Case Report

Left ventricular non-compaction; an unusual presentation with supraventricular tachycardia in the emergency department

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1. Introduction

Left ventricular non-compaction (LVNC) is a rare genetic cardiomyopathy affecting the ventricular wall anatomy [1]. The condition is characterized by the presence of prominent left ventricular trabeculae, a thin compacted layer, and deep intertrabecular recesses that are continuous with the left ventricular cavity and separated from the epicardial coronary. Left ventricular non-compaction can present with acute heart failure, arrhythmias, or sudden cardiac death. We present a case of a common cardiac arrhythmia in the emergency department with a work up consistent with LVNC being the underlying etiology.

2. Case report

A 46 year old male with a past medical history of hypertension, presents to the emergency department with shortness of breath on exertion, cough, and palpitations for 2 days. On physical examination the vital signs were: blood pressure of 150/107 mmHg, heart rate of 162 beats per minute (bpm), respiratory rate of 18 breaths per minute, temperature of 37.1 °C, and a pulse oximetry of 99% on room air. The patient was oriented to person, place, and time, while appearing anxious, dyspneic, and uncomfortable. The rest of the patient's exam was grossly unremarkable aside from a tachycardia. An electrocardiogram (ECG) was performed, which revealed SVT with a rate of 157 bpm and evidence of left ventricular hypertrophy (LVH). After unsuccessful conversions with Valsalva maneuvers and 6 mg of adenosine, the patient was successfully converted to normal sinus rhythm with 2 doses of 12 mg.

Given ECG findings and presentation, lab work was done and was notable for a white blood cell count of 15.8 × 10^9/L, sodium of 134 mmol/L, carbon dioxide level of 20 mmol/L, B type Natriuretic Peptide of 312 pg/mL and D-dimer of 4030 ng/mL. Chest radiograph showed cardiomegaly and findings consistent with acute congestive heart failure. However, given the elevated D-dimer and presentation of a tachycardia, a computed tomography angiography of the chest was ordered, which was ultimately negative for pulmonary embolism. Point-of-care ultrasound (POCUS) echocardiogram performed in the emergency department team demonstrated hypokinesis of the LV. Given the evidence of new onset heart failure with an atypical bedside echocardiogram, the patient was admitted with Furosemide, Aspirin, and a cardiology consultation. 2D echocardiogram was done with findings of an ejection fraction of 34%, with severe global hypokinesis, demonstrated in Fig. 1.

The interpreting cardiologist expressed concern for isolated LVNC. Cardiac magnetic resonance imaging (MRI) was recommended to help articulate the findings, and implantable cardioverter defibrillator (ICD) placement was recommended to help prevent ventricular tachycardia. Unfortunately, the patient refused these procedures and chose to leave the hospital against medical advice.

3. Discussion

This case illustrates a unique presentation of LVNC, with an initial arrhythmia of SVT in the ED. To our knowledge, this is the first report of LVNC presenting as SVT in the ED. While it is not expected that EM physicians make a diagnosis of LVNC, one should consider utilizing POCUS to assess patients in SVT whom otherwise have no significant risk
factors. Having some index of suspicion for underlying cardiomyopathies, such as LVNC, should be considered when assessing patients with SVT, as the treatment plan deviates greatly from other etiologies.

Left ventricular non-compaction is a rare subset among cardiomyopathies, and is known to have serious consequences such as arrhythmias, sudden cardiac death, systemic thrombosis, and heart failure \[3\]. The majority of patients diagnosed with LVNC will eventually require systolic support or heart transplantation \[2\]. Given this morbidity, emergency clinicians should be aware of LVNC and how it presents. The prevalence of LVNC is uncertain, but one study by Ritter et al. determined a prevalence of 0.05% among all adult echocardiographic examinations at a large institution \[4\]. Despite its rarity, LVNC is important to identify, since as many as 47% of adults with LVNC die within 6 years of presentation. This is due to a variety of arrhythmias, including life-threatening arrhythmias such as ventricular tachycardia and ventricular fibrillation. In addition, Steffel et al. found that supraventricular tachyarrhythmias (Atrial flutter, atrial fibrillation, etc.) were inducible in only 29% of the patients studied, with only 1 (4%) of the patients having an undifferentiated SVT \[5\]. For these reasons, earlier recognition could play a vital role reducing morbidity and mortality in this patient population.

The case we have presented is an example of a young, otherwise healthy adult who was seen in the ED with a common tachyarrhythmia. Given the utility of POCUS, the patient was identified as having findings concerning for an underlying cardiomyopathy, which initiated further workup by the inpatient cardiology team. Early recognition of the possibility of LVNC is crucial, as it can lead to fatal arrhythmias and treatment requires ICD placement. Left ventricular non-compaction is a rare but serious form of cardiomyopathy that should be in strong consideration, along with other cardiomyopathies, when assess patients who present to the ED with otherwise common tachyarrhythmias, such as SVT.

Funding
None.

Declaration of Competing Interest
None.

Acknowledgements
We would like to thank the William Beaumont Hospital (Royal Oak, MI) Emergency Department, as well as all the nurses and physicians that played a role in this patients care.

References