a heart frequency of 135 beat per minute(bpm). A complete blood analyses was carried out in the patient including complete blood count test, electrolytes which showed lymphopenia at 640/µL, C-reactive protein (CRP)at 43.5mg/l with negative values of D-Dimers and ultrasensitive troponines whilst INR ratio was at 2.8 (respective therapy range).

TTE showed a heavily calcified mitral valves predominantly at their free leaflet sides extending to the bases with commissural fusion associated with severe mitral stenosis with a mitral surface area of 0.8cm^2, severe mitral regurgitation (Regurgitation surface area 42cm^2 and regurgitation volume of 60ml) and transvalvular gradient of 24mmHg. The ventricular ejection fraction was lightly reduced estimated at 49% Simpson bipolar plane associated with a severe tricuspid regurgitation with an estimating resting systolic pulmonary pressure of 131mmHg (Fig. 5 B) and dysfunction right ventricle. Ectasia of the left atrium (LA) with a surface area volume of 616ml/m^2 indexed volume and 102cm^2 LA surface area and spontaneous contrast (Fig. 5 A). CT-scan was performed in patient which showed an alveolo-interstitial syndrome associated with mild right plural effusion probable of infectious origin. PCR-COVID 19 test conducted was negative for the virus.

The patient was put under medical therapy based on restriction hydric intake associated with enzyme converter inhibitor (coversyl 5mg daily), mineralocorticoid (Aldactone 50mg ½ tablet a day), high dose loop diuretic (lasilix intravenously) for decongestion and betablocker (Cardensiel; initial dose of 1.25mg daily titrated) for rhythm control. Patient died 2 weeks later, during her hospitalization due to complicated cardio-renal syndrome.

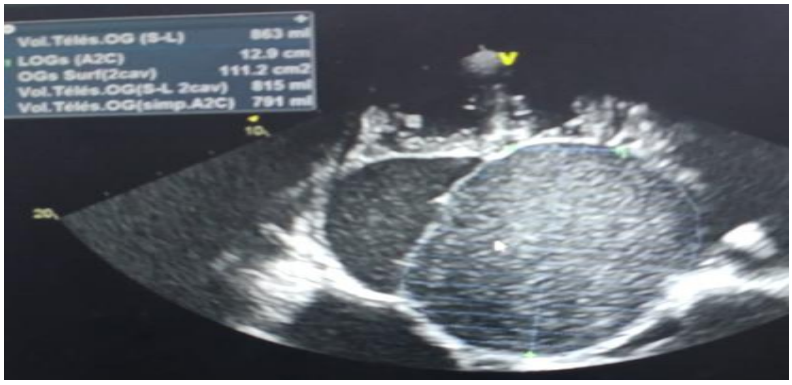
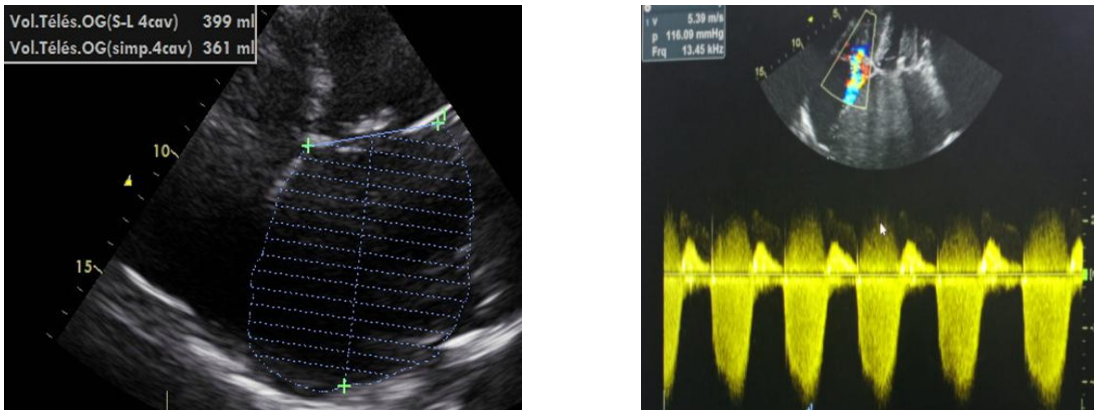


Fig. 4. TTE: 4 chambers apical view: showing an ectasia of the left atrium with a surface volume of 863ml associated with severe mitral stenosis, heavy calcified valves and spontaneous contrast in the left atrium



A **B**

Fig. 5 A. TTE: 4 chambers apical view: showing an ectasia of the left atrium with a surface volume of 399ml associated with rheumatic mitral valve stenosis
Fig. 5 B. TEE: 4 chambers apical view of the eldest patient: CW doppler flux of tricuspid valve regurgitation with systolic pulmonary pressure estimated at 131mmHg

3. DISCUSSION

Rheumatic mitral stenosis (MS) remains the most common type of valvular heart disease worldwide yet there are few studies on optimal timing of intervention in asymptomatic patients. Postulated benefits of intervention before symptom onset include prevention of left atrial dilation, atrial fibrillation (AF) and pulmonary hypertension leading to fewer thromboembolic events, less heart failure [4]. Dilatation of the left atrium with mitral lesion is caused by an increase in compliance of the left atrium due to chronic valvular disease. This compensatory role of the left atrium is considered to reduce the effect of pulmonary congestion and to be beneficial for circulatory function in the case of mitral lesion [5]. Giant left atrium can be really challenging when it comes surgical therapy reduction. Transthoracic echocardiography is first tool used in heart valve evaluation and evaluating heart remodeling during advanced stage valve diseases like dilatation of the left atrium [6,7,8]. In our case a 2D transthoracic echocardiography was used to evaluate the severity of the mitral stenosis and measurement of left atrium surface area and volume.

Right Heart catheterization is also the only method available to directly measure absolute pressure within the cardiac chambers, and pulmonary vascular resistance can be calculated which may be important in decision making of patients with severe pulmonary hypertension to estimate the risk of surgery [9]. However, in our case, right heart catheterization was not possible for several reasons.

Although valvular surgery is the fundamental operation in some series, left atrial plication (LAP) may have an ancillary role, and some studies showed that it can be performed at least without a detrimental effect [5]. In our cases, only one patient was operated without left atrial

reduction (LAR) which led to a rapidly increase in diastolic volume pressure from the left atrium non supported by her left ventricular muscles. It's important to note without LAR during mitral valve replacement in a left giant atrium heart, the left ventricle muscles contraction can easily be exhausted because of the important diastolic pressure volume from the left atrium and secondly with the presence of irreversible pulmonary hypertension, immediate post operated patients have a slim chance of living [5].

Irreversible pulmonary hypertension is an important determinant factor in these cases as its crucial in decision making as patients in spite of valve replacement, LAR will remain symptomatic without reducing re-hospitalization rate for heart failure and death, thus absolutely necessary for such patients to undergo right heart catheterization before surgery.

Other methods like percutaneous balloon mitral valvuloplasty (PBMV) or transcatheter mitral valve lithotripsy (TMVL) cannot be use especially if giant left atrium is involved but could be use if irreversible pulmonary hypertension is present and high surgical mortality risk in patient [10]. Heart team evaluation is highly recommended in such profile patients [9] and expectations of the patient should well enlighten before consent for surgery.

The prognosis of patients diagnose lately for rheumatic mitral stenosis with giant left atrium (ectasia) depends largely on some factors; pulmonary hypertension, enlarged left atrium with necessity reduction. In this series, El Maghraby [11] et al. found that the incidence of giant left atrium (GLA) in rheumatologic valve disease was about 0.6%, which is 11–12 times more common than non-rheumatological valve diseases. Other methods of reduction of giant left atrium could be considered like modified volume reduction technic [12], plasty reduction [13], Pomerantzeff's technique [14] or partial cardiac auto transplantation [15] reduction of the left atrium or new methods like inflow artifact reduction [16]. These factors should always be highlighted during heart discussion before any therapy strategy and patients should be always enlighten of the worse outcomes if surgery is done in the presence of irreversible pulmonary hypertension.

4. CONCLUSION

Giant left atrium secondary to rheumatic mitral valve disease can be of poor prognosis in the presence of irreversible pulmonary hypertension and right heart catheterization is important before any decision of surgical treatment. In the case of possible reversible pulmonary hypertension, left atrium reduction by any suitable methods could be practice as some studies shows their efficacity in atrium remodeling. Early heart valve screening is important in rheumatic disease patients to reduce morbi-mortality. Cardiac MRI is an important tool which can add precision to the diagnose of left atrium surface area and volume.

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

CONSENT

As per international standard or university standard, patient(s) written consent has been collected and preserved by the author(s).

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COMPETING INTERESTS

Authors have declared that no competing interests exist.

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