Case Report

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Marons Posterior Type Hypertrophic Cardiomyopathy

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Abstract

Hypertrophic cardiomyopathy (HCM) is one of the most common inherited cardiomyopathy. The identification of patients with HCM is sometimes still a challenge. Moreover, the pathophysiology of the disease is complex because of left ventricular hyper-contractile state, diastolic dysfunction, ischemia and obstruction which can be coexistent in the same patient. We report a case of 35 year old male presenting with effort dyspnea NYHA class II with ECG showing deep arrowhead T wave inversion in anterolateral leads and TTE revealed it to be HCM with thickened LV posterior wall (26mm) as compared to septum (12 mm) which belongs to Marons Type IV type, rarest across HCM phenotypes.

Keywords: Cardiomyopathy; Classification; Septum.

Introduction

Hypertrophic cardiomyopathy (HCM) is clinically defined in presence of left ventricular (LV) hypertrophy in the absence of hypertension and valve disease. HCM occurs in approximately 1:500 of the general population [1-3]. HCM is usually diagnosed as LV thickness at septum and free wall level being ≥15 mm with characteristic asymmetrical septal hypertrophy with septal to free wall thickness ratio > 1.3 - 1.5 but Marons posterior type is an exception as here the reverse occurs i.e LV posterior wall is thicker as compared to septum.

Case Report

We report a 35 year old male presenting to the Cardiology OPD of AIIMS, Bhubaneswar with effort dyspnea NYHA class II, he was normotensive, clinical examination was unremarkable except the presence of LVS_4, ECG revealed deep arrowhead like T wave inversion in anterolateral leads and echocardiography revealed thickened LV posterior wall i.e 26 mm as compared to the interventricular septum i.e. 12 mm. LVOT gradient was 6 mmHg and ejection fraction was supernormal i.e. 75% with presence of grade 1 diastolic dysfunction.

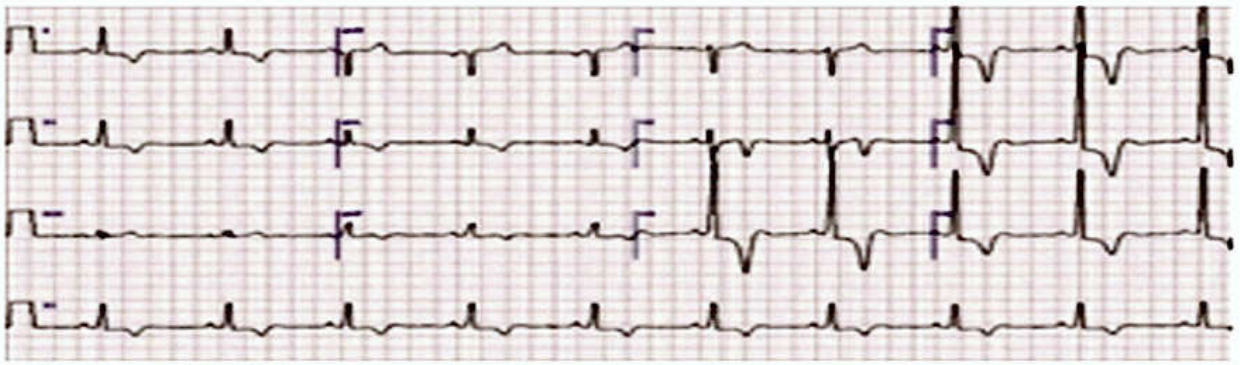

Fig. 1: ECG

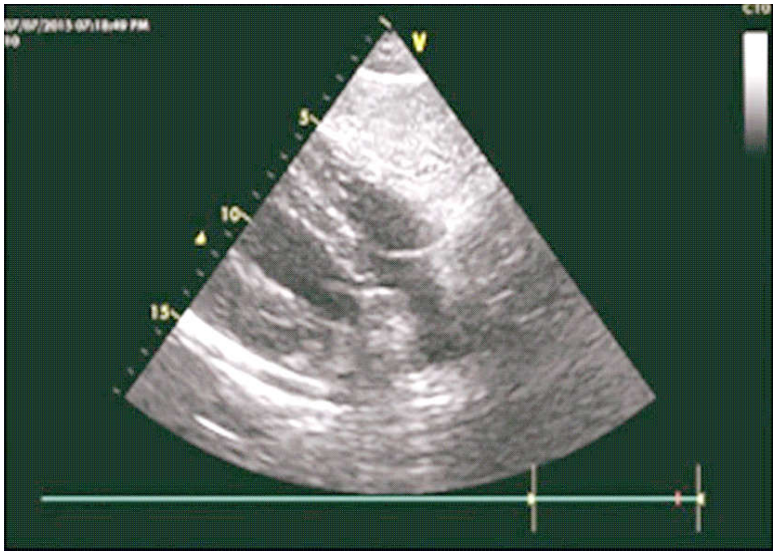

Fig. 2: Thickened LV posterior wall compared to IVS (Modified PLAX view)

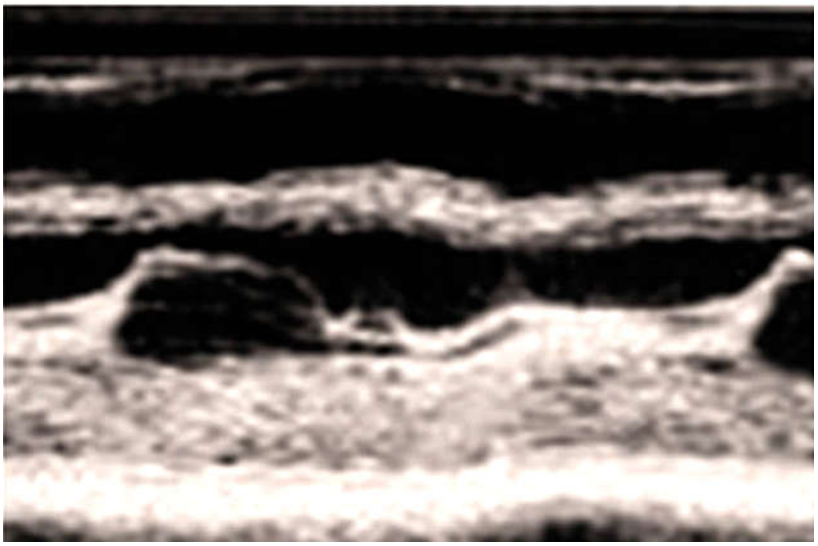

Fig. 3: M –mode echocardiography showing thick LV posterior wall

The patient had no history of syncope nor any history of sudden cardiac death in family. Holter analysis was advised and it revealed occasional ectopics. Patient was advised to take metoprolol 50 mg twice daily with periodic follow up.

Discussion

BJ Maron in 1981 provided morphological classification of HCM as follows [4,5].

Type I- Antero septum

Type II- Whole septum

Type III- Septum + at least a part of LV free wall (anterior/posterior/lateral)

Type IV- Other location (Posterior/Apical/Lateral)

Out of all four subtypes Type I is commonest type (47%) and type IV type is rarest (7%). *Hence our case belongs to the rarest phenotype of HCM.* Type I patients are usually elderly, whereas Type IV patients are younger than other groups. Family history of HCM and sudden cardiac death are significantly more common in Type II and type III (51 and 38 %, respectively) as compared to Types I and IV. *Patients with Type IV hypertrophy exhibit less dyspnea at exertion than the three other groups, and less frequently undergo medical treatment.* Type III pattern exhibit more frequently maximal LV hypertrophy > 30 mm with higher LV filling pressures. Type III patients also exhibit significantly lower exercise tolerance and are more likely to present with abnormal blood pressure responses to exercise. Type IV patients rarely suffer NSVT or paroxysmal atrial fibrillation and also rarely require ICD implantation as compared to other

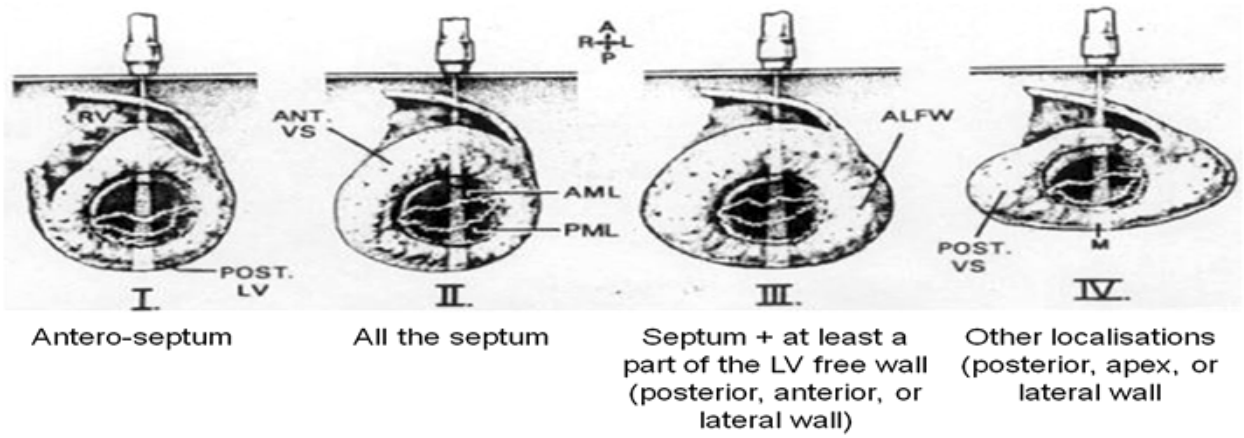

Fig. 4: Bulls eye plot of Marons HCM types

phenotypes [6]. Although Maron initially described 17 subtypes of HCM, at least recognizing these four phenotypes one can prognosticate the HCM patients as in our case being *least malignant* and rarest of the phenotype.

Conclusion

We report the rarest phenotype of hypertrophic cardiomyopathy i.e. Marons posterior type. Physicians while evaluating a suspected hypertrophic cardiomyopathy should not ignore the thickened posterior wall; otherwise they will miss this rare variety landing the patient into sudden cardiac death one day. This rare case report teaches us eye ball assessment of whole left ventricle is required, not only the asymmetrical hypertrophy of septum to diagnose the dangerous hypertrophic cardiomyopathy.

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