

A Late Diagnosis of Left Ventricular Non-Compaction Cardiomyopathy



Viral Desai MD, Neha Nidhi MD,
Marcus Stoddard MD

University of Louisville, Louisville, Kentucky

BACKGROUND

Left ventricular non-compaction (LVNC) cardiomyopathy is characterized by a bilayered myocardium consisting of prominent trabeculations, most commonly at the apex and deep intra-trabecular recesses in the left ventricle.

CASE

A 64-year-old African American male with a history of tobacco use, myocardial infarction, congestive heart failure secondary to dilated cardiomyopathy with AICD placement with an ejection fraction of 25%, paroxysmal atrial fibrillation, ventricular tachycardia and hypertension presented with worsening dyspnea, lower extremity swelling and orthopnea.

Physical examination: tachypneic with bilateral basilar lung crackles, oxygen saturation of 93% on room air, and pretibial pitting edema

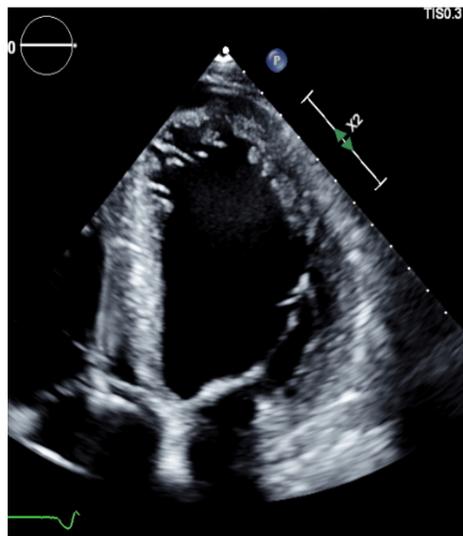
Admission labs & imaging: elevated BNP of 3171 pg/ml, chest x-ray showing pulmonary vascular congestion.

This patient's presentation was concerning for acute decompensated heart failure

Transthoracic echocardiogram (TTE) (Image 1 & 2):

- Severe global hypokinesia of the left ventricle (LV)
- Calculated LV ejection fraction of 23%
- Severely dilated LV
- Marked trabeculation of the LV apex (ratio of end diastolic noncompacted to compacted layer thickness >2) consistent with noncompaction dilated cardiomyopathy was noted

Underwent aggressive diuresis and was discharged with follow-up in advanced heart failure clinic to assess for possible heart transplant



Early diagnosis of LVNC using available institutional resources will allow for use of life saving interventions such as implantable cardioverter-defibrillator, anticoagulation therapy, and heart transplants and thus prevent adverse outcomes in these patients

For more information, scan the QR code or email:

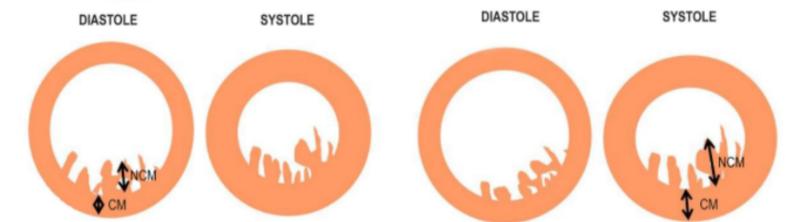
viral.desai@louisville.edu
neha.nidhi@louisville.edu

DISCUSSION

Our case describes a patient in whom LVNC was detected years after initial diagnosis of non-ischemic cardiomyopathy (DCM) – with prior echocardiograms not revealing the classic prominent trabeculations of the LV.

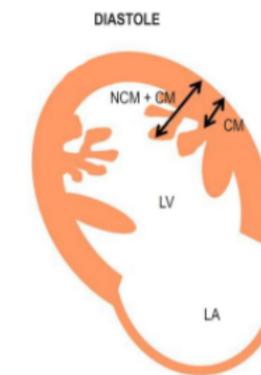
Despite widely used two-dimensional imaging modalities such as TTE and cardiac MRI (CMR), LVNC remains poorly diagnosed.

The wide range of diagnostic criteria and the lack of a 'true' diagnostic gold standard in a complex disease phenotype such as LVNC contributes to its variable prevalence rate.



PATEREK CRITERIA (Wisconsin) = $NCM/CM > 2$
Measured in End Diastole

JENNI CRITERIA (Zurich) = $NCM/CM > 2$
Measured in End Systole



CHIN CRITERIA (California) = $CM/(NCM + CM) \leq 0.5$
Measured in End Diastole



Image 2

The current diagnostic criteria (eg. Chin criteria, Jenni and Peterson criteria) of this rare disease based on studies with extremely small homogenous cohorts and thus has limited external validity.

Given the uncertainty around the diagnosis there is a need for the development and acceptance of a more precise and universal diagnostic criteria for LVNC.

DISCLOSURES

No disclosures