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Case Report

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A Rare Case of Multivalvular Prolapse

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Abstract

Acute rheumatic fever has been the most frequent cause of cardiac disease in underdeveloped and developing countries. The mitral valve is the most frequently affected followed by the aortic valve. Tricuspid and pulmonary valve involvement is rarely seen. We report a case of multivalvular involvement during the first attack of rheumatic carditis with prolapse as the predominant cause of regurgitation.

Keywords

Acute rheumatic fever ; Carditis; Valvular prolapse

Key Messages

In patients with multivalvular prolapse particularly in the children and young adults, acute rheumatic fever has to be considered as one of the differential diagnoses. Early diagnosis and management can reduce the morbidity and mortality.

Introduction

Acute rheumatic fever (ARF) is the leading cause of acquired heart disease in children and young adults worldwide. The mean incidence of ARF globally is 5-50/100000 [1]. Initiated by a pharyngeal infection with group A beta hemolytic streptococci (GAS), after a latent period of two to three weeks, the illness is characterized by acute inflammation of the heart, joints, subcutaneous tissue and central nervous system. The theory of molecular mimicry holds that the GAS pharyngitis triggers an autoimmune response to epitopes in the organism that cross react with similar epitopes in the heart, brain and joints [2]. In rheumatic carditis, mitral and aortic valves are commonly involved followed by tricuspid and pulmonary valves. We report a case of all four valves involvement with prolapse as the predominant cause of pathologic regurgitation along with review of literature.

Case history

An 18year old male was referred for evaluation of a cardiac murmur to the cardiology department. He had a history of fever and joint pains of 2 weeks duration. The fever was of low grade, intermittent mitigated with medication. The arthralgia were of migratory and fleeting type involving initially lower limbs followed by upper limbs. There was no history of sore throat. There was no history of similar complaints in the past. There was no history of dyspnea, chest pain, syncope and palpitations. On examination, he was conscious and coherent. The joints were tender on movement. The pulse was 84 per minute, regular with no special character, blood pressure was 110/70mm Hg in left upper limb. On cardiac auscultation, S1 was soft, S2 normal, a pansystolic murmur of IV/VI grade over mitral area and a II/IV grade short early diastolic murmur heard over the neo-aortic area. Rest of the systemic examination was normal.

His biochemical profile showed normal hematological parameters except for the raised erythrocyte sedimentation rate of 85mm in first hour and C-reactive protein of 8.26mg/L. In view of the symptoms pointing towards ARF, the antistreptolysin (ASO) titre was done which was found to be 618 IU/ml. His Electrocardiogram (ECG) showed prominent notched P wave and increased Left ventricular (LV) forces suggestive of left ventricular hypertrophy. The chest X-ray showed cardiomegaly with straightening of left heart border. Transthoracic echocardiography showed dilated left atrium, anterior mitral leaflet prolapse with severe mitral regurgitation, aortic leaflet prolapse with moderate aortic regurgitation, tricuspid valve prolapse with moderate tricuspid regurgitation and mild pulmonary regurgitation (Figures 1 and 2). There was mild thickening of leaflets with no mitral commissure involvement. The left ventricular function was normal. There was no pericardial effusion. Patient was kept on NSAIDS, ACE inhibitors and corticosteroids along with penicillin prophylaxis. His symptoms subsided after a week of treatment and was on follow-up.

Discussion

In 2015, American heart association (AHA) -Revised jones criteria [3] for diagnosis of acute rheumatic fever had been proposed. The major criteria include carditis, arthritis, chorea, erythema marginatum and subcutaneous nodules. The minor criteria include arthralgias, fever(>38.50C), ESR >60mm in the first hour and/or CRP>3.0mg/dl and prolonged PR interval. Our patient had 2 major (carditis and arthritis) and 3 minor criteria (arthralgias, fever and raised ESR) making a definite diagnosis of ARF.

Incidence of carditis in ARF varies with the age of the patient. Compared to 90-92% of children under 3 years of age, only 15% in adults will be affected with first attack of acute rheumatic fever [2]. While traditionally rheumatic carditis is described as a pancarditis affecting the pericardium, myocardium, and endocardium, the predominant manifestation is endocarditis i.e., valvulitis [4]. Valvulitis most commonly affects the mitral and aortic valves manifesting as regurgitation. Less commonly the tricuspid and very rarely the pulmonary valves can be affected but almost never in isolation. Typically the valvulitis is usually evident at the time of presentation; however, on occasion it can be of low grade and its onset may be delayed by 2–6 weeks [5]. Valvulitis during the initial presentation of ARF is often mild and the majority of patients have mild disease. Around 5–10 % of patients who present with severe carditis have significantly worse outcomes and many will require cardiac surgery and/or succumb [6]. In our case, all the four valves were involved during the acute stage with mitral valve being the most severely affected. The prolapse is the major cause for the pathologic regurgitation in these valves. The pulmonary valve was not adequately visualized in our patient, however there is mild regurgitation.

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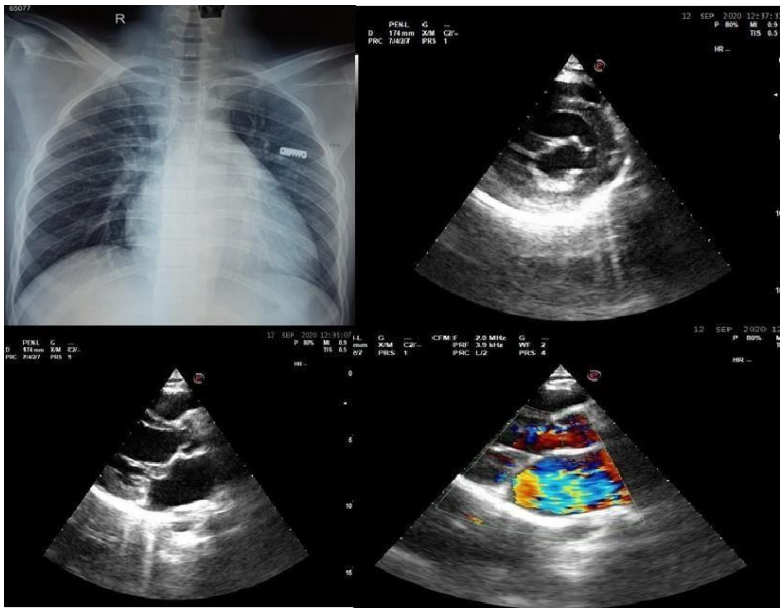

Figure 1: Upper left, Chest X-ray showing straightening of left heart border and cardiomegaly with Left ventricular apex. Upper right, Parasternal short axis view still 2D image at the mitral valve level showing beaded appearance. Lower left, Parasternal long axis still 2D image showing anterior mitral leaflet prolapse. Lower right, Parasternal long axis view color doppler showing severe mitral regurgitation.

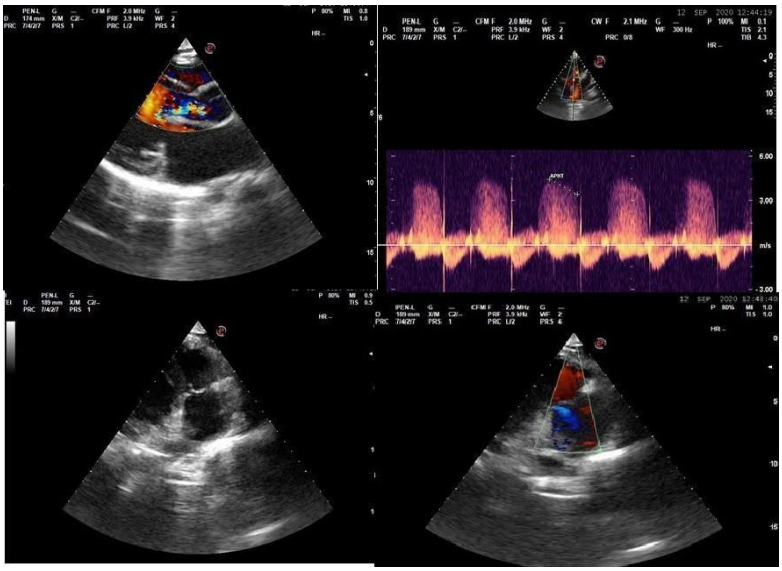

Figure 2: Upper left, Parasternal short axis view color Doppler at the aortic valve showing moderate aortic regurgitation. Upper right, Apical 5 chamber view with continuous wave (CW) Doppler showing moderate aortic regurgitation. Lower left, Right ventricular inflow view showing tricuspid leaflet prolapse. Lower right, Right ventricular inflow view color Doppler showing moderate tricuspid regurgitation.

In a study by Vijaya et al [7], echocardiographic evaluation of patients with ARF showed mitral valve prolapse in 83.69% and tricuspid valve prolapse in 12.6% of patients. A chronic rheumatic activity leads to the development of pulmonary hypertension with secondary involvement of pulmonary and tricuspid valves. However in our case, there are no other findings suggestive of pulmonary hypertension on transthoracic echocardiography.

In a case report by Güvenç O et al [8], all four valves were involved, however prolapse of only mitral valve was seen and rest of the valves had evidence of valvulitis. Multivalvular prolapse as a predominant cause of pathologic regurgitation has been rarely documented in the literature which was seen in our patient. On follow up, the degree of tricuspid and pulmonary regurgitation was reduced, however the mitral and aortic regurgitation persisted.

Conclusion

In patients with multivalvular prolapse particularly in the children and adolescent age group, acute rheumatic fever has to be considered as one of the differential diagnoses. ARF has to be ruled out even in patients without classic rheumatic fever presentation. Routine

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echocardiographic evaluation of patients with acute rheumatic fever even with normal cardiovascular examination can detect the mild carditis, so that early guideline-directed therapy can prevent long-term morbidity and mortality.

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