Case Report

Left Ventricular Noncompaction Cardiomyopathy (LVNC) with Left Ventricular Apical Thrombus– A Case Study

Hasnat MA[1], Hossain MI[2], Kafee MA[3], Uddin MB[4]

Abstract

Left ventricular non-compaction (LVNC) is a rare congenital cardiomyopathy, with or without LV dysfunction, characterized by prominent trabeculations and associated deep recesses which communicate with the ventricular cavity rather than the coronary circulation. LVNC affects all age groups and can occur in isolation or association with other cardiac and systemic anomalies, especially with neuromuscular disorders. Patient may be asymptomatic or present with ventricular arrhythmias, thromboembolism, heart failure and sudden cardiac death. Echocardiography is the most common tool for diagnosis of LVNC. Contrast ventriculography, computed tomography (CT) and magnetic resonance imaging (MRI) are other useful diagnostic tools. Due to increasing awareness and improvement in imaging methods, LVNC is being diagnosed frequently in patients now a day.

Key-words: Trabeculations, left ventricular noncompaction cardiomyopathy, congestive heart failure.

Introduction

LVNC or spongy myocardium is a rare congenital cardiomyopathy which can be diagnosed at any age. The histology of persistent spongy myocardium with embryonic blood supply was first discussed in the literature[1] in 1975. This is a genetic defect and can run in family or occurs in sporadic form. Family screening is required to find out the asymptomatic cases[2,3]. Genetic defect can overlap with other cardiomyopathic phenotypes including hypertrophic cardiomyopathy[4,5]. Prominent trabeculations and intertrabecular recesses are the characteristic features of LVNC which communicates with the ventricular cavity rather than the coronary circulation[6,7]. During intrauterine life the compaction of myocardial fibers and meshwork occurs. This important compaction process is arrested during intrauterine life in case of LVNC patient but there are some controversies that the condition can also be acquired[7]. Patient may be asymptomatic or can present with the features of heart failure, thromboembolism, ventricular arrhythmias, and sudden cardiac death. Most of the time, they are recognized as a differential diagnosis of patients with heart failure.

Case Report

In February 2017, a 60 years old man was admitted in Kurmitola General Hospital with the features of congestive heart failure for previous one month. He had no history of chest pain, palpitations, or syncope. Physical examination revealed bilateral basal crepitations in both lungs with lower limbs edema. Other systemic and neurological findings were unremarkable. An electrocardiogram (ECG) showed features of Left bundle branch block (LBBB). Patient was anaemic with a haemoglobin level of 9.9 gm/dl. All other routine biochemical tests were found normal. In transthoracic echocardiography, the trabeculae were visible on the posterolateral wall of the left ventricle (Fig-1 and Fig-2). The blood flow was visualized into the intertrabecular recesses by color Doppler (Fig-1). At the end of systole, ratio of non-compacted myocardium to compact myocardium was >2:1. The patient also had moderate systolic dysfunction with an ejection fraction of 34% with moderate mitral and tricuspid regurgitation. Heart valves morphology were normal, no coexisting congenital anomaly was found. Small thrombus trapped in between the trabeculae was visualized in the apical region (Fig-3). These findings lead to the diag-nosis of LVNC. Contrast CT scan (Fig-4) and contrast echocardiography (fig-5) were done for further confirmation. During hospital stay, the patient had no significant arrhythmias. After medical management with loop diuretic, spirolactone, β-blockers, angiotensin-converting-enzyme inhibitor (ACEI), digoxin and warfarin, he was asymptomatic and improved clinically at the 3 months follow-up examination.

Discussion

LVNC is considered as a rare disease found in 0.05% in adult population, although it's exact prevalence is unknown[7]. It's prevalence is 3-4% in patients with HF, as expected which is higher[8,9]. Both sexes are equally affected but it's prevalence is higher among the black population compared to the white population[10]. Its prevalence is increasing with time due to the awareness, medical literature, and improving diagnostic techniques.

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In early embryonic phase, the fetal myocardium consists of a meshwork of loosely woven myocardial fibers which are separated by deep recesses. The coronary circulation develops between 5th and 8th weeks of intrauterine life, and the meshwork of loosely woven myocardium gradually compacts from epicardium to inward and the recesses are reduced in size to form capillaries[1,2]. This normal endomyocardial morphogenesis is arrested in LVNC and the other congenital heart diseases and coronary artery abnormalities may be associated with it[7]. Usually left ventricle is uniformly affected, while in less than half of the patients, right ventricular involvement is also reported. Non-compaction can be found in both sporadic and familial forms. Various genes are involved in the familial form of the disease, including mutations in G4.5 gene on Xq28 and cardiac specific CSX gene[11,12]. Screening and genetic counseling is recommended in asymptomatic first degree relatives due to such genetic linkage. Hypertrabeculation also occurs in physiological condition like pregnancy. It is also found in other conditions that lead to heart failure like- hypertension, congenital heart defects, severe anaemia, hyperthyroidism.

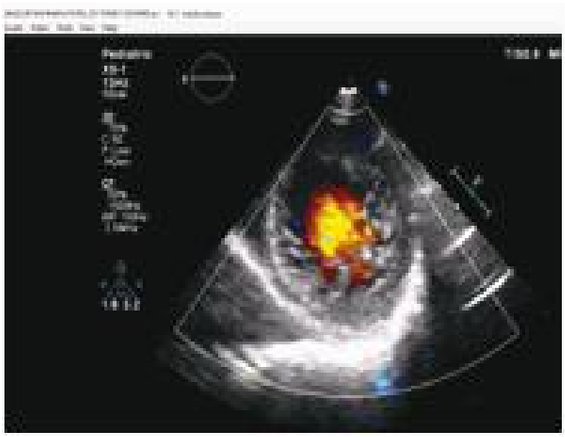
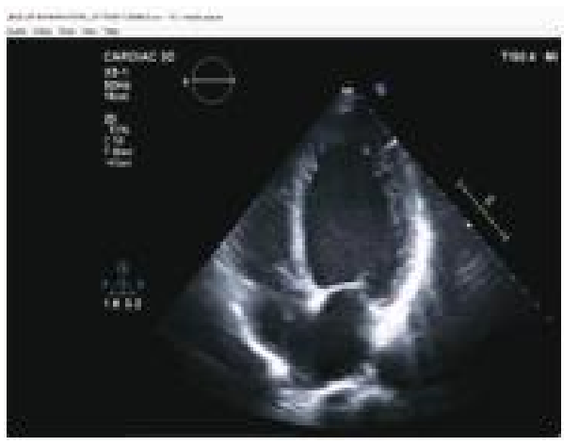


Fig: 01



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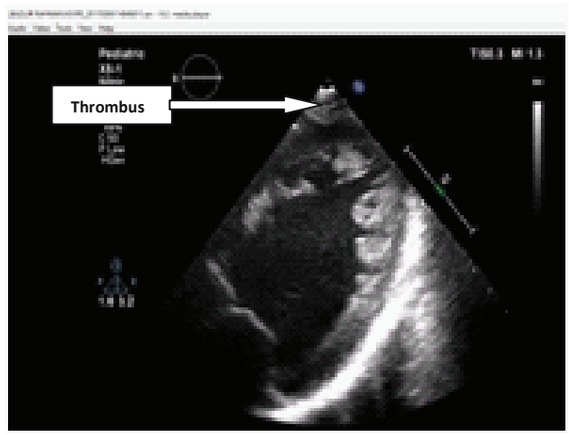

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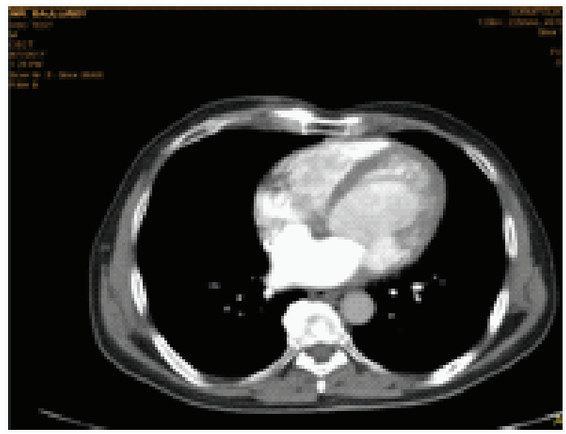

Fig: 04

Fig: 05

For the diagnosis of LVNC currently available tools are echocardiography, contrast ventriculography, magnetic resonance imaging (MRI) and computed tomography (CT). Among the several proposed criteria for the diagnosis of LVNC, the criteria proposed by Jenni et al are commonly used[13] which are as follows:

a) Absence of coexisting cardiac abnormalities.

b) Two layers segmental thickening of left ventricular myocardial wall: a thick endocardial layer with prominent trabeculations and deep recesses and a thin epicardial layer. Ratio of non-compacted myocardium to compact myocardium at the end of systole is >2:1.

c) The trabeculae are usually located on the apical/lateral, middle/bottom walls of the left ventricle. Most non-compacted segments are hypokinetic. The flow between the intertrabecular recesses can be identified by using the color Doppler method.

Treatment options for LVNC patient depends on an individual presentation. Mild case can be managed by medical management but in patient with end stage or refractory symptoms heart transplantation may be needed. To manage systolic and diastolic dysfunction, beta blockers, ACE inhibitors, diuretic, digoxin can be used[14]. So far, heart transplantation is only reported in six cases of LVNC[15]. Automated implantable cardioverter defibrillator (AICD) placement and biventricular pacemakers may help to reduce sudden cardiac death and have a role in patients with reduced ejection fraction and prolonged intraventricular conduction[14]. Long term anticoagulation is required in every case regardless of symptoms or the presence of intracardiac thrombus. Prognosis of LVNC depends on the degree and progression of heart failure, presence of thromboembolic events and arrhythmias. Whether the diagnosis is incidental or as a consequence of familial screening, follow up of asymptomatic patients is encouraged to avoid of possible future complications like- heart failure, thromboembolic events.

Conclusion

Although LVNC is a relatively rare cause of heart failure, this case suggests that it should be included in the differential diagnosis in patients who presented with first time symptoms of heart failure. In recent years, LVNC is increasingly diagnosed in clinical practice due to improvements in echo-cardiographic techniques and use of cardiac imaging. Hypertrabeculation also occurs in conditions like pregnancy and heart failure due to other causes which may be falsely diagnosed as LVNC. Therefore, a suspected case of myocardial non-compaction should be carefully evaluated by imaging methods to avoid inappropriate and exaggerated diagnoses.

References

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