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Case Report

Primary Restrictive Nonhyperthrophied Cardiomyopathy: Radiological Definition of Cardiomyopathy

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ABSTRACT

Primary restrictive nonhyperthrophied cardiomyopathy is a rare type of cardiomyopathy with poor prognosis and highest rate of cardiac sudden death. We describe a rare case of this type of cardiomyopathy that suffered a intrahospital sudden death during assessment.

Keywords: Restrictive nonhyperthrophied; Cardiomyopathy; Radiological definition

INTRODUCTION

Restrictive Cardiomyopathy (RCM) is a uncommon type of cardiomyopathy with poor prognosis and highest rate of Sudden Cardiac Death (SCD) [1]. The exact incidence of RCM remains unknown but some epidemiological study reported that RCM accounts for 5% of cardiomyopathic disorders [2]. Based on underlying etiology the RCM is categorized to two types of primary and secodary cardiomyopathy [3]. Primary restrictive nonhyperthrophied cardiomyopathy is the least common type of restrictive cardiomyopathy. It is associated with biatrial enlargement, a normal or small sized LV and RV and ventricular wall thickness. The patients primary symptoms and signs are those of pulmonary venous obstruction and right heart failure. Respiratory symptoms are prominent sudden death may occur [4]. We aimed to report a young patient who came to our heart center with presentation of primary restrictive nonhyperthrophied cardiomyopathy.

CASE REPORT

A 19-year old lady presented with a 6 months DOE functional class 2-3 and reduced exercise tolerance her familial history of sudden cardiac death and CMP was negative. She had no prior medical history 12-leads ECG showed sinus rhythm with p-pulmonale, normal QT interval, narrow QRS and without repolarization change. Routine blood test was normal initially. Transthoracic study showing (Figure 1) Normal LV size with mild LV systolic dysfunction , LVEF: 45% ,Increase LV filling pressure, Diastolic dysfunction grade 3 (PEV: 54 cm/s, PAV: 19 cm/s, DT: 99m/s, S: 18 cm/s, D: 44 cm/s, eseptal: 4 cm/s, elateral: 5 cm/s, E/e: 13.5), Normal RV size with moderate RV dysfunction, severe LA and RA enlargement, at least moderate TR, sPAP: 70 mmHg. Based on above data RCM considered for her diagnosis, thus non-invasive diagnostic approach was done. She had normal serum free light chain and serum and urine protein electrophoresis with immuno fixation. There wasn't extra cardiac organ involvement. For better evaluation of RCM cause, she was referred to do CMR exam. CMR findings (Figure 2 and 3) showed small LV cavity size (EDV index: 32.8 ml/m2), LVEF: 36%, RVEF: 35%, LV mass: 52 g, Normal LV wall thickness, RVH, severe biatrial enlargement, no septal bounce, Cardiact[2]*: 40 ms, no myocardial inflammation in STIR scene, normal pericardial thickness in T1W scene, In late GAD scene, there was Late gad enhancement at LV attachment to ventricular septum

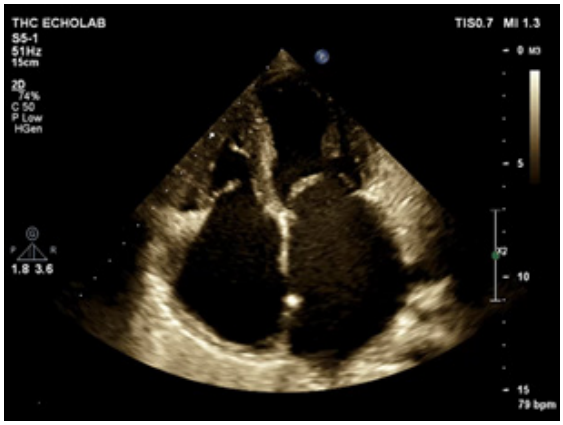

Figure 1: Normal LV size with mild LV systolic dysfunction, LVEF: 45%, normal RV size with moderate RV dysfunction, severe LA and RA enlargement, at least moderate TR, sPAP: 70 mmHg.

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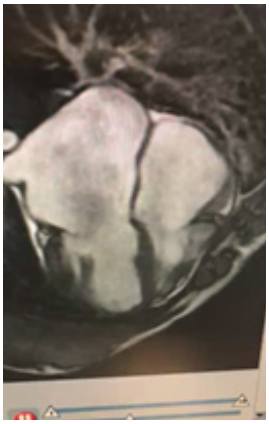

Figure 2: Small LV cavity (EDV index: 32.8 ml/m2), LV mass: 52 g, normal LV wall thicknes, RVH, Cardiac t2*: 40 ms, no myocardial inflammation.

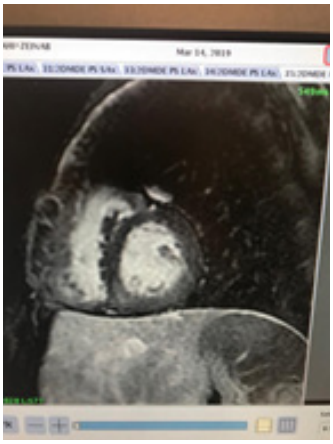

Figure 3: Normal pericardial thickness late gad enhancement at LV attachment to ventricular septum.

DISCUSSION

Genetics and breeding

Primary restrictive nonhyperthrophied cardiomyopathy is the least common type of cardiomyopathy [1]. Owing to its rarity diagnosis is challenging the case was diagnosed based on careful assessment. Essential feature of this patient was restrictive filling pattern, based on above data we exclude amyloidosis, sarcoidosis, hemochromatosis (3 major causes of RCM) and other rare cause of RCM for instance idiopathic diabetic, CMP, scleroderma, myofibrilar myopathies, pseudoxanthoma elasticum, sarcomeric protein disorder, werners syndrome (appearance of premature aging) for exact diagnosis. We had planned to do right heart catheterization and endomyocardial biopsy, but the patient refused and we lost her in short period due to SCD. However in spite of the biopsy failed and contraversy in its exact definition, by excluding other causes of RCM the primary cardiomyopathy was proposed but based on radiological findings we introduced this term for this presentation. The LVEF was difference between echocardiography and cardiac MRI findings and the reason that justify this difference is that in CMR the LVEF is measured by volume and because of taking diuretic and consequently reduced preload, this difference was likely two reasons justify the reduced LVEF, one of them is pulmonary hypertention and the other one is advanced disease. According to this definition there were two special points in this case. One of them is the extracardiac involvement was absent and the other one is that she had no family history. However these two points had no role in diagnosis because these are not diagnostic criteria based on this case report the authors suggest that cardiologists should be considered this diagnostic term for this presentation.

The authors have no conflict of interest to declare.

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