CASE REPORTS

Left Ventricular Non-Compaction with Viral Myocarditis: A Rare Presentation of a Rarer Disease

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Abstract
We report an unusual case of a 22-year-old male who was incidentally diagnosed with isolated noncompaction of the left ventricle (LV) when he was admitted with viral myocarditis. Left ventricular noncompaction (LVNC) is a congenital cardiomyopathy which presents with ventricular dysfunction, cardioembolic manifestations or with arrhythmias. A diagnosis can be made with the commonly available modality of echocardiography but is still often overlooked. There is no specific treatment directed at isolated noncompaction. Treatment is focused on the cause of presentation, with medication aimed at improving ventricular dysfunction, as well as treating and preventing thrombosis and arrhythmia. This is, we feel, the first case report of isolated LVNC presenting with viral myocarditis.

Background
Congenital heart defects may be diagnosed before birth, right after birth, during childhood or not until adulthood. Adults presenting with cardiac disease are generally diagnosed with structural (valvular, septal) defects. There is subset of conditions like congenital cardiomyopathy which can also cause cardiovascular disease in the young. Here we describe a case of LVNC, which is a result of abnormal myocardial development during embryogenesis. It is associated with significant morbidity and mortality. Acute presentation due to viral myocarditis in LVNC is as yet not reported and a positive association between the two conditions has not been established. Nonetheless, this entity should be kept in mind while treating patients who present with compromised LV function in young age.

Case Report
A 22-year-old male presented with fever and malaise 10 days ago which lasted for 5 days and progressive breathlessness and mild precordial pain since 2 days. The chest pain was non-pleuritic in nature. He denied having cough or haemoptysis. He was a non-smoker and non-alcoholic. He did not report any symptoms of migratory polyarthritis, involuntary movements, subcutaneous nodules or evanescent maculopapular rash anytime in the past or present.

On initial evaluation, he was afebrile, had tachycardia and a normal blood pressure. Respiratory and cardiac system assessment did not yield any evidence of consolidation, effusion, airway disease, pulmonary oedema or any valvular pathology. There was no organomegaly on abdominal examination.

Complete haemogram was normal, as were other routine biochemical investigations. The chest radiograph revealed mild cardiomegaly but did not suggest any pleural or parenchyma pathology. His ECG showed Left Bundle Branch Block (LBBB) with 3mm
ST elevation in concordant leads. Based on the clinical features and ECG findings, coronary ischaemia was suspected for which Troponin- T was assayed qualitatively in the blood. It was weakly positive and thus confirmed our doubts about myocardial necrosis.

Patient was started on enoxaparin, anti-platelet agents and injectable frusemide. He showed marginal improvement in symptoms by the next day. His 2D-Echocardiogram was suggestive of moderately dilated LV with globally reduced systolic function (ejection fraction, EF = 20%) In addition, there was evidence of marked trabeculation and recesses of the left ventricle consistent with LVNC.

On the third day of hospitalisation, he developed severe precordial pain and worsening breathlessness. He was markedly breathless at rest and orthopnoeic. On auscultation a pericardial rub was heard and there were bibasal rales. Myopericarditis (Post myocardial infarction Dressler’s syndrome) was suspected and keeping in mind the risk of haemopericardium, anticoagulation was withdrawn. This kind of a baffling clinical picture, in which a young male in his early twenties presented with features of myocardial and pericardial involvement and an abnormal 2D-Echo finding, prompted us to search for non-ischaemic causes for his condition. With this in mind, we ordered for a Cardiac MRI.

Cardiac MRI showed evidence of myocarditis with global hypokinesia, extensive trabeculations and recesses in LV with reactive pericardial effusion. Anticoagulation was restarted and digitalis, ACE-inhibitors were added. He showed symptomatic improvement within in 6 days. Patient was investigated for the cause of myocarditis (HIV, Hepatitis) but no aetiology could be found out. Once haemodynamically stable and symptom free, he was discharged with advice to follow-up after 6 weeks for repeat cardiac imaging.

**Discussion**

**Introduction:** Noncompaction of the left ventricle (LVNC), also known as spongiform cardiomyopathy is a rare disorder, classified as a primary genetic cardiomyopathy by the American Heart Association.1 Use of non invasive diagnostic technologies has led to better delineation and visualisation of myocardium and increased diagnosis of LVNC. But, a lack of understanding of its pathophysiological origins, and consistent diagnostic criteria, has led to this condition being under-diagnosed and frequently missed.

LVNC may occur in isolation or in association with other anomalies like pulmonary atresia, anomalous origin of coronary artery and hypoplastic left ventricles. It may manifest itself at any time from infancy to adulthood but most commonly occurs in adulthood, with men and women being affected equally frequently.

**Embryology:** Trabeculations in the human embryo emerge at the end of the fourth week of gestation. Trabecular remodelling starts after completion of ventricular septation. Increase in ventricular volumes results in compression of the trabeculations with an increase in the thickness of the compacted myocardium.2

Some of the trabeculations coalesce to produce the anterior and posterior papillary muscles of the mitral valve and apical trabeculations transform into fine honeycomb-like reliefs on the inner ventricular surface. The compaction process coincides with vascularisation of the myocardium. It gradually progresses from the epicardium to the endocardium, from the base to the apex and from the septum to the free wall in the LV, and is more pronounced in the LV than in the right ventricle. Thus, the time of arrest of the normal embryonic myocardial maturation determines the severity and extent of LVNC; the ventricular apex is always involved as the compaction process concludes in the ventricular apex. In the majority of patients, it is the LV that is involved. Right ventricular involvement is difficult to be certain of, as prominent trabeculation may be a normal variant.3

**Genetics:** LVNC can be either sporadic or familial. In various reports, 12 to 50 percent of those with LVNC had a family history of LVNC.4 Autosomal dominant inheritance is the common mode of inheritance.5 Mutations of various genes like Tafazzin, Alpha-dystrobrevin (DTNA), Sarcomeric protein genes have been studied and found to be contributory for LVNC.6

**Clinical Manifestations:** The major clinical manifestations of LVNC are heart failure, atrial and ventricular arrhythmias, and thromboembolic events, including stroke. The electrocardiogram abnormalities that can be seen include left or right bundle branch
block, fascicular block, atrial fibrillation, and ventricular tachycardia.

**Diagnosis**: The diagnosis of LVNC is usually established by echocardiography. Cardiovascular magnetic resonance (CMR) imaging, computed tomography, and left ventriculography are other imaging modalities that may be diagnostic or raise the initial clinical suspicion. Commonly used criteria include the identification of excessive (more than three) and/or prominent (more than 2 mm diameter) trabeculae with inter-trabecular recesses that penetrate deeply into the myocardium, from which blood flows directly into and out of the ventricular cavity.

**Prognosis**: Published series have found that LVNC is associated with high rates of morbidity and mortality in adults. Complications may arise due to CHF, thromboembolism, or ventricular arrhythmias. The probability of survival at five years was 58 percent.

**Management**: Data on treatment of LVNC are limited, and there is no specific therapy. Medical management varies with the clinical manifestations, left ventricular ejection fraction (LVEF), the presence or absence of arrhythmias, and perceived risk of thromboembolism.

Patients with LVNC should receive implantable cardioverter defibrillator (ICD) therapy according to standard indications for ICD therapy in patients with nonischaemic cardiomyopathy. Patients with history of sustained ventricular tachycardia or sudden cardiac arrest (SCA) should receive an ICD therapy for secondary prevention of SCA. ICD implantation for primary prevention is indicated in patients with LVNC with an LVEF ≤ 35 percent and NYHA class II to III HF.

Successful cardiac transplantation in patients with end-stage heart failure with LVNC has been reported.9

**Conclusion**

This case highlights the importance of being vigilant to the diagnosis of LV noncompaction in younger individuals presenting with myocardial dysfunction. We also implore our readers to study and report any association of LVNC with myocarditis.

**References**