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# Left Ventricular Noncompaction: A Rare Cause of Heart Failure in a HIV Patient

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***Authors' contributions***

*All the four authors contributed to the conception, design, analysis and preparation of the manuscript. All authors read and approved the final manuscript.*

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***Case Study***

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## ABSTRACT

**Background:** Heart failure in patients with *human immunodeficiency virus (HIV)* is often from dilated cardiomyopathy as a result of HIV itself, drug myotoxicity, secondary infections, or drug-induced atherosclerosis. Left ventricular noncompaction (LVNC) is a rare cardiac congenital abnormality which occurs due to early arrest of endomyocardial morphogenesis.
**Case:** A 47- year-old female patient with HIV presented with sudden onset shortness of breath and symptoms of congestive heart failure. Echocardiography showed noncompacted endocardium with reduced left ventricular function. She was subsequently diagnosed with LVNC.
**Discussion:** Multiple etiologies have been implicated in cardiomyopathy among HIV patients. LVNC is a rare cause of left ventricular failure, particularly in this population. Echocardiography plays a pivotal role in the diagnosis.
**Conclusion:** It is often challenging to identify the underlying cause of cardiomyopathy in a patient with HIV. While LVNC is a rare cause of left ventricular failure, typical findings on echocardiography can obviate the need for a more complex evaluative strategy.

*Keywords: Left ventricular noncompaction; Human immunodeficiency virus infection; heart failure; Echocardiography.*

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## 1. BACKGROUND

The Joint United Nations Program estimated 35 million people were living with *human immunodeficiency virus* (HIV) infection at the end of 2012 [1]. Heart failure in this population is often a result of dilated cardiomyopathy from HIV itself, drug myotoxicity, secondary infections, nutritional deficiencies or drug-induced atherosclerosis. Left ventricular noncompaction (LVNC) is a rare cardiac congenital abnormality, which occurs due to embryological arrest of endomyocardial morphogenesis between 4-8 weeks. We present a rare cause of cardiomyopathy due to LVNC in a 47-year-old woman with HIV infection.

## 2. CASE

A 47 year-old Caucasian woman with a seven year history of HIV infectionpresented to the emergency department withseven hours of worsening shortness of breath, paroxysmal nocturnal dyspnea, and orthopnea unrelieved with albuterol nebulization. She also complained of malaise and palpitations, but denied fever, chills, chest pain, or diaphoresis.

Her past medical history was significant for HIV infection, hypertension (diagnosed a few months prior to the presentation), anemia, and history of poly-substance abuse. Medications included emtricitabine-tenofovir, darunavir, ritonavir, metoprolol, promethazine, and amlodipine. She was a current half pack per day smoker, and had regularly consumed three beers a week since age 21. Shelast used cocaine two days previously, and methamphetamine onemonth ago, (she reported these as sporadic usage, drug screens from other admissions were negative). Family history was negative for premature coronary artery disease or congestive heart failure.

Physical examination was significant for heart rate of 105 beats/minute, cachetic appearance, conjunctival pallor, bibasilar crackles, and a 2/6 systolic murmur, most prominent in the mitral area.

Laboratory tests revealed elevated brain natriuretic peptide (BNP) at 492 pg/ml (normal <100 pg/ml), and negative cardiac enzymes with troponin levels consistently below 0.02 ng/ml (normal 0.00-0.03 ng/ml). Hemoglobin was 7.6 grams/dL, MCV of 65 fL, MCH 20.3 pg, MCHC 31.1 g/dL and known iron deficiency from internal hemorrhoids for which she was not taking her iron. A lipid panel was within normal limits. CD4 count of 230 cells/uL. Electrocardiogram (ECG) revealed sinus tachycardia without any ST-T changes suggestive of ischemia or infarction. Chest radiograph showed cardiomegaly with pulmonary vascular congestion. In the emergency department, spiral computer tomographic angiography was negative for pulmonary embolism.

A 2D-echocardiogram with perflutren lipid microsphere echo contrast (DEFINITY®) showed left ventricular (LV) ejection fraction of 25-30% with moderate to severe global hypokinesis of the left ventricle. The left ventricle appeared mildly dilated, with prominent sinusoidal trabe culations to the apex suggestiveof LVNC (Figs. 1, 2 and 3). Right ventricular function was normal. She was also incidentally noted to have mild mitral regurgitation.

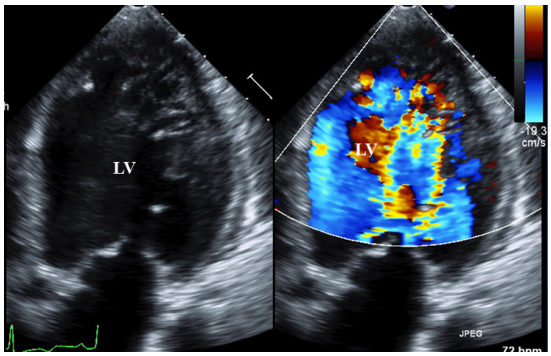


**Fig. 1. 2D echocardiogram of the left ventricle in the apical two chamber view with and without color flow Doppler. This view demonstrates the flow of blood between the trabeculae. [LV-left ventricle]**

She was medically managed with an angiotensin converting enzyme inhibitor, beta blocker, loop diuretic, fluid restriction and sodium restriction. Her HIV is well managed without history of opportunistic infections. HerECG, biomarkers did not suggest any ischemia or infarction. The diagnostic echocardiography findings obviated the need for an extensive evaluation for other causes of her cardiomyopathy. She was discharged within three days, and has not required additional hospitalizations.

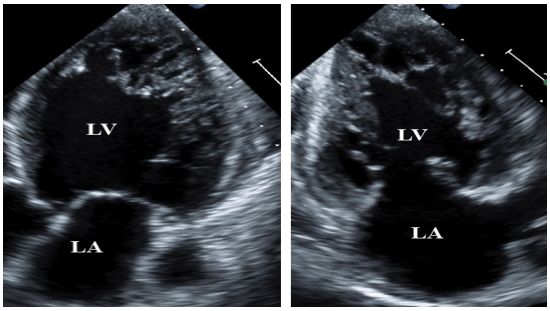


**Fig. 2. Apical three and two chamber view demonstrating noncompaction of the left ventricle. [LA-left atrium, LV-left ventricle]**

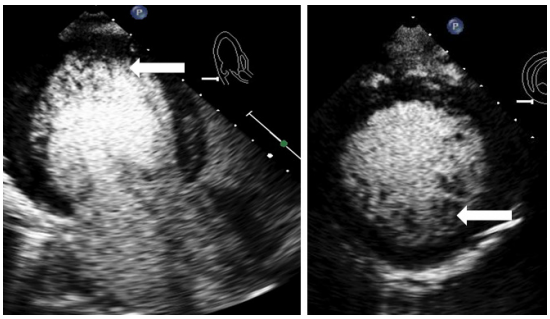

**Fig. 3. 2D echocardiogram with perflutren lipid microsphere echo contrast (DEFINITY®) of the left ventricle in the apical three chamber and parasternal short axis view showing compacted and non-compacted myocardium (arrow)**

## 3. DISCUSSION

The HIV virus itself is asignificant cause of dilated cardiomyopathy, late in the course of infection.With the advent of highly active antiretroviral therapy, cardiomyopathy has become less common among patients with HIV, falling 30% [2] from a baseline annual rate of 15.9/1000 [3] patients annually in developed nations.

Other causes of cardiomyopathy in this population are myriad including: drug myotoxicity; secondary infection; coronary artery disease from medication-induced hyperlipidemia; and an autoimmune process triggered by HIV, coxsackievirus, cytomegalovirus, or Epstein-Barr virus [4]. Understandably, then, the diagnostic work up for a patient with HIV and new onset congestive heart failure is often complex.

Left ventricular noncompaction (LVNC), previously known as spongy myocardium or hypertrabeculation syndrome, is a rare cause of cardiomyopathy in the general population. One of several primary genetic cardiomyopathies, it is generally inherited in an autosomal dominant fashion [5]. It usually affects the left ventricle, but right ventricularinvolvement has been reported. It is an under-recognized phenomenon, but lately there has been greater interest in LVNC due to the technological advances and increased availability of the imaging modalities.

The pathophysiology of ventricular dysfunction in LVNCis not well understood. One group has suggested that subendocardial hypoperfusion and microcirculatory dysfunction related to the structural changes lead to ventricular dysfunction and arrhythmias [6]. Alternatively, the persistent presence of the trabecular layer may interfere with the effective ejection of blood from the left ventricle because of changes in laminar flow [7,8]. LVNC has been associated with mitochondrial myopathy and othergenetic mutations that cause myofibrosis, which in turn may lead to LV dysfunction [9,10]. As one of our authors previously elucidated in their previous report the late presentation is mostly related to the process of myocardial fibrosis which takes several years leading to the LV dysfunction [11].

Patients with LVNC may be asymptomatic, or may present with heart failure or arrhythmias. In one case series, the most common manifestations at diagnosis were: dyspnea (79 %) and New York Heart Association class III or IV heart failure (35 %) [12].

Diagnosis is usually made based upon echocardiography. The most widely used criteria are noncompaction to compaction ratio (NC:C) > 2, and visualization of blood flowing directly from the ventricular cavity into the deep inter-trabecular recess [12]. Computed tomography, positron emission tomography, and magnetic resonance imaging may also be used [13-17].

LVNC has a poor prognosis. Early studies reported 35% mortality with mean follow-up of 3.5 years [12]. More recently, mortality has been reported at 2-15% annually [18]. Patients should be treated aggressively according to standard heart failure guidelines, including the use of implantable cardiac defibrillators (ICD) to prevent sudden cardiac death. While there have been severalreports of unsuccessful defibrillation with ICDs in LVNC, it is unclear if this is related to the underlying disease [19,20]. Heart transplant may be considered if medical therapies fail.

Our patient received optimalmedical therapy. On follow up, she did not meet criteria for ICD placement.

## 4. CONCLUSION

It is often challenging to identify the underlying cause of cardiomyopathy in a patient with HIV. While LVNC is a rare cause of left ventricular failure, typical findings on echocardiography can obviate the need for a more complex evaluative strategy.

## CONSENT

All authors declare that 'written informed consent was obtained from the patient (or other approved parties) for publication of this case report and accompanying images.

## ETHICAL APPROVAL

Not applicable.

## COMPETING INTERESTS

Authors have declared that no competing interests exist.

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