ORIGINAL RESEARCH PAPER

General Medicine

A RARE CASE OF HEART FAILURE SECONDARY TO LEFT VENTRICULAR NON COMPACTION CARDIOMYOPATHY IN A YOUNG MALE

KEY WORDS: left ventricular non compaction, cardiomyopathy, Heart failure.

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ABSTRACT

Left ventricular non-compaction (LVNC) is a rare congenital cardiomyopathy in which failure of left ventricle to compact properly resulting in prominence of endomyocardial trabeculations during cardiac embryogenesis. Here we present a rare case of isolated Left ventricular non compaction Cardiomyopathy presented with Heart failure in a young male.

INTRODUCTION

Derangements in the development of ventricular chamber due to embryonic cardiac trabeculation and its association with congenital heart disease, namely left ventricular (LV) non-compaction (LVNC), also termed as cardiac hypertrabeculation and LV Non Compaction cardiomyopathy, are characterized by abnormal protrusions of ventricular myocardium with a thin layer of properly compacted myocardium. While advancement in cardiovascular imaging have improved detection of LV trabeculations, LVNC cardiomyopathy remains a remarkably rare entity with a prevalence less than 0.02%. The anatomical organization seen in LVNC, consisting of prominent trabeculations with intertrabecular recesses that are continuous with the LV lumen overlying a thin compacted layer. This may also be present in the right ventricle (RV), either in a biventricular or isolated unilateral RV pattern.The presence of Left Ventricular trabeculations across different cardiomyopathies, both familial and sporadic, major national and international cardiology societies have accepted various classifications of the disease itself.. LVNC affects both children and adults. The incidence of LVNC in infants is 0.81 per 100,000 infants/year, in children is 0.12 cases per 100,000 children/year and a prevalence of 0.014% in adults. However, in adults presenting with heart failure, this has been rises to as much as 3%. The prevalence of isolated LVNC in adults remains unclear, although in observational studies, LVNC has been found in 0.01– 0.26% of all adults referred to an echocardiography laboratory. Alternatively, the prevalence of LVNC among general population has not been published yet, and the diagnosis is presumably often missed, because the disease is relatively unknown among physicians. The diagnosis of LVNC in adults has been reported with increasing frequency because of improved imaging modalities such as echocardiography and cardiac magnetic resonance imaging (MRI), but still little is known among physicians.

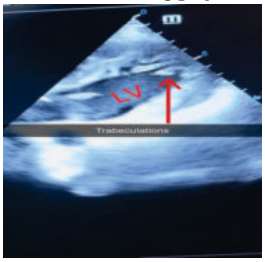

Figure 1& 2: 2D echo s/o trabeculations in left ventricle

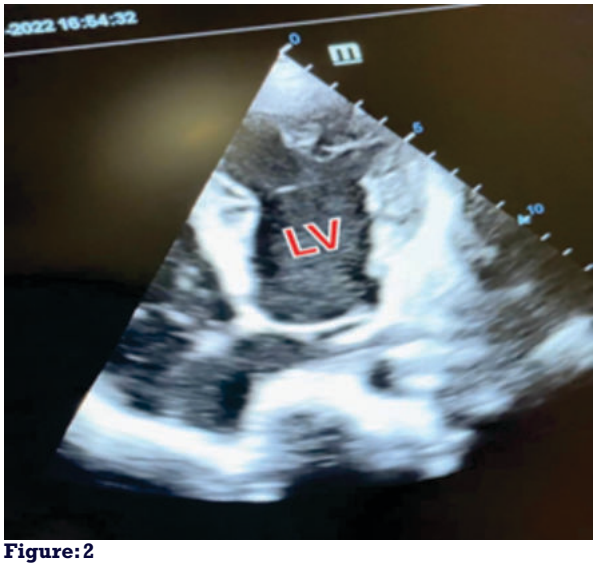

Figure: 2

Echocardiography is the most important modality for diagnosing LVNC. Characteristic LVNC echocardiographic features include (1) a thick, bilayered myocardium composed of non-compacted and compacted layers; (2) prominent trabeculations; and (3) deep endomyocardial recesses . Criteria for diagnosis have been proposed by three groups

CASE STUDY

A 31 year old hindu married male patient presented to medicine opd for chief complaints of gabharaman, palpitation, chest pain and dyspnea on exertion which was NYHA grade 2. These all symptoms started 6 hours since the presentation to opd. Chest pain was left sided, non radiating and diffuse type. No similar complaints in the past. Patients vital was within normal limits, except for tachycardia and systemic examination revealed nothing significant.

Personal History: Patient is chronic alcoholic since 10 years. Last intake was 1 day before the onset of symptoms.

Past history: History of pulmonary Tuberculosis 4 years back for which patient had completed the course of AKT for 6 months.

ECG with rhythm strip was done which was suggestive of

LBBB. Patients was given Loading dose of Aspirin, Clopidogrel and Atorvastatin. Patient was admitted in ward and basic investigation was done with Provisional diagnosis of ?Alcoholic Dilated Cardiomyopathy.Following Investigations were done for further diagnosis.

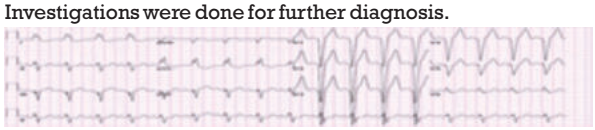

Figure 3: ECG of the patient s/o LBBB

Chest x-ray pa view: NAD
Trop I level: 2.16 (WNL)

Hb: 11.2	alt:90	
Pcv: 33.2	t.B. 1.0	sgot: 63.45
Mcv: 67	d.B. 0.3	alp: 219
Plt: 150000	albumin: 2.3	
Wbc: 4400	sodium: 140	
M.P: no parasites	potassium: 3.71	Magnesium: 2.7
CK-MB 22 (WNL)	creatinine: 0.7	

Hiv/ hep B, C: non reactive
Lipid profile: normal

Cardiac MRI

- Normal sized left ventricle showing mild global hypokinesia with resultant mildly reduced systolic function, with LVEF = 42%
- Severely reduced wall thickness with abnormal late gadolinium enhancement and akinesia involving the basal infero-septal, basal inferior, basal infero lateral and basal antero-lateral segments of left ventricle, most likely represents congenital hypoplasia of myocardium.
- Prominent non-compacted myocardium involving the mid & apical parts of left ventricle.
- Above Described features are most likely s/o Left Ventricular Non-Compaction Cardiomyopathy (LVNC) with Congenital developmental hypoplasia of the mitral subvalvular basal walls of left ventricle as described.

CONCLUSION

LVNC is being increasingly diagnosed in clinical practice, although there are still many unanswered clinical questions. The diagnosis of LVNC is an real issue. However, multimodality imaging approaches and genetic testing may improve diagnostic accuracy. It can be made with echocardiogram once the clinician has elicited a thorough family history coupled with a high degree of clinical suspicion.. Current diagnostic criteria alone may be inadequate to obtain the diagnosis. The clinical presentation of LVNC is highly variable; it may be asymptomatic or it may lead to severe HF, sudden death, and thrombotic events, depending on the case. There is no specific therapy for LVNC, and management depends on the clinical manifestations Treatment includes managing heart failure, preventing sudden cardiac death with AICD and anticoagulation.

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