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CASE REPORT

Left Ventricular Non-compaction Cardiomyopathy: A Case Report and Literature Review

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Abstract

Left Ventricular Non-Compaction Cardiomyopathy (LVNC) is a rare myocardial disorder characterized by abnormal myocardial tissue formation in which the left ventricular wall appears to be trabecular with prominent intertrabecular recesses. The diagnosis of LVNC is predominantly reliant on cardiac imaging, namely thoracic echocardiography, however, cardiac MRI is indicated in conditions in which echocardiography is inconclusive. Diagnostic criteria for both echocardiography and cardiac MRI differ, however, the general principle of diagnosis is a comparison of the thickness of non-compacted to compacted myocardial tissue. The management of LVNC is nearly identical to that of Heart Failure with reduced Ejection Fraction (HFrEF), however, anticoagulation is an additional measure of management to the thrombogenic nature of non-compacted myocardial tissue. Here, we discuss a case of LVNC and the current data on its management.

Keywords: Cardiomyopathy, Myocardial trabeculations, Intertrabecular recesses, Hearth failure

1. Introduction

Left ventricular noncompaction cardiomyopathy, formerly known as spongy myocardium syndrome, is a rare disorder characterized by myocardial trabeculations and intertrabecular recesses.[1]

LVNC has long been regarded to be a congenital disorder, however, there has been some discussion on whether such cases can be acquired. The etiology of congenitally acquired LVNC is unclear, however current literature describes the pathogenesis to arise from the intrauterine arrest of a "compactive" process that occurs during fetal development.[1,2] There also has been some data suggesting a genetic basis for acquiring LVNC which may involve abnormalities of various sarcomere proteins involved in the myocardial architecture. Due to its morphology, individuals with LVNC are at risk of developing multiple complications including heart failure, thromboembolisms, ventricular arrhythmias, and Sudden Cardiac Death (SCD). Due to its trabecular architecture, turbulent blood flow in the regions of the myocardium promotes thrombi formation.[2,3] In addition, it is believed that heart failure predominantly occurs to the inefficient contraction of the left ventricular wall. Finally, ventricular arrhythmias and SCD may arise to the abnormal electrical circuitry within the non-compacted ventricular wall.[3]

Here, we present the case of a male who presented with his first signs and symptoms of heart failure at the age of 42 years old. We further discuss our management of his case and discuss some of the current data on LVNC.

2. Case presentation

Our patient is a 42 -year-old male with no reported past medical history who presented to the ED with the complaint of worsening shortness of breath for a few weeks prior to presentation. In addition to

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dyspnea, he also complained of substernal, moderate in intensity chest pain, that was dull in quality, worsened with exertion, and relieved with rest. He denied any cough, anosmia, changes in taste, fever, nausea, vomiting, radiation of the pain, diaphoresis, palpitations, or dizziness. He reported exposure to COVID-19 via a cousin whom he lived with. The patient was unvaccinated. He endorsed occasional alcohol intake; marijuana use and 10 pack-year smoking history. He had quit 4 years prior to the presentation. He denied any family history of coronary artery disease or similar symptoms as his.

In the ED, he was saturating 100% on room air and the rest of the vitals were normal. On physical examination, the patient was in mild distress due to chest pain and dyspnea, no JVD was appreciated, and regular rate and rhythm with no murmurs, rubs, or gallops on cardiac auscultation. Bilateral clear lung fields on auscultation without wheezing, crackles, or rhonchi, no cyanosis or peripheral edema. His EKG showed V1, V5–V6 T wave inversions. (These changes were new compared to a prior EKG in 2016). Initial troponin was 0.11 and he tested positive for COVID-19.

The patient was then admitted and started on aspirin 81 mg daily, Lipitor 40 mg at bedtime, and ibuprofen as needed for pain. Since his saturations were normal and he had no COVID-19 symptoms it was deemed unnecessary to treat COVID-19. Further evaluation showed that his troponin trended down to 0.09. CT angiogram was negative for pulmonary embolism. His A1c was 5.3 and his lipid panel was unremarkable. 2D-Echocardiogram showed left ventricular (LV) ejection fraction <20%, increased left ventricular chamber size, trabeculated myocardium, especially in the lateral and inferior wall with non-compacted to compacted myocardium ratio more than 2:1 making the diagnosis of left ventricular non-compaction (Video 1 [https://scholarlycommons.gbmc.org/cgi/editor.cgi?article=1168&window=additional_files&context=jchimp]). It also showed moderate mitral and tricuspid valve regurgitation and moderately reduced right ventricular function.

The patient was then started on lisinopril 5 mg daily, metoprolol succinate 25 mg daily, and furosemide 20 mg daily. Metoprolol succinate dose was later increased to 50 mg daily. The patient was also started on Eliquis 5 mg bid to prevent thromboembolism in the setting of LV non-compaction and given the patient's COVID status. A Life Vest was arranged for the patient in the meantime. His symptoms significantly improved after 3 days. He was then discharged to follow up on an outpatient basis for cardiac MRI and genetic evaluation.

3. Discussion

Left ventricular noncompaction (LVNC) is a rare congenital disorder prevalent in less than 0.02% of the population.[1] LVNC has an autosomal dominant and/or sporadic mode of inheritance with a mutation in sarcomere genes – differentiated via family screenings. These genes are similar to the ones in hypertrophic and dilated cardiomyopathy.[2] During embryogenesis, the myocardium gets compacted, forming smooth and firm muscle instead of trabeculated. That trabecular compaction usually begins at the base of the heart and developments toward the apex. In LVNC, this process is interrupted, forming thick and spongy muscles. Thus, the epicardial layer is compacted and the endocardial layer shows intertrabecular recesses.[3] There is also evidence of hypoperfusion in LVNC due to the discrepancy between the number of capillaries and myocardial mass.[3] This hypoperfusion leads to reduced ejection fraction and systolic disturbance. The increased amount of trabeculations further leads to diastolic disturbance due to a restrictive filling pattern.[4]

The clinical presentation of these patients varies from asymptomatic to symptoms of heart failure, arrhythmias, thromboembolic events, and sudden cardiac death and predominantly affects children and middle-aged adults up to the age of 50s. 84% of these patients report the typical symptoms of heart failure such as shortness of breath, lightheadedness, unexplained weight gain, swelling, and syncope while 38% report thromboembolic complications.[3]

LVNC can be diagnosed via computed tomography, MRI, contrast ventriculography, and most accurately, echocardiography. Although this cardiomyopathy is "unclassified" via the World Health Organization, there are two criterion types, Jenni and Chin, that are used today. The most accepted is the Jenni criterion which encompasses a clear-cut echocardiographic standard to diagnose LVNC. The criteria are as follows: 1) Absent coexisting cardiac abnormalities. 2) A two-layer structure showing a compacted thin epicardial layer and a thicker non-compacted endocardial layer of trabecular meshwork with deep endomyocardial spaces. An end-systolic ratio of non-compacted to compacted layers of >2.3) Pathology localized to mid-lateral, apical, and mid-inferior areas. 4) Deep perfused intertrabecular recesses seen on color doppler.[5] In inconclusive cases, other techniques such as contrast enhancement, speckle tracking, and tissue doppler imagining can be used; however, echocardiography is the gold standard.[6]

There is no specific therapy for patients with LVNC other than the management of complications such as heart failure. Asymptomatic patients must

be monitored as symptoms may appear later.[7] If indicated, all patients should be put on antiarrhythmic and congestive heart failure therapy.[8] If the patient has an ejection fraction <40%, a history of thromboembolism, or atrial fibrillation, anticoagulation is warranted.[9] The prevention of systemic embolism is the main goal of therapy. If oral anticoagulation is contraindicated, risk assessment based on CHADS2/CHADS2-Vasc scores would be necessary.[8] Patients with sustained ventricular fibrillation may need a cardioverter-defibrillator implantation as they are at a higher risk for sudden cardiac death.[10] The progress in patients with LVNC is determined by the improvement in their symptoms of heart failure.[11]

Conflict of interest

There is no conflict of interest.

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