Left Ventricular Noncompaction Cardiomyopathy (LVNC): A Case Report

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Abstract

Left ventricular noncompaction is a rare congenital cardiomyopathy. It can occur in isolation or coexist with other cardiac anomalies. Clinical presentation varies from patients to patients. LVNC is commonly diagnosed by echocardiography. We descried the case of a man with heart failure for the first time at 55 years of age. Transthoracic echocardiography shows trabeculation and intertrabecular recesses communicate with the ventricle cavity. The patient was relatively well after medical management. This case is reported for clinical awareness and share experience.

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Introduction

LVNC is a rare congenital cardiomyopathy. It is a disease of endomyocardial trabeculation, characterized by prominent trabeculations and intertrabecular recesses that communicate with the ventricular cavity.[1,2] The American Heart Association classified it as a primary genetic cardiomyopathy 18% to 42% of left ventricular noncompaction cases are familial first-degree relatives of affected individuals, to identify potential risks and the possibly causative genes clinical presentations varied ranging from patients to patients and develop ventricular arrhythmias, thromboembolism, heart failure, and sudden cardiac death.[3-6] Here, we described the case of a man who presented with heart failure for the first time at 55 years of age. We discussed the diagnosis of this case.

Case Report

In October 2020, a 55-year-old man was admitted to our hospital because of congestive heart failure during the previous twelve days. He had no chest pain, palpitations, or syncope. Physical examination revealed bilateral crackles in the lower-lung fields and edema of lower extremity. Neurologic findings were unremarkable. The results of routine biochemical tests were within normal limits. An electrocardiogram showed ST-T changes and (LBBB) (Figure 1). Chest x-ray showed Cardiomegaly (Figure 2). The trabeculae were located on the posterolateral wall of the left ventricle (Figure 3). The blood flow into the intertrabecular recesses could be visualized by color Doppler (Figure-4). The ratio of non-compacted myocardium to compact myocardium at the end of the systole was > 2:1 (Figure 5). The patient also had mild systolic dysfunction and an ejection fraction of 40% (Figure 6). Moderate mitral and tricuspid regurgitation and Mild pulmonary hypertension were noted (peak systolic pulmonary artery pressure, 40 mmHg). Morphologically, the heart valves were normal, and no coexisting congenital anomaly was found.

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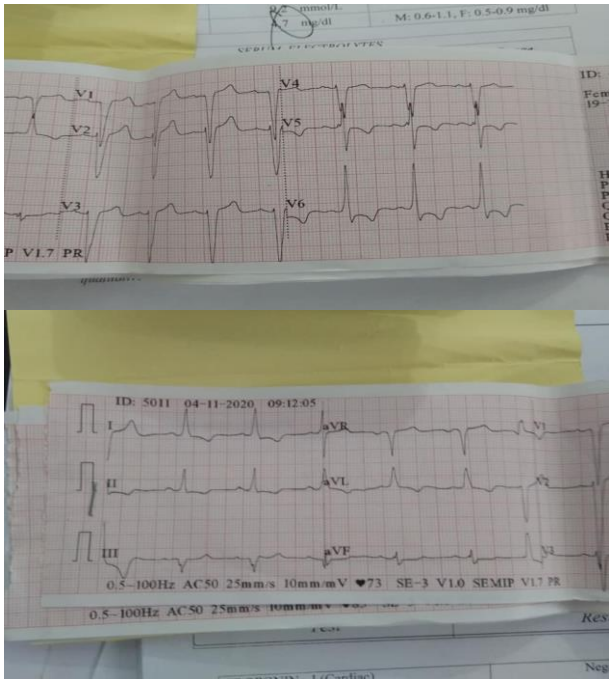
These findings led to the diagnosis of LVNC. The patient had no significant arrhythmias during his hospital stay. After medical management with β-blockers, ACEI, a loop diuretic, spironolactone and digoxin, he was asymptomatic at the 1-month follow up examination.

ECG (Figure 1): LBBB with non-specific ischemic changes



CX- Film 2 PA (Figure 2): Cardiomegaly

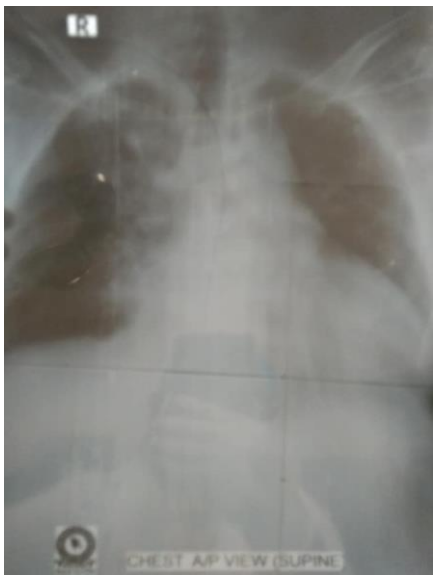


Figure 3: Left ventricular with more than 3 trabeculations protruding from the posterolateral wall visualized by two-dimensional echocardiography.

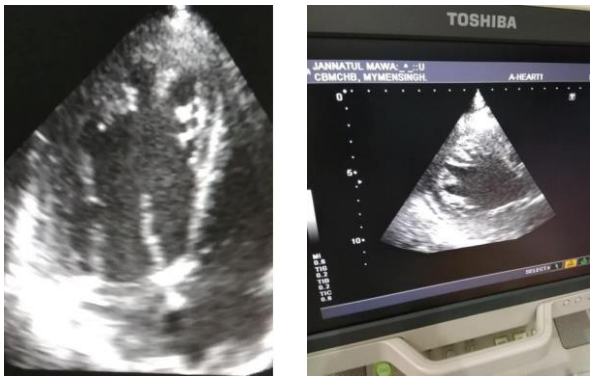


Figure 4: Left ventricular trabeculations and blood flow into the intertrabecular recesses visualized by Color Doppler flow imaging.

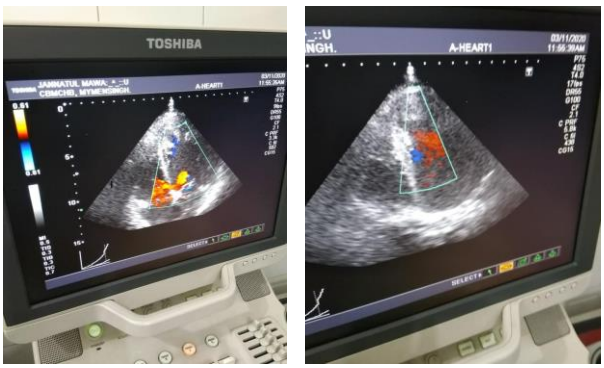


Figure 5: The ratio between noncompacted and compacted walls was more than 2 in the diastole.

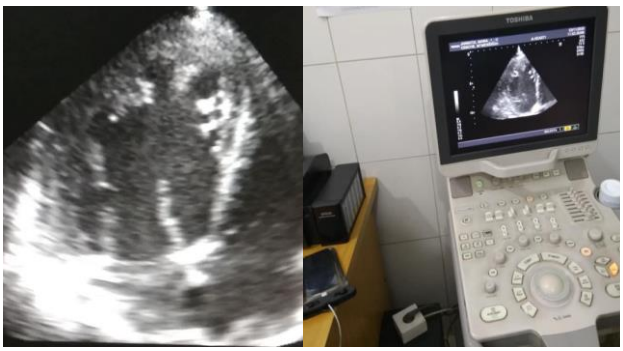
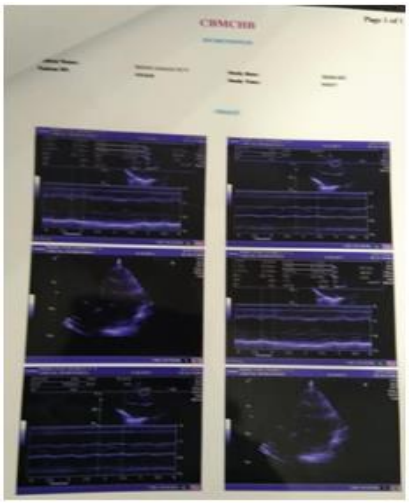


Figure 6: The result of the left ventricular ejection fraction measured indicated that the systolic function was impaired.



Discussion

LVNC is a structural abnormality of the left ventricular (LV) myocardium of uncertain etiology and mechanism but is generally considered genetic in nature. In some patients, neuromuscular disorders and chromosomal defects are noted.[7,8] LVNC cardiomyopathy (OMIM No. 604169) was first described at the time of autopsy because current imaging modalities were not available. Feldt *et al*.[9] The first report of an echocardiographically diagnosed case of LVNC was by Embedding and Bender in 1984.[10] In 1990, the term noncompaction was first introduced by Chin *et al.*[1] In 1997, cardiac magnetic resonance imaging (CMR) was initially used to identify LVNC by Hany *et al.*[12] The etiology of LVNC is uncertain, but it is speculated that it results from a disturbed compaction process during early development of the LV myocardium.[13] It is believed that LVNC result from a failure of the final phase of cardiac development, the myocardial compaction process. Failure of the compaction process results in deep intertrabecular recesses between the abnormal trabeculations. Mutations in the murine genes Casz1,[14] LVNC have been considered to be rare. However, Andrews *et al* [11] reported that 9% of 104 children with primary cardiomyopathy had LVNC. In adults referred for echocardiography, the prevalence of LVNC reportedly ranged from 0.01% to 0.3%.[15-17] We have described the diagnosis of LVNC in a 55-year-old man. The patient had been asymptomatic until mild symptoms of congestive heart failure developed during twelve days before initial presentation. He responded well to medical management of this relatively rare cause of heart failure. Tools for the diagnosis of LVNC are echocardiography, contrast ventriculography, computed tomography (CT) and magnetic resonance imaging (MRI).[18] Widely accepted diagnostic criteria by Jenni *et al.* were as follows:[19,20]

(1) Absence of coexisting cardiac abnormalities.
(2) Segmental thickening of myocardial wall of left ventricle with two layers: a thin pericardial layer and a thick endocardial layer with prominent trabeculations and deep recesses. The ratio of non-compacted myocardium to compact myocardium at the end of systole is > 2:1.
(3) The trabeculae are usually located on the apical/lateral, middle/bottom walls of the left ventricle. Most non-compacted segments are hypokinetic.
(4) The flow between the intertrabecular recesses can be identified by using the color Doppler method.

In patients with LVNC, treatment options vary on an individual basis, ranging from medical management in mild cases to heart transplantation in patients with refractory symptoms besides treatment for congestive heart failure and antiarrhythmic therapy, if

indicated, anticoagulation therapy is recommended because of the risk of systemic embolism.[2,4] Monitoring asymptomatic patients is encouraged, whether diagnosis is incidental or the consequence of familial screening, because of possible future complication in those individuals.[3,14] Interestingly, LVNC can be identified with overlapping features of other cardiomyopathy, as well as with congenital heart disease.[2,4,5] The diagnosis may be made at any age and in either sex.[17] The prognosis of patients with LVNC is determined by the degree and progression of heart failure, presence of thromboembolic events and arrhythmias.[9] Our patient's mild symptoms responded well to medical therapy.

Conclusion

Diagnosis of spongy myocardium is challenging, echocardiography is routinely used initially other imaging modalities which are being increasingly used these days for diagnosis are computed tomography or cardiac magnetic resonance imaging. We suggested that the diagnosis of suspected myocardial noncompaction should be carefully evaluated by imaging methods to avoid inappropriate and exaggerated diagnoses.

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