A New Diagnosis of Left Ventricular Non-Compaction in a Patient Presenting with Acute Heart Failure

Aakash Bavishi[1*], Kaitlin Lima[2], Lubna Choudhury[3]

1. Department of Internal Medicine, Northwestern University Feinberg School of Medicine, Chicago, Illinois, USA

2. Department of Internal Medicine, Northwestern University Feinberg School of Medicine, Chicago, Illinois, USA

3. Division of Cardiology, Department of Medicine, Bluhm Cardiovascular Institute, Northwestern University Feinberg School of Medicine, Chicago, Illinois, USA

* **Correspondence:** Aakash Bavishi M.D., Department of Internal Medicine, Northwestern University Feinberg School of Medicine, 251 E. Huron Street Galter Suite 3-150, Chicago, Illinois, USA
(*a-bavishi@northwestern.edu*)

Radiology Case. 2018 July; 12(7):10-15 :: DOI: 10.3941/jrcr.v12i7.3351

ABSTRACT

Left ventricular non-compaction is an overall rare cardiomyopathy; however, it is increasingly being recognized with advances in imaging technology. We present the case of a 47-year-old man with new diagnosis of heart failure and left ventricular non-compaction. We review the literature regarding diagnostic imaging criteria and management of this condition.

CASE REPORT

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A 47-year-old African American male with history of hypertension presented with significant progressive dyspnea, orthopnea, and paroxysmal dyspnea. One month prior to admission, he had symptoms consistent with upper respiratory infection including cough and rhinorrhea. His mother had a vague history of heart failure and the patient was not taking any medications.

On examination, he had tachycardia with an S3 gallop. ECG showed sinus tachycardia with left atrial enlargement and left ventricular hypertrophy. His peak Troponin I was 0.45 ng/dl and Brain Natriuretic Peptide was elevated at 951 pg/ml. TSH was within normal limits.

Posteroanterior chest film displayed moderate cardiomegaly with prominent left ventricle (Figure 1). Transthoracic Echocardiography showed dilated left ventricle with global hypokinesis, ejection fraction of 15 percent, and prominent trabeculations of the left ventricular myocardium (Figure 2). Cardiac MRI (Figure 3) confirmed the diagnosis of left ventricular non-compaction cardiomyopathy (LVNC) with focal myocardial infarcts likely consistent with coronary embolism (Figure 4, 5). Specifically, he met Petersen criteria for diagnosis of LVNC with ratio of noncompacted to compacted myocardium of ≥2.3:1 at end-diastole in the cardiac MRI images (Figure 3). Coronary angiography showed normal coronary arteries.

He was started on carvedilol and sacubitril/valsartan as well as warfarin. He was sent home on wearable defibrillator with plan for follow up in the heart failure clinic and subcutaneous ICD placement if his systolic function failed to improve after 3 months of maximally tolerated guideline directed medical therapy. After 6 months, patient had significantly improved symptomatically and had recovery of his left ventricular function to normal level so ICD was deferred.

DISCUSSION

Etiology & Demographics:

Left ventricular non compaction (LVNC) is a cardiomyopathy characterized by prominent left ventricular trabeculae, deep intertrabecular recesses, and thin compacted layer [1]. The etiology of LVNC is presumed to be an arrest of the normal process of intrauterine endomyocardial morphogenesis although this theory is not universally held

[2,28]. LVNC occurs more commonly in men and can be diagnosed at any age. Median age of diagnosis in adults is 40 years where as in children it is 7 years [3]. There is very limited data on incidence and prevalence of LVNC. Estimated incidence is <0.05% in adults and even lower in children[3] With advances in ECHO and cardiac MRI technology, LVNC is increasingly being recognized in heart failure patients, with prevalence reported at 3 to 4%[4].

There is still debate whether LVNC is a distinct cardiomyopathy or a morphologic trait shared by different cardiomyopathies. Whereas the American Heart Association classifies LVNC as a genetic cardiomyopathy, the European Society of Cardiology labels LVNC as an unclassified cardiomyopathy [1,5]. LVNC trait may be inherited or sporadic and there is evidence that there may be some genetic overlap between LVNC and hypertrophic cardiomyopathies [6]. The spectrum of morphologic variability in LVNC is also broad with varying extremes of compaction and trabeculations found in different hearts [7].

Imaging Findings:

There is no gold standard for diagnosis of LVNC. Echocardiography is the first tool in establishing the diagnosis of LVNC. In most patients, it is necessary to image the left ventricle with atypical views to detect prominent trabeculae in apex or with Doppler flow to highlight intertrabecular recesses [7]. Specific echocardiographic criteria including the Jenni, Chin, and Stollberger criteria have been established using small population cohorts [8,28]. There is concern that these echocardiographic criteria may have lower specificity in African American patients and athletes [9]. Furthermore, there is higher interobserver variability in making the diagnosis of LVNC, with disagreement in up to 35% of patients [10].

As demonstrated in our patient, MRI is a helpful adjunct to echocardiography as it is superior in assessing the extent of non-compaction [11]. Our patient had a noncompacted to compacted myocardial thickness ratio of > 2.3 at end diastole (Figure 3). This ratio is 99% specific for LVNC and confirms his diagnosis [12].

Differential Diagnosis:

The differential diagnosis of left ventricular non-compaction is broad and includes the following: cardiomyopathies including dilated, restrictive, and particularly hypertrophic (HCM), aberrant chordae tendineae, and hypertensive heart disease. LVNC must also be differentiated from trabeculations seen in younger, healthy individuals particularly athletes and African Americans. With excellent spatial resolution of all segments of left ventricle, especially the apex, cardiac MRI is particularly useful in distinguishing LVNC from hypertrabeculations seen on echocardiogram in younger, healthy individuals [13]. The distinction between HCM and left-ventricular non-compaction can be difficult to make on echocardiogram as well. Certain clues such as increased number of trabeculations, thinner maximal wall thickness, and lower ejection fraction are more suggestive of LVNC [14]. A noncompacted/compacted ratio of >2.0 in end-systole is highly specific for LVNC and is particular useful is differentiating from hypertrophic cardiomyopathy [8]. Cardiac MRI is particularly useful in distinguishing LVNC from other cardiomyopathies given the ability to absolutely quantify the percentage of non-compacted myocardium and definitively assess if the ratio of non-compacted to compacted myocardium meets the criteria for LVNC [15]. Advanced cardiac MRI modalities and functional data – including delayed gadolinium enhancement of trabeculae, high intensity endocardial T2 signals, and hypokinesis of noncompacted segment can also help support the diagnosis of LVNC [7,16].

Treatment:

As there are no specific therapy or guidelines for LVNC, treatment is focused on management and prevention of certain key clinical features. These include heart failure, sudden cardiac death, arrhythmia, and thromboembolic events. Patients such as ours with impairment of left ventricular ejection fraction are treated according to standard heart failure guidelines. LVNC patients are at increased risk of sudden cardiac death and limited data suggests that ICD devices are indicated in LVNC patients with reduction in ejection fraction [17]. LVNC patients may also qualify for ICD based on standard indications for nonischemic cardiomyopathy. Furthermore, LVNC patients are at increased risk for atrial and ventricular arrhythmias and may benefit from Holter monitoring [18]. With or without atrial fibrillation, LVNC patients are at increased risk of thromboembolism including stroke [19]. Despite overall increased risk, there is no specific guideline for anti-coagulation in LVNC patients and standard guidelines for atrial-fibrillation and cardio embolic events are followed. Given that our patient had infarcts consistent with coronary embolism, he was anti-coagulated with warfarin given no proven efficacy of Novel Oral Anti Coagulants (NOACs).

It is important to remember a possible hereditary genetic component to LVNC. The current recommendation is for careful family history of at least three generations and screening – including echocardiography- of first degree relatives of patients with LVNC [20].

Prognosis:

For both children and adults, mortality and morbidity of LVNC are high. A systematic review of 241 adults with isolated LVNC with mean follow-up of 39 months found a mortality rate of 14%, with half of the deaths from sudden cardiac death [21]. Mortality rate of children has been estimated between 12-14% after 3-4 year in multiple single center studies [22, 23]. At an average follow-up time of 44 months in a 34 patient cohort of adults diagnosed with LVNC, there were 12 deaths (34%) and major complications: heart failure in 18 patients (53%), thromboembolic events in 8 patients (24%), and ventricular tachycardias in 14 patients (41%) [24]. The presence of higher diastolic diameter of left ventricle, New York Heart Association Class III-IV heart failure, atrial fibrillation, and low ejection fraction were associated with poorer prognosis. Interestingly, the ratio of non-compacted to compacted myocardium does not correlate with prognosis [26]. 3 year mortality in adults similar to that of patients with non-ischemic dilated cardiomyopathy survival rate of 85% [27].

TEACHING POINT

Left ventricular non-compaction (LVNC) is an increasingly recognized cause of heart failure with specific echocardiographic and MRI criteria for diagnosis. The characteristic left ventricular trabeculae, deep intertrabecular recesses, and thin compacted layer that define LVNC lead to unique clinical manifestations that require specific management.

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FIGURES

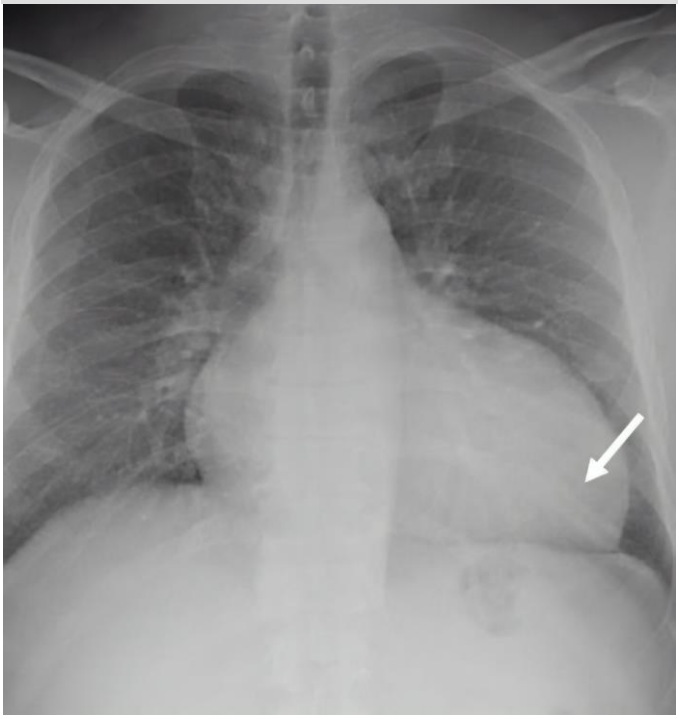

Figure 1: 47-year-old male presenting with new onset heart failure due to left ventricular non-compaction.
Findings: Posterior anterior chest film displaying enlargement of cardiac silhouette, vascular congestion, and prominent left ventricle (arrow).
Technique: Posterior anterior chest x-ray of patient

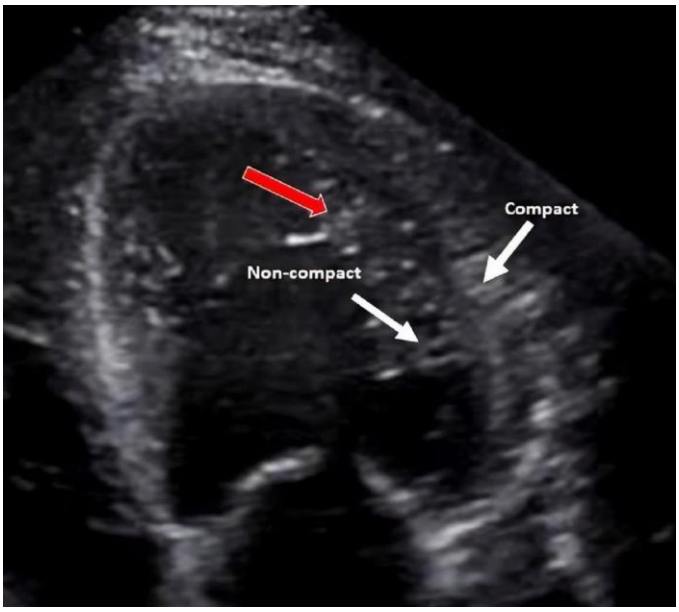

Figure 2: 47 year old male presenting with new onset heart failure due to left ventricular non-compaction.
Findings: Apical 4 chamber view of left ventricle showing dilated left ventricle with trabeculations noted on the lateral wall (red arrow). Non-compact and compact myocardium displayed on lateral wall.
Technique: 2 Dimensional Transthoracic Echocardiography.

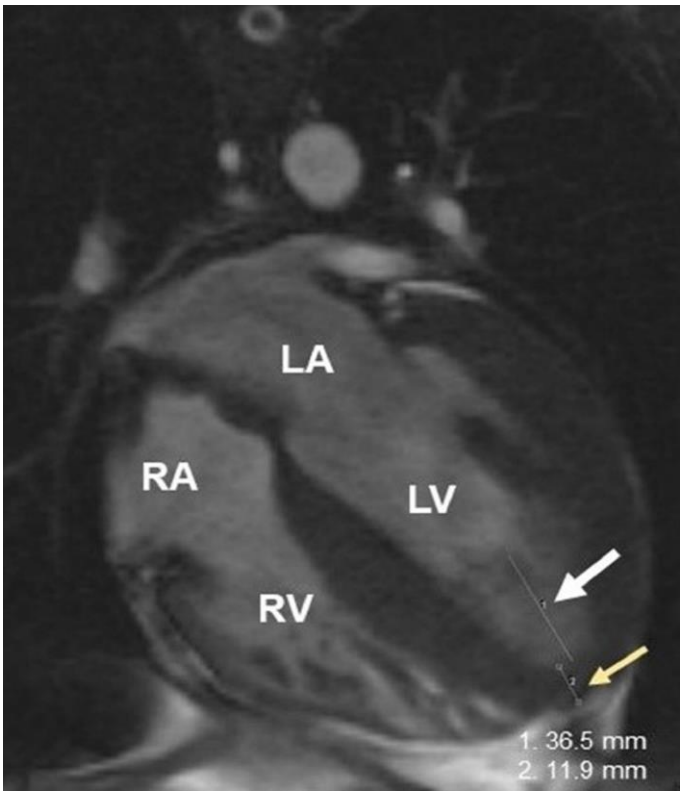

Figure 3: 47 year old male presenting with new onset heart failure due to left ventricular non-compaction.
Findings: 4 chamber still frame cardiac MRI showing a layer of non-compact myocardium (white arrow) in the left ventricular apex compared with compact myocardium (yellow arrow) with a ratio of non-compact:compact myocardium >2.3, RA= right atrium, RV= right ventricle, LA= left atrium, LV= left-ventricle
Technique: Still frame of steady state free precession axial 4 chamber view of left ventricle.

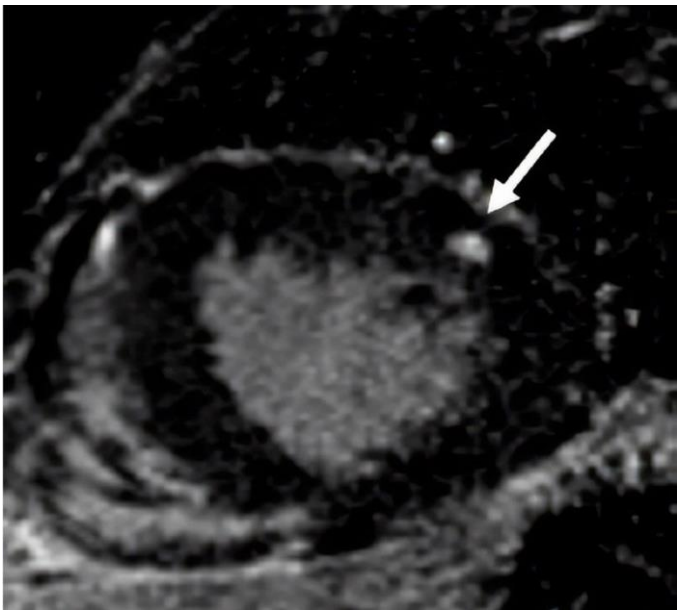

Figure 4: 47 year old male presenting with new onset heart failure due to left ventricular non-compaction.
Findings: Short axis cardiac MRI view showing focal mid myocardial infarct delayed contrast enhancement (arrow) likely representing an embolic infarct in the anterolateral wall of left ventricle.
Technique: T1 weighted phase sensitive inversion recovery with delayed contrast enhanced viability sequence. 20 mL Gadavist administered at dose of 0.2 mmol/kg.

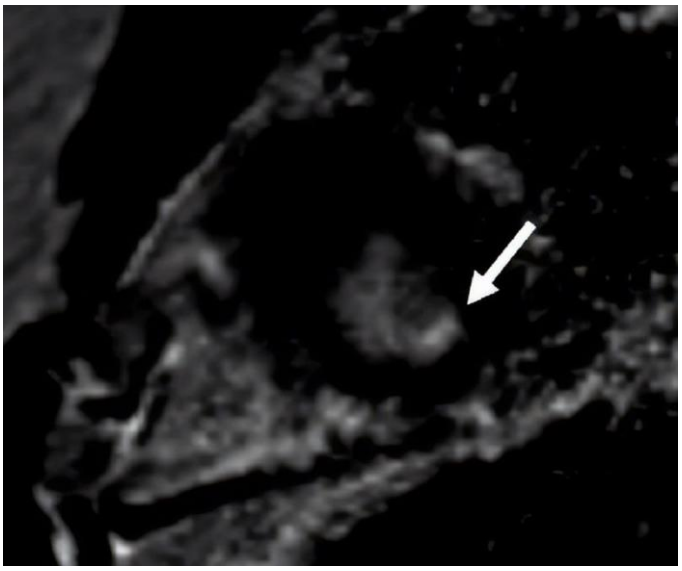

Figure 5 (right): 47 year old male presenting with new onset heart failure due to left ventricular non-compaction.
Findings: Short axis cardiac MRI view showing focal subendocardial infarct in the inferolateral wall of left ventricle.
Technique: T1 weighted phase sensitive inversion recovery with delayed contrast enhanced viability sequence. 20 ml Gadavist administered at dose of 0.2 mmol/kg.

Etiology	Likely due to intrauterine arrest of compaction of fetal myocardium in utero; can be sporadic or familial
Incidence	Overall- not much data • Incidence estimated to be 0.05% in adults ; <.01% in children ages 0-10 • Prevalence: 0.01 to 1.3% in patients undergoing echocardiography. 3-4% in patients with heart failure.
Gender Ratio	Men are most commonly affected, accounting for 56-82% of cases
Age Predilection	Can be diagnosed at any age with mean diagnosis of 40 years for adults and 7 years for children
Risk Factors	• Family History of LVNC
Treatment	No specific management guidelines for LVNC • Standard Guideline Directed Heart Failure Therapy (Beta-blocker, ACE –I for reduced ejection fraction) • ICD placement according to Heart Failure and Non-Ischemic Cardiomyopathy Guidelines • Anti-coagulation for arrhythmia or systemic thromboembolism • Family Counseling and Screening
Prognosis	• Overall, high mortality and morbidity due to heart failure, thrombo-embolic events and ventricular arrhythmias

Table 1: Summary of Left Ventricular Non-Compaction.

	Chest X-ray	**Transthoracic Echocardiography**	**Cardiac MRI**
LVNC	Non-specific; can show cardiomegaly and pulmonary edema if patient in heart failure	• Thickened left ventricular wall with thin compacted epicardial layer • Numerous trabeculations within thickened endocardial layer • Lower ejection fraction • Noncompacted to compacted myocardium > 2:1 at end-systole	• Non-compacted to compacted myocardial ratio of > 2.3 in diastole • Delayed gadolinium enhancement of trabeculae • High intensity endocardial T2 signals • Hypokinesis of non-compacted segment
Hypertrophic Cardiomyopathy	Non-specific; cardiomegaly may be seen along with left atrial enlargement	• Unexplained increased LV thickness ≥15mm or ≥13mm • Frequently have systolic anterior motion of the mitral valve	• Can identify areas of segmental LV hypertrophy not reliably visualized by echocardiography(apex and anterolateral wall) • Late gadolinium enhancement indicating myocardial fibrosis
Healthy, Young Individual	Normal	• Extensive left ventricular trabeculations may be seen at end-diastole	• Quantification shows that patient does not meet criteria for LVNC

Table 2: Differential diagnosis table for Left-Ventricular Non-Compaction.

ABBREVIATIONS

ECG = electrocardiogram
HCM = Hypertrophic Cardiomyopathy
LVNC = Left ventricular noncompaction
MRI = Magnetic Resonance Imaging
TSH = Thyroid Stimulating Hormone
TTE = transthoracic echocardiograpy

KEYWORDS

Case Report; MRI; Left Ventricular Non Compaction; Heart Failure; Non-Ischemic Cardiomyopathy; Petersen criterion

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