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Case Report

Rare Case of Isolated Right Sided Dilated Cardiomyopathy

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ABSTRACT

In dilated cardiomyopathies biventricular involvement is the usual case though left ventricular dysfunction is more apparent. In RVDCM, as the name suggests right ventricle is solely involved and left ventricular function is normal

The other causes of right heart failure are tricuspid valve disease (infective endocarditis, rheumatic, Ebsteins anomaly), right ventricular infarction and rarely pulmonary regurgitation without pulmonary hypertension. The diagnosis was made on echocardiography which revealed dilated, hypokinetic right ventricle, with normal left ventricular dimensions and ejection fraction and no evidence of pulmonary artery hypertension with normal pulmonary artery size and normal pulmonary valve with dilated right ventricle outflow tract

Keywords: Heart Failure, Right Sided Cardiomyopathy, Cardiomegaly

INTRODUCTION

Dilated cardiomyopathy involves the left ventricle with some dysfunction of the right ventricle. Isolated right dilated cardiomyopathy presents as right sided cardiac failure or ventricular tachycardia and sometimes as sudden cardiac death[1]

There is a paucity of clinical findings in Right sided heart failure and the patient is asymptomatic when the rhythm is not irregular. The diagnosis can be missed. Here we discuss a case where the presentation was with syncope, bilateral pedal edema, abdominal distension, nausea, anorexia and general debility. The diagnosis was made on echocardiography which revealed dilated, hypokinetic right ventricle, with normal left ventricular dimensions and ejection fraction and no evidence of pulmonary artery hypertension with normal pulmonary artery size and normal pulmonary valve with dilated right ventricle outflow tract[2]

CASE HISTORY

A 34-year-old female presented to G.K. General Hospital in emergency department with complains of bilateral leg swelling, abdominal distension, generalized weakness, anorexia and vomiting.

On physical examination we noticed engorged neck veins, tachycardia with heart rate 120beats/min, blood pressure was 90/60 mm Hg, bilateral pedal edema which was pitting in nature and ascites.

She had enlarged, pulsatile liver with tenderness and hepatojugular reflux was positive,

On cardiovascular examination apex beat was shifted towards left fifth intercostal space in mid axillary line, left parasternal heave grade 3/3 was present, on auscultation grade 4/6 murmur of tricuspid regurgitation was heard, p2 was normal, mitral valve aortic valve and pulmonary valve clinical examination was normal

Respiratory system examination was not significant. Per abdominal examination: Ascites was elicited. Patient also had peripheral cyanosis which did not respond to oxygen therapy. ECG showed (Figure 1) qrs axis +110, sinus tachycardia with normal p wave

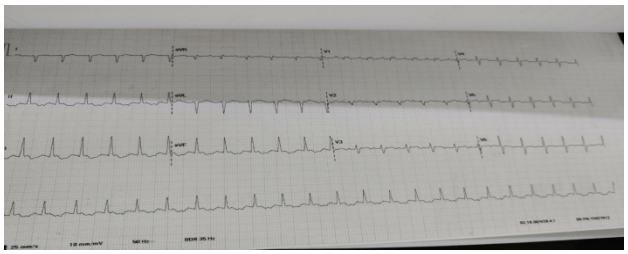

Figure 1: ECG

Chest X-ray PA view showed gross cardiomegaly with mild pleural effusion on left side, there was no pulmonary plethora and lung parenchyma was normal.

Echocardiography showed (Figure 2) RV size of 4.5 cm with paradoxical motion of IVS, LV size was hypo plastic, mitral, aortic and pulmonary valves were normal, (Figure 3) right ventricular outflow tract was hugely dilated, Color Doppler, (Figure 4) pulse wave Doppler and continuous wave Doppler suggested severe TR with normal pulmonary arterial pressure, flow velocity of TR was 120 cm/second. There was reduced motion of the right ventricular anterior wall, (Figure 5) RA was grossly dilated, structure of the tricuspid valve was normal except during systole the closure was not proper due to the dilatation of TV ring, interatrial septum was tilted towards the left atrium, no clots were seen in right atrium, left atrium was totally normal.

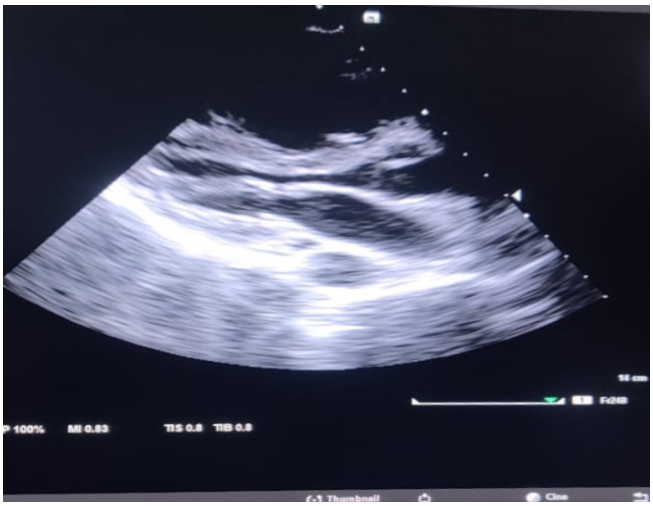

Figure 2: Dilated RV, Paradoxical IVS, Hypoplastic LV and LA, Normal MV

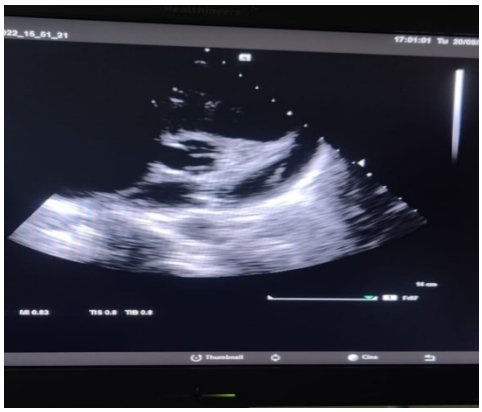

Figure 3: Grossly Dilated RVOT

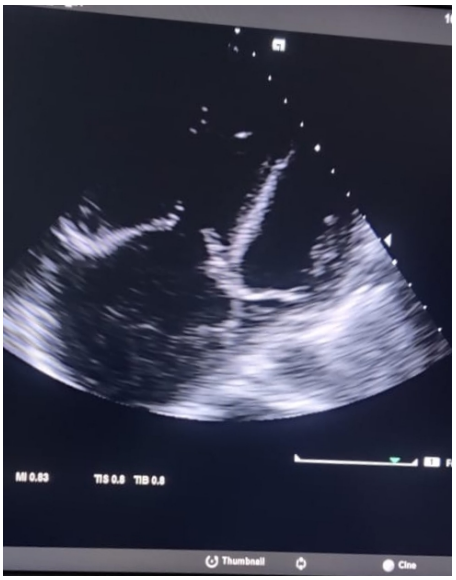

Figure 4: Dilated RV and RA

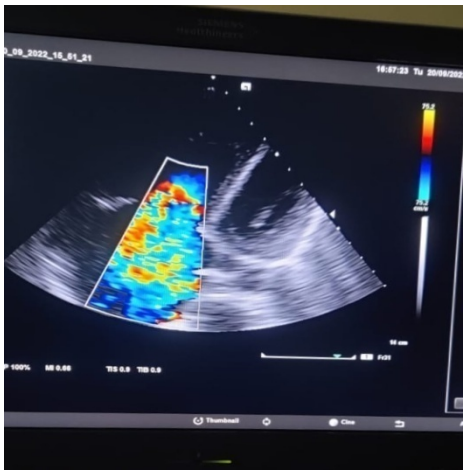

Figure 5: Severe TR Jet in RA

DISCUSSION

In dilated cardiomyopathies biventricular involvement is the usual case though left ventricular dysfunction is more apparent. In RVDCM, as the name suggests right ventricle is solely involved and left ventricular function is normal.[3]

The other causes of right heart failure are tricuspid valve disease (infective endocarditis, rheumatic, Ebsteins anomaly), right ventricular infarction and rarely pulmonary regurgitation without pulmonary hypertension. RVDCM most often announces itself with ventricular tachycardia and rarely as congestive cardiac failure. A male preponderance is seen and the most common clinical presentation is syncope or sudden death in young healthy male, thus unexplained ventricular tachycardia should raise the possibility of RVDCM[4]

A recent study of sudden cardiac death among healthy young athletes right ventricular cardiomyopathy was the most common cause followed by hypertrophic cardiomyopathy and coronal artery anomalies. The ventricular tachycardia has left bundle branch block pattern (ectopic focus in right ventricle) and the sinus rhythm electrocardiogram shows low voltage complexes reflecting the extent of right ventricular dilatation[5]

Cardiomegaly with right ventricular involvement is seen on chest radiograph and echocardiography reveals no abnormality with left ventricle, with poorly contractile and grossly dilated right ventricle. On myocardial biopsy hypertrophy and fibrosis are mostly seen.

Two conditions which closely resemble RVDCM are arrhythmogenic right ventricular dysplasia and right ventricular endomyocardial fibrosis. Arrhythmogenic right ventricular dysplasia which also presents as ventricular tachycardia is now considered a separate entity from RVDCM. The former is characterised by infundibular dyskinesia, aneurysm of the inferior or lateral wall, diastolic bulging below the tricuspid valve and highly refractile papillary muscle.

In endomyocardial fibrosis there is fibrotic obliteration of right ventricular apex and extension of the fibrosis back along the inflow tract to encase the papillary muscles. Uhl's anomaly is a form of RVDCM which presents as cyanosis, congestive cardiac failure and death in infants. Here the myocardiumis hypo plastic or aplastic (parchment heart of Osler).

The etiology of RVDCM is ill understood. Heredofamilial basis has been established. In experimental studies Coxsackie virus infection can produce RVDCM. No causal relationship has been linked with alcohol or any specific dietary deficiency.

RVDCM in our country usually present with congestive symptoms and supraventricular tachycardia unlike the British variety in which ventricular tachycardia is more common. This case was atypical for the Indian series and presented with ventricular tachycardia.

Treatment of RVDCM is towards prevention of ventricular arrhythmias and sudden death. This is in contrast to other cardiomyopathies where afterload reduction and improvement in systolic function is aimed for. Prophylactic antiarrhythmic drugs which are useful in RVDCM are propranolol and amiodarone. Internal defibrillator implantation and electrode ablation of ectopic focus are employed for recurrent and resistant ventricular tachyarrhythmia's. Yamaguchi et al from Japan had detected selective apical hypertrophic cardiomyopathy, so such isolated cardiomyopathy entities involving only segmental chambers of the heart have been detected.

CONCLUSION

This was a case of rare isolated right ventricular dilated cardiomyopathy presenting with right sided heart failure rather than with ventricular arrhymias or syncope, there were characteristic echocardiography finding of dilated right atrium and ventricle with tricuspid regurgitation without evidence of pulmonary hypertension.

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