Journal of Diabetic Association Medical College 2017;1(1)37-39

Case Report

Lutembacher Syndrome

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Abstract

Lutembacher syndrome a rare complex heart disease comprises ASD (secundum) with mitral stenosis. ASD (Secundum) usually congenital but may be iatrogenic during IAS puncture during PTMC procedure. MS usually rheumatic origin. Our Patient Mrs. Fulmala, 60 yrs. old, housewife, non hypertensive, non diabetic admitted in DAMCH cardiac unit on 10.11.12 with the complaints of progressive respiratory distress for ten years, cough for 5 yrs, chest pain for 2 yrs. Initially shortness of breath was in NYHA class II & at present it in NYHA class-III. She had a previous history of rheumatic fever in early childhood. There was history of recurrent attack of rheumatic fever. Subsequently she developed MS from rheumatic carditis. ASD was congenital in origin. If diagnosis could be done earlier, surgical closure of ASD with replacement of mitral valve bears a good prognostic value. Our patient is in elderly age & already developed pulmonary hypertension, so operative procedure is not suitable. So, the patient should be kept in conservative treatment.

Key words: Lutembacher syndrome, atrial septal defect, mitral stenosis, septum primum, septum secundum.

Introduction

Cross finger Lutembacher syndrome- is defined as a combination of mitral stenosis & left to right shunt at the atrial level. Typically the left to right shunt is an atrial septal defect (ASD) of the ostiumsecundum variety[1]. Both these defects ASD & MS, can be either congenital or acquired. In 1916, Lutembacher described his first case of this syndrome involving a 61 years old woman. In the current era of mitral valvuloplasty for acquired mitral stenosis, residual iatrogenic ASD secondary to transseptal puncture is more common than congenital ASD, as is the combination of ASD & mitral stenosis[2]. Incidence of ASD in-patient with MS is 0.6 - 0.7%[3]. Syndrome is more common in females than males. Lutembacher syndrome can present at any age. Cases have been diagnosed in the seventh decade of life. Heart disease comprises- congenital heart disease & acquired heart disease. There are many heart diseases which are very complex form that is combination of congenital & acquired in origin. Lutembacher syndrome is one of the complex heart diseases & its incidence is very rare[4].

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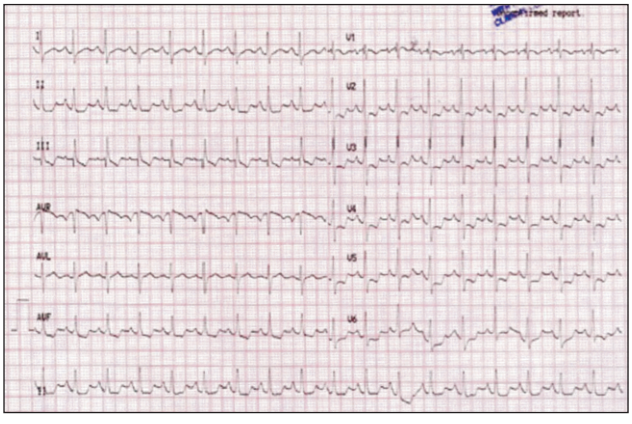
Case

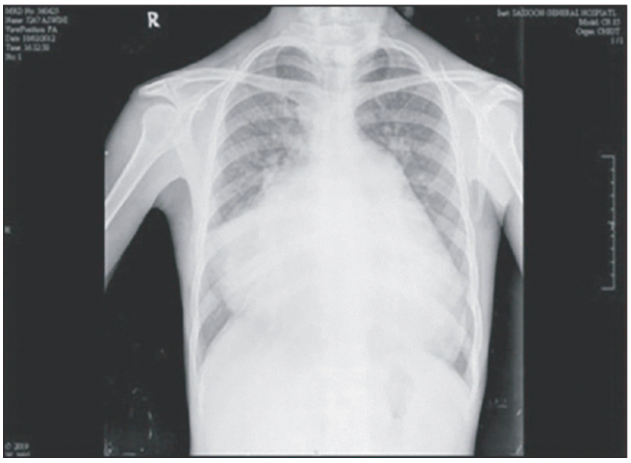
Mrs.Fulmala, 60 yrs. old, house wife, Hindu, non-hypertensive, non-diabetic hailing from Gholmazi, charmuguria, Madaripur, admitted in DAMCH- cardiac unit, Faridpur on 10.11.12 with the complaints of progressive respiratory distress for ten years, cough for 5 years, chest pain for two years. Initially shortness of breath developed after moderate exertion (NYHA- class II). But now it appears after minimal exertion (NYHA- class III). Patient also complaints recurrent attack of cough, frothy sputum, chest pain with radiation to left arm. She came from poor social-economic status. On examination she is ill looking, on propt up position, mild anemic, engorged pulsatile neck vein, palpable tender liver, leg odema, CVS examination reveals pulse-110/m, regular, BP- 125/80 mm. Hg, apical impulse in left 6th intercostal space, left parasternal heave present, P_2 palpable, S_1- louder, P_2- louder, mid diastolic murmur in mitral area, systolic murmur in upper leftpara sternal area, respiratory system examination reveals- crepitation in both lung bases. Other systemic examination reveals- no abnormalities.

Investigations findings

ECG findings-



- Sinustachycardia.
- Incomplete RBBB.
- T' wave invertion in L_{II}, L_{III}, AVF, V_1-V_4



X-ray chest PA view-

- Cardiomegaly.
- Pulmonary conus full.
- Features pulmonary congestion.

Laboratory investigation:

a) Blood sugar - FBS-5.5 mmol/l
b) Serum creatinine - 0.9 mg/dl
c) Lipid profile:
Cholesterol - 300 mg/dl
HDL - 45 mg/dl
LDL - 130 mg/dl
TG - 260 mg/dl

ECHO cardiogram (1-3)

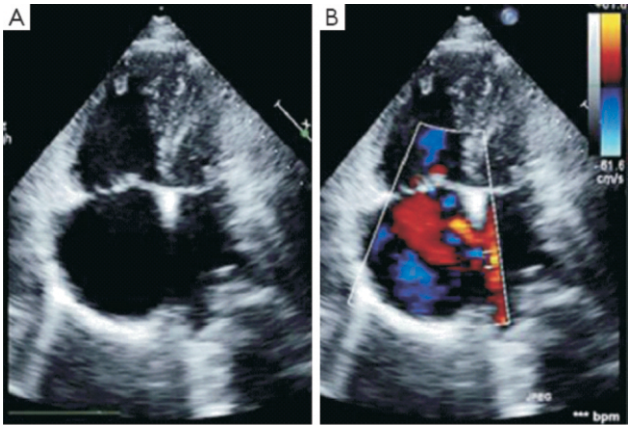


Fig. 1

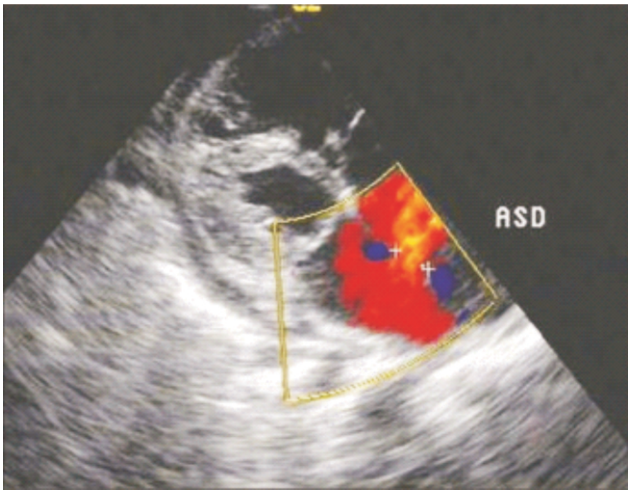


Fig. 2

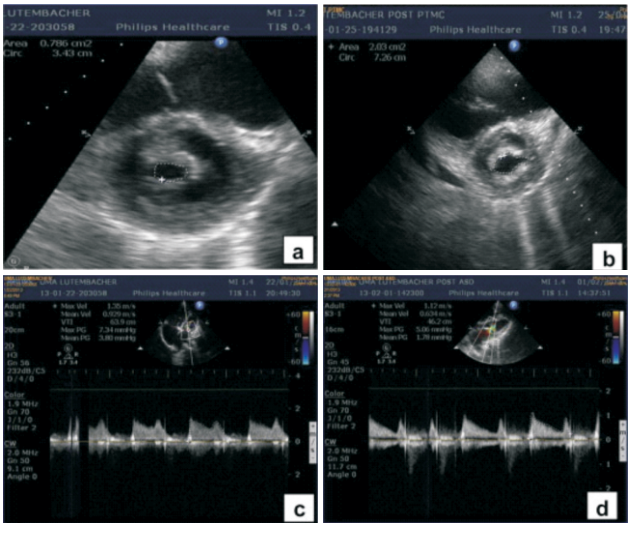


Fig. 3

MV- Thickened, calcifled, reduced orifice.
MVA- 0.8 cm^2.
IAS Deficit.

Color flow mapping
MV- Mosaic flow seen from LA to LV.
ASD - Seen. Mosaic flow seen from LA to RA, then to RV in a butterfly fashion.

Final impression- MS (Severe) with ASD (Secundum) with PH.

Diagnosis: Lutem Bacher Syndrome

Discussion

Incidence of ASD in-patient with MS is 0.6 0.7%. It is found more in female than males. Syndrome can present at any age. Cases have been diagnosed in the seventh decade of life.

Lutembacher's original case was a 61 years old woman who had been pregnant 7 times. Early diagnosis& surgical treatment bears a good prognostic value. If a patient is diagnosed at late stage, pulmonary hypertension & heart failure develops & the prognosis is bad[5]. If the patient is diagnosed earlier before the development of PH & heart failure, ASD closure with mitral valve replacement bears a good prognosis & prolongs survival. Our patient, Mrs. Fulmala is an elderly patient & already develops pulmonary hypertension, so operative treatment is not possible & the patient is kept under conservative treatment, optimizing medical therapy with adequate control of heart failure.

Conclusion

Lutembacher's syndrome is a rare, complex heart disease-constellation of both congenital & acquired condition of the heart. Early diagnosis & operative treatment has a good prognostic value but late diagnosis & development of heart failure bears bad prognosis. Most of the patient dies subsequently due to heart failure, cardiac arrhythmias & thrombo-embolic cerebrovascular disease[6]. Early diagnosis & management can reduce morbidity & mortality.

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