Unmasking Left Ventricular Noncompaction Cardiomyopathy: NSTEMI in a 37-Year-Old Woman with Hypertensive Crisis

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INTRODUCTION

- A rare familial cardiac condition with initial presentation as NSTEMI (Non-ST-Elevation Myocardial Infarction).

EPIDEMIOLOGY

- Prevalence reported between 0.014% and 1.3% in cohorts undergoing echocardiography; often quoted ~0.05% in echo series.
- Familial in 12–50% of cases; mostly autosomal dominant with ~66 implicated genes reported.

COMMON CLINICAL PRESENTATIONS

- Dyspnea (~60%), palpitations (~18%), chest pain (~15%), syncope/presyncope (~9%), prior stroke in ~3%.
- Complications: heart failure, ventricular arrhythmias, thromboembolism.

CASE PRESENTATION

Case summary: 37-year-old woman with tobacco dependence presenting with 12 hours of crushing chest pain and hypertensive crisis, ultimately found to have features suggestive of LVNC with NSTEMI (demand ischemia).

➢ Demographics & risk factors

- Age: 37 years, female
- Past medical history: no significant PMH reported (other than chronic tobacco dependence)
- Risk factors: cigarette smoking, family history of heart disease, sedentary lifestyle

➢ Presenting complaint

- 12 hours of classical crushing chest pain (anginal in quality)

➢ Vitals / exam on arrival

- Blood pressure: BP - 220/120mmHg (hypertensive emergency)
- Heart rate: 51 bpm, sinus bradycardia on ECG
- Chest X-ray: no acute findings

➢ Key laboratory results

- Serial troponin: 5,000 → 21,387
- proBNP: 3,304

➢ Initial working diagnosis & acute management

- Working dx: Acute NSTEMI (type 2 / demand ischemia likely related to hypertensive emergency and myocardial strain)
- ACS protocol initiated; cardiology consulted; decision for early invasive strategy

➢ Coronary angiography / cath

- Left heart cath: mild disease of nondominant right coronary artery — ~40% proximal stenosis
- Left heart pressures: normal
- LVEF (angiography estimate): 60%
- Valves: mild MR, trace AR

➢ Echocardiography

- Transthoracic echo (TTE): LVEF 60%, prominent trabeculations consistent with suspected LVNC

➢ Additional imaging / plan

- Cardiology recommended cardiac MRI for diagnostic confirmation and further characterization (CMR is gold standard for morphology/quantification)

➢ In-hospital treatment

- Dual antiplatelet therapy (DAPT) initiated
- Apixaban (Eliquis) started for thromboprophylaxis (concern for thromboembolism risk with LVNC)
- BP control: Losartan-HCTZ + Amlodipine
- Atorvastatin started (secondary prevention)

IMAGING

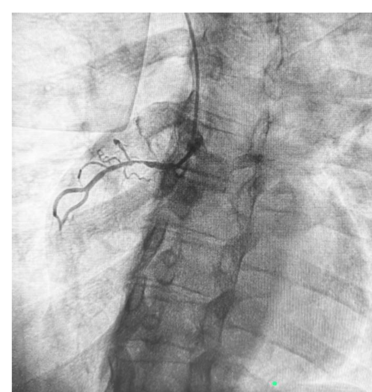

Fig 1: Right Coronary System

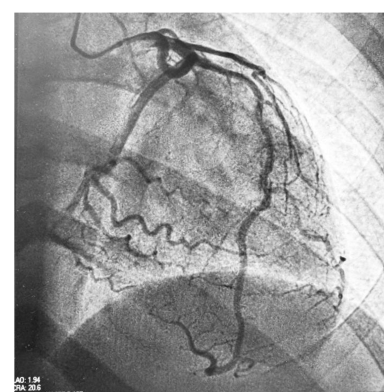

Fig 2: Left Coronary System

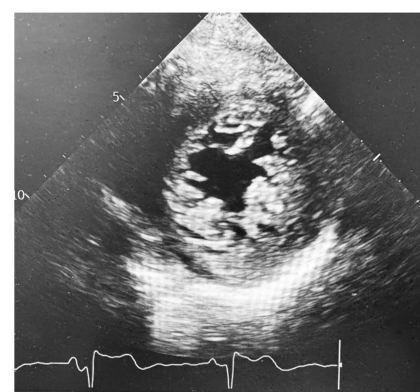

Fig 3: Short axis of the left Ventricle showing trabeculation

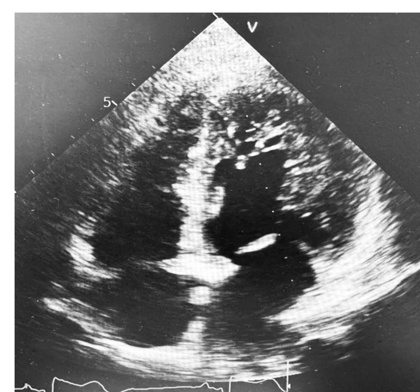

Fig 4: Apical 4-chamber view showing trabeculation in LV apex

CONCLUSION

This case emphasizes the need to consider left ventricular noncompaction cardiomyopathy (LVNC) in young patients with acute coronary syndrome-like symptoms but no significant coronary artery disease. Early detection through echocardiography and cardiac MRI is vital for accurate diagnosis and management, helping to prevent complications like heart failure, arrhythmias, and thromboembolism.

DISCUSSION

- Left ventricular noncompaction (LVNC), also known as isolated ventricular noncompaction or left ventricular hypertrabeculation, is a rare unclassified cardiomyopathy characterized by an altered myocardial wall. This condition results from an intrauterine arrest of compaction in the loose interwoven meshwork that forms the fetal myocardial primordium.
- LVNC has been identified in approximately 0.05 percent of patients undergoing echocardiography. It can be either sporadic or familial, with the latter predominantly following an autosomal dominant inheritance pattern involving around 66 implicated genes.
- Patients with LVNC often present with heart failure, thromboembolism, or ventricular arrhythmias. Our index patient was a young woman with no past medical history, though she had risk factors including cigarette smoking, a family history of heart disease, and a sedentary lifestyle.
- This case report illustrates a rare cause of cardiomyopathy that predisposed the patient to a hypertensive emergency, ultimately leading to myocardial strain and NSTEMI type 2 (demand ischemia).
- The diagnosis of LVNC is typically established by identifying morphologic diagnostic criteria on TTE, which includes a non-compacted to compacted myocardium ratio greater than 2:1 at end-systole. Cardiac MRI is recommended for most patients with known or suspected LVNC.

References

1. Christen H Attenhofer Jost, MD et al, Isolated Left Ventricular Noncompaction in adults: Clinical manifestations and diagnosis Isolated left ventricular noncompaction in adults: Clinical manifestations and diagnosis - UpToDate
2. Marilyn Weigner, MD, FACC, James P Morgan, MD, PhD, Causes of Cardiomyopathy Causes of dilated cardiomyopathy - UpToDate