Case Reports

A rare malady with a rarer complication

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ABSTRACT

Isolated ventricular noncompaction (IVNC) is an unclassified cardiomyopathy characterized by prominent intraventricular trabeculations separated by deep intertrabecular recesses. Although microvascular dysfunction is known, myocardial infarction is rare and usually seen as a consequence of coincidental coronary artery disease. We report the case of a 19-year-old male patient who presented to us with symptoms and signs of heart failure. Echocardiography revealed IVNC with severe left ventricular dysfunction. He was put on medical treatment including oral anticoagulants. Six months later, he came to our emergency department with anterior wall STEMI. Coronary angiogram revealed thrombotic occlusion of distal LAD, which resolved completely with tirofiban and heparin. Coronary thromboembolism due to blood stasis in the left ventricular cavity has not been previously documented in IVNC.

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1. Introduction

Isolated left ventricular noncompaction (IVNC) is a rare hereditary cardiomyopathy characterized by persistence of fetal trabeculations in the LV.[1,2] The main clinical manifestations of this disorder are heart failure, arrhythmias and, to a lesser extent, systemic embolism.[3] While no abnormalities in epicardial coronary arteries have been demonstrated, myocardial ischemia could be present in patients with IVNC due to microcirculatory dysfunction. STEMI in IVNC is rare and is usually due to coexistent atherosclerosis. In this report, a 19-year-old male with previous diagnosis of noncompaction with no apparent risk factors for ischemic heart disease presented with anterior wall myocardial infarction. Coronary angiogram documented thromboembolic occlusion of distal LAD, which later resolved with medical management.

2. Case report

A 19-year-old male from India with no coronary risk factors presented to us with complaints of insidious onset gradually progressive exertional dyspnea, which progressed from NYHA class II–IV in over 4 months. There were frequent episodes of paroxysmal nocturnal dyspnea. There was no family history of significant cardiac diseases. Clinical examination revealed features of congestive cardiac failure in the form of bilateral pitting pedal edema, tender hepatomegaly, raised jugular venous pressure, cardiomegaly, and a left ventricular third heart sound. Patient was admitted to intensive care and started on diuretics. EKG showed normal sinus rhythm. A transthoracic echocardiogram showed all four chambers to be dilated. There was global hypokinesia of left ventricle with ejection fraction of 20%. Left ventricle showed numerous
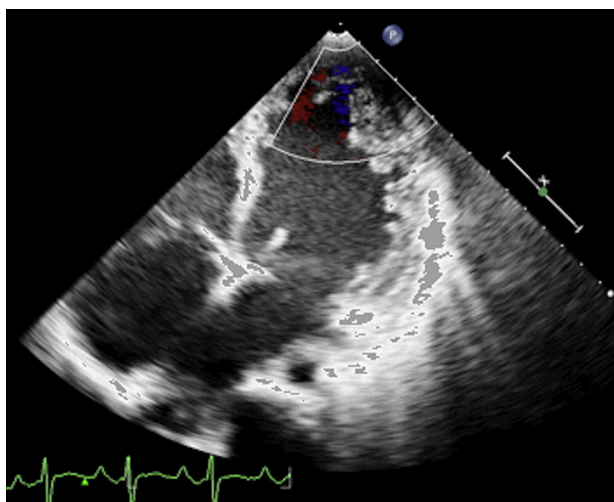
Fig. 1 – Transthoracic echocardiogram apical four-chamber view showing prominent trabeculations in apical and midlateral segments with deep intertrabecular recesses filled by direct blood flow from ventricular cavity on color Doppler.
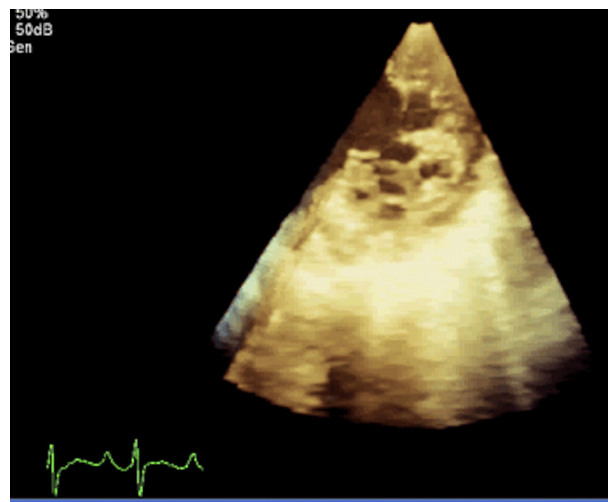

Fig. 2 – Three-dimensional short-axis view of left ventricle showing bilaminar structure with the ratio of noncompact subendocardial layer to compact subepicardial layer in systole of 3 in the apicolateral segment.

prominent trabeculations with deep intertrabecular recess, predominantly in the apical and midventricular areas of both the inferior and the lateral walls (Fig. 1). Parasternal short axis view showed two-layered structure of the myocardium with a thin, compacted outer (epicardial) band and a much thicker, noncompacted inner (endocardial) layer consisting of trabecular meshwork with deep endocardial spaces (Fig. 2). The maximal endsystolic ratio of the noncompacted endocardial layer to the compacted myocardium was 3. Color Doppler showed evidence of flow into these deep recesses. The patient was stabilized with parenteral diuretics and discharged on ramipril, carvedilol, torsemide, spirinolactone, and warfarin. He was lost to follow-up. Six months later, the patient presented with typical angina pain and EKG showed normal sinus rhythm with evidence of anterior wall myocardial infarction. Transthoracic echocardiogram confirmed features of IVNC with no evidence of clot in left ventricle. Coronary angiogram revealed thrombus in distal LAD with distal TIMI II flow. Other vessels were normal. The patient was given tirofiban bolus followed by infusion for 24 h and continued on heparin. Repeat angiogram after 3 days showed resolution of thrombus and good distal flow in LAD (Fig. 3). Patient was discharged on ramipril, carvedilol, torsemide, spirinolactone, and warfarin with target INR kept between 2 and 3. He was later referred for cardiac transplantation in view of recurrent heart failure.
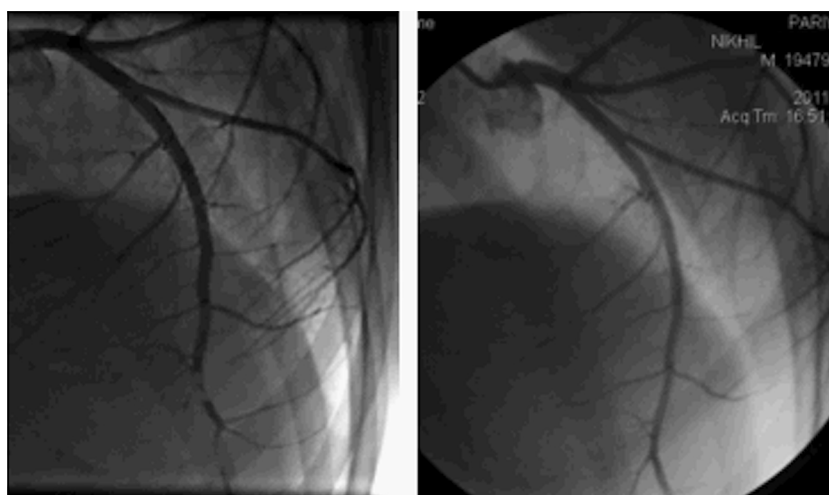

Fig. 3 – Coronary angiogram at presentation (on left) showing thrombus in distal LAD, which resolved completely with tirofiban and heparin, as shown in angiogram done after 3 days (on right).

3. Discussion

IVNC is a genetic disease caused by intrauterine arrest of compaction of the myocardial fibers, leading to the persistence of numerous deep trabeculations communicating with the ventricular cavity.[1] Although described as rare (with a prevalence in adults estimated at 0.014%), recent studies show that it is underdiagnosed.[2] The classical triad include heart failure symptoms, arrhythmias (supraventricular or ventricular), and arterial embolic events. Both familial (autosomal dominant/X-linked inheritance) and sporadic forms of noncompaction have been described.[3]

The diagnosis of IVNC can be made by two-dimensional and color Doppler echocardiography, as well as magnetic resonance imaging.[4,5] On transthoracic echo, multiple (>3) hypertrabeculations of the myocardium with deep intertrabecular recesses communicating directly with the left ventricular cavity are seen, resulting in two layers of myocardium with a ratio of noncompacted to compacted myocardium >2. The affected segments are usually located in the apical, midlateral, and midinferior regions and are often hypokinetic due to subendocardial hypoperfusion, diminished coronary flow reserve, and microcirculatory dysfunction.[6] In cardiac MRI, a diastolic quotient of more than 2.3 between the noncompact and compact layers is required for the diagnosis.[7] Prognosis remains poor for symptomatic patients with impaired systolic left ventricular function. Oechslin et al. in the largest data series followed 34 patients with IVNC over a mean period of 44 months; over this time, 53% developed heart failure, 41% developed a ventricular tachycardia, and 24% had thromboembolic events.[8] Management of patients with IVNC is similar to that of patients with other cardiomyopathies and should therefore include appropriate treatment for heart failure, management of arrhythmias including implantation of an ICD in high-risk patients, and oral anticoagulation. In a study of patients among whom a large percentage (60%) were anticoagulated, the frequency of thromboembolic complications was only 4%,[9] while the reported event rates in other studies were as high as 38%.[8]

Cardiac ischemia in IVNC is due to microvascular dysfunction. Jenni et al.[10] have shown that IVNC patients exhibit decreased coronary flow reserve in both compacted and noncompacted segments. Approximately half the cases of IVNC have reversible ischemia on myocardial perfusion scans. There are some case reports that have shown an association between IVNC and CAD.[11,12] Panduranga and Mukhaini[11] reported IVNC predominantly involving septum and associated with two-vessel coronary artery disease. In another report, a patient with IVNC presented with acute inferolateral STEMI but angiogram revealed normal epicardial coronary arteries.[13] Güvenç et al.[14] reported myocardial infarction in a young IVNC patient without any identifiable cause, speculating thromboembolism as the probable cause. The current one is a rare case of STEMI with documented coronary thromboembolism in a prospectively followed up patient with IVNC.

Conflicts of interest

The authors have none to declare.

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