

CASE REPORT

Restrictive Cardiomyopathy: A Rare Case Report**Bilal Bin Abdullah^{*}, Mehboob.M.Kalburgi, Sahana Shetty
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Abstract: We report a 28 years old male presenting with heart failure. A thorough clinical evaluation directed us towards restrictive heart disease. Doppler echocardiographic study was used as a main modality of diagnosis and cardiac catheterization confirmed the diagnosis of idiopathic restrictive cardiomyopathy. We express the contribution of clinical findings and appropriate diagnostic measures in approaching a case of Restrictive cardiomyopathy (RCM).

Introduction

Restrictive cardiomyopathy (RCM) refers to either idiopathic or systemic myocardial disease which is characterized by restrictive filling, normal or reduced left ventricle (LV), right ventricle (RV) volumes with preserved systolic function. Abnormal ventricular diastolic compliance and impaired ventricular filling constitute the central pathophysiologic components. Congestion and elevated diastolic pressure are the major clinical and hemodynamic manifestations [1]. It has a rare incidence of 6 in 1 lakh population with elderly females more affected than males (F: M ratio 1.5: 1) [2]. Endomyocardial fibrosis variety of RCM is commonly reported from Kerala in India [3]. Here we report a rare case of IRCM in a young adult male and the diagnostic approach.

Case History

A 28 year old male presented to the out patient department with breathlessness on exertion, fatigability, pedal edema and abdominal discomfort since 2 years, aggravated since last 2 months .O/E- Pulse rate 88/min, irregularly irregular low volume .Blood pressure 100/70 mm Hg, respiratory rate 18/min, bilateral pedal edema present, jugular venous pressure (JVP) raised (10cm H₂O) with prominent “y” descent. CVS examination revealed apical impulse well defined in left fifth intercostal space 1 cm lateral to mid clavicular line, variable S₁, loud P₂, systolic murmurs in mitral and tricuspid areas. Respiratory system revealed clinically no abnormalities. Per abdomen showed congestive hepatomegaly with ascites. On investigations, ECG showed normal voltage complexes with atrial fibrillation, right axis deviation with ST –T changes (reverse tick sign) (Fig 1). Chest x ray showed enlarged cardiac silhouette with pulmonary congestion (fig 2). Echocardiography showed marked biatrial enlargement, mildly hypertrophied but normal sized ventricles, moderate MR, severe TR, mild PAH (SPAP 40 mmHg), fair biventricular function, no intracavitary clots, with minimal pericardial effusion. Echocardiography measurements LA 64mm, IVS 10 mm, LVID(d) 45mm, LVID(s)

35mm, PWD 8 mm, EF 50 % (fig 3). Cardiac catheterization revealed grade III MR, severe TR severe PAH, mild RV systolic dysfunction, no apical obliteration, no endocardial calcification, fair LV systolic dysfunction with normal coronaries. With these evidences a diagnosis of restrictive cardiomyopathy was made.

Fig: 1 ECG

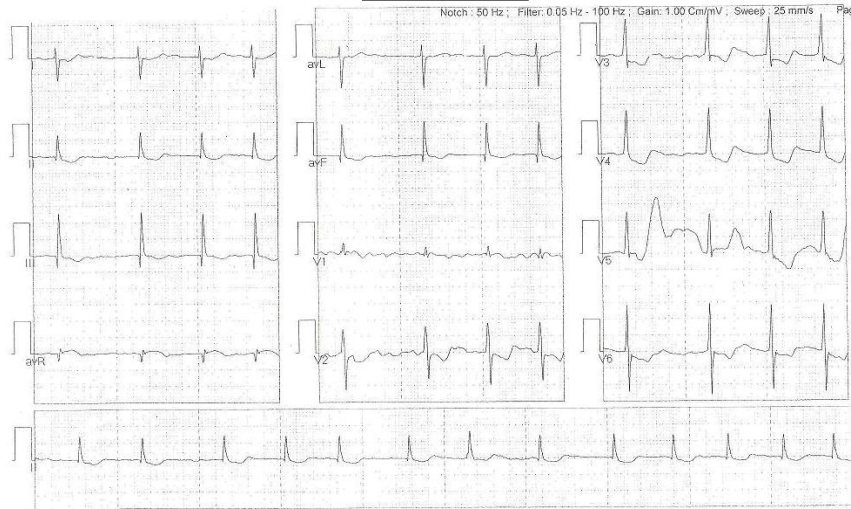


Fig 2: Chest X-ray PA view

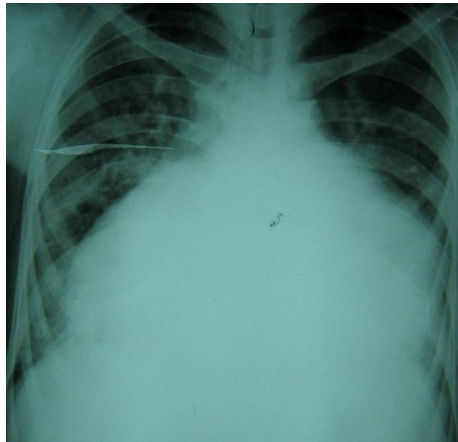
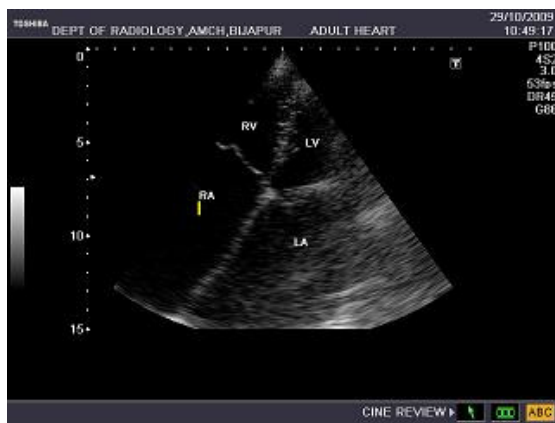


Fig 3: Echocardiography four chamber view



Discussion

This patient of heart failure presented with symptoms of systemic and pulmonary congestion was evaluated and significance was given to the raised JVP with prominent “y” decent suggestive of restrictive heart disease. Clinical signs of mitral regurgitation (MR), tricuspid regurgitation (TR) and pulmonary arterial hypertension (PAH) with atrial fibrillation (AF) were significant. ECG showed normal QRS

voltage with AF, right axis deviation and ST-T changes (suggestive of digoxin toxicity). Presence of AF with negative “y” descent made us to evaluate regarding restrictive right heart diseases. Echocardiography showed biatrial dilatation, restrictive Doppler flows, dilated inferior vena cava, preserved systolic function and atrio ventricular regurgitations which were characteristic of RCM. Echocardiography also helped us in excluding endomyocardial involvement as seen in endomyocardial fibrosis by the absence of obliteration of ventricular cavities and right ventricular outflow tract dilatation. This was confirmed by cardiac catheterization which revealed markedly elevated ventricular filling pressure with characteristic restrictive haemodynamic pattern and no obliteration of cavities. Endomyocardial biopsy was not performed as studies have shown its limitation reserved in excluding infiltrative disorders and being a blind procedure [4]. Idiopathic restrictive cardiomyopathy is characterized by clinical syndrome of restrictive heart disease of unknown etiology without pericardial and endocardial pathology.

Evaluation of IRCM starts with thorough clinical examination which shows pulmonary and systemic congestion, with normal and palpable left ventricular impulses, third heart sound and systolic murmurs of MR, TR. ECG may be nonspecific but usually show rhythm disturbances like atrial fibrillation, premature beats and conduction delays with normal voltage criteria. Chest x ray may be near normal in early stages but symptomatic patient shows moderate to mark cardiomegaly due to biatrial enlargement and pulmonary venous congestion. Echocardiography with Doppler studies has now become revolutionized modality of diagnosis which shows the characteristic morphology of non dilated, non hypertrophied ventricles, preserved LV function, atrio ventricular (AV) regurgitations and restrictive flow. The typical restrictive pattern can be recognized by increased mitral E velocity, increased ratio of mitral early to late filling (E/A ratio >2) and shortened deceleration time (typically less than 150mmsec) [5]. Cardiac catheterization is advised in patients with strong consideration of RCM to document the diagnosis, assess severity and in some patient to establish the etiology by means of endomyocardial biopsy [6]. Restrictive cardiomyopathy has to be differentiated from secondary myocardial involvement with amyloid, sarcoidosis, hemochromatosis, glycogen deposition, EMF, scleroderma, hypereosinophilic disease ,following mediastinal irradiation, neoplastic infiltration and myocardial fibrosis of diverse causes. These are differentiated by the presence of substantial concomitant endomyocardial involvement, partial obliteration of ventricular cavity by fibrous tissue and thrombi, on echo and CT angiogram [2]. Few cases of pseudo xanthoma elasticum are attributed with restrictive cardiomyopathy. Studies have shown poor prognosis in IRCM and was related adversely with age, male sex, NYHA functional class, left atrial diameter (>60mm) [2].

Hence IRCM should be considered in patients presenting with diastolic heart failure, with non hypertrophied and non dilated ventricles. Comprehensive echocardiography Doppler assessment has now become an important non invasive technique of detecting the pathophysiology, morphology and prognosis of RCM [7].

References

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