



A RARE CASE OF COMBINATION OF CONGENITAL AND ACQUIRED HEART DISEASE : LUTEMBACHER'S SYNDROME

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ABSTRACT Any combination of ASD (congenital or iatrogenic) and MS (congenital or acquired) is referred to as Lutembacher's syndrome (LS)¹. Incidence of LS is 0.001 per million populations, while the proportion of iatrogenic LS stands at 11-12%². A 26-year multiparous woman presented with complaints of progressive pedal edema and dyspnea on exertion. Diagnosed as chronic rheumatic heart disease during first pregnancy. On examination, she had facial puffiness, bilateral pedal edema, raised JVP, a displaced apex beat in the 6th intercostal space, palpable first heart sound, palpable pulmonary component of second heart sound and parasternal heave. On auscultation, she had a loud first heart sound and a mid-diastolic murmur in the mitral area. We suspected the patient to have secondary pulmonary arterial hypertension due to mitral stenosis and right-sided heart failure along with acyanotic congenital heart disease like ASD and confirmed by 2d ECHO.

KEYWORDS : Atrial Septal Defect, Mitral Stenosis, Lutembacher Syndrome.

INTRODUCTION:

Lutembacher syndrome (LS) was first described in a letter by anatomist Johann Friedrich Meckel in 1750^{1,3}. The definition of lutembachers's syndrome(LS) has evolved over time since it was first discovered. Currently, any combination of atrial septal defect (ASD) (congenital or iatrogenic) and mitral stenosis MS (congenital or acquired) is referred to as LS¹. In the current era of percutaneous balloon mitral valvuloplasty (BMV) for acquired MS, residual iatrogenic ASD secondary to trans-septal puncture is more common than the congenital ASD. Physicians refer to this as iatrogenic LS⁴. Clinical features and hemodynamic effects of LS depend on the balance of effects of the MS and the ASD.

CASE REPORT :

A case of 26-year-old multiparous woman presented to the medicine OPD with complaints of progressive pedal edema and dyspnea on exertion. She was diagnosed with chronic rheumatic heart disease on routine evaluation during her first pregnancy. All her pregnancies were uneventful.

On general examination, she had facial puffiness, B/L pedal edema, raised JVP.

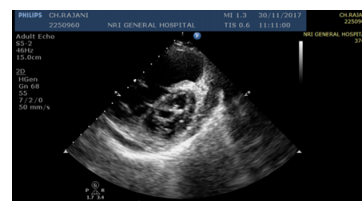
On cardiovascular examination, she had a displaced apex beat (lateral) in the 5th intercostal space, precordial pulsations, epigastric pulsations, palpable first heart sound(S1), palpable pulmonary component of second heart sound(P2), Lt parasternal heave. On auscultation, she had a loud first heart sound(S1) and a mid-diastolic murmur in the mitral area. She had a loud P2 and fixed split of second heart sound in the pulmonary area.



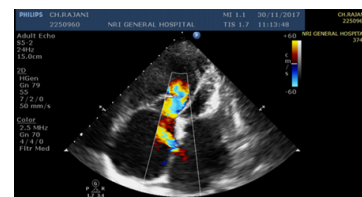
Ecg showed heart rate of 98/min, normal sinus rhythm, right axis deviation.

CHEST X-RAY (POSTERIOR-ANTERIOR VIEW):

Cephalization of vessels, Prominent mid and lower zone vascular markings, prominent pulmonary trunk, moderate cardiomegaly, straightening of left heart border.



SHORT AXIS VIEW : FISH MOUTH APPEARANCE OF MITRAL STENOSIS



FOUR CHAMBER VIEW : LEFT TO RIGHT SHUNT OF ASD

DISCUSSION:

The exact prevalence of LS is unknown. It is likely to be more in developing countries of Southeast Asia, where rheumatic heart disease is more common. The incidence rate of congenital ASD in patients with MS is 0.6% to 0.7%. Incidence of LS is 0.001 per million populations. Prognosis is influenced by several factors - pulmonary vascular resistance, right ventricle (RV) compliance, size of ASD and MS severity. Echocardiography remains the gold standard for diagnosis and evaluation of LS. Timely diagnosis is critical for modifying the natural course, by allowing patients to benefit from currently available percutaneous trans-catheter therapies with favo-

rable effects on the outcome . the occurrence of secondary pulmonary hypertension and congestive heart failure is commonly associated with poor outcome . Reverse LS has also been described as a predominant pulmonary-to-systemic or right- to left shunting of blood in the context of an ASD and severe tricuspid stenosis.

CONCLUSION:

This is a case of LUTEMBACHER SYNDROME (ASD + MS) . Echocardiography remains the gold standard for diagnosis and evaluation of LS. with 3D echo and TEE further helpful in excluding co-existent cardiac pathologies. Planimetry by Doppler echo remains the best method for assessing MVA. Whilst open heart surgery is frequently the treatment modality of choice in case of concurrent cardiac malformations, LS is currently an example of an exception, with new percutaneous trans-catheter therapies, combining PBMV with Inoue balloon technique for MS and the Amplatz Septal occluder for ASD closure.

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