



Lutembacher Syndrome from the State of Bihar, India: A Case Report

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ABSTRACT

Lutembacher syndrome refers to a combination of an atrial septal defect and a mitral stenosis (MS). It is a rare disease in which both the pathologies intermix and cause changes to the hemodynamics and resultant disease course. This is to report a case of Lutembacher syndrome from the state of Bihar.

Keywords: Congenital heart disease, Lutembacher syndrome, Mitral stenosis.

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INTRODUCTION

Lutembacher syndrome is defined by the occurrence of a congenital atrial septal defect and mitral valve stenosis together.¹ The valvular stenosis is usually acquired, mostly rheumatic in origin. It is a rare entity with the incidence of atrial septal defect (ASD) in MS patients being 0.6 to 0.7 percent.² This is to report one such case of a 25-year-old lady who was a typical example of the interplay between both the lesions which changes the course of the disease. As far as known this is the first case report from this part of the world.

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CASE REPORT

A 25-year-old lady presented to the outpatient department with complaints of breathlessness, palpitation, lower leg and abdominal swelling and abdominal discomfort which were present for the past 1 month. Breathlessness was gradual in onset but had worsened over the last one month so much so that she had problems with ordinary activity (New York Heart Association grade 3). There was no associated cough or fever and no history of similar complaints. The leg and abdominal swelling had come around the same time as the breathlessness worsened.

There was no history suggestive of a renal or hepatic pathology. The patient had had two event free pregnancies with the youngest child being 2 years old. On examination, the positive findings were bilateral pitting pedal edema, an irregularly irregular pulse with a heart rate of around 104/minute, a blood pressure of 108/58 mmHg and an elevated jugular venous pressure. Systemic examination revealed a left parasternal lift, loud s1, loud p2, a Mid-Diastolic murmur typical of mitral stenosis and a right-sided s3. Abdominal exam revealed ascites and tender hepatomegaly. Till this point, the diagnosis was a case of rheumatic heart disease, mitral stenosis and right ventricular failure with atrial fibrillation. She was started on diuretics, beta blockers, and digoxin and advised investigations.

However, the fact that even with this level of heart failure, the patient had had two uncomplicated pregnancies made us suspicious. Amongst the investigations, the ECG showed atrial fibrillation with controlled ventricular rate (Fig. 1). X-ray was suggestive of mitral stenosis. The lung fields were plethoric (Fig. 2).

It was the echocardiographic findings that clinched the diagnosis. It showed a large 29 mm septum secundum type of ASD accompanying the severe (0.9 cm²) mitral stenosis. There was accompanying right atrial and ventricular dilatation.

The left ventricular function was normal with an EF of 65%.

Echo Report

Left Ventricle

EDD 40 mm

IVS 08 mm intact

EDD 23 mm

EF 65%



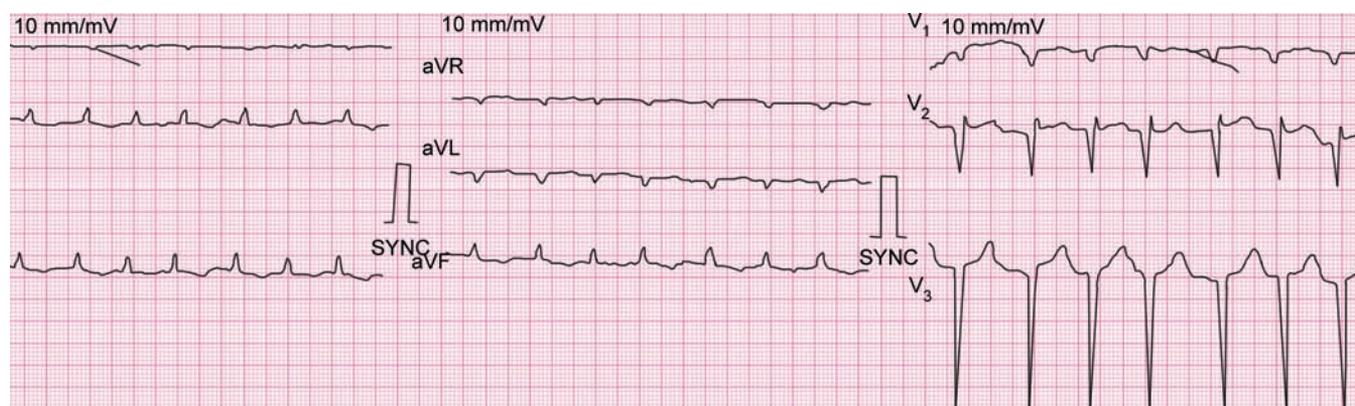


Fig. 1: ECG showing atrial fibrillation

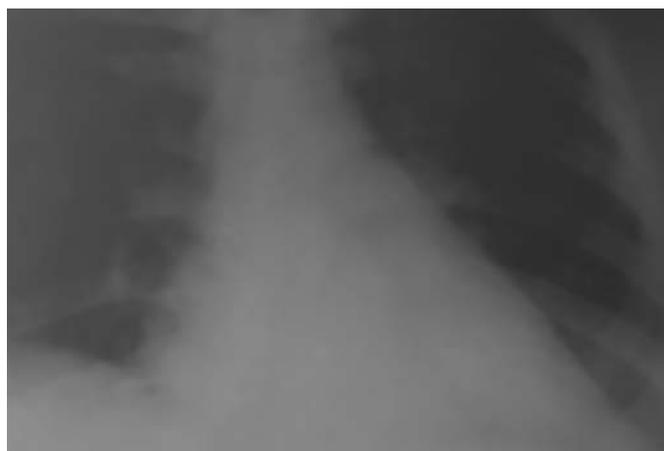


Fig. 2: X-ray showing signs of mitral stenosis

LEFT ATRIUM 48mm
 RIGHT VENTRICLE: Dilated
 RIGHT ATRIUM: Dilated
 IAS: Large Secundum type ASD of size 29mm with left to right shunt
 Mitral Valve: thickened, PMI restricted and paradoxical, doming MVA of 1.0cm²
 MVA by planimetry 0.9 cm²
 Velocity across mitral valve: E 2.04 A 1.44
 Gradient: Peak 16 mean 9

Routine hematological and biochemical parameters including complete blood count, kidney function tests, liver function tests, random blood sugar, and urine examination were within normal limits.

Clinical and echocardiographic findings the patient was diagnosed as a case of Lutembacher syndrome. She was treated for congestive heart failure and referred for surgical correction to All India Institute of Medical Sciences Delhi.

DISCUSSION

Lutembacher syndrome as originally described was an ASD with an MS by a French physician Rene Lutembacher in 1916.³ The incidence of the syndrome has gone down in the western world to a great extent due to the elimination

of rheumatic heart diseases. However, it continues to be reported from developing countries like India, where rheumatic heart disease is still common.

The most important thing to consider here is the hemodynamics that changes with these two lesions coming together. ASD provides a second outlet for the blood in the left atrium which is usually at high pressure in MS and hence results in a left to right shunt across the ASD. This shunt reduces the symptoms of pulmonary congestion and breathlessness. Development of pulmonary hypertension is also delayed. However, as a result of the left to right shunt right ventricle gets overloaded and eventually fails as happened in this patient.

To summarize, the symptoms and signs of MS including mid-diastolic murmur loud S1 and signs of pulmonary hypertension are modified due to the decompression of LA across the ASD.⁴ However right ventricular volume overload and later right ventricular failure occur is the result. The patient presented in this case report experienced two event-free pregnancies even in the presence of severe MS but eventually got symptomatic due to right ventricular overload.

Treatment^{5,6} for Lutembacher syndrome involves medical treatment for right heart failure by diuretics and atrial fibrillation treatment by beta blockers, calcium channel blockers and/or digoxin followed by closure of ASD either by percutaneous device closure or surgical closure and relief of MS either by balloon valvuloplasty or open commissurotomy. There have been many case reports of Lutembacher syndrome from India.^{7,8} To our knowledge this is the first case report from Bihar, India.

CONCLUSION

Lutembacher syndrome is a rare entity, and it should be kept in mind as a differential diagnosis when mitral stenosis presents atypically. The diagnosis rests on echocardiographic findings and the patient should be referred for surgical/non-surgical correction (ASD closure and mitral valvotomy) promptly.

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