Concomitant Tuberculous Meningitis and Lutembacher Syndrome with Multiple Atrial Septal Defects and Infective Endocarditis

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INTRODUCTION

Lutembacher syndrome is a rare cardiac condition comprising of congenital or acquired atrial septal defect (ASD) and mitral stenosis. Symptoms depend on the size of ASD, extent of mitral stenosis and degree of changes in the pulmonary circulation. Presentation can be due to cardiac failure, atrial arrhythmias, dyspnoea, exercise intolerance, paradoxical emboli or other disease related complications like pulmonary hypertension and infective endocarditis. Tuberculous meningitis is a chronic infection due to haematogenous dissemination of tubercle bacilli from lungs. It can lead to complications like cranial nerve palsies, hydrocephalus, cerebral oedema or focal neurological deficits presenting as stroke. The treatment should include anti-tuberculous therapy for one year and corticosteroids for initial 4 – 6 weeks depending on the symptoms of the patient. This report describes the concomitant occurrence of all these conditions at a time in a 45 years old lady.

CASE REPORT

The patient was a 45 years old female, presented to the Emergency with the complaint of low grade intermittent fever with evening rise for one month. The fever had become high grade and continuous a week before presentation. She also had generalized headache and diplopia for 2 days, followed by progressive loss of consciousness for one day. There was nothing significant on systemic enquiry and the only past history of note was that she had pulmonary tuberculosis 4 years ago for which she took anti-tuberculous therapy for only 3 months. Since then she was having dry cough, but she never expectorated any blood or sputum.

On examination, she was pale looking, thin and emaciated. Her pulse was 98 beats per minute and it was regular, blood pressure was 100/70 mmHg and temperature was 100°F. There was clubbing and central cyanosis. Her JVP was raised 5 cm above clavicle in propped-up position. Her Glasgow Coma Scale was 11/15. There was right sided hemiparesis with brisk reflexes and extensor plantar response in right lower limb. Signs of meningeal irritation including neck stiffness and Kernig’s sign were also present.

Examination of precordium revealed apex beat in left 5th intercostal space lateral to mid clavicular line and left parasternal heave. On auscultation, there was a loud first heart sound at apex of heart and a systolic murmur along left sternal border. In respiratory system, there was a patch of bronchial breathing with coarse crepitations and increased vocal resonance at apex of right lung. On auscultation of abdomen there was mild splenomegaly.

Her ESR was 55 mm after first hour, haemoglobin was 9.0 g/dl; TLC and platelet counts were normal. Her liver function tests, renal and coagulation profiles were normal. Chest X-ray showed cardiomegaly and fibrosis of right lung in apical region. There was sinus tachycardia and left axis deviation in ECG. Fundo-
scopy was done, which was normal, therefore, lumbar puncture was performed that showed normal opening pressure. Gross examination of CSF showed cob-web formation due to raised proteins. Biochemical and cytological examination of CSF showed high protein content, low glucose and TLC of 200 cells per cubic millimeter with predominance of lymphocytes. CSF culture did not show growth of any microorganism initially but culture for *Mycobacterium tuberculosis* later came positive. Her CT brain (plain and contrast) showed meningeal enhancement and communicating hydrocephalus (Figure 1). Since she had past history of incomplete treatment for pulmonary tuberculosis, she was diagnosed as a case of tuberculous meningitis (TBM) as suggested by raised proteins, low glucose and predominant lymphocytes on CSF analysis and positive CSF culture for *Mycobacterium tuberculosis*. Her blood cultures came out to be positive for *Streptococcus viridans*. So she was put on injection vancomycin, anti-tuberculous therapy, injectable dexamethasone and mannitol infusion. Within next 2 days her sensorium began to improve and she was sent for echocardiography which showed two atrial septal defects, the primum was about 11.9 mm and other in the mid septum was 11.0 mm with bidirectional flow (Figure 2). Her mitral valve was stenosed with valve area of 1.5 cm² (calculated by pressure half time) and there was loss of E and A waves on M-mode (Figure 3). There was no evidence of vegetation on valves or along defects. She had severe tricuspid regurgitation with pressure gradient of 90 mmHg across the tricuspid valve. She had severe pulmonary hypertension and her estimated pulmonary artery pressure was 105 mmHg.

She was labelled as a case of concomitant TBM and Lutembacher syndrome with multiple ASDs complicated by infective endocarditis keeping in view the physical, laboratory and radiological findings. She remained hospitalized for 3 weeks and during this period her clinical condition remarkably improved. Her temperature settled and she began to walk with support. Since she had severe pulmonary hypertension, surgical correction of ASD was deferred.

**DISCUSSION**

Lutembacher syndrome is an uncommon cardiac syndrome and this case was particularly unusual as she had two ASDs along with moderate mitral stenosis and pulmonary hypertension but she neither complained of any symptom suggestive of cardiac involvement in the past nor her presenting complaints favoured any cardiac problem. Her symptoms were exclusively pertaining to involvement of nervous system and diagnosis of tuberculous meningitis was not only supported by one month history of low grade fever and clinical signs but her CSF picture and brain imaging as well. The reasons for hemiparesis in this case are brain infarctions as a result of vasculitis involving vessels of brain or vasospasm of cerebral vessels leading to impaired blood flow resulting in stroke.

When mitral stenosis and ASD both co-exist, one lesion affects the dynamics of other and clinical signs depend on the dominant lesion causing major changes in cardiac chambers and pulmonary vasculature. In normal people, left side of heart is stronger than the right. In presence of an ASD, high pressures on left side cause shunting of blood from left to right. As a result, pressure on right side of heart increases causing increased pressure in pulmonary artery and eventually pulmonary hypertension. If uncorrected, it can cause damage of small pulmonary arteries and they get narrower and thicker making gaseous exchange difficult. Therefore, many areas of lung remain less oxygenated and anaerobic conditions prevail. This can flourish growth of those bacteria requiring absence of oxygen for their growth. But contrary to this fact is the growth of *Mycobacterium tuberculosis*, which is aerobic bacteria and cannot multiply in anaerobic conditions. However, after exposure to tubercle bacilli either the infection is concealed as a dormant focus or active pulmonary tuberculosis develops. If not treated in time it can disseminate to other organs, as this patient developed tuberculous meningitis as a result of non-compliance for treatment of pulmonary TB in the past. Patients of Lutembacher syndrome are usually at risk of developing infective endocarditis (unlike those with ASD alone) so...
prophylaxis of infective endocarditis is mandatory in such cases. 7 Although many cardiologists recommend transcatheter treatment of Lutembacher syndrome, 8,9 comprising of balloon mitral valvuloplasty and percutaneous closure of ASD as a bridge to surgical treatment, but outcomes are variable. However, early diagnosis and treatment to avoid complications like pulmonary hypertension is the key to best management. 10

REFERENCES