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Case Report

Acute Rheumatic Fever Complicating Tetralogy of Fallot

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This is a unique case of a patient aged 35 years developing rheumatic carditis secondary to acute rheumatic fever. The patient developed acute mitral regurgitation yet tolerated it relatively well because of coexistent Tetralogy of Fallot. The hemodynamics in this patient was significantly altered by the coexistence of these two conditions. This is the first case of its kind when acute rheumatic fever has been documented in a patient of Tetralogy of Fallot.

1. Introduction

The prevalence of rheumatic heart disease (RHD) in India reportedly ranges from 0.5 to 0.67 per 1000 in recent times [1]. RHD in patients with congenital heart defects is known to occur [2, 3]. Higher incidence of RHD has been reported in patients with congenital heart disease as compared to the general population [2, 4]. Tetralogy of Fallot (TOF) is the commonest cyanotic congenital heart disease encountered in adults [5]. The aortic and pulmonary valves in a patient of TOF have been reported to be predisposed to infective endocarditis [6]. There have been reports of mitral stenosis due to RHD coexisting with TOF [7, 8]. We present a unique case of acute rheumatic fever with carditis complicating TOF.

2. Case Presentation

A thirty-year-old female was referred for evaluation of recently worsened breathlessness. She had a history of mild breathlessness on exertion since last ten years. Earlier on, she used to get breathless on strenuous activity. Up to the past week, she had been unable to perform activities of daily routine. There was no history of fever, joint pain, any involuntary movements, or any other complaint. General physical examination

revealed small-built individual with mild anemia and fluctuation and softening of nail bed corresponding to grade I clubbing. The pulse was regular with a rate of 100 beats per minute.

The blood pressure was 100/60 mmHg. The apex beat was palpable in the sixth intercostal space, lateral to the midclavicular line. A soft first heart sound and a single loud second heart sound were heard. A grade III ejection systolic murmur was heard at the base, and a grade III pansystolic regurgitant murmur was heard at the apex, which radiated to the axilla. The lungs were clear. There was no hepatic or splenic enlargement.

On room air, the patient maintained oxygen saturation of 85%. Chest X-ray revealed enlarged heart, pulmonary oligemia, and no evidence of pulmonary venous hypertension. The coeur en sabot typical of TOF was not present. Electrocardiogram revealed sinus rhythm, increased PR interval [0.24 seconds], low voltage, and QRS axis of 90 degrees.

An echocardiogram revealed a large malaligned ventricular septal defect (VSD) with 50% aortic override (see Figure 1). There was a net right to left shunt. There was moderate grade, predominantly pulmonary valvular stenosis with peak gradient across the pulmonary valve being 55 mmHg (see Figure 2). There was no poststenotic dilatation of pulmonary trunk. The left and right pulmonary arteries were confluent. There was moderate pericardial effusion without any evidence of tamponade. The mitral valve was thickened. There was severe mitral regurgitation with a posteriorly directed jet (see Figure 3). Mitral valve

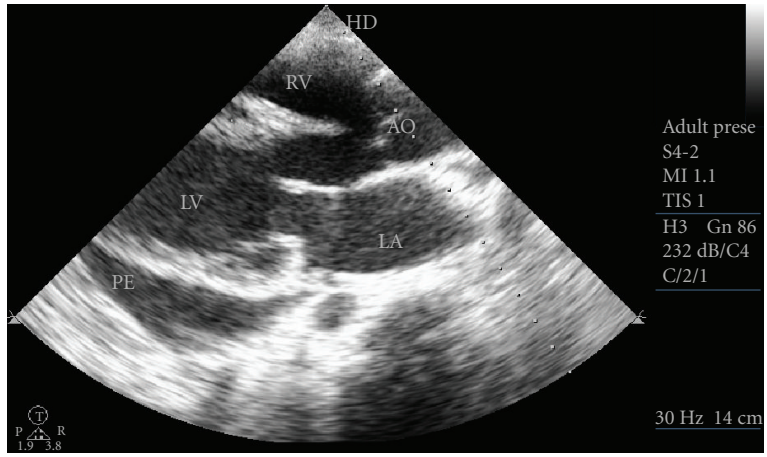

Figure 1: Parasternal long axis view depicting nonrestrictive, subaortic VSD, thick mitral valve leaflets and pericardial effusion.

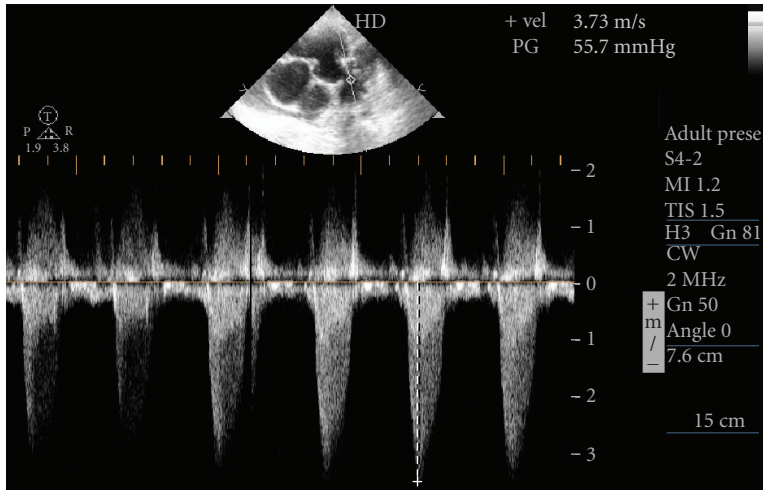

Figure 2: Parasternal short axis view depicting moderate grade pulmonary stenosis with gradient across the valve as 55 mmHg.

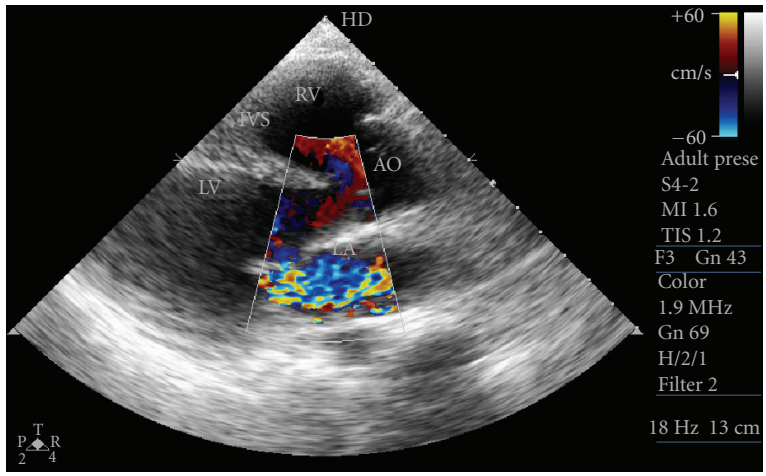

Figure 3: Parasternal long axis view depicting nonrestrictive VSD with moderate to severe mitral regurgitation.

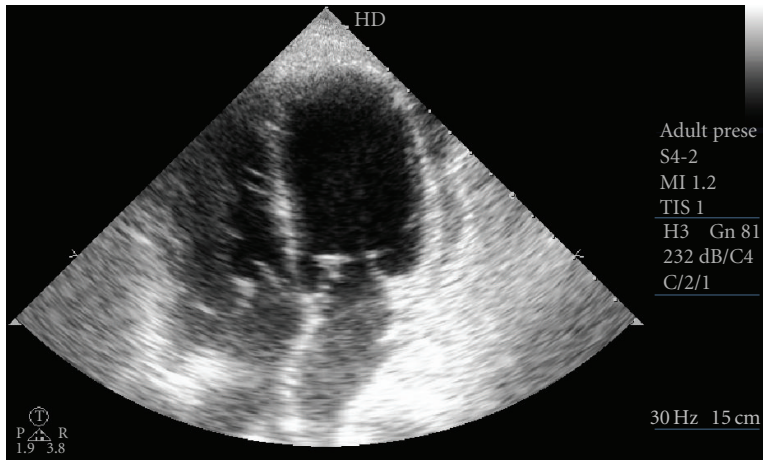

Figure 4: Apical four-chamber view depicting thick mitral and tricuspid valve and a rim of pericardial fluid.

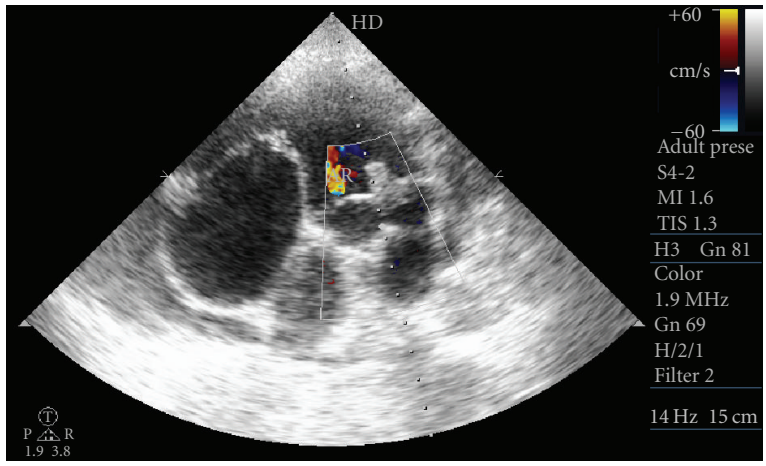

Figure 5: Parasternal SAX view depicting mild aortic regurgitation.

area was 3.2 cm^2 as measured by planimetry. The tricuspid and aortic valve leaflets were also thickened with moderate tricuspid regurgitation and mild aortic regurgitation (see Figures 4, 5).

A repeat echocardiogram after a few days showed a decrease in the amount of pericardial fluid. The serum anti-streptolysin O titer was elevated above the normal reference range; C-reactive protein and erythrocyte sedimentation rate were elevated, and hemoglobin was 12 gm/dl. Blood cultures were negative.

The presence of malaligned nonrestrictive VSD with aortic override and moderate pulmonary stenosis with net right to left shunt confirmed the diagnosis of TOF although we did not find significant infundibular stenosis.

Pericardial effusion with thickening of mitral, aortic, tricuspid valves, and valvular insufficiency was the evidence of acute rheumatic carditis.

The final diagnosis of Tetralogy of Fallot and rheumatic carditis secondary to acute rheumatic fever was made.

3. Conclusion

This patient fulfilled the Jones criteria of acute rheumatic fever. Although the occurrence of acute rheumatic fever at this age is unusual, she may be having a recurrence with the previous episode being unrecognized. An alternative hypothesis is that TOF predisposed her to the development of acute rheumatic carditis even at a later age.

The occurrence of acute severe mitral regurgitation and mild aortic regurgitation imposed volume overload on the left ventricle. This was relatively well tolerated in this patient without the development of overt pulmonary edema although there was slight elevation in pulmonary artery pressure with a peak systolic pressure of 45 mmHg (ventricular pressure = 100 mmHg/gradient = 55 mmHg) possibly because of low pulmonary flow situation in a patient of TOF. However, the occurrence of these lesions would substantially complicate further management in this patient. The risk of developing infective endocarditis would

substantially increase. Given the multifactorial etiology of both congenital and rheumatic heart disease, it is quite possible that congenital heart disease predisposes an individual to the development of rheumatic heart disease. The present case highlights this association. This is the first reported case of documented acute rheumatic activity in a patient of TOF with severe mitral regurgitation, moderate tricuspid regurgitation, mild aortic regurgitation, and pericardial effusion. We need to think about primary prophylaxis against acute rheumatic fever in patients with congenital heart disease especially in endemic areas.

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