Volume : 3 | Issue : 6 | June 2014 • ISSN No 2277 - 8179

Research Paper

Medical Science

KEYWORDS :

Case Report On Lutembacher Syndrome

Dr Surendra Verma	Associate Professor, Department of Medicine, SPMC Medical College Bikaner, House no H/2 PBM Hospital campus , Bikaner Rajasthan
Dr Rahul Singla	Resident 1st year, Deparmtmeent of medicine, Room no 66/B PBM Hospital campus, Bikaner Rajasthan
Dr Aniket Mule	Resident 1st year, Deparmtmeent of medicine , Room no 66/B PBM Hospital campus, Bikaner Rajasthan
Dr Nimbaram	Resident 1st year, Deparmtmeent of medicine , Room no 6/B PBM Hospital campus, Bikaner Rajasthan
Dr Rajendra Tanwar	Associate professor, Department of Medicine , SPMC Bikaner
Dr B.K.Gupta	Senior Professor, Department of Medicine , SPMC Bikaner
Dr.V.B.Singh	Senior Professor, Department of Medicine, SPMC Bikaner

ABSTRACT

Lutembacher's syndrome refers to a congenital atrial septal defect (ASD) complicated by acquired mitral stenosis (MS).[1]. Inter-atrial septum develops from two sources-septum primium and septum secundum. If defect is in the formation of septum primium-it leads to the formation of ASD (Primium) and if defect is in the formation of septum secundum it forms ASD (secundum). Here, we will discuss about a case who was diagnosed to have Lutembacher's syndrome.

CASE REPORT

A 20 year old female was referred to P.B.M. Hospital as a registered case from a MGM Government Hospital Hanumangarh. She had complaints of breathlessness on exertion and on lying down since 6 days prior to admission to our hospital. She also complained of exertional dyspnoea (New York Heart Association - NYHA Class II) which was gradually progressive, and orthopnoea. Patient also complained of pain in right hypochondrium with distension of abdomen, swelling of feet and palpitations. There was no history of accompanying chest pain, cough, fever, or symptoms of upper respiratory infections. There was no preceding or accompanying vaginal bleeding. There was no past history of joint pains with throat infection, or any long term medications for any premorbid condition. She was married since 8 months, without consanguinity and nulliparous. (Obstetric score $G_0P_0L_0A_0$) with irregular menstrual cycles.

On examination in our hospital, she was afebrile with low volume irregularly irregular pulse of 110 beats/min and blood pressure 98/60 mm of Hg, with respiratory rate of 22 breaths/min. She was pale and showed bipedal oedema. In cardiac examination, there was loud mitral S_1, and wide fixed split pulmonary S_2 with opening snap in mitral area. There was IV/IV mid diastolic murmur in mitral area and early systolic murmur, and non-radiating short murmur in pulmonary area. There was parasternal heave and thrill. Respiratory system examination revealed bilaterally decreased air entry in basilar region with bilateral lower zones end-inspiratory fine crackles. Abdominal examination revealed massive abdominal distension with massive hepatomegaly.

Her investigation reports were as follows:

- Haemoglobin (Hb)-9.8 gm%;
- White blood cells (WBC) - 10,400/cu mm of blood;
- Polymorphs - 68%;
- Lymphocytes - 25%;
- Eosinophils - 02%;
- Platelet count - 1.91 lakh;
- Random blood sugar - 85 mg/dl;
- Blood urea – 35.99 mg/dl; [normal value→20 – 40 mg/dl]
- Serum creatinine - 0.84 mg/dl; [normal value→0.8 -1.6 mg/dl]
- Serum sodium - 138 mEq/L; [normal value→135 – 145 mEq/L]
- Serum potassium - 3.5 mEq/L. [normal value→3.5 – 5.5 mEq/L]

Urine Routine and Microscopic exam was Normal.

Electrocardiogram [Figure 1]→ Irregularly irregular ventricular rhythm; Right Axis Deviation; ST depression with T-wave inversion in Inferior leads..

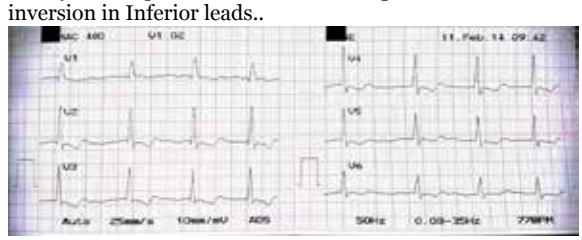


Figure 1

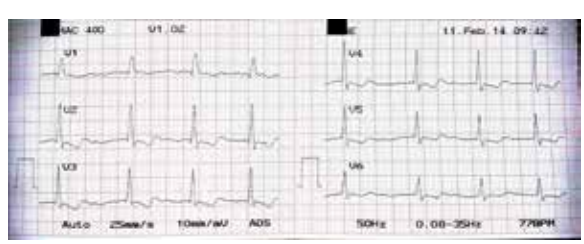
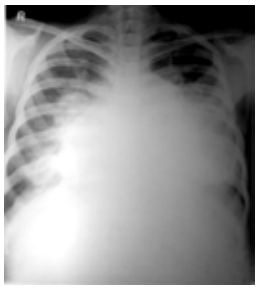


Figure 2

Electrocardiogram[Figure 2] of patient of Lutembacher's syndrome suggestive of Right Ventricular Hypertrophy

Chest X-ray PA view [Figure 3] showed markedly increased CT ratio with area of heaziness along the right heart border s/o Cardiomegaly with pericardial effusion and fluffy ill defined opacities in B/L mid-lower lung fields s/o Pulmonary edema

Volume : 3 | Issue : 6 | June 2014 • ISSN No 2277 - 8179



Electrocardiogram of patient of Lutembacher's syndrome suggestive of Right Ventricular Hypertrophy

2D-Echocardiography [Figures [Figures 3 and and 4] → Severe Mitral stenosis; Moderate Mitral regurgitation; mitral valve area 0.8 cm^2. Large Ostium Secondum atrial septal defect With Left To Right Shunt; Severe pulmonary arterial hypertension.

Figure 3

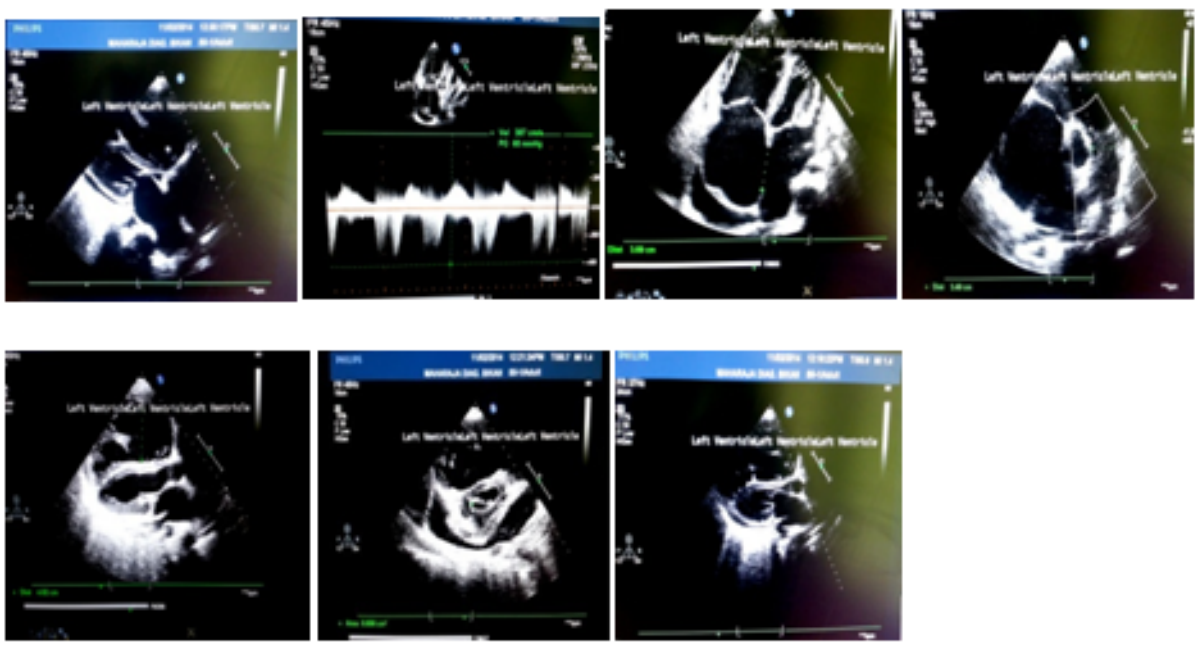


Figure 4

Transthoracic echocardiogram of patient of Lutembacher's syndrome showing thickened mitral leaflets with doming of anterior mitral leaflet with mitral stenosis and atrial septal defect

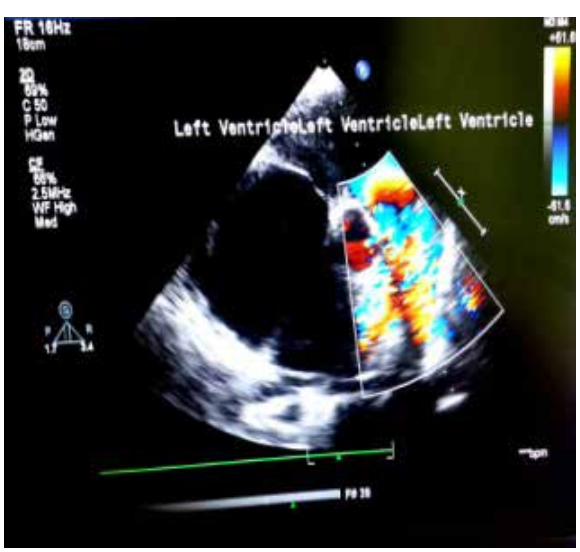


Figure 4

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

Other Valves- Normal. No Clots/No Effusion/No Vegetations.

With the above history and findings, we came to a final diagnosis of Lutembacher syndrome and patient was started with the diuretics and the standard prophylaxis for Rheumatic Fever with Benzathine Penicillin 12 lakh units deep intramuscular (After Skin Test Dose) once every 3 weeks. She was advised surgical intervention at a later date, after Trans-Oesophageal Echocardiography. She was discharged in a haemodynamically stable condition.

DISCUSSION

In Lutembacher's syndrome, initially, high left atrial pressure due to mitral stenosis was thought to stretch open the patent foramen ovale, causing left-to-right shunt and providing another outlet for the left atrium. Now ASD in this syndrome, like mitral stenosis, is recognized as being either congenital or acquired, as already described. The haemodynamic effects of this syndrome are a result of the interplay between the relative effects of ASD and mitral stenosis. In its initial description, the ASD was typically large in Lutembacher syndrome, thus providing another route for blood flow. The direction of blood flow is determined largely by the compliance of left and right ventricles. Normally, the right ventricle is more compliant than the left ventricle. As a result, in the presence of mitral stenosis, blood flows to the right atrium through the ASD instead of going backward into the pulmonary veins, thus avoiding pulmonary congestion. This happens at the cost of progressive dilatation and, ultimately, failure of the right ventricle and reduced blood flow to the left ventricle. 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.

The incidence of this condition is very rare. In one study published in American Heart Journal in 1997, it is found that the incidence of Lutembacher's syndrome is- 0.001/10,00000.[2] The ameliorating role of the ASD in MS was evident in Lutembacher's original report of 1916; the patient was a 61-year-old woman who had been pregnant seven times.[3] An earlier case report in the literature in 1880 (and referred to by Perloff)[1] was of a 74-year-old woman. Survival to advanced age has also been reported;[4] in one instance an 81-year-old woman experienced no symptoms related to her heart disease until she reached 75 years of age.[4] These favourable reports, however, should not obscure the fact that the long-term natural history of ASD is unfavourably influenced by MS, which augments the left-to-right shunt and predisposes to atrial fibrillation and right ventricular failure.[5] The presence of MS, especially when accompanied by mitral regurgitation, increases susceptibility to infective endocarditis, in contrast to the low incidence of infective endocarditis in uncomplicated ASD,[1] just like in our case. Early diagnosis and surgical treatment bears a good prognostic value. If patient is diagnosed at late stage, pulmonary hypertension and heart failure develops and the prognosis is bad.[6] If the patient is diagnozed earlier before the development of pulmonary hypertension and heart failure, - ASD closure with mitral valve replacement bears a good prognosis and prolongs survival.

REFERENCE

1. Perloff JK. The clinical recognition of congenital heart disease. 4th ed. Philadelphia: Saunders; 1994. pp. 323–8. | 2. Ali SY, Rahman M, Islam M, Barman RC, Ali MY, Islam MMSU, et al. Lutembacher's Syndrome- A Case Report. Faridpur Med Coll J. 2011;6:59–60. | 3. Lutembacher R. De la sténose mitrale avec communication interauriculaire. Arch Mal Coeur.1916;9:237–60. | 4. Rosenthal L. Atrial septal defect with mitral stenosis (Lutembacher's syndrome) in a woman of 81. Br Med J. 1956;2:1351. [PMC free article] [PubMed] | 5. Cheng TO. Coexistent atrial septal defect and mitral stenosis (Lutembacher syndrome): An ideal combination for percutaneous treatment. Cathet Cardiovasc Intervent. 1999;48:205–6. [PubMed] | 6. Levin AR, Spach MS, Boimeau JP, Canent RV, Jr, Capp MP, Jewett PH. Atrial Pressure flow dynamics and atrial septal defects (Secundum type) Circulation. 1968;37:476–88. [PubMed]