Case study

GIANT LEFT ATRUIM IN RHEUMATIC MITRAL VALVE DISEASE, WHAT PROGNOSIS? (About 3 cases).

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 pulmonary hypertension, possible LAP associated with other comorbidities.

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

Keywords : Giant left atrium (ectasia), Mitral stenosis, Pulmonary Hypertension, LAP

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 rhematic mitral stenosis with giant (ectasia) left atrium and high pulmonary hypertension who in spite of surgical valve replacement shortly died because of persistent pulmonary hypertension due to lack of left atrial remodeling and left ventricular adaptation.

Patients and Method

We report the cases of 3 young female patients aged 48, 42 and 58 years respectively, with a

Comment [OAG1]: 1)This seciton should be called as : CASE DESCRIPTION or CINICAL CASES 2) I recommend to describe each case as separate as Case 1, Case 2, and Case 3. In this way, a better understanding for the audience can be reached.

history of non-surgical rheumatic mitral valve disease evolving over 20years in the first patient, 17 years in the second patient and 15 years in the third patient respectively under acenocoumarol (Sintrom) tablets daily without regular fellow-up. Only the third patient was menopaused whereas the other patients had neither other personal, family medical history nor surgical history. The eldest and youngest patients first consulted at the emergency department of cardiology at the University teaching Hospital, Ibn Rochd of Casablanca for worsen dyspnea and altered general health state whereas the younger patient consulted at our outpatient cardiology department for medical follow-up referred by another hospital in the sub-region. The clinical examination of the eldest and younger patients found patients in altered health state with stage IV NYHA dyspnea, edema of both lower limbs rising up to knees with no acute articular disease. At auscultation, bilateral midfield crackles, irregular heartbeat, diastolic rolling at the mitral focus and burst of B2 sound at the pulmonary valve focus was found in the eldest and youngest patients whereas the younger patient had irregular heartbeat and diastolic rolling at the mitral focus. The patients admitted at the emergency department of cardiology had an ambient air oxygen saturation of 96% and 98% respectively, blood pressure of 150/90mmHg and 130/70mmHg whereas the younger patient's BP was normal at the out-patient consultation. The routine ECG of the three patients showed atrial fibrillation (AF) with an accelerating rhythm of more than normal in the first patient 110bpm in the hospitalized patients (Figure 1).

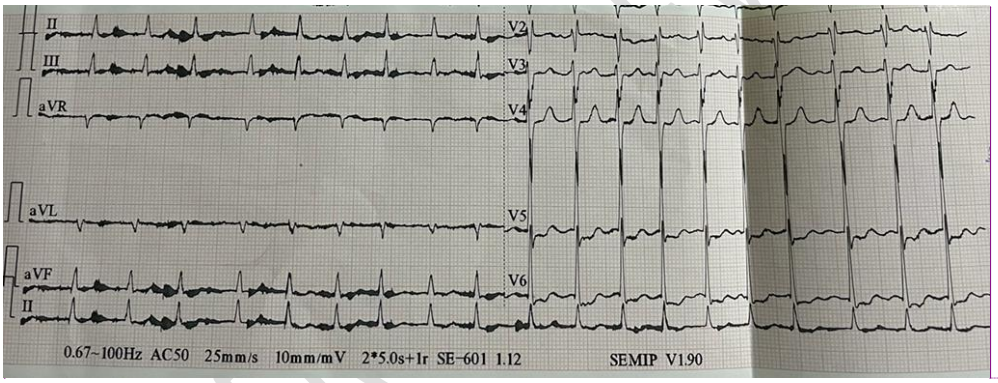

Figure 1: Electrocardiogram of the 58 years old patient: Accelerated atrial fibrillation (AF).

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A complete blood analyses was carried out in the patients including complete blood count test, electrolytes which showed lymphopenia at 640/µL, C-reactive protein (CRP)at 43.5mg/l in the eldest patients, whereas the youngest patient had high white blood count at 12000/µL with predominant neutrophil. The INR ratios were normal in all three patients 2.5, 3.14, 2.8 respectively. Transthoracic echocardiography was performed in all three patients which confirmed a severe mitral stenosis with mitral surfaces of 0.6cm^2, 0.9cm^2 and 0.8cm^2 respectively and transvalvular gradient of more than 8mmHg. The mitral valves where heavily calcified at their free leaflet sides with commissural fusion in all three patients. Only the two younger patients have mild mitral regurgitation whereas the eldest had a severe mitral regurgitation, with a high pulmonary hypertension estimating the systolic pulmonary pressure at 131mmHg whilst the younger had a systolic pulmonary pressure at 80mmHg. Only the youngest patient had a less elevated pulmonary pressure of 35mmHg. Their ventricular ejection fraction was normal except the eldest patient with an EF of 49%. Ectasia of the left atrium with the highest volume and atrial surface area was found in the younger patient 980ml/m^2 indexed volume and 129cm^2 left atrial (LA) surface area whilst the eldest patient had 616ml/m^2 indexed volume, 102cm^2 LA surface area, the youngest patient had 390ml/m^2 indexed volume, 80cm^2 LA surface area respectively (Figure 2a, b, c). Severe tricuspid regurgitation in the younger and eldest patients (Figure 3) while the younger had a mild to moderate tricuspid regurgitation.

There were no other abnormal cardiac valves.

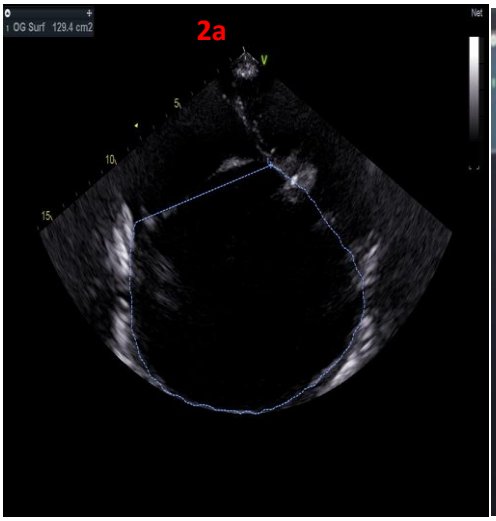
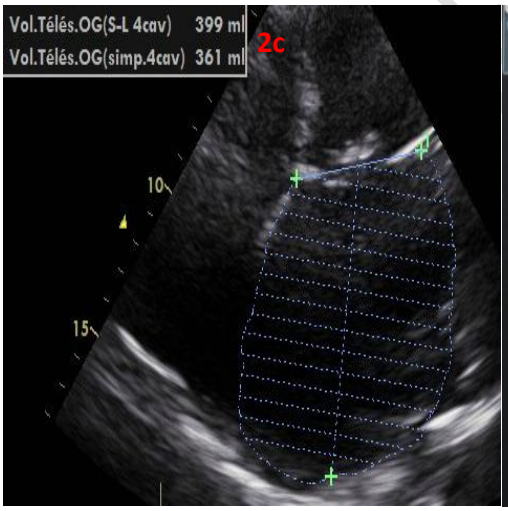


Figure 2a: Transthoracic echocardiography (TTE): 4 chambers apical view: showing a giant left atrium (ectasia) occupying the whole monitor screen with a surface area of 129.4cm^2.

Figure 2b: 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.



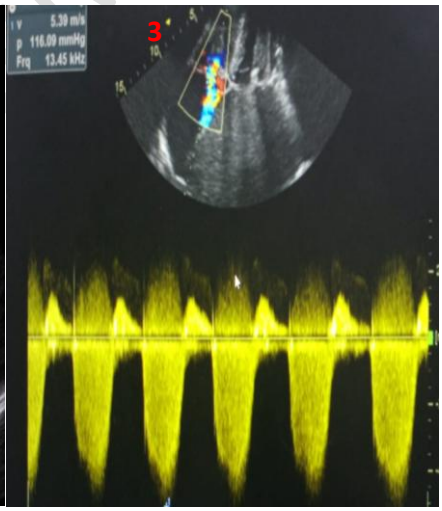


Figure 2c: TTE: 4 chambers apical view: showing an ectasia of the left atrium with a surface volume of 399ml associated with rheumatic mitral valve stenosis

Figure 3: TEE: 4 chambers apical view of the eldest patient: CW doppler flux of tricuspid valve regurgitation with systolic pulmonary pressure estimated at 131mmHg.

CT-scan was performed in the eldest 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 patients were put under medical therapy based on

restriction hydric intake for the younger and eldest patients 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. The youngest patient was only put on enzyme converter inhibitor, betablocker and small dose of loop diuretic. Patients were later programmed for surgery, mechanic valve replacement and tricuspid plasty was performed in the younger patient who died 24hours after surgery due to pulmonary hypertension as the surgery was only based on mechanic mitral valve replacement without left atrium plication (LAP) (Figure 4), the youngest refused surgery and the eldest died later due to renal failure. Right heart catheterization was not done our operated patients.

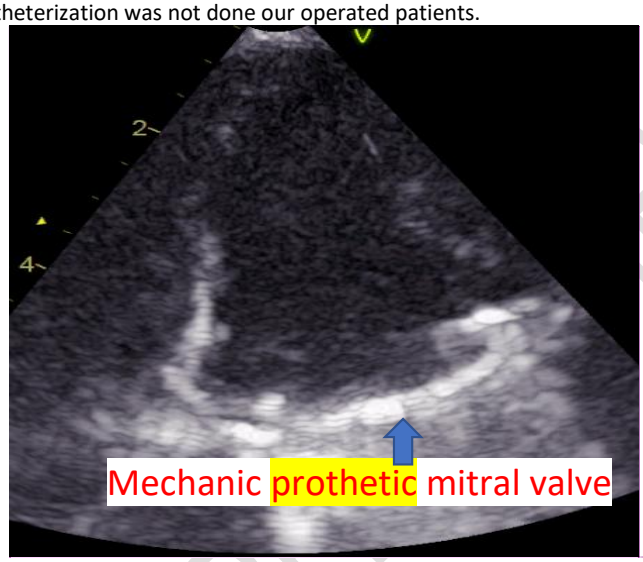


Figure 4: TTE: 4 chambers apical (operated patient) view zoomed at the mechanical mitral prosthetic valve replacement with shadows of rings in an ectasia atrium.

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 due to long-term evolution of rheumatic mitral valve disease, approximately 10years after diagnosis of MS can be really challenging when it comes surgical therapy. 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 Cardiac 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 clinical decision making in patients with severe pulmonary hypertension to estimate the risk of surgery (9). However, in our case, right cardiac catheterization was not done in the patients especially the operated patient because of lack of patient consent of the patient and pressure to operating even her surgical risk was very high.

Comment [OAG3]: Mechanical prosthesis mitral valve

Comment [OAG4]: Please, support all comments with REFERENCE citations.

Comment [OAG5]: This pharagraph should be eliminated. It is assumed GLA is due to long term MV disease, whatever the etiology.

Comment [OAG6]: It would be more proper just mentioning that « in our three cases, right catheterizarion 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 our results show that it can be performed at least without a detrimental effect (5). In our cases, only one patient was operated without LAP which led to a rapidly increase in diastolic volume pressure from the left atrium non supported by her left ventricular muscles. Its important to note without LAP 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 with the presence of irreversible pulmonary hypertension, immediate post operated patients have a slim chance of living.
Irreversible pulmonary hypertension is an important determinant factor in these cases as its crucial in decision making as patients in spite of valve replacement, LAP will remain symptomatic without reducing re-hospitalization rate for heart failure and death thus absolutely necessary for such patients to undergo right cardiac catheterization before surgery.
Other methods like percutaneous balloon mitral valvuloplasty (PBMV) or transcatheter mitral valve lithotripsy (TMVL) cannot be use especially if LAF 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, possible LAP. 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.

Comment [OAG7]: Support this statement with REFERENCE citation.

Comment [OAG8]: Support this method with REFERENCE citaiton

Comment [OAG9]: What does it mean this term ?

Comment [OAG10]: The aurhors are including only LAP, but missing the LEFT ATRIAL REDUCTION as a method to treat GLA. It should be included into this manuscript.

Conclusion

Giant left atrium secondary to rheumatic mitral valve disease can be of poor prognosis in the presence of irreversible pulmonary hypertension and right cardiac catheterization is important before any decision of surgical treatment. In the case of possible reversible pulmonary hypertension, LAP 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.

Comment [OAG11]: The current Conclusions cannot be accepted as it stands now. Other possible surgical techniques such as LA reduction, etc must be included. Also, the possible association with an increase likelihood for thrombus formation, LA appendage involvement, as well as the role of the MRI in the diagnosis for these cases.

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