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Case Report

A rare case of lutembacher syndrome in a young female: a case report from a rural population of Western Uttar Pradesh, India

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ABSTRACT

Lutembacher syndrome is a rare entity presenting with a combination of congenital atrial septal defect with acquired mitral stenosis. Lutembacher syndrome is reported to be more prevalent in developing countries where the incidence of rheumatic fever is high. We also came across with a young female with the similar clinical presentation in our hospital situated in a rural area in Western Uttar Pradesh, India. Keeping in mind its rare occurrence, we are presenting an overview of this syndrome including its various aspects and the problems faced by the patients in rural scenario.

Keywords: Lutembacher syndrome, Mitral stenosis, atrial septal defect, Rural population

INTRODUCTION

A 23 years old female presented to the Medicine OPD with complaints of chest discomfort on exertion. Past history revealed that she was communicated by her treating physician about two years back that she was suffering from heart disease which required surgery. However she decided to be on medical management as she could not afford to go for specialized urban hospital for her treatment due to financial constraints. Here in our new rural based multispecialty hospital, we investigated her thoroughly. 2D echocardiography revealed the combination of rheumatic mitral stenosis (MS) with atrial septal defect (ASD) and a diagnosis of Lutembacher syndrome (LS) were considered. Further she was referred to the Department of Cardiac Surgery of our institute. Surgical correction of the cardiac lesions was advised but she was not willing for it due to financial constraints and remains on medical treatment and regular follow-up.

CASE REPORT

A 23 years old female presented to the Medicine OPD of UP RIMS & R, Saifai, Uttar Pradesh, India with complaints of excessive chest discomfort on exertion and fatigue for the last 2 years and it had progressed gradually in the last 15 days. She also had mild unproductive cough for 7 years. Past history revealed that she was communicated by her treating doctor about her heart disease, the details of which were not known to the patient. According to her, surgery was advised during that time, but she did not undergo surgery due to financial constraints.

On physical examination, she was of moderate built and vitals were within normal limit. Cardiovascular examination showed diastolic murmur in mitral area. Respiratory examination was within normal limit. Initial working diagnosis as severe MS was kept and was taken for echocardiography for further assessment. 2D echocardiography showed thickened and doming anterior

mitral leaflet with thickened posterior mitral leaflet with restricted mobility. Left atrium, right atrium and right ventricle were dilated. These features were suggestive of severe MS. Along with it, she was also found to have an echogenic defect in interatrial septum (ostium secundum type) of the 24mm size with left to right shunt (Figure 1 and 2). All other valves morphology was within normal limit.

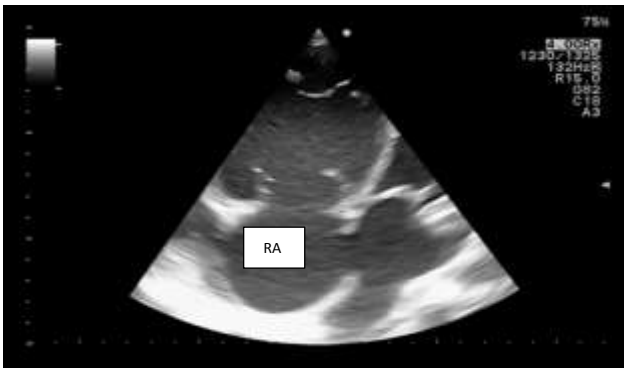

Figure 1: 2D Echocardiogram showing dilated right heart chambers with atrial septal defect.

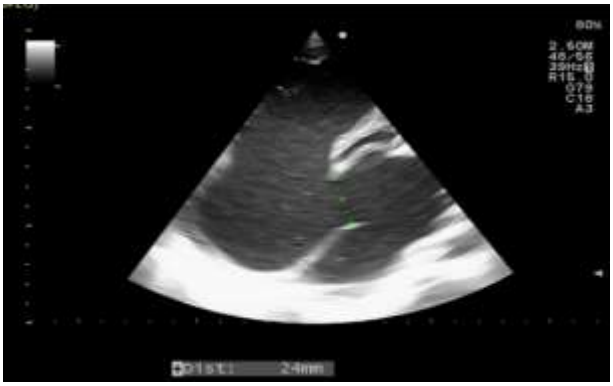

Figure 2: 2D Echocardiogram showing atrial septal defect – ostium secundum type (subcostal view).

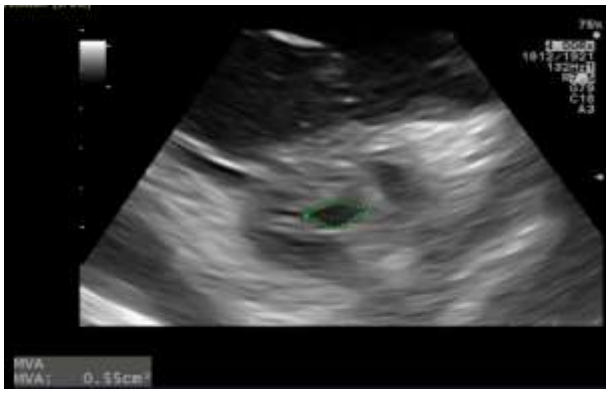

Figure 3: 2D Echocardiography measurement of mitral valve area by planimetry.

Mitral valve area was measured by planimetry as the color Doppler study at mitral level would not be reliable (Figure 3 and 4). Pulmonary pressure was on the higher side suggesting pulmonary artery hypertension. Left ventricular systolic function was reduced (44%) with normal right ventricular systolic function. This all showed features of severe MS accompanied with ASD, hence the diagnosis of Lutembacher syndrome was considered. ECG showed right ventricular enlargement with strain pattern with mild right axis deviation (Figure 5). Chest X ray showed cardiomegaly with right ventricular enlargement, left atrium enlargement with prominent bronchovascular markings (Figure 6).

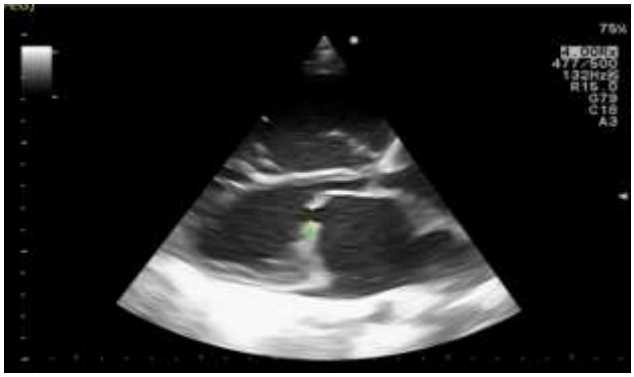

Figure 4: 2D Echocardiography depicting a stenosed mitral valve with dilated right ventricle in left parasternal long axis view.

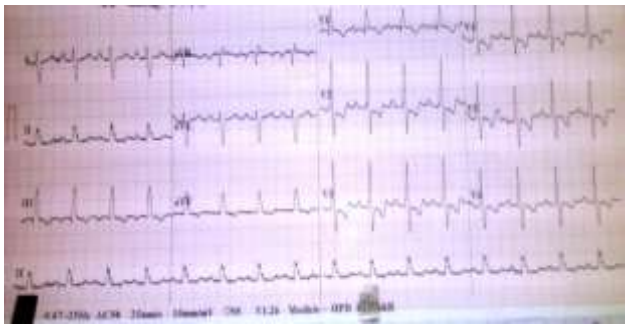

Figure 5: Electrocardiogram showed prominent R in V1-V2, notching of R in leads II, III, avf with mild right axis deviation.

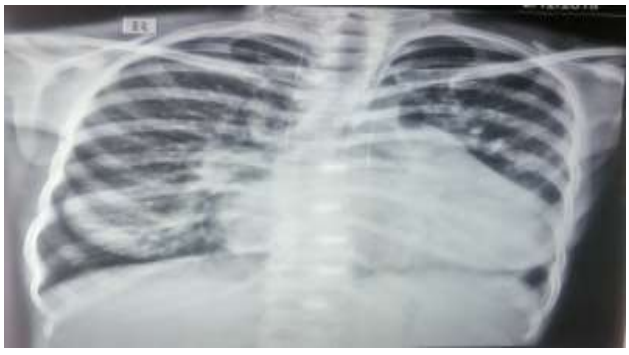

Figure 6: Chest X-ray showed cardiomegaly with right ventricular enlargement, left atrial enlargement with prominent bronchovascular markings.

She was further referred to the cardiac surgery department in our hospital for further plan of management where she was advised for surgical repair of the lesions. She opted for medical management presently and asked for the time to decide on the surgical intervention. The probable reason would probably being the same as financial constraints.

DISCUSSION

Corvisart first described the association of MS with ASD in 1811. Renè Lutembacher, a French physician born in 1884, first described the combination of rheumatic MS and ASD (usually of patent foramen ovale type) and published the first data described as LS in 1916.[1,2] ASD can be congenital (most commonly ostium secundum) or iatrogenic. MS can be congenital or acquired. Congenital MS is rare. The current consensus is that LS consists of a congenital defect in the atrial septum upon which acquired MS is imposed[4]. Usually MS is rheumatic in nature (probably due to recurrent attacks of rheumatic carditis) and its prevalence depends on the prevalence of rheumatic fever in that geographical area.[3] In underdeveloped countries, history of rheumatic fever has been reported in 40% of patient with LS.[4] Hence Lutembacher's syndrome defect occurs due to septum secundum defect and recurrent rheumatic carditis.[3] The syndrome is seen more commonly amongst females as also seen in our case. The incidence of MS in patient with ASD is 4% while the incidence of ASD in MS is 0.6 – 0.7%.[4] In contrast to this classic LS, another condition of iatrogenic LS has also been noticed. Iatrogenic LS occurs in the setting of percutaneous mitral balloon valvotomy for MS as atrial septal puncture is required to access the mitral apparatus. This condition can be well differentiated by echocardiography as explained below.

Hemodynamics – The syndrome presents with a different hemodynamic pattern that alter the clinical findings in these patients. The association of both ASD and MS has effect on each other. The presence of ASD decreases the gradient across the stenosed mitral valve and hence decreased murmur intensity. The classic opening snap and presystolic accentuation with sinus rhythm are not commonly seen. As a result of significant right ventricle enlargement, the left ventricle is shifted posteriorly leading to significant underestimation of auscultatory features of MS. Progressive left to right shunt initially helps to decompress the left atrium in MS, subsequently leads to right ventricle dilation, right ventricle pressure overload and volume overload and further progressive pulmonary hypertension. In severe cases that may have been left undetected for long, may have right to left shunt caused by severe pulmonary hypertension with dramatic worsening of symptoms. LS is sometimes associated with continuous murmur. The presence of severe MS and ASD of relatively small size are necessary to produce the continuous murmur.[5]

Role of echocardiography in evaluation of LS – Transthoracic echocardiography establishes the diagnosis of LS. It is helpful in identifying the type and size of ASD and degree of MS. ASD in LS should have a diameter of more than 1.5 cms.[2] Mitral valve area is best calculated by planimetry, PISA and continuity equation in these cases. Pressure half time is unreliable as it gives false low value due to the simultaneous flow across ASD. ASD is also best assessed by subcostal window and thus avoids echo dropouts in apical 4-chamber view.

The characteristic Doppler flow pattern across the atrial septum shows continuous or late systolic and holodiastolic left to right flow produced by the high gradient in the left atrium caused by MS.[2] Echocardiography can also be helpful in differentiating the classical form from iatrogenic LS. In iatrogenic LS, the ASD diameter ranges from 0.5-1.0 cm. Other differentiating features are small left atrium, mitral valve calcification and small increase in mitral valve area. In contrast to classic LS where ASD is large and non-restrictive, the acquired form has restrictive ASD. The accurate assessment of planimetric mitral valve area and subvalvular apparatus should be done in these patients before treatment.[6]

Treatment of LS – These cases are better managed by early diagnosis and surgical treatment and are associated with good outcome. However the presentation amongst our rural population is usually late. The prognosis tends to worsen with the onset of pulmonary hypertension and heart failure.[3] The size of ASD is also crucial before the therapeutic intervention. An ASD of more than 38mm are usually ineligible for percutaneous therapy but rather open heart surgery.[7] Many corrective surgery options are available now. Percutaneous transcatheter therapy has become the most widely accepted therapy using balloon mitral valvuloplasty for MS (the Inoue balloon being most widely used) and the Amplatzer atrial septal occluder for closure of an ASD. Percutaneous correction is preferred to surgical correction in view of decreased morbidity and faster recovery. In advanced cases, the mortality is increased due to heart failure, cardiac arrhythmia (most commonly atrial fibrillation), thromboembolic cerebrovascular disease. The classical LS can be corrected satisfactorily whereas the acquired LS usually need early surgical intervention as they are more prone to deteriorate with the development of severe pulmonary hypertension and right heart failure.[8]

CONCLUSIONS

LS are a rare clinical entity. In patients with MS who lack typical clinical findings, the possibility of LS should be kept under consideration. Echocardiography is a helpful tool in diagnosing this condition. Early diagnosis helps a patient by undergoing only percutaneous correction of the condition as prognosis become quite worse in the advanced cases.

Few published case reports show these patients to survive long without any symptoms as reported by Kulkarni SS. An 81 year old woman became symptomatic at 75 years of age. The female with LS can also well survive multiple pregnancies, as reported by Perloff, who was a 74 years old lady with 11 pregnancies.[9]

Also in the rural areas in our country, patients are deprived of proper diagnosis of their illness due to unavailability of advanced investigations within their reach. So with the opening of new advanced centers/hospitals in the rural areas would definitely help majority of such patients. Our case also got benefitted in the similar way and thus it became possible to plan a definite management for her. Along with it the burning issue of relatively higher cost of the surgical procedures also makes these patients deprived of the interventions even though a proper diagnosis is made. Hence a need to draft a definite policy is certainly required worldwide.

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