Case study

Lutembacher's syndrome: a case report from hospital IBN Rochd of Casablanca, Morocco

ABSTRAT

Lutembacher's syndrome (LS) refers to the rare combinaison of acquired mitral stenosis (MS) with congenital atrial septal defect (ASD). Other forms described in the literature include iatrogenic LS and reverse LS. LS is a very rare entity with a female predominance, it has been either overdiagnosed or misdiagnosed. The prognosis is good before the onset of pulmonary hypertension and right heart failure. This condition is usually treated surgically by mitral valve operation with concomitant closure of the atrial septal defect. We report the observation of a 62-year-old female presented with a one year history of progressive shortness of breath. Cardiac ultrasound lead to the diagnosis of this rare clinical syndrome. The patient was referred for mitral valve replacement with ASD closure.

Keywords: Mitral stenosis (MS); Atrial septal defect (ASD); Lutembacher'syndrome (LS); Morocco.

INTRODUCTION

Most cardiovascular conditions are either acquired or congenital in origin, but in rare instances, a combination of both is found. Lutembacher syndrome (LS) refers to the uncommon combinaison of acquired mitral stenosis (MS) and congenital atrial septal defect (ASD) . Other forms described in the literature include iatrogenic LS and reverse LS (1). Mitral stenosis refers to mitral valve leaflets (or valve flaps) sticking to each other making the opening for blood to pass from the atrium to the ventricles very small(2). With the valve being so small, blood has difficulty passing from the left atrium into the left ventricle. Septal defects that may occur with Lutembacher's syndrome include: Ostium primum atrial septal defect or ostium secundum which is more prevalent(3) . Lutembacher's syndrome affects females more often than males(4). It can affect children or adults; the person can either be born with the disorder or develop it later in life. The syndrome was first described by René Lutembacher (1884–1968) of Paris in 1916(5).

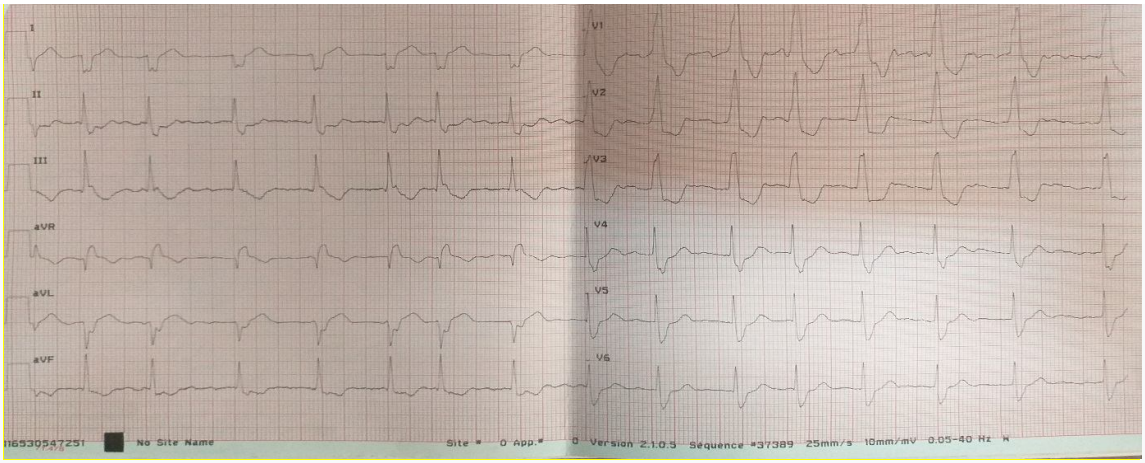
For most people, they will remain asymptomatic (experience no symptoms) but when symptoms are shown, they are due mainly to ASD and will vary depending on the size of the hole in the atria. If the patient has a large ASD, pulmonary congestion (blood or fluid buildup in the lungs) will happen later but if the patient has a small ASD, symptoms will appear early in the disorder. Major symptoms of Lutembacher's syndrome as a result of ASD and MS can range from heart failure to pulmonary congestion(6).The prognosis is good before the onset of pulmonary hypertension and right heart failure. Surgical and percutaneous trans-catheter therapies with balloon valvuloplasty and septal closure using an Amplatzer closure device have proven to be beneficial (7).

We report the case of an adult female who presented with this rare clinical syndrome in casablanca's hospital.

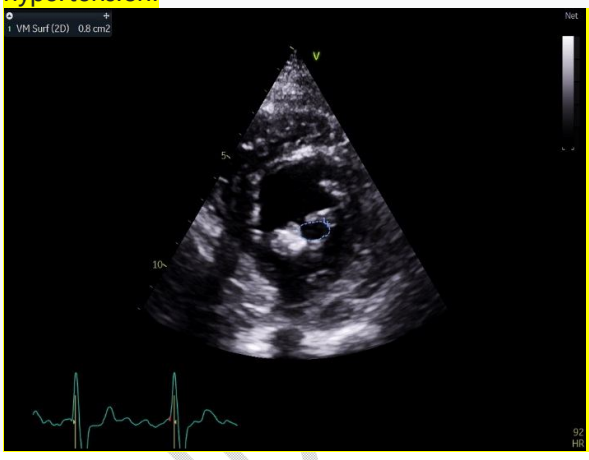
CASE REPORT

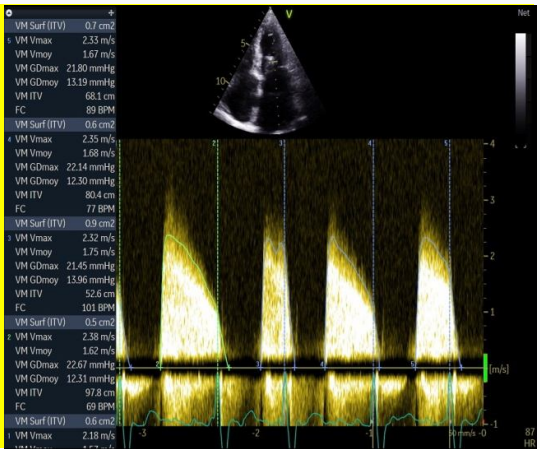
A 62-year-old woman presented for consultation for exertional dyspnea that had been evolving for about a year. She reports that for about a month before her consultation, dyspnea would occur with less and less significant efforts, and would be accompanied by physical asthenia. The interrogation did not find any antecedents of acute articular rheumatism in childhood. On examination, she was afebrile with a pulse of 81 beats/min and blood pressure of 130/70 mm Hg, with a respiratory rate of 20 breaths/min. On cardiac examination, there was a 4/6 systolic murmur at the mitral focus, associated with a mitral opening click. At the pulmonary focus, there was a burst of B2. Lung auscultation revealed equal bilateral normal breath sounds with bilateral end-inspiratory crackles in the lower areas.

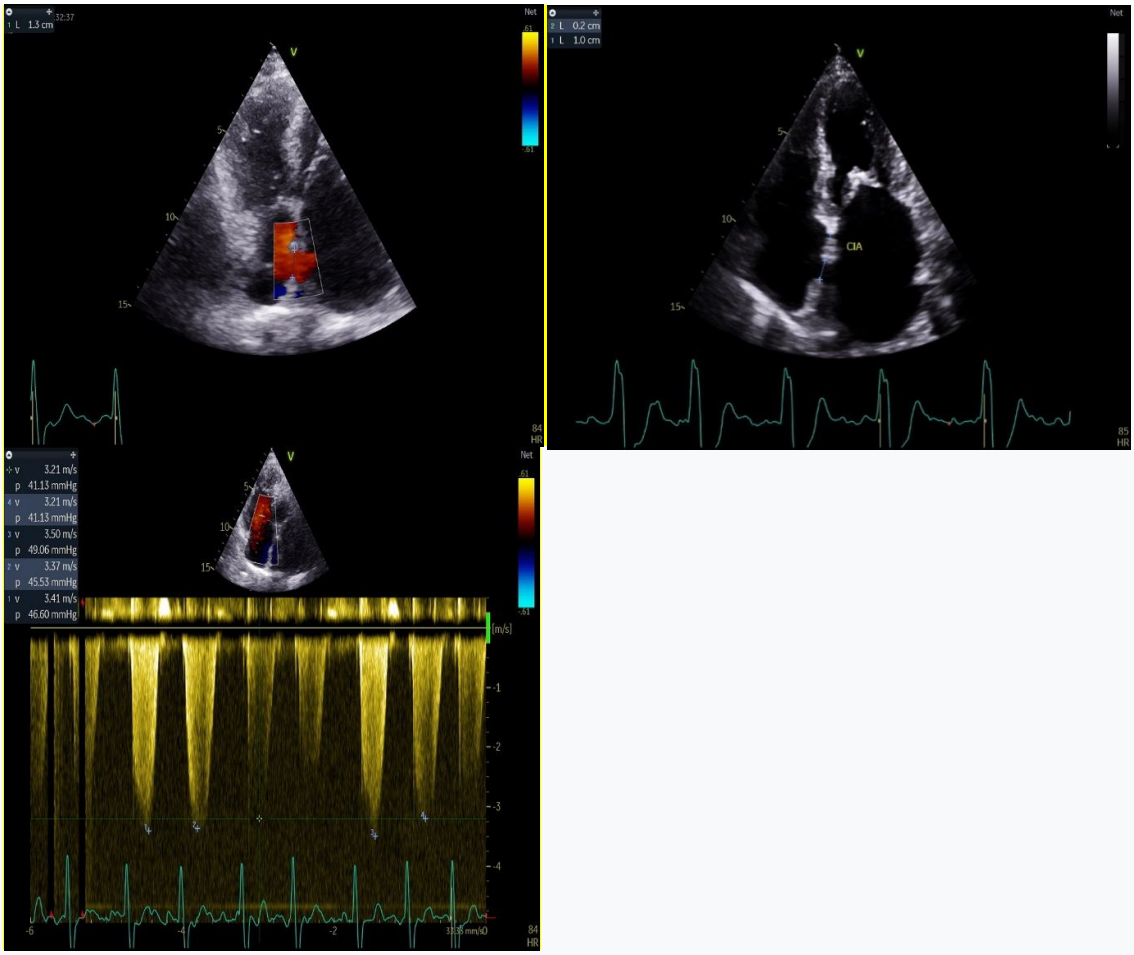
Electrocardiogram [Figure 1]→ Atrial fibrillation Rhythm; Incomplete Right Bundle Branch Block (RBBB); Left Atrial Enlargement; Normal Qtc.



2D-Echocardiography [Figures [Figures 2 and 3,4,5,6] → Very Tight Mitral stenosis; Mild Mitral regurgitation; mitral valve area 0.70-0.80 cm2 ; mean averaged transmitral gradient: 13 mmHg.Large Ostium Secondum atrial septal defect (10 mm) with Left to Right Shunt; Moderate pulmonary arterial hypertension.







Other Valves- Normal. No Clots/No Effusion/No Vegetations.

The patient is put on diuretics, antiarrythmics and anticoagulants.

She was referred for mitral valve replacement with ASD closure.

DISCUSSION

Classically, Lutembacher's syndrome(LS) is defined as the association a left-right interatrial shunt and a mitral stenosis (MS). As a general rule, the shunt is related to congenital ostium secondum atrial septal defect (ASD), and MS is most often of rheumatic origin. Some authors have proposed to broaden the definition of this syndrome, to the association of ASD and mitral valve damage: insufficiency, stenosis, and mitral disease. Rare cases have also been reported as the association of an acquired rheumatic MS and an atrioventricular canal[8].

In developing countries, LS remains certainly more important because of the prevalence of rheumatic fever[9]. The incidence of this condition is very rare, with female predominance. In one study published in American Heart Journal in 1997, it is found that the incidence of Lutembacher's syndrome is- 0.001/10,00000[10]. Like us, in Morocco, Nassour et al, reported the case of a female patient who presented with rheumatic mitral stenosis associated with ASD[11].

Usually, the attack predominates in the young subject, but it remains possible to find it in older patients. The reported case is about a 61-year-old woman. Sophie Monin et al, in France, also described the case of a patient diagnosed during her sixtieth year of life [12].

The hemodynamic effects of this syndrome are the result of interactions between the effects of mitral stenosis and atrial septal defect. As a result, in the presence of mitral stenosis, blood flows to the right atrium through the ASD instead of going backward into the pulmonary veins, thus avoiding pulmonary congestion[13]. This happens at the cost of progressive dilatation and, ultimately, failure of the right ventricle and reduced blood flow to the left ventricle. Development of Eisenmenger syndrome or irreversible pulmonary vascular disease is very uncommon in the presence of large ASD and high left atrial pressure because of mitral stenosis[14].

Early diagnosis and surgical treatment bears a good prognostic value. If patient is diagnosed at late stage, pulmonary hypertension and heart failure develops and the prognosis is bad. In our clinical case, the patient consulted one year after the onset of symptoms, and presented at the stage of pulmonary hypertension. In Africa, the problem of access to health care is particularly complex, marked by a weak of the supply of care, the shortage of human resources, the inadequacy of the quality of care and the widespread absence of basic medical coverage.

However, if the patient is diagnozed earlier before the development of pulmonary hypertension and heart failure, ASD closure with mitral valve replacement bears a good prognosis and prolongs survival[15].

Cardiac glycosides, beta blockers and calcium channel blockers may be used for rate control while drugs like amiodarone, besides rate control, also help in achieving and maintaining a normal sinus rhythm. diuretics such as furosemide are generally used to ameliorate symptoms of right heart failure[16]. Open heart surgery had been the preferred method of treatment of patients with LS

involving ASD closure and mitral commisurotomy or valve replacement [17]. Recently, progress in interventional cardiology has significantly changed the treatment of LS with trans-catheter therapies (in eligible patients) with impressive success rates[18].

CONCLUSION

Lutembacher's syndrome (LS) is a rare clinical entit, and a challenge for clinical diagnosis due to the masking of the signs and symptoms of mitral stenosis by shunt through the defect of the interatrial septum. Echocardiography assessment is the current diagnostic modality of choice with 3D echo and TEE further helpful in excluding co-existent cardiac pathologies. Planimetry by Doppler echo remains the best method for assessing MVA. If diagnosed early, patients benefit from surgical or percutaneous trans-catheter therapy. The outcome is better if treated before the onset of heart failure and pulmonary hypertension. However, surgical and percutaneous trans-catheter therapy is costly and not readily available in low-income settings in developing countries.

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