

Myocardial infarction in a patient with left ventricular noncompaction: a case report

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Abstract: We describe a 73-year-old male patient with left ventricular noncompaction (LVNC) who was diagnosed with acute myocardial infarction (MI), three-vessel coronary artery disease, a fresh intraventricular thrombus, and mitral regurgitation. He was treated with full anticoagulant therapy, coronary artery bypass grafting, and mitral valve repair. This case adds to a small but growing literature showing association between LVNC and MI and/or coronary artery disease. We suggest that patients with LVNC could be considered at heightened risk for MI, and the two conditions might have a common genetic underpinning in some cases.

Keywords: myocardium, development, cardiomyopathy, echocardiography

Introduction

Left ventricular noncompaction (LVNC) arises during development of the myocardium after week 18 of gestation, and is sometimes associated with other congenital cardiac abnormalities.^{1,2} It is characterized by deep endothelium-lined trabeculations, identifiable by colored Doppler imaging,³ that are continuous with the lumen of the left ventricle and give the ventricular wall a two-layered appearance (noncompacted and compacted). Echocardiography showing recesses in the wall, especially around the apex, is diagnostic for this type of cardiomyopathy, though magnetic resonance imaging is useful when no echocardiogram is available.^{4,5} Nevertheless, LVNC can be mistaken for other conditions, such as dilated, hypertrophic, and apical hypertrophic cardiomyopathies,⁶ or infiltrative disease such as cardiac amyloidosis,⁷ and LV opacification might help to confirm the diagnosis.⁷ The incidence of LVNC is uncertain,³ but has been estimated at 0.12 per 100,000 in children younger than 10 years.⁸

As with other cardiomyopathies, depressed systolic and diastolic functions are typical, and tachyarrhythmias are common; there is a predisposition to arterial embolism.⁸ Thus, patients are likely to complain of breathlessness, fatigue, and peripheral edema. Electrocardiogram features include extreme QRS voltages, isolated T waves, and evidence of Wolff–Parkinson–White syndrome, with premature contraction of both atria and ventricles.⁹

LVNC is familial and is now considered a primary genetic cardiomyopathy, though phenotypic features can differ among members of the same family. Patient cohorts show a high frequency of mutation in the gene *Cypher/ZASP*, which encodes a component of the Z line of cardiac muscle; histologically, individuals with *Cypher/ZASP* mutations show disordered muscular cytoskeletons.¹⁰ Other genes reported to be mutated in cases of LVNC include those encoding myoadenylate deaminase,¹¹

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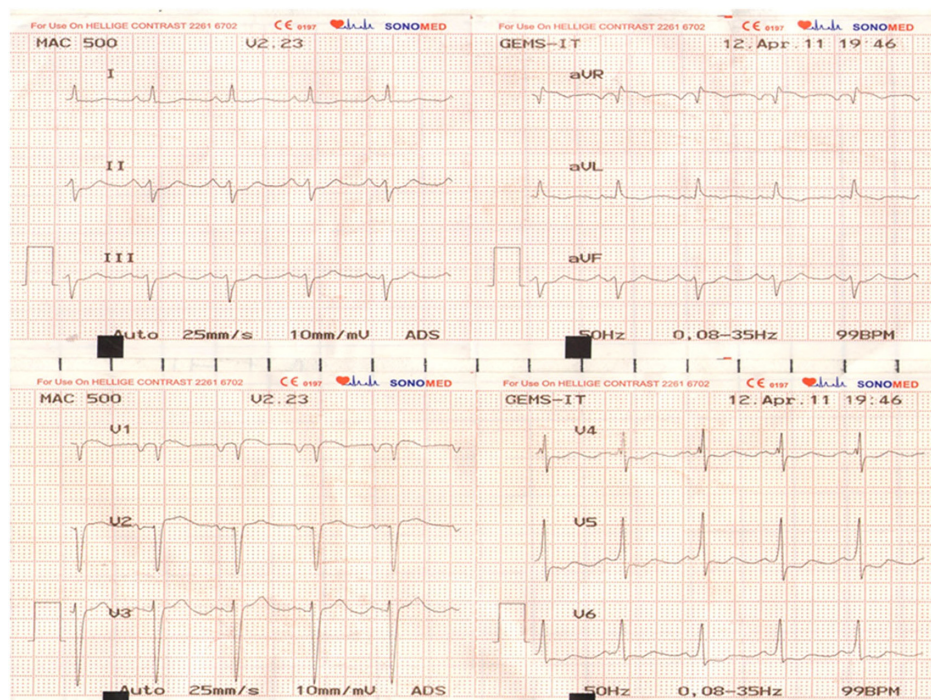


Figure 1 Electrocardiogram of the patient with nonspecific changes; left axis deviation of QRS, ST-segment elevation in augmented vector right (aVR) and V₁, poor R-wave progression in right precordial leads, ST-segment depression in left precordial leads as evidence of ischemia.

cytochrome P450 2C9,¹² cardiac α -actin,¹³ troponin T,¹⁴ and other sarcomere-related proteins.^{15–17}

We describe a case of LVNC associated with myocardial infarction (MI) in a 73-year-old man.

Case report

A 73-year-old man was admitted with typical chest pain and exertional dyspnea (New York Heart Association functional

class III), with the diagnosis of acute MI. Clinical examination revealed an apical systolic murmur. Figure 1 shows an electrocardiogram of the patient, demonstrating nonspecific changes: left-axis deviation of QRS, ST elevation in aVR and V₁, poor R-wave progression in right precordial leads, ST depression in left precordial leads as evidence of ischemia. Transthoracic echocardiography revealed apical left ventricular hypertrabeculation characteristic of left ventricular

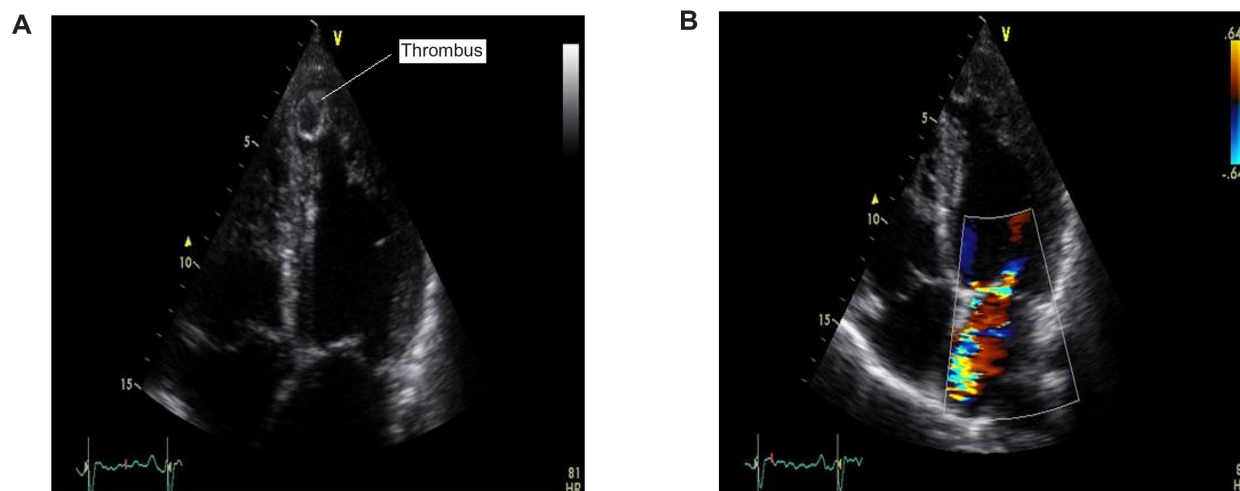


Figure 2 Transthoracic echocardiography showing apical akinesis with increased trabecularization and thrombus (A), and color Doppler echocardiography revealing severe mitral regurgitation (B). Apical thrombus is shown by arrow in A.

noncompaction, akinesis in a small area of the apex, and fresh thrombus (Figure 2A), and color Doppler echocardiography showed partially flail mitral valve with severe mitral regurgitation (MR) (Figure 2B). Coronary angiography showed three-vessel coronary artery disease; the patient received full anticoagulant therapy. Follow-up transthoracic (Figure 3A) and transesophageal (Figure 3B) echocardiography 10 days later showed resolving apical thrombus.

The patient underwent coronary artery bypass grafting and mitral valve (MV) repair. The patient received three

grafts, and the MV was repaired by insertion of an annuloplasty ring in MV annular position. He was discharged 8 days after operation with residual mild MR, and left ventricular ejection fraction of 35% indicating a good surgical result.

Discussion

LVNC was first identified in a 33-year-old woman in 1984 by Engberding and Bender,¹⁸ who supposed that the trabeculae were sinusoids. It was long thought to be predominantly a disease of children, but by the late 1990s there were

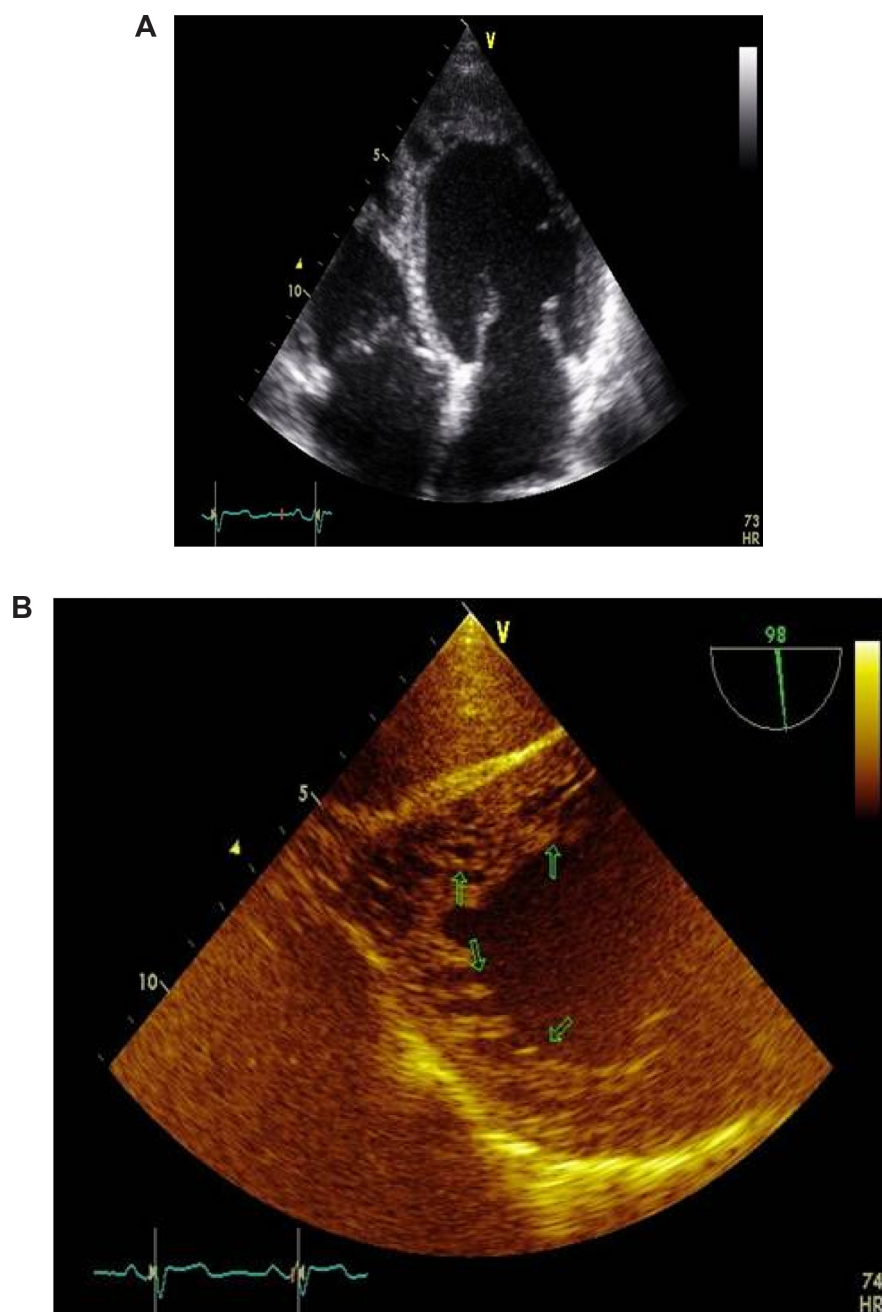


Figure 3 Follow-up transthoracic (A) and transesophageal (B) echocardiography showing resolution of apical thrombus.

increasing numbers of reports in adults.¹⁹ Some authorities now suppose it could be more common than hitherto suspected because it may have been misdiagnosed in the past.⁴ Magnetic resonance imaging (MRI) is now considered the gold standard for the diagnosis of this entity. However, MRI is usually used when results of other imaging modalities are not definitive.²⁰ In the present report, LVNC was diagnosed based on the echocardiographic assessment; therefore, MRI was not applied. Moreover, LVNC is usually managed in the same way as other cardiomyopathies; beta-blockers, aspirin, and angiotensin-converting enzyme inhibitors are customarily used; installation of a pacemaker is an option if the risk of arrhythmia is high.⁸ In the case of our patient, these approaches to management were deemed unnecessary.

LVNC can be associated with other cardiac and neuromuscular pathologies.^{21,22} Mitral regurgitation and coronary heart disease have been reported in some patients.²³ Although association of LVNC with coronary heart disease is said to be rare,²⁴ we found four recent reports of association between LVNC and subclinical or acute MI,^{25–28} and it is possible that this association has previously been undiagnosed. The recognized pathogenic consequences of LVNC could predispose to MI, and our patient may exemplify such predisposition. Moreover, LVNC may affect the progression of remodeling in patients with MI and may worsen their prognosis. Further studies are recommended to focus on this hypothesis.

It is worth considering the possibility of common genetic underpinning of LVNC and MI in some cases. For instance, the myocardial adenylate deaminase C34T genotype predicted mortality in some patients with histories of MI,²⁹ and there are suggestions of association between MI and both cytochrome P450 2C9 activity and sarcomere-related proteins such as the calcium-binding protein S100A.^{30,31} One may suggest that when LVNC is diagnosed and the genes for one or more of these proteins are mutated, an increased risk for MI should be suspected. However, the present report lacks the genetic evaluation, and such workup is recommended in future reports of MI in patients with LVNC.

Disclosure

The authors report no conflicts of interest in this work.

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