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Non-Compaction Cardiomyopathy Presented with Atrial Fibrillation: A Case Report and Literature Review

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Abstract

Background—Left ventricular non-compaction cardiomyopathy (LVNC) is a rare congenital cardiomyopathy characterized by increased trabeculation in one or more segments of the ventricle. LVNC presented with non-specific symptoms and highly variable clinical presentation ranging from asymptomatic to progressive heart failure and recurrent or life-threatening arrhythmias.

Case presentation—54-year-old Black man with a history of hypertension, diabetes and end-stage renal disease presented with one day palpitations and lightheadedness following a dialysis session. He denied any dyspnea or syncope. On examination, blood pressure was 175/91 mmHg with irregular pulse. No murmur, rubs or gallops were appreciated. Laboratory were unremarkable except increased creatinine and mild anemia with normal thyroid function test. Electrocardiogram (ECG) revealed atrial fibrillation with normal ventricular rate. Transthoracic echocardiogram revealed mildly increased left ventricular (LV) wall thickness with prominent trabeculation and ejection fraction of 55–60 percent, a pseudo-normal LV filling pattern, with concomitant abnormal relaxation and increased filling pressure, suggestive of LVNC. The patient was switched to apixaban. Genetic testing was recommended for family members.

Conclusions—LVNC is rare congenital cardiomyopathy with non-specific symptoms and should be considered among the possible diagnosis in patients presenting with arrythmia patients. Echocardiographic and cardiac magnetic resonance imaging can be utilized to establish diagnosis.

Keywords

Cardiac arrythmia; Non-compaction Cardiomyopathy; Trabeculation; spongiform

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1. Introduction

LVNC, a rare congenital cardiomyopathy, is characterized by prominent trabeculae, deep intertrabecular recesses, and thickened myocardium with two distinct layers (compacted and noncompacted) [1]. LVNC is encountered in 0.045% to 0.26% of adult patients referred for echocardiographic examinations and 0.01% to 1.3% in the general population with a male predominance [2]. The rate of familial involvement appears to vary from 18 to 33% [3]. Mutations in several genes with different inheritance patterns has been found related to LVNC, with MYH7 and MYBPC 3 genes estimated to cause up to 30 percent of cases. Most of the genes are involved in sarcomeric or cytoskeletal protein productions. The following vignette highlights a case of LVNC initially presented with atrial fibrillation.

2. Case Report

A 54-year-old Black Male with a history of hypertension, diabetes and end-stage renal disease presented with one day history of palpitations and lightheadedness following a dialysis session. Patient has never had palpations in the past, denied chest pain, lightheadedness, shortness of breath, syncope. Family history was negative for arrythmias or sudden cardiac death. No significant social history was appreciated. On physical exam, BP was 175/91 mmHg with irregular heart rate 65–72 bpm. Cardiovascular exam showed irregular pulse with no murmur, rubs or gallops appreciated. Laboratory was unremarkable except creatinine 9.2 mg/dL(normal range 0.7–1.3 mg/dL), hemoglobin 11g/dL with normal thyroid function test. [Table 1] Electrocardiogram showed atrial fibrilization with a ventricle rate of 76 bpm [Figure 1]. Heparin drip was initiated initially. Later, 2D-transthoracic echocardiogram revealed mildly increased left ventricular (LV) wall thickness with prominent trabeculation and ejection fraction of 55–60 percent, a pseudo-normal LV filling pattern, with concomitant abnormal relaxation and increased filling pressure [Figure 2]. The spongiform appearance of LV was suggestive for non-compaction cardiomyopathy and considered as the possible etiology for arrythmia. The patient was switched to apixaban for outpatient treatment. Genetic testing and echocardiogram was recommended for first-degree relatives to rule out a familial variant. Further follow-ups are scheduled.

3. Discussion

LVNC describes a LV wall anatomy characterized by the presence of three key morphologic elements- prominent trabeculae, a thin compacted layer, and intertrabecular recesses that are continuous with the LV cavity and separated from the epicardial coronary arteries [4]. Noncompaction can affect both ventricles or the right ventricle only. LVNC can be regarded as an isolated entity or as some of the traits that may recur in cardiac and noncardiac disease [5].

The clinical presentations are highly variable, ranging from asymptomatic to progressive heart failure and recurrent or life-threatening arrhythmias. Ventricular and supraventricular arrhythmias are prominent clinical components of LVNC, including atrial fibrillation (adult), atrioventricular accessory pathways/ Wolff-Parkinson-White syndrome(children) and ventricular tachycardia [6].

No ECG findings were thought to be specific for LVNC. Diagnosis is usually established by identifying morphologic diagnostic criteria on echocardiography with Jenni criteria [7,8,9,10] being the most commonly used criteria with the aid of cardiovascular magnetic resonance imaging [11,12,13] measuring ratios between noncompacted (NC) and compacted(C) layers of the LV wall(NC/ C ratio usually ≥ 2) when echocardiography findings are inconclusive [10]. [Table 2]

A serial clinical evaluation and symptom monitor were recommended for LVNC patients. For patient develop heart failure symptoms or asymptomatic LV systolic dysfunction, treatment should adhere the standard treatment. However, some managements are unique to LVNC patients, including anticoagulation due to increased risk of thromboembolism and primary prevention of sudden cardiac death (SCA). Patient with atrial fibrillation should be on anticoagulation if they meet standard criteria for anticoagulation. The risk of sudden death are associated with LV size, systolic function and presence of arrhythmias [6]. Implantable cardioverter-defibrillator was warranted for LVNC patients with ejection fraction less than 35 percent and New York Heart Association class II to III heart failure as primary prevention and patients who survive an episode of sustained ventricular tachycardia or SCA as secondary prevention [14].

LVNC is associated with high rates of morbidity and mortality with adverse prognostic factors being low ejection fraction, severe disease at presentation and other comorbidities. Genetic evaluation, including both counseling and genetic testing, is recommended for patients with LVNC and first-degree relatives.

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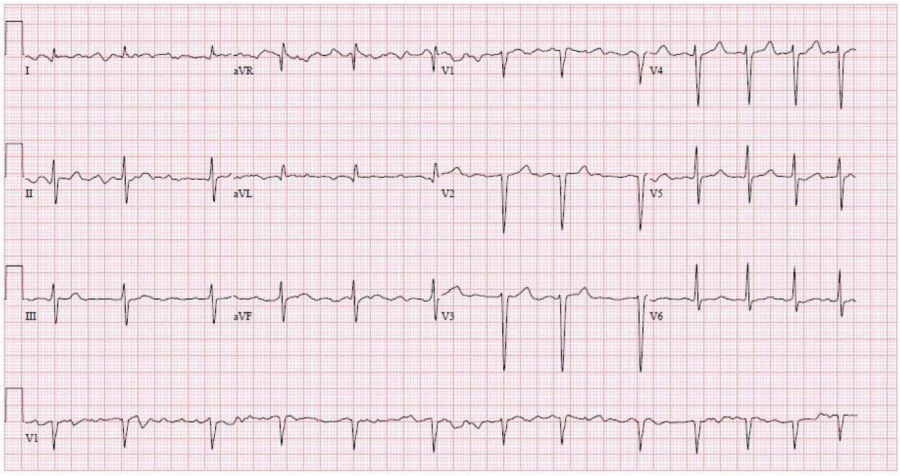

Figure 1.
Electrocardiogram showed atrial fibrilization with a ventricle rate of 76 bpm

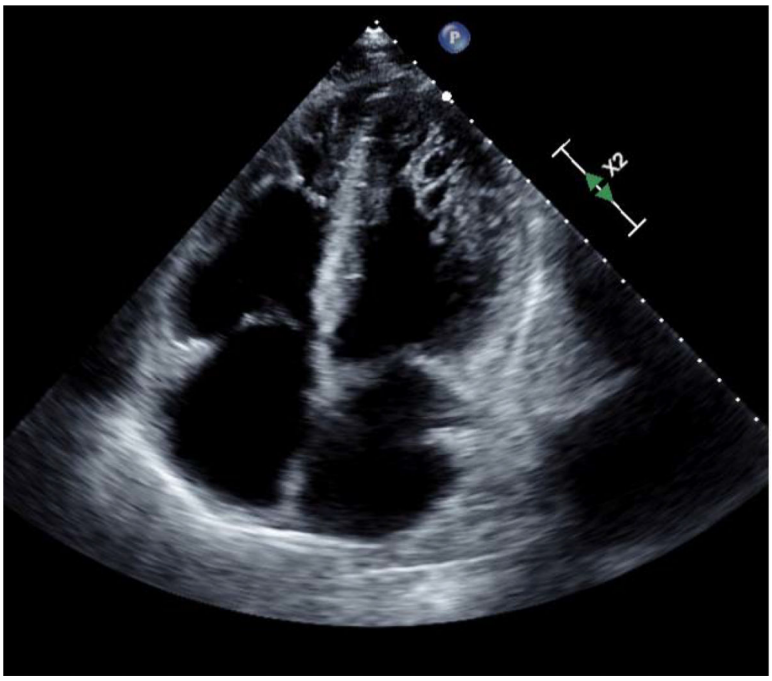

Figure 2.

Echocardiogram demonstrated mildly increased left ventricular wall thickness with prominent trabeculation[red arrow], ejection fraction of 55–60% and a pseudo-normal left ventricular filling pattern, with concomitant abnormal relaxation and increased filling pressure

Table 1.

Laboratory Data on Initial Presentation

Serum	Patient	Reference
Na	140	136–146 (mmol/L)
K	4.2	3.5–5.5 (mmol/L)
Cl	104	98–106(mmol/L)
Glucose	85	70–99(mg/dL)
Blood urea nitrogen	41	6–20(mg/dL)
Creatinine	9.2	0.4–12(mg/dL)
Protein total	6.5	6.0–8.5(g/dL)
Albumin	3.86	2.8–5.7(g/dL)
Alkaline phosphatase	78	25–125(U/L)
AST	18	10–35(U/L)
ALT	23	0–31(U/L)
Calcium	9.1	8.4–10.3 (mg/dL)
Magnesium	2.3	1.9–2.7(mg/dL)
Phosphorus	4.4	2.5–5.0(mg/dL)
Total Bilirubin	0.7	0.0–1.2(mg/dL)
Troponin	0.07	0.0–0.15(ng/dL)
Thyroxine	6.7	5.2–10.5 ug/dL
Thyroid Stimulating Hormone	3.07	0.38–4.70 uIU/mL
T4 free	0.98	0.71–1.85 ng/mL
Hemoglobin	11.9	12.0–16.0(g/dL)
WBC	4.95	4.5–109(cells/mm^3)
Platelets (K/uL)	136	130–400(K/mm^3)

Table 2.

"Jenni Criteria" - Four Morphological Echocardiographie Diagnostic Criteria for LVNC [7]

	Four Morphological Diagnostic Criteria for LVNC based on Echocardiogram
(1)	Coexisting cardiac abnormalities were absent (by definition)
(2)	A two-layer structure was seen, with a compacted (C) thin epicardial band and a much thicker non-compacted (NC) endocardial layer of trabecular meshwork with deep endomyocardial spaces. A maximal end systolic ratio of NC/C layers of >2 is diagnostic.
(3)	The predominant localization of the pathology was to midlateral, apical, and mid-inferior areas. The pathological preparations confirmed the echocardiographic findings.
(4)	There was color Doppler evidence of deep perfused intertrabecular recesses.