Predominant RV Endomyocardial Fibrosis Masking Rheumatic Mitral Stenosis

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Abstract

We report a 30 year female with rheumatic mitral stenosis, presenting with progressive right heart failure without overt left heart symptoms. Rapid progression to severe right heart failure with moderate pericardial effusion prompted the diagnosis of associated restrictive cardiomyopathy. A MRI scan of the heart revealed diffuse subendocardial scarring of LV and RV explaining patient’s rapid downhill course.

Introduction

Rheumatic mitral stenosis remains most common cause of right heart failure in adult females in India. Rapid progression of symptoms should alert one to associated underlying pathologies which may be responsible for patient’s symptoms.

Case Report

A 30 year female, presented with complaints of swelling of both lower limbs and fatigue increasing over a period of 3 years. Her symptoms had progressed over last 6 months with increasing oedema feet and abdominal distension. She was admitted to hospital with generalised oedema and decreased urinary output. Examination revealed pulse of 110/minute, low volume; blood pressure of 90/60 mm of Hg, raised JVP and muffled heart sounds.

Electrocardiogram showed sinus rhythm, low voltage complexes (Figure 1). An emergent 2D echocardiogram revealed dilated RA and LA, with moderate to large effusion. Also noted was thickened and doming mitral valve with a mean gradient of 10 mm of Hg, severe free TR, mild mitral regurgitation and no pulmonary artery hypertension. No diastolic RA or RV collapse was noted. Careful echocardiography after pericardiocentesis revealed obliterated RV apex with good function and severe free TR (Figures 2, 3). Pericardial fluid examination showed normal cytology and biochemistry. Routine investigations showed leucocytosis, raised creatinine and liver enzymes. In view of disproportionately dilated RA, MRI of heart was done which showed bialtral enlargement (hugely dilated RA), with normal sized LV and RV. Diffuse subendocardial enhancement involving left and right ventricle with obliteration of RV apex was noted suggesting endomyocardial fibrosis. Along with these features of endomyocardial fibrosis; thickened doming mitral valve was noted seen consistent with rheumatic mitral stenosis (Figures 4, 5).

A diagnosis of rheumatic mitral stenosis with endomyocardial fibrosis was made.

Patient was managed conservatively with poor response to treatment.

Discussion

Rheumatic mitral stenosis presents with symptoms of right heart failure in advanced cases. Onset of right heart failure in patients with mitral stenosis is associated with amelioration of breathlessness with fatigue and oedema setting in. Absence of this typical natural history (as in our case) should make one suspicious...
of associated underlying right heart disease. Normal right ventricular systolic function with diated RA was another clue to the diagnosis.\(^1\) Presence of moderate to large pericardial effusion in absence of infection is unusual in isolated mitral stenosis with right heart failure. Our patient satisfied two major criteria (obliteration of RV apex and severe tricuspid valvular regurgitation due to adhesion of valvular apparatus to ventricular wall) and one minor criteria (Enlarged atrium with normal ventricles) for diagnosis of EMF.\(^2\)

The hallmark feature of RV apical obliteration could be appreciated only after the effusion was tapped. Predominant involvement of right ventricle with raised RA pressures might have precluded the development of pulmonary venous congestion associated with mitral stenosis. Presence of enormously dilated right atrium (6 cm) is another clue to diagnosis.\(^3\) Although both ventricles were involved in our patient, predominant RV involvement was obvious. On Magnetic resonance imaging the presence of delayed enhancement of LV and RV subendocardium helped in further confirming the diagnosis.

EMF is a disease of tropical and subtropical countries. EMF is the commonest restrictive cardiomyopathy reported in India.\(^1\) Isolated RV involvement has been reported more commonly in southern India, whereas biventricular involvement was common in a study in northern India.\(^1,4,5\) It affects young adults and adolescents, with majority of patients presenting with class III symptoms. Eosinophilia has been traditionally regarded as a risk factor, although it is absent in many cases.\(^6,7\) In a retrospective Indian study neither eosinophilia nor eosinophilic organ infiltration was identified among angiographically diagnosed cases of EMF.\(^1\) In the setting of clinically severe MR or TR, EMF can be suspected if there is echocardiographic evidence of a small or normal-size ventricle with a grossly dilated corresponding atrium.\(^3\)
Both rheumatic heart disease and endomyocardial fibrosis are common in tropical countries and have predilection for young patients. It has also been reported in Middle East countries. Eosinophilia (primary or secondary), tapioca ingestion and cerium have been implicated in pathogenesis of EMF. Commonly pathogenesis of rheumatic heart disease starts earlier; and EMF alters natural history of rheumatic heart disease by accelerating the process of fibrosis, with patient presenting earlier or it can delay the diagnosis by masking the signs of left heart failure. In absence of apical obliteration (a hallmark feature of EMF); diagnosis of EMF may be masked when both occur concurrently. MRI can be exceedingly helpful in establishing diagnosis in absence of biopsy and/or cardiac catheterisation in unstable patients.

Presentation with either LV or RV failure is common for both. However isolated RV failure with no preceding history of LV failure should prompt one to think of EMF as one of differentials in patient with established rheumatic heart disease. Concurrent occurrence of rheumatic heart disease and endomyocardial fibrosis is known. EMF itself affects AV valves (where posterobasal myocardium and papillary muscles are almost invariably involved); however posterior mitral leaflet affection is more common and severe then anterior causing predominantly regurgitant lesions. In a necropsy study from Africa, concurrent occurrence of rheumatic heart disease and endomyocardial fibrosis was 5 times more common than expected. This may indicate that in endemic region rheumatic heart disease may increase susceptibility to endomyocardial fibrosis and may itself be involved in pathogenesis. (Although no direct evidence as such is available to us). As hypothesised by Shaper almost 4 decades ago, there may be a common predisposition factor for EMF and RHD.

Decline in incidence of RF as well as that of EMF in Kerala points to a common predisposition. This decline parallels improving socioeconomic condition in the region, which may be a major factor in the control of this enigmatic disease.

References