Lutembacher's Syndrome: A Case Report from Hospital IBN Rochd of Casablanca, Morocco

ABSTRACT

Lutembacher syndrome (LS) is a rare clinical entity, associated with acquired mitral stenosis (MS), congenital atrial septal defect (ASD). In the literature, other forms have been described including iatrogenic LS and reverse LS. LS is a condition with a female predominance, and, over-diagnosed, is badly suffering, making its particularity. The prognosis for this syndrome is best before the onset of pulmonary hypertension and right heart failure. LS is usually treated surgically by mitral valve surgery with concomitant closure of the atrial septal defect. We report in this case report, the observation of a 62-year-old woman consulting for dyspnea evolving for about a year, in a context of physical asthenia. Cardiac ultrasound led 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's Syndrome (LS); Morocco.

1. INTRODUCTION

"The majority of cardiovascular conditions are of acquired or congenital origin, but in rare cases an association of the two is found" [1]. "Lutembacher syndrome (LS) refers to a rare combination of acquired mitral stenosis (EPS) and congenital atrial septal defect (ASD). Mitral stenosis is a narrowing of the mitral valve that obstructs blood flow from the left atrium to the left ventricle" [2]. The ASD is a solution of continuity between the two atria (left and right), consequence of a deficit at the level of their common wall. ASD ostium secundum type is the pathological type most commonly associated with this syndrome, than other entities [3]. Women are more affected by this syndrome than men [4]. It can affect children or adults, just like, be congenital or acquired. The syndrome was first described by René Lutembacher (1884-1968) from Paris in 1916 [5].

The clinical picture varies from patient to patient. The bulk of patients are asymptomatic, but when symptoms do occur, they are primarily due to ASD and vary depending on the size of the defect between the atria. The larger the defect, the later the pulmonary congestion will occur, on the other hand the smaller the defect, the earlier the symptoms will appear. The main symptoms of Lutembarcher syndrome resulting from ASD and MS can range from heart failure to pulmonary congestion [6]. "Surgical and percutaneous catheter therapies with balloonvalvuloplasty and septal closure using an Amplatzer closure device have been shown to be beneficial" [7].

We report the case of an adult woman who presented with this rare clinical syndrome at the Casablanca hospital.

2. CASE REPORT

A 62-year-old womanpresented for consultation for exertionaldyspneathathad been evolving for about a year. She reports that for about a monthbeforeher consultation, dyspneawouldoccurwithless and lesssignificant efforts, and wouldbeaccompanied by physicalasthenia. The interrogation did not

findanyantecedents of acute articularrheumatism in childhood. On examination, shewasafebrilewith a pulse of 81

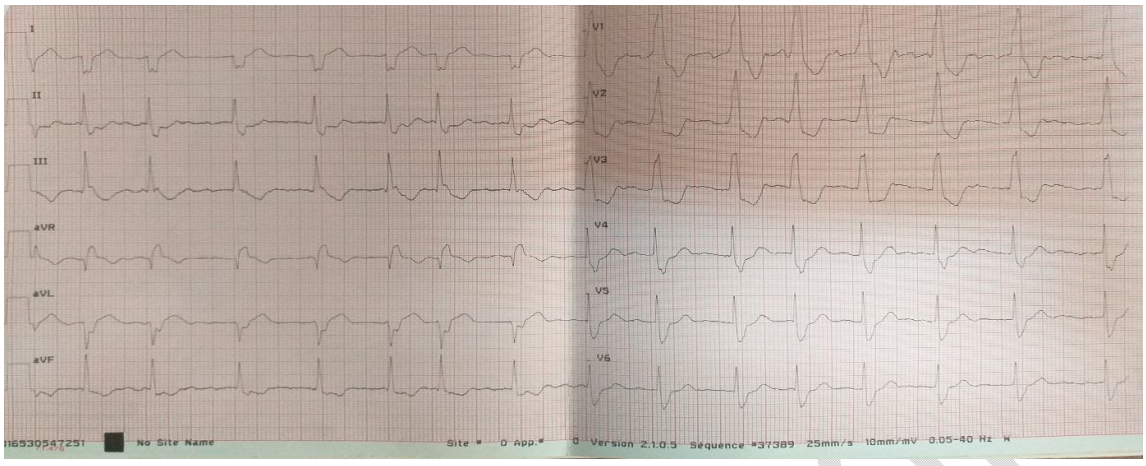

Fig. 1. Electrocardiogram : Atrial fibrillation rythm; Incomplete right bundle branch block (RBBB) ; Left atrial enlargement; Normal QTC.

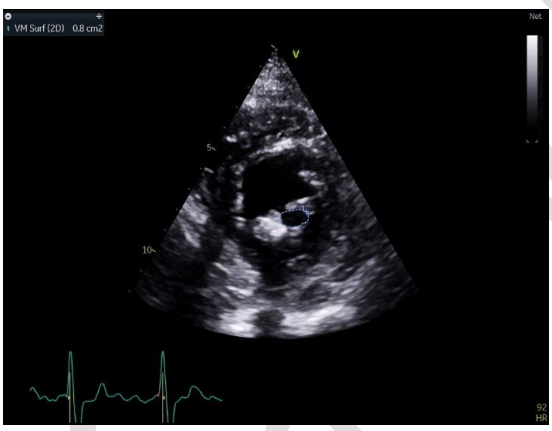

Fig. 2. Verytight mitral stenosis ; Mitral valve area : 0,70-0,80 cm²

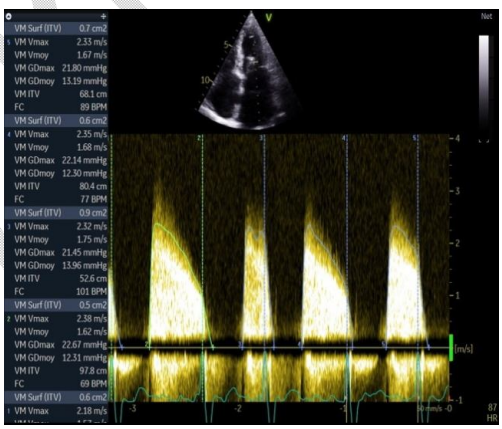

Fig. 3. Meanaveragedtransmitral gradient: 13 mmHg.

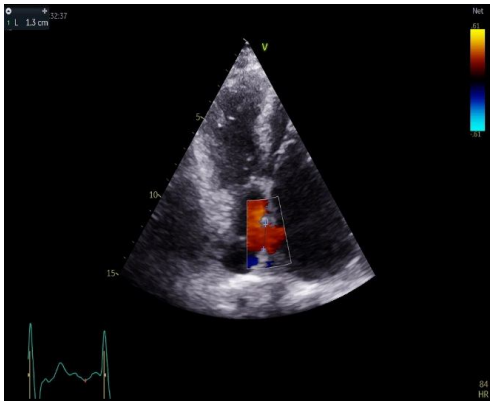

Fig 4. Large ostium secondum atrial septal defect (10 mm) withleft to right shunt

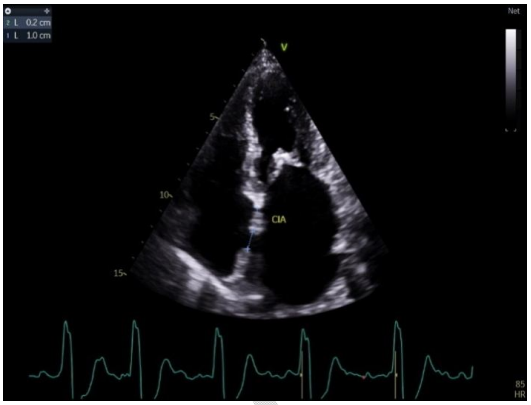

Fig. 5. Large ostium secondum atrial septal defect (10 mm) withleft to right shunt

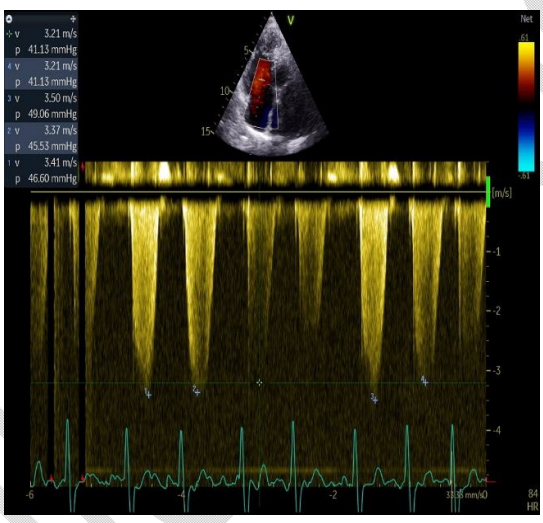

Fig. 6. Moderatepulmonaryarterial hypertension

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beats/min and blood pressure of 130/70 mm Hg, with a respiratory rate of 20 breaths/min. On cardiacexamination, therewas a 4/6 systolicmurmur at the mitral focus, associatedwith a mitral opening click. At the pulmonary focus, therewas a burst of B2. Lung auscultation revealedequalbilateral normal breathsounds.

Other valves were normal looking. No clots ; No effusion ; No vegetations.

The patient is put on diuretics, antiarrythmics and anticoagulants.
After diagnosis, the patient was referred for surgery for mitral valve replacement with ASD closure.

3. DISCUSSION

Classically, Lutembacher's syndrome (LS) isdefined as the association a left-right interatrial shunt and a mitral stenosis (MS). As a generalrule, the shunt isrelated to congenital ostium secondum atrial septal defect (ASD), and MS ismostoften of rheumaticorigin. Someauthors 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 acquiredrheumatic MS and an atrioventricular canal [8].
In developing countries, LS remainscertainly more important because of the prevalence of rheumaticfever[9]. The incidence of this disease is low with a female predominance. Its incidence

was around 0.001/1000000 in a study published in the American Heart Journal in 1997 [10]. Like us, in Morocco, Nassour et al, reported the case of a female patient whopresentedwithrheumatic mitral stenosisassociatedwith ASD [11].

Usually, the attackpredominates in the youngsubject, but itremains possible to findit in older patients. The reported case is about a 61-year-old woman. Sophie Monin et al, in France, alsodescribed the case of a patient diagnosedduringhersixtiethyear of life [12].

"The hemodynamic manifestations of this syndrome are the consequence of interactions between the effects of mitral stenosis and atrial septal defect. Indeed, following the presence of a mitral stenosis, the blood flows towards the right atrium through the ASD instead of going up in the pulmonary veins, thus avoiding pulmonary congestion" [13]. Secondarily, "there will be progressive dilation, failure of the right ventricle, and reduced blood flow to the left ventricle. However, the development of Eisenmenger syndrome or irreversible pulmonary vascular disease is very rare in the presence of a large ASD and high left atrial pressure due to mitral stenosis" [14].

The earlier the diagnosis and the surgical treatment, the better the prognosis. Unfortunately, when the patient is diagnosed at an advanced stage, pulmonary arterial hypertension and heart failure develop, making the prognosis bad. In ourclinical case, the patient consulted one yearrafteryheonset of symptoms, and presented at the stage of pulmonary hypertension. In Africa, the problem of access to health care isparticularlycomplex, marked by a weak of the supply of care, the shortage of humanresources, the inadequacy of the quality of care and the widespread absence of basic medicalcoverage.

Proper management of this rare syndrome is based on early diagnosis, ie before the installation of pulmonary arterial hypertension, followed by closure of the ASD with replacement of the mitral valve. This would improve prognosis and prolong survival [15].

"Cardiac glycosides, beta-blockers and calcium channel blockers will be used to control heartrate; while medications like amiodarone, in addition to rate control will also help achieve and maintain normal sinus rhythm. Diuretics such as furosemide are generally used to relieve symptoms of right heart failure" [16]. Initially, "the preferred method of treatment for patients with LS was open-heart surgery, involving closure of the ASD and mitral commissurotomy or valve replacement" [17]. Recently, "with advances in interventional cardiology, the treatment of LS has changed dramatically using trans-catheter therapies (in eligible patients) with impressive success rates"[18].

4. CONCLUSION

Lutembacher's syndrome (LS) is a diagnostic challenge due to the masked nature of signs and symptoms of mitral stenosis by shunt across the atrial septal defect. Echocardiographic assessment is the current diagnostic modality of choice, with 3D echo and trans-oesophageal ultrasound also useful in excluding coexisting cardiac pathologies. When patients are diagnosed early, they benefit from surgical or percutaneous catheter treatment. The result is better if the treatment is done before the onset of heart failure and pulmonary hypertension. However, surgical and percutaneous transcatheter therapy is expensive and not available in low-income settings in developing countries.

CONSENT

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

ETHICAL APPROVAL

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

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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