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Case Report

Improvement in systolic function in left ventricular non-compaction cardiomyopathy: A case report

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ABSTRACT

This is a case of a 48-year-old man who presented with a pulmonary embolism and was found to have left ventricular non-compaction cardiomyopathy. Initial echocardiograms demonstrated prominent apical trabeculations with reduced biventricular function. These findings were further confirmed and characterized by cardiac magnetic resonance imaging. He met all major criteria used to identify left ventricular non-compaction cardiomyopathy. He underwent medical management for heart failure and during follow-up was noted to have significant improvement in left ventricular systolic function and symptoms. While most management attention is focused on rhythm disturbances or embolic risk, particular attention should also be exercised to ensure that heart failure medical therapy is optimized. While many with left ventricular non-compaction cardiomyopathy have irreversible dysfunction, this case highlights that there may be some who will respond well to aggressive medical therapy. The diagnosis and medical management of left ventricular non-compaction cardiomyopathy are reviewed in light of our patient and his clinical course.

<**Learning objective:** Historically, left ventricular non-compaction cardiomyopathy (LVNC) has been associated with significant morbidity and mortality. Discussion often focuses on sudden cardiac death and prevention of embolisms. Many of the initial reports and case series were written in an era when standard medical therapy for congestive heart failure was not yet defined. While many do not respond as this case did, this case emphasizes that optimal medical therapy can make a substantial difference, even for LVNC.>

Introduction

Isolated left ventricular non-compaction cardiomyopathy (LVNC) is an uncommon myocardial disorder characterized by prominent trabeculae and deep intertrabecular recesses forming thickened myocardium consisting of a thin compacted epicardial layer and a thickened non-compacted endocardial layer [1]. Initial case series reported significant morbidity and mortality from heart failure, ventricular arrhythmias, thromboembolism, and sudden cardiac death among individuals affected with LVNC [2,3]. Given this association with adverse outcomes, several imaging criteria have been proposed to identify those with LVNC. As cardiac imaging techniques have improved, it has become more common for cardiac imaging to identify trabeculations, and at times, it is more difficult to separate the normal from the pathological. More difficult still is the management of those with LVNC.

Case report

The patient was a 48-year-old Caucasian male with a history of hyperlipidemia and peripheral vascular disease who presented to the emergency department with a 4-day history of shortness of breath, cough, hemoptysis, and bilateral swelling of the legs. He was found to have a pulmonary embolism via computed tomography angiography imaging. He also complained of intermittent shortness of breath and symptoms of orthopnea for the past year. He was still able to carry out his job as a construction worker, albeit with reduced workload tolerance. His vital signs on presentation showed a heart rate of 78 beats per minute with a blood pressure of 148/102 mmHg. His oxygen saturation was 94%.

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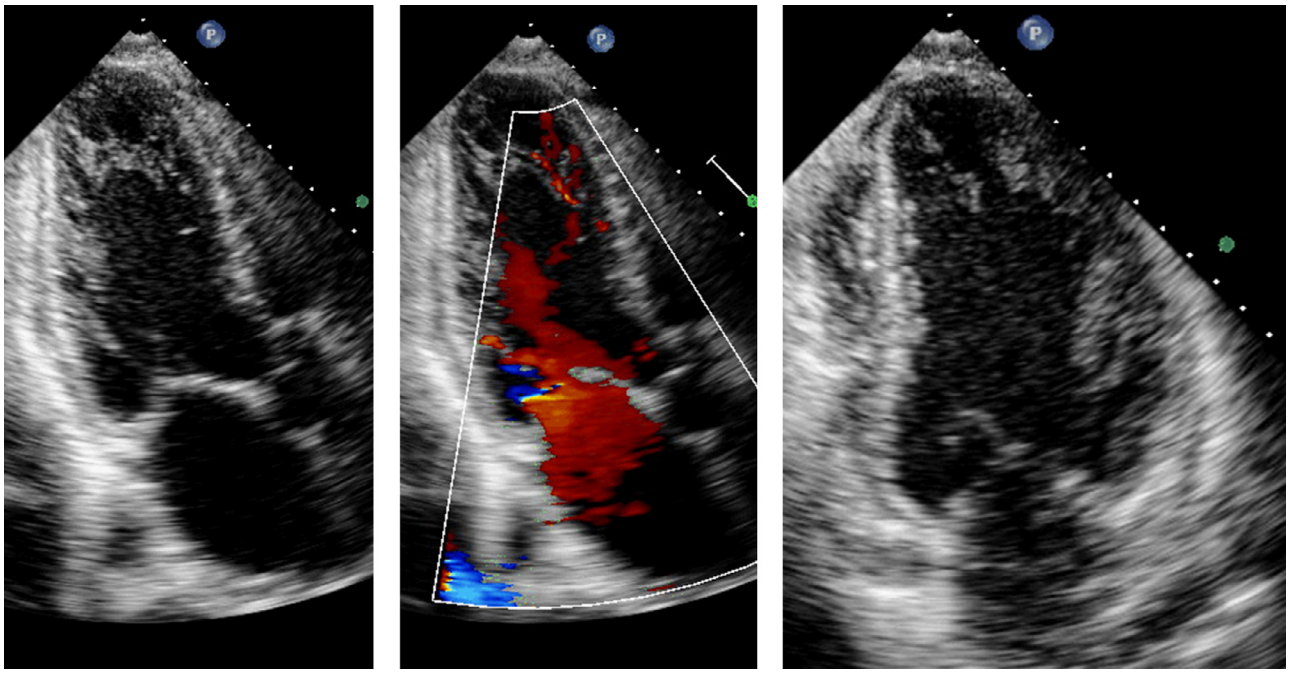

Fig. 1. Initial echocardiogram with apical 3-chamber view presented with (center) and without (left) color flow Doppler to demonstrate blood flow within the trabecular recesses. Apical 2-chamber view shown to demonstrate trabeculations (right).

On physical examination, heart sounds were normal. There was mild tachypnea, but the lungs were clear to auscultation. Symmetric bilateral lower extremity edema and tenderness to right lower extremity were also present. His 12-lead electrocardiogram showed non-specific anterior T-wave abnormalities.

The initial transthoracic echocardiogram showed prominent trabeculations in both the left and right ventricular apex with severely reduced left ventricular ejection fraction (LVEF) of 15–20% as well as color flow Doppler in the trabecular recesses (see Fig. 1 and Movie 1). The ratio of non-compacted to compacted myocardium by echocardiography was 2.4 at end-diastole and 2.1 (normal: <2) at end-systole from the short-axis apical slices. Cardiac magnetic resonance imaging (MRI) demonstrated a moderately dilated left ventricle with a severely reduced LVEF of 11% (see Fig. 2 and Movie 2). Trabecular mass to total mass ratio was 50% (normal: <20%), the ratio of non-compacted to compacted myocardial thickness was 2.6 at end-diastole from the apical short axis view (normal: <2.3), and the ratio of non-compacted to compacted myocardial thickness was also 2.3 at end-systole from the same view (normal: <2). In reviewing the long-axis views, the average of the end-diastolic non-compacted to compacted ratios from the 3 long-axis views was 3.3 (normal: <2.3). These measurements confirmed the diagnosis of LVNC. There was no late gadolinium enhancement seen during the MRI study, nor was

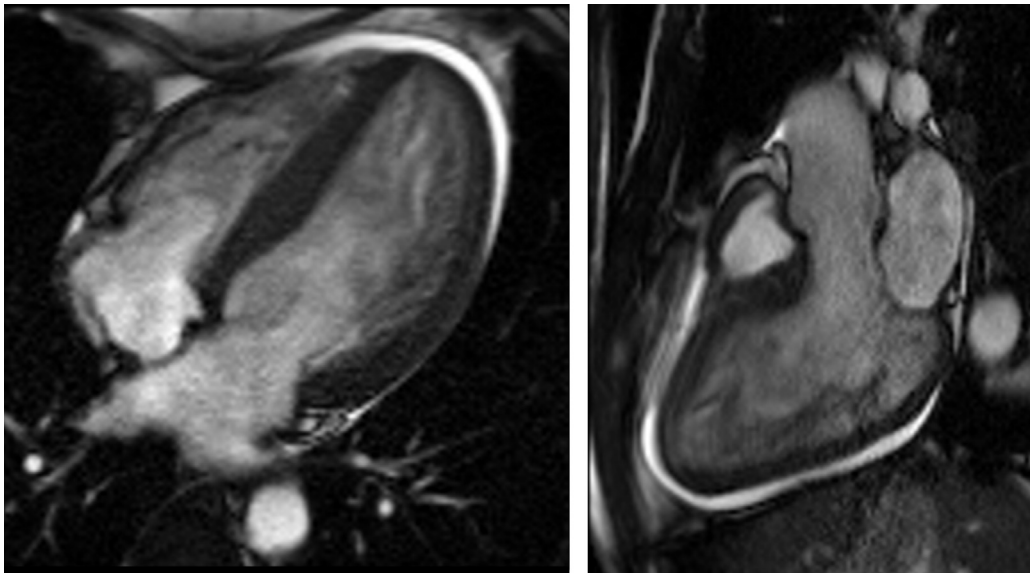

Fig. 2. Cardiac magnetic resonance imaging of 4-chamber view (left) and 3-chamber view (right) to demonstrate trabeculations.