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Clinical case report based study

Biventricular noncompaction presented with symptomatic complete heart block – Report of a case and review of literature

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ABSTRACT

Ventricular noncompaction has been recognized as a distinct form of rare cardiomyopathy characterized by numerous, prominent ventricular trabeculations and deep intertrabecular recesses and is caused by a disorder of endomyocardial morphogenesis. Concomitance of either valvular pathologies or complete atrioventricular block with biventricular noncompaction has rarely been reported. Herein, we present a case of 67 years old male presented with syncopal attack and congestive heart failure due to biventricular noncompaction with significant left ventricular dysfunction associated with complete atrioventricular block. He was formerly diagnosed as dilated cardiomyopathy for last 2 years. Review of literatures of all reported cases has been discussed.

1. Introduction

Noncompaction of the ventricular myocardium (NCVM) is an unclassified congenital cardiomyopathy resulting from an arrest in normal endomyocardial embryogenesis characterized by an excessively prominent trabecular meshwork with deep intertrabecular recess.[1] It frequently complicated with heart failure, malignant arrhythmias and embolic events.[2] The clinical manifestations of this entity are variable, from patients who are asymptomatic with a preserved LV ejection fraction to patients that have various degrees of systolic ventricular dysfunction and eventually develop heart failure, systemic embolism and various types of cardiac arrhythmias.[3] We are reporting a patient with NCVM who presented with symptomatic complete heart block.

2. Case history

A 67 years old male, non-diabetic, non-hypertensive, non-smoker and had history of dyspnea on exertion of New York Heart Association (NYHA) class II since last two years. He was previously admitted twice due to congestive heart failure (CHF) in the past and was on treatment with oral enalapril, digoxin, diuretics and carvedilol. This time he admitted in our hospital with a history of syncopal attack and features of CHF. At admission, his heart rate was 20 beats/min (bpm), blood pressure was 130/90 mm oh Hg with engorged neck veins and pedal edema. Systemic examination revealed pansystolic murmur of grade III/VI at apex and left lower parasternal border, early diastolic murmur at neo aortic area along with bilateral basal crepitations and altered sensorium. Electrocardiogram (25 mm/s, 10 mm/mV) was done at admission which showed complete heart block (CHB) with ventricular escape rate of 20 bpm (Fig. 1). Patient was immediately put on bedside temporary pacemaker through internal jugular vein and rate was fixed at 80 bpm. Blood investigation showed renal dysfunction (blood urea-70, serum creatinine-1.6), normal electrolytes and normal complete blood counts. Transthoracic echocardiography revealed non-compaction of ventricular myocardium involving apical portions of both ventricles and lateral & inferio-posterior wall of left ventricle (Fig. 2). The ratio of noncompacted-to-compacted myocardium was >2 in the maximal thickened wall, measured at the end systole. On the color Doppler echocardiography, these recesses were filled with blood from the ventricular cavity. All the four cardiac chambers were dilated with biventricular global hypokinesia was present. The calculated ejection fraction by modified biplane was only 30%. On color Doppler echocardiography, there was presence of aortic, mitral and tricuspid regurgitation of moderate severity. Aortic cusps and mitral valve leaflets were calcified and thickened. Papillary muscles were normal. Doppler echocardiographically

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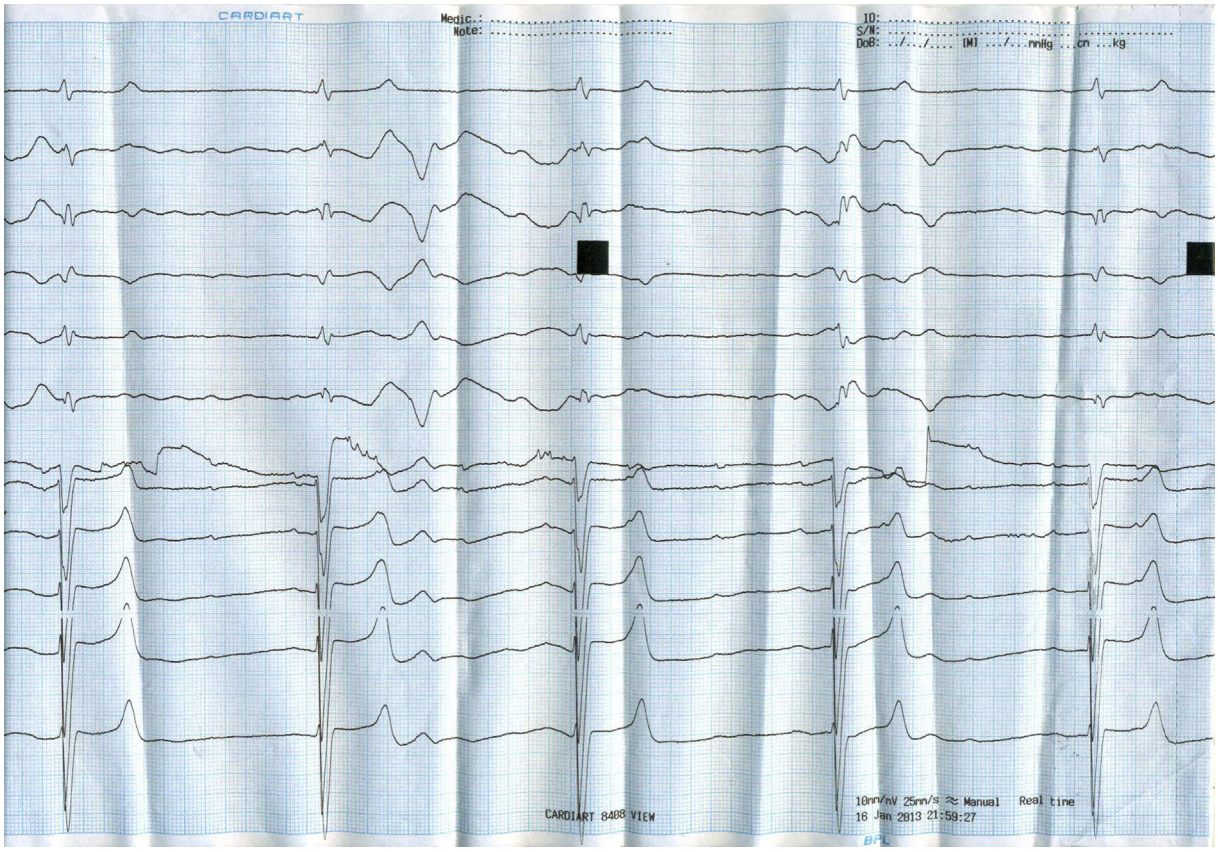

Fig. 1. ECG showing complete heart block with ventricular escape at 20 bpm.

measured pulmonary artery systolic pressure over tricuspid insufficiency was 64 mm Hg. Patient was on temporary pacemaker along with conservative treatment for CHF. His sensorium and symptoms of cardiac failure improved after four days. Though he was offered dual chamber pacemaker for the management of CHB along with severe biventricular dysfunction, but due to financial constrain, he was put on VVIR pacemaker and was discharged to home after 10 days of hospital admission. He was put on angiotensin converting enzyme inhibitor (ACEI); loop diuretic, aldactone and acetyl salicylic acid.

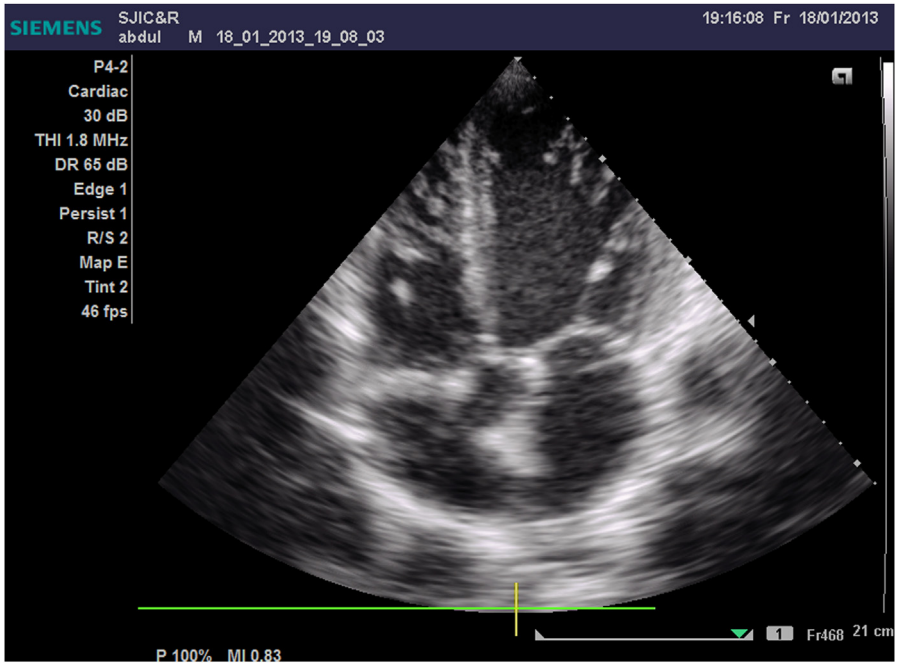

Fig. 2. Biventricular noncompaction of apical & lateral wall of LV & apical wall of RV.