Left Ventricular Non-Compaction in an Adult with Patent Ductus Arteriosus

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Abstract

Introduction: Left ventricular non-compaction (LVNC) is a rare form of cardiomyopathy that may occur in isolation or with an associated cardiac anomaly. It presents with a wide array of manifestations, prompting early recognition to be imperative to prevent progression of symptoms.

Case presentation: We report a case of a 46-year-old male complaining of palpitations for 10 years who survived sudden cardiac arrest on the same year as symptom onset. Consult was advised but was not done until he had heart failure symptoms. Carvedilol, furosemide and digoxin were given. Initially, some improvement was noted but he later developed dyspnea on exertion prompting consult at our institution. Pertinent physical examination findings include a dynamic precordium, apex beat at sixth left intercostal space-anterior axillary line (LICS AAL), right ventricular heave, distinct heart sounds, normal rate, irregularly irregular rhythm, a grade 4/6 continuous murmur heard best at the left upper sternal border, suggestive of patent ductus arteriosus (PDA), and a grade 3/6 holosystolic murmur at the apex radiating to the axilla, suggestive of mitral regurgitation. Transthoracic echocardiography confirmed presence of a PDA (0.8cm) with left to right shunt and Qp/Qs of 2.7:1. Incidental finding of LVNC was noted characterized by prominent ventricular trabeculations and deep intertrabecular recesses. Optimal medical treatment for heart failure was given with symptomatic relief. Surgical closure of the PDA was contemplated after hemodynamic studies can confirm the absence of irreversible pulmonary hypertension.

Discussion: Patients with LVNC may be asymptomatic or may present with heart failure, sudden cardiac death or arrhythmias. The diagnosis of LVNC poses a diagnostic challenge. Echocardiography is a cost-effective diagnostic tool that will allow early diagnosis. Cardiac magnetic resonance (CMR) imaging is an alternative diagnostic modality. Once the diagnosis has been confirmed, prompt initiation of guideline-directed medical treatment for heart failure may prevent progression of disease.

Conclusion: Left ventricular non-compaction may occur in isolation or in association with other congenital heart diseases such as patent ductus arteriosus. Closure of a PDA is indicated in the presence of a significant shunt and with confirmation of acute reversibility in the presence of pulmonary hypertension to prevent the possibility of decompensation in a patient with heart failure.

Keywords: left ventricular non-compaction, patent ductus arteriosus, cardiomyopathy, sudden cardiac death

Introduction

Noncompaction of ventricular myocardium is a recently recognized congenital cardiomyopathy characterized by a distinctive ("spongy") morphological appearance of the left-ventricular (LV) myocardium. It has an estimated incidence of 0.05%. This involves predominantly the distal (apical) portion of the LV chamber with prominent ventricular trabeculations and deep intertrabecular recesses due to arrest in the normal embryogenesis. LV noncompaction (LVNC) may be an isolated finding or may be associated with other congenital heart anomalies. The natural history of this condition is largely unresolved. One may be asymptomatic or present with LV systolic dysfunction and heart failure, thromboembolism, arrhythmias, and sudden death. Diagnosis can be made with two-dimensional echocardiography, cardiac magnetic resonance (CMR) imaging, or LV angiography.[1-4]

To date, this is the first reported case of LVNC in our institution and only a few cases have been described worldwide, especially among Asians. The combination with patent ductus arteriosus (PDA), makes it rarer. This report aims to increase awareness of this type of cardiomyopathy which may be missed on echocardiography without a heightened index of suspicion.

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Case Presentation

We report a case of a 46-year-old male who has been complaining of palpitations for 10 years. He had history of cardiac arrest on the same year as the onset of symptoms. Cardiopulmonary resuscitation was done for five minutes followed by return of spontaneous circulation. Unrecalled medications were given. Patient continued to have on and off palpitations but condition was tolerated. Cardiology consult was advised but did not comply until five years ago when he had palpitations associated with shortness of breath, dyspnea on exertion and bipedal edema. Consult was done and he was given carvedilol, furosemide and digoxin with some improvement. Ten months prior to admission, patient sought consult at our institution due to palpitations and dyspnea even on mild exertion.

Pertinent physical examination findings at time of consult include dynamic precordium, apex beat at the sixth left intercostal space-anterior axillary line (LICS AAL), right ventricular (RV) heave, distinct heart sounds, normal rate, irregularly irregular rhythm, a grade 4/6 continuous murmur heard best at the first and second left intercostal space (LICS) parasternal border, suggestive of patent ductus arteriosus, with and a grade 3/6 holosystolic murmur at the apex radiating to the axilla, suggestive of severe mitral regurgitation (MR). Chest x-ray showed an enlarged heart with RV prominence, prominent main pulmonary artery and increased pulmonary vascularity. ECG showed atrial fibrillation with controlled ventricular response, normal QRS axis, left ventricular hypertrophy by Sokolow-Lyon criteria. 2D echocardiography confirmed the presence of a congenital heart disease, patent ductus arteriosus (0.8cm) with left to right shunt and Qp/Qs of 2.7:1 by LVOT/RVOT as seen in Figure 1. Incidental finding of dilated left ventricular dimension (LVEDD 7.2, LVESD 5.6) with non-compaction, generalized hypokinesia and depressed global systolic function with increased filling pressure were noted (Figure 2). Non-compaction was further investigated by 3D echocardiography (Figure 3). It was confirmed that the patient indeed had LVNC and met all the diagnostic criteria for LVNC as seen in Figures 4 to 6. The rest of the chambers were dilated (LA, RA, RV). Other findings include a dilated MV annulus with severe MR, dilated tricuspid valve annulus with severe tricuspid regurgitation and severe pulmonary hypertension. Cardiac MRI was not done since patient met the echocardiographic criteria for the diagnosis of LVNC.

He was managed as a case of heart failure, given losartan, metoprolol, aldactone, rosuvastatin and aspirin. His CHADS2-VASc score was one, warranting oral anticoagulation, but financial constraints and inability to follow-up on a regular basis lead to the decision to give an anti-platelet instead. Surgical closure of the patent ductus arteriosus was contemplated after hemodynamic studies can confirm the absence of irreversible pulmonary hypertension. As of this writing, patient's functional status has improved.

Discussion

Left ventricular noncompaction is a rare form of cardiomyopathy that may be an isolated finding or may be associated with other congenital heart anomalies[1] such as the case presented. It affects eight to 12 individuals for every one million individuals per year.[6] Another study reported a prevalence of 0.24% (n=26) among 10,857 patients who underwent echocardiography.[7]

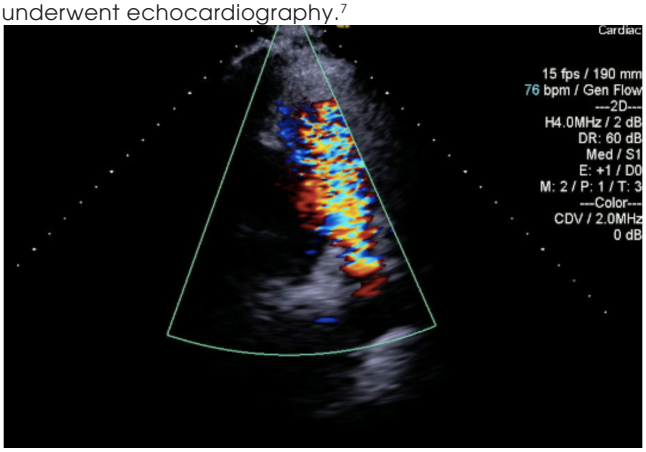

Figure 1. Parasternal short axis view at the level of the great vessels demonstrating continuous color flow from the proximal descending aorta to the main pulmonary artery. Systolic gradient = 101mmHg. QpQs = 2.7:1.

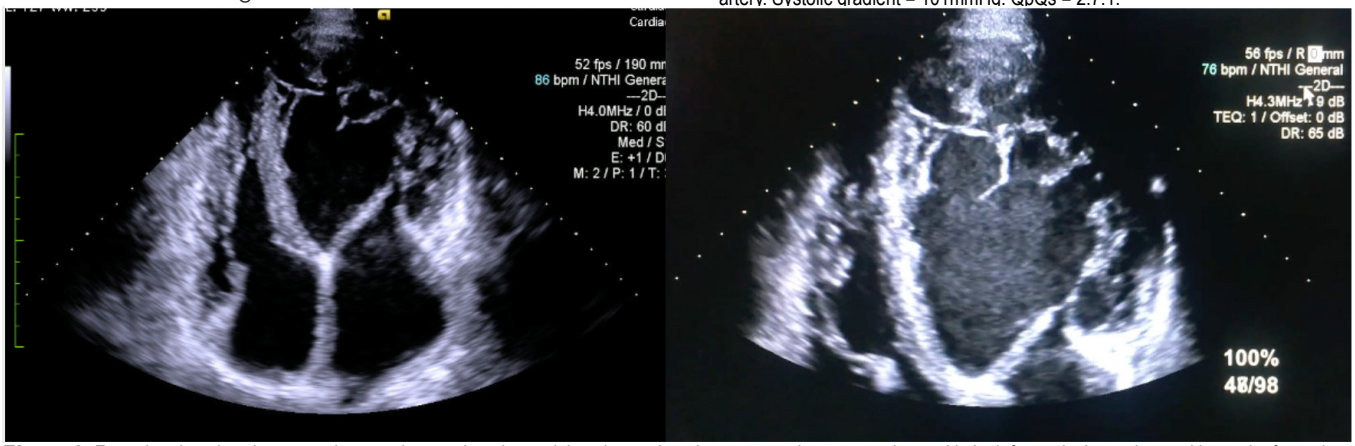

Figure 2. Four-chamber view demonstrating prominent trabeculae and deep intertrabecular recesses that communicate with the left ventricular cavity resulting to the formation of a compacted epicardial layer with a loose, interwoven "spongy" meshwork.

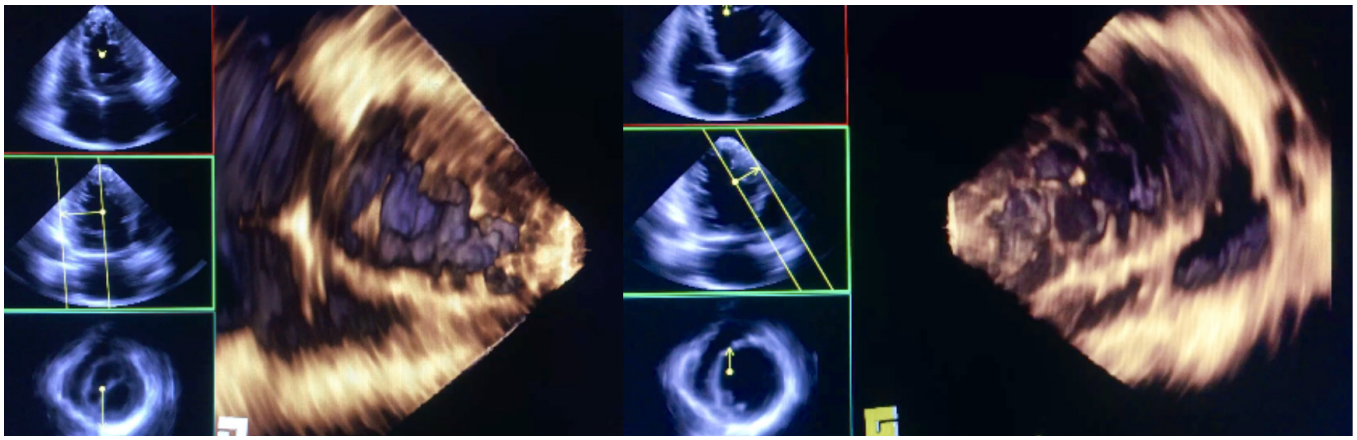

Figure 3. Two-chamber view in 3D echocardiography showing prominent trabeculae and deep intertrabecular recesses.

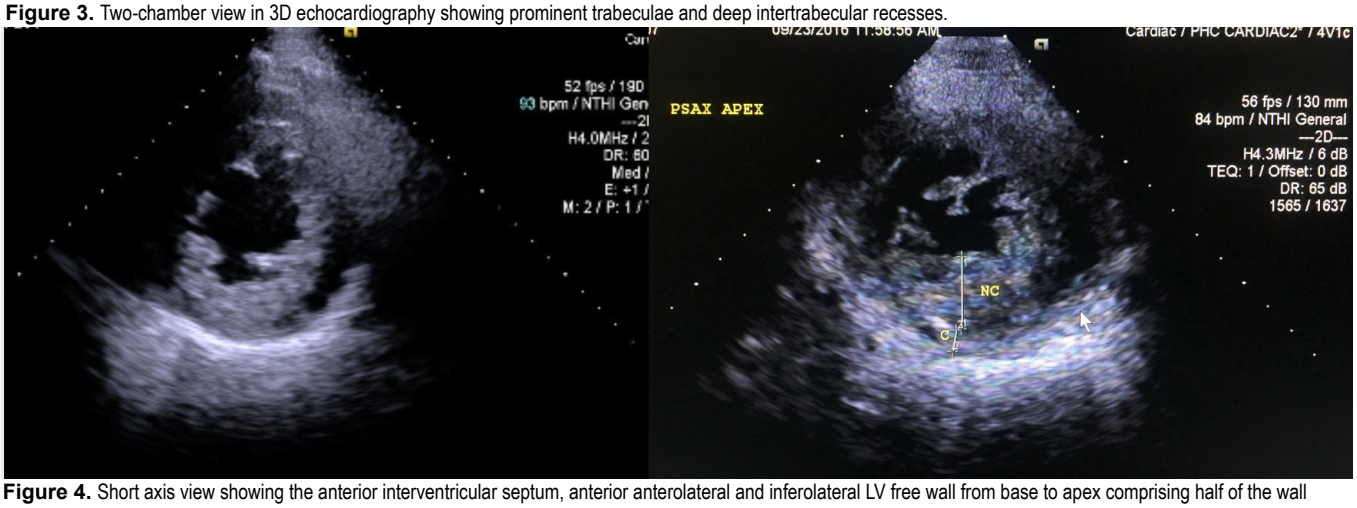

Figure 4. Short axis view showing the anterior interventricular septum, anterior anterolateral and inferolateral LV free wall from base to apex comprising half of the wall thickness with non-compacted to compacted ratio of 3.4 at mid SAX view during end-systole. (JENNI Criteria)

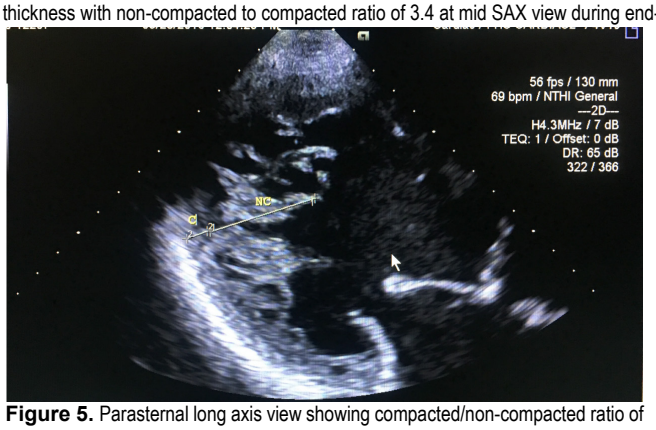

Figure 5. Parasternal long axis view showing compacted/non-compacted ratio of 0.20 on apical SAX view during end-diastole (CHIN Criteria)

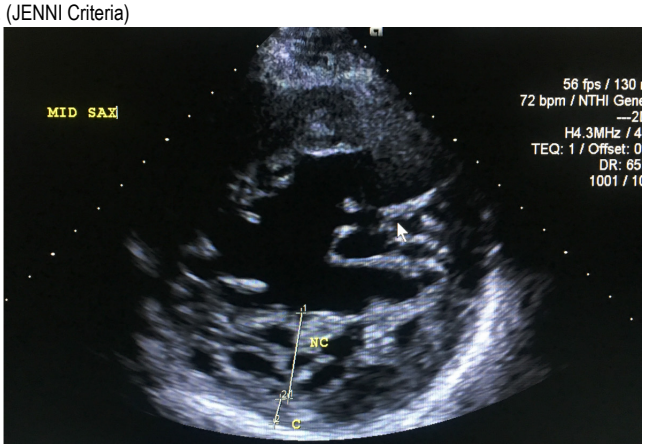

Figure 6. Short axis view showing compacted/non-compacted ratio of 3.4 (PATERICK Criteria)

Patients with LVNC present with a wide array of clinical manifestations such as: sudden cardiac arrest which the patient survived and heart failure as presented. LVNC in this case was discovered incidentally on transthoracic echocardiography which met the LVNC criteria by Paterick[3], Chin[4,] and Jenni[5] as shown in Figures 4-6.

Jenni[5] published the most widely used criteria which deserves much attention:

1. An excessively thick myocardial wall structure in two differing layers-a thin, compacted epicardial layer and a thicker, noncompacted endocardial layer
2. A characteristic end-systolic ratio of >2:1 for noncompacted-to-compacted wall thickness
3. Prominent multiple, chiefly intracavitary trabeculae, with color-Doppler echocardiographic evidence of communication between the deep intertrabecular recesses and the ventricular cavity.

Cardiac magnetic resonance (CMR) imaging was no longer done since there was good echocardiographic window and superior image quality. CMR is advised

to validate the diagnosis most especially if there are technical limitations or poor echocardiographic window on transthoracic echocardiography. To compound the case, our patient had a large PDA which has the potential to worsen any case of heart failure. Cardiac catheterization was advised, mainly to confirm severity of pulmonary hypertension that will determine eligibility for surgery. This will also determine if the patient's PDA has a role to the patient's heart failure. In the absence of irreversible severe pulmonary hypertension and predominant left-to-right shunting of blood, closure of the PDAis indicated. Ventricular arrhythmias are more common than atrial fibrillation as in the patient's case. It is possible that he may also be having ventricular arrhythmias which can be confirmed with holter monitoring.

There is a paucity of evidence in the management of LVNC. As of this writing, there is no treatment for LVNC other than to follow international guidelines on heart failure. Primary prevention for thromboembolic events with oral anticoagulation is imperative. The long-term follow-up of adults diagnosed with LVNC by echocadiography was studied by Oechslin et al. After a mean follow-up of 44 months, 12 out of 34 adult patients expired. Sudden cardiac death and heart failure were the most common causes of death.[8]

Conclusion

The diagnosis of LVNC poses a diagnostic challenge. Echocardiography is essential in the diagnosis of LVNC. Cardiac MRI can be done if findings on echocardiography are equivocal. Earlier diagnosis can prompt the attending physician to start management of heart failure to prevent or retard its progression. Closure of a patent ductus arteriosus is indicated if significant and if a patient is still operable to prevent the possibility of decompensation due to heart failure.

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