

Case Report

Lutembacher's Syndrome - A Case Report

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Abstract

Lutembacher's Syndrome is a rare heart disease first described by Lutembacher, is a combination of a congenital atrial septal defect (ASD) and acquired mitral stenosis (MS). Traditionally Lutembacher syndrome has been corrected by surgical treatment. If diagnosis could be done earlier, surgical closure of ASD with replacement of mitral valve bears a good prognostic value. Cases were reported where patients treated percutaneously with a combined Inoue balloon valvuloplasty and septal defect closure using the Amplatzer septal occlusion device. Our patient Mrs. Kohinoor Begum, 26 years old, non diabetic, non hypertensive, admitted in Khwaja Yunus Ali Medical College & Hospital (KYAMCH) on 03.04.2013 with the complaints of respiratory distress and chest pain for 3 years. There was history of recurrent attack of fever in childhood. Subsequently she developed MS from Rheumatic carditis. ASD was congenital in origin. After thorough perioperative evaluation pericardial patch closure of ASD with open mitral commissurotomy was done. Postoperative follow-up 6 months after operation showed no residual shunt with adequately functioning mitral valve.

Keywords: Lutembacher's syndrome, Atrial Septal Defect, Mitral stenosis.

Introduction

Lutembacher syndrome, first described by Lutembacher in 1916, is a combination of a congenital atrial septal defect (ASD) and acquired mitral stenosis (MS)¹. Mitral stenosis serves to augment the left-to-right atrial shunt through the ASD, but decompression of the obstructed left atrium by the ASD attenuates the symptoms of MS². The condition is usually treated surgically³, however, both abnormalities are amenable to percutaneous transcatheter therapy⁴⁻⁸. Inter-atrial septum develops from two sources-septum primum and septum secundum. If there is defect in the formation of septum primum, it forms ASD (Primum) and If the defect is in the formation of septum secundum it forms ASD (secundum). Mitral stenosis is an acquired heart disease develops due to recurrent attack of Rheumatic carditis. So, for the development of Lutembacher's syndrome defect in the formation of septum secundum and recurrent Rheumatic carditis are required.

It is very rare. It is found that the incidence of Lutembacher's syndrome is-0.001/10,00000. The incidence of MS in patients with ASD is 4% and conversely, the incidence of ASD in patients with MS is 0.6- 0.7% (10). It occurs predominantly in women. It usually presents in young adults but may present in elderly patients.

Case Report

Mrs. Kohinor Begum aged 26 years, non diabetic, normotensive, housewife, hailing from Enayetpur, Sirajgonj was admitted in Khwaja Yunus Ali Medical College & Hospital on 04.03.2013 with the complaints of respiratory distress on exertion and Palpitation for 3 years. Respiratory distress is progressive in nature and is more in lying position and also during exertion. Within last one year she developed several episodes of respiratory distress. She also complaints of easy fatigability.

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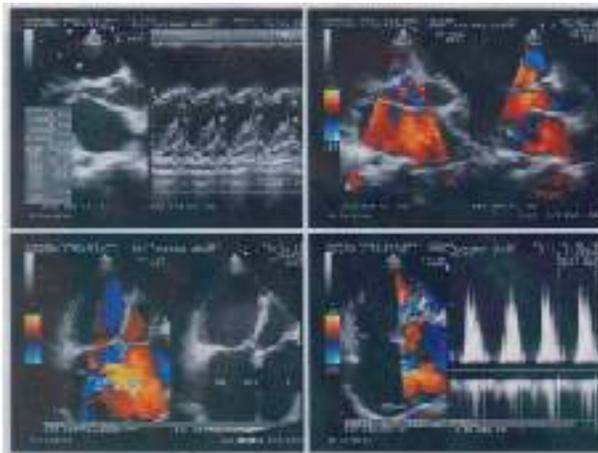
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She has history of repeated attack of fever in childhood. On examination in our hospital, pulse was 70 beats/min, irregularly irregular and of small volume. BP -100/60 mm of Hg, Jugular Venous Pressure (JVP) was raised, no ankle oedema. Precordial examination revealed accentuation of the first heart sound with an opening snap and a grade 3/6 mid-to-late diastolic rumbling murmur at the apex. Additionally, there were features suggestive of ASD, such as fixed splitting of the second heart sound with a grade 3/6 ejection systolic murmur at the pulmonary area. The exercise capacity was New York Heart Association (NYHA) class II. Respiratory system examination revealed bilaterally equal normal breath sounds with bilateral lower zones end-inspiratory fine crackles. Abdominal examination revealed no hepatomegaly. Other systemic examination reveals no abnormality.



Pre Operative echocardiogram

An electrocardiogram showed atrial fibrillation, incomplete right bundle branch block, and right atrial hypertrophy. A chest radiogram revealed cardiomegaly with increased pulmonary vascularity, a prominent main pulmonary artery. Transthoracic echocardiography showed a large-sized ASD secundum and moderately severe mitral stenosis. The mitral valve area was 1.3 cm^2 by planimetry and Doppler methods. Both AML & PML are minimally thickened, both commissures were partially fused. Mild to moderate calcification present in both leaflets. Moderate subvalvular changes were noted. The diameter of the ASD was estimated to be 25 mm. Color flow mapping revealed a left-to-right shunt across the ASD and trivial mitral regurgitation with moderate pulmonary hypertension (PASP-40 mmHg). Peak pressure gradient (PPG) across the mitral valve was 24.8

mmHg. Her all other investigation reports for preoperative assessment and anesthesia fitness were within normal limit.

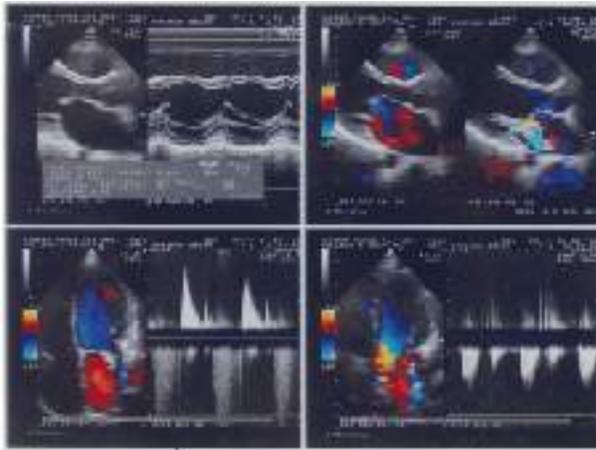
Surgical procedure

After obtaining informed written consent it was planned to do pericardial patch closure of ASD with open mitral commissurotomy as the valve morphology was suitable and patient is in child bearing age group. After establishment of all invasive and non-invasive monitoring lines patient was put under general anesthesia. Chest cavity was opened through Median sternotomy. Pericardiotomy done & pericardium was harvested. After full heparinization, Standard Cardiopulmonary bypass (CPB) was established with bicaval venous and aortic cannulation.

Aortic cross clamp was applied & Heart was arrested with antegrade warm blood root cardioplegia. Right atrium was opened & Secundum type of ASD was identified. Mitral valve was then assessed & found minimally thickened leaflets and commissures was partially fused. Posterior papillary muscle found to be shortened and splitting of the head of papillary muscle done. Open Mitral commissurotomy (OMC) was carried out and pericardial patch closer of ASD was done. Right Atrium wound was then closed. Deaeration procedures were carried out and cross clamp was released. Patient came off CPB smoothly. Heparin effect was reversed with Protamine infusion. Haemostasis was secured. Chest closure done after keeping two drain tubes (mediastinal) and single ventricular pacing wire in situ. Patient was then shifted to ICU in stable condition. Patient was extubated after 3 hours of ventilation and kept on inotropic support for 24 hours. Patient was shifted from ICU to cabin on 2nd postoperative day. She was discharged on 8th postoperative day with stable haemodynamic condition.

Follow-up

6 months after operation postoperative follow-up with Echocardiographic evaluation was carried out. Transthoracic echocardiography showed intact patch in situ with no intracardiac residual shunt. The mitral valve area was 2.3 cm^2 by planimetry and Doppler methods. There was trivial mitral regurgitation. Both AML & PML were minimally thickened, both commissures were partially fused. Mild calcification present in both leaflets. Mild subvalvular changes were noted.



Post operative echocardiogram at 6 month

Discussion

The original case describing Lutembacher syndrome involved a 61-year-old woman who had been pregnant 7 times¹. Female predominance has been noted in both ASD and MS, and thus Lutembacher syndrome has a predilection for females. The incidence of MS in patients with ASD is 4% and conversely, the incidence of ASD in patients with MS is 0.6-0.7%¹⁰. In Lutembacher syndrome, MS augments the left-to-right shunt through the ASD, while the non-restrictive ASD decompresses the left atrium, reducing the diastolic mitral pressure gradient¹¹. Planimetry and the Doppler continuity equation methods yield an accurate mitral valve area in the Lutembacher syndrome^{12,13}. The experience with transcatheter treatment of the Lutembacher syndrome is small^{2,3,9,14-17}.

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 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 times¹. An earlier case report in the literature in 1880 (and referred to by Perloff) was of a 74-year-old woman who had endured 11 pregnancies¹⁰. Survival to advanced age has also been reported¹². In one instance an 81-year-old woman experienced no symptoms related to her heart disease until she reached 75 years of age¹².

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 failure¹³. 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,¹⁰ just like in our case.

In Bangladesh, a case of Lutembacher syndrome involving inoperable elderly patient was reported from Faridpur medical college hospital¹⁸. 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 bad¹⁴. 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 of the patient.

Summary and Conclusion

Lutembacher's syndrome is a rare, complex, congenital heart disease. Early diagnosis and operative treatment has a good prognostic value but late diagnosis and development of heart failure bears bad prognosis. Most of the patients die subsequently due to heart failure, cardiac arrhythmias and thrombo-embolic cerebrovascular diseases. Early diagnosis and management can reduce morbidity and mortality.

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