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Lutembacher's syndrome in Rhuematic Heart Disease (Mitral stenosis and regurgitation) presenting with Eisenmenger syndrome - A rare entity.

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Abstract

Lutembacher's syndrome is very rare. In a study published in American Heart Journal in 1997, it was found that the incidence is- 0.001/10,00000.[1] It is a combination of mitral stenosis(MS) and atrial septal defect(ASD), which can be either congenital or acquired. Mitral stenosis is generally acquired in this syndrome as ramification of rheumatic heart disease. Atrial septal defect can be congenital and can be iatrogenic, secondary to cardiac interventional procedures like mitral valvuloplasty.

Keywords: Lutembacher's syndrome, Atrial septal defect, Mitral stenosis, Eisenmenger complex,Transthoracic echocardiography.

Introduction

Atrial septal defect (ASD) embraces one of the commonest congenital cardiac anomalies. Among them, the ostium primum defects are unfailingly associated with congenital mitral valvular (MV) malformation, However, MV disease that involves non-primum atrial septal defects, especially the secundum type is rare.[2] The incidence of mitral stenosis(MS) in patIents with ASD is 4 to 6%, where as the incidence of ASD in patients with MS is 0.6 to 0.7%.[3] Development of Eisenmenger syndrome is very uncommon in the presence of large ASD and high left atrial pressure because of mitral stenosis.[4] We are reporting a case of this rare syndrome with congenital atrial septal defect and acquired rheumatic heart disease with Eisenmenger syndrome in a patient of rheumatic heart disease.

Case Report

A 25 year male presented with a history of difficulty in breathing, for last 3 months, which was experienced more during his routine work (NYHA GIII) and gradually progressed to grade IV in 3 months. Dyspnoea was not associated with chest pain, cough and upper respiratory tract infection but was associated with palpitations on rest. On examination, patient was conscious thin built without any chest deformity. His blood pressure was 90/60mmHg and pulse 96bpm, respiratory rate 20 cycles per minute. No pallor, icterus, oedema, splinter haemorrhages, subcutaneous nodules or rashes. Clubbing and cyanosis were present.

JVP was elevated at 12 cm of water. Oxygen saturation was 88% on room air. On precordial examination the apex impulse was in left 5th intercostal space in anterior axillary line, with hyperdynamic character. P2 was palpable with thrill present in left 2nd intercostal space. Parasternal heave and epigastric pulsations were present. In mitral area loud S1 with grade II/IV mid-diastolic rumbling murmur was present along with systolic murmur radiating to axilla and inferior border of scapula and pansystolic murmur of grade IV/IV along the left lower parasternal border. Also loud P2 with fixed split was heard in pulmonary area. Respiratory system examination revealed bilateral basal crepitations.

Abdomen was soft, nontender, liver was palpable 6cm below the right costal margin. A provisional diagnosis of rheumatic heart disease with active carditis, congestive cardiac failure with mitral stenosis and regurgitation, tricuspid regurgitation with pulmonary arterial hypertension without infective endocarditis was made. Atrial septal defect with mitral stenosis with Eisenmenger complex was kept as remote possibility due to cyanosis and clubbing.

Labortary investigations revealed a haemoglobin 13.2 gm%, total leucocyte count 6,500/cumm, neutrophils 56%, lymphocyte 36%, eosinophil 1%, erythrocyte sedimentaon rate of 58 mm at the end of first hour, random blood sugar was 107mg/dl, serum sodium 138 meq/L and serum potassium 3.5 meq/L. LFT and RFT were normal. X-ray chest revealed plethoric lung fields and cardiomegaly with a C/T ratio 0.80 and right ventricular hypertrophy with prominent pulmonary vein and right atrial enlargement. Electrocardiogram (Figure 1) showed rsR complex in II, III, and AVF leads suggestive of right axis deviation and right bundle branch block.

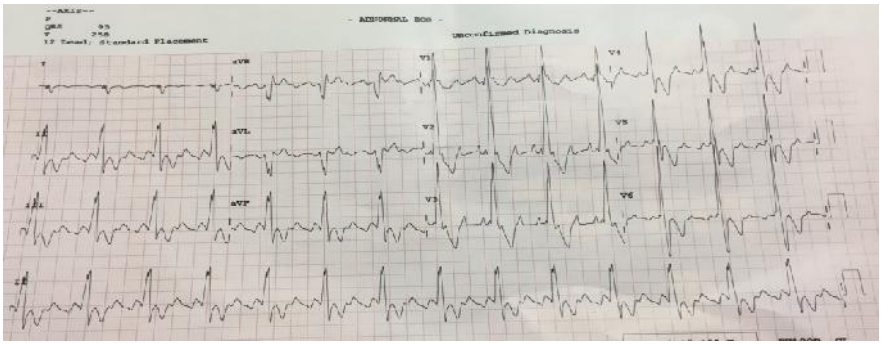

Figure 1 ECG showing right axis deviation with right bundle branch block

Transthorasic echocardiography (Figure 2,3,4,5) revealed a large ostium secundum atrial septal defect (3.9cm) with left to right shunt, mild mitral stenosis with mitral valve orifices 1.6-1.7cm with anterior mitral leaflet(AML) and posterior mitral leaflet(PML) thickened, AML doming, PML fixed, right atrium and right ventricle grossly dilated, grade 3 tricuspid regurgitation with gradient of 44 mmHg, moderate pulmonary hypertension with normal left ventricular systolic function with LVEF of 53%. The patient was treated with diuretics, antibiotic coverage, aspirin and digoxin. Due to poor socioeconomic status patient was not willing for surgical intervention and is under medical therapy for follow-up.

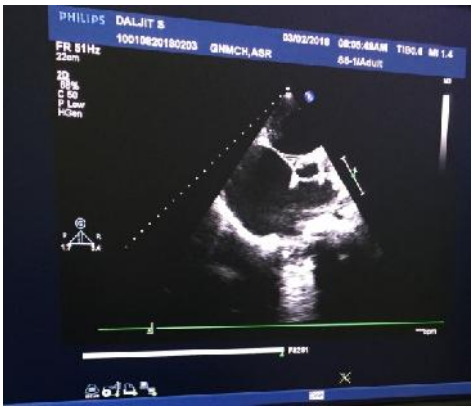

Figure 2 Transthoracic echocardiogram of patient of Lutembacher's syndrome showing large ostium scundum type defect with mitral stenosis.

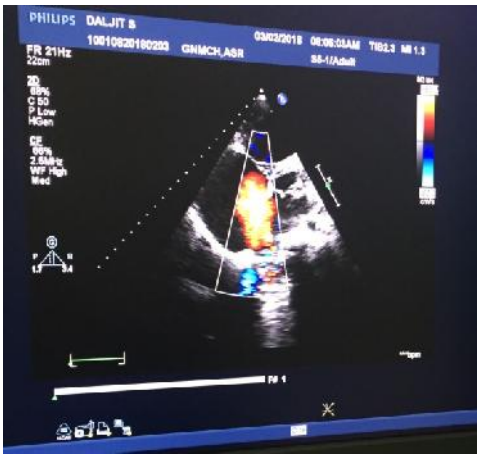

Figure 3 Transthoracic echocardiogram colour doppler of patient of Lutembacher's syndrome showing flow from left atrium to right atrium through atrial septal defect.

Figure 4 Transthoracic echocardiogram of patient of Lutembacher's syndrome showing diastolic dooming of anterior mitral leaflet.

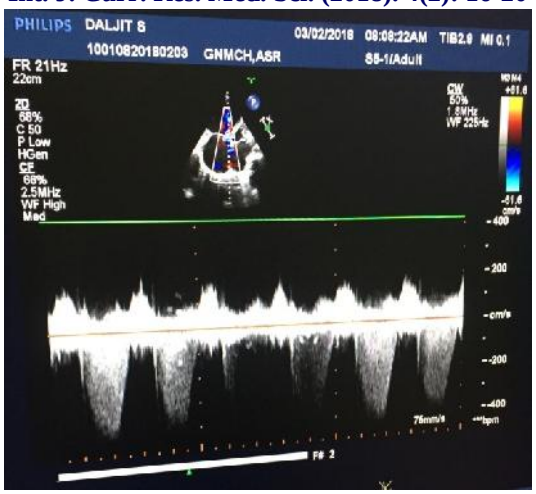

Figure 5 Transthoracic echocardiogram of patient of Lutembacher's syndrome showing tricuspid regurgitation.

Discussion

Inter-atrial septum develops from two sources-septum primum and septum secundum. If defect is in the formation of septum primum, it leads to the formation of ASD (Primum) and if defect is in the formation of septum secundum, it forms ASD (secundum). The role of the ASD in MS was evident in Lutembacher's original report of 1916 when the patient was a 61-year-old woman who had been pregnant seven times.[5] Survival to advanced age has also been reported;[6.] In one instance an 81-year-old woman experienced no symptoms related to her heart disease until she reached 75 years of age.[6] Lutembacher's syndrome has high female preponderance, which can be explained by increase prevalence of ostium scundum atrial septal defects and rheumatic heart disease in females.[7] But our patient was a male patient which is unusual.

Lutembacher believed that the MS was congenital, but current consensus supports the opposite view, that is, the stenosis is acquired and congenital mitral stenosis is a rare phenomenon. It is almost always labelled and presumed to be rheumatic, especially in developing countries like India with a high rheumatic disease burden.[8]

In Lutembachers syndrome, atrial septal defect and mitral stenosis modifies the hemodynamics and clinical expressions of each other, and these modifications depend upon the severity of mitral stenosis rather than on the size of atrial septal defect because ostium secundum defects are typically non restrictive. In uncomplicated atrial septal defect, left to right shunt determined principally by the relative resistances to flow from the left atrium into the left ventricle or from the left atrium through the defect into the right ventricle. Increased right ventricular distensibility favours left to right shunting. Mitral stenosis increases the resistance to blood flow from left atrium into left ventricle and, in so doing, augments the left to right shunt. In a non-restrictive atrial septal defect, the magnitude of augmentation varies directly with the severity of mitral obstruction.[9] Patients may be asymptomatic for decades or they can suffer from severe hemodynamic instability and heart failure early on.[10]

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.[4] As in our patient, Eisenmenger component was present since our patient had developed cyanosis and clubbing which is usually absent in isolate Lutembacher's syndrome. The presence of MS, when accompanied by mitral regurgitation, increases susceptibility to infective endocarditis, as compared to the low incidence of infective endocarditis in uncomplicated ASD.[9]

Definitive percutaneous treatment of a patient with Lutembacher's syndrome was successfully accomplished using the Amplatzer septal occluder to close a secundum atrial septal defect and the Joseph mitral balloon catheter to dilate rheumatic mitral valve stenosis. Transcatheter therapy is an effective alternative to surgery in selected patients with Lutembacher's syndrome.[11]

Early diagnosis and surgical treatment is having a good prognostic value. If patient is diagnosed at late stage, pulmonary hypertension and heart failure develops and the prognosis is bad. If the patient is diagnosed earlier before the development of pulmonary hypertension and heart failure, - ASD closure with mitral valve replacement bears a good prognosis and prolongs survival.[12]

Conclusion

Although Lutembacher syndrome is rare, but it must be diagnosed correctly at appropriate time via echocardiography to provide adequate medical and surgical therapies. Early diagnosis can prevent the onset of pulmonary hypertension and heart failure, thus improves the survival rates.

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