

CARDIOMYOPATHY

Delayed Enhancement Cardiac MR Imaging in Noncompaction of Left Ventricular Myocardium

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ABSTRACT

A 49-year-old female with a history of paroxysmal atrial fibrillation, presented with worsening dyspnea on minimal exertion. During the follow-up period, transthoracic echocardiography and cardiac magnetic resonance imaging (CMR) were consistent with the diagnosis of noncompaction of the left ventricle. Delayed-enhancement CMR demonstrated hyperenhancement of the prominent trabeculations located at the mid and apical portions of the left ventricle, suggesting areas of fibrosis. Although previous cases of left ventricular noncompaction diagnosed with CMR have been described in the literature, this is the first case to describe the utility of delayed-enhancement imaging in the pathohistological confirmation of myocardial fibrosis and scarring in the hypertrabeculated myocardium.

CLINICAL SUMMARY

A 44 year-old female with a history of paroxysmal atrial fibrillation, presented with worsening dyspnea on minimal exertion. She denied symptoms of orthopnea, palpitations, syncope, infection or any risk factors for thromboembolism.

The patient was afebrile and normotensive and had no signs of system infection. The cardiorespiratory examination was remarkable for an elevated jugular venous pressure at the angle of the jaw, a left-sided third heart sound, and no murmurs. There was no calf or thigh swelling or tenderness.

The complete blood count, electrolyte level, liver function results, cardiac enzyme level, coagulation parameters, and urinalysis were within normal limits. Electrocardiography revealed normal sinus rhythm with a normal chest radiograph.

Transthoracic echocardiography revealed normal left ventricular dimensions, mildly reduced ejection fraction of 50%, and heavy trabeculations extending from the midcavity region to the apex (Fig. 1A). Color Doppler imaging of the apical region demonstrated blood flow through the deep intramyocardial recesses and trabeculations (Fig. 1B), confirming the diagnosis of noncompaction of the left ventricular myocardium (LVNC).

Cardiac cine-MRI technique showed prominent trabeculations in the mid and apical left ventricular levels further supporting the diagnosis (Fig. 2A). Delayed-enhancement inversion recovery MRI was performed after 10 minutes of 0.2 mmol/kg injection of Gadolinium (Gd-DTPA) and interestingly demonstrated hyperenhancement of the prominent trabeculations at the mid and apical portions of left ventricle, suggesting areas of fibrosis (Fig. 2B).

In follow-up, the patient demonstrated sustained ventricular tachycardia on an outpatient Holter monitor, and subsequently had implantation of an intracardiac defibrillator in the setting of LVNC.

DISCUSSION

Isolated noncompaction of the left ventricular myocardium is a rare diagnostic entity and remains as an unclassified cardiomyopathy by the World Health Organization Task Force (1). Clinical presentation of this rare cardiomyopathy varies,

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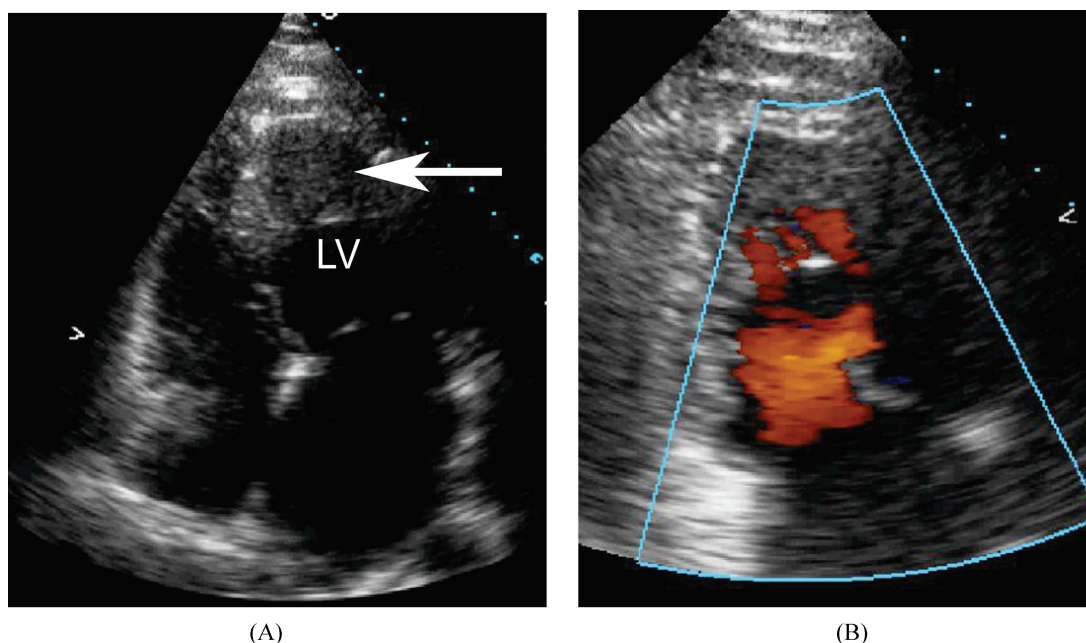


Figure 1. (A) Apical 4-chamber view on transthoracic echocardiography illustrating the heavily trabeculated mid to apical LV cavity. (B) Color Doppler imaging of the left ventricle in an apical 2-chamber view demonstrating color flow within the deep intratrabecular recesses of the noncompacted left ventricle.

ranging from asymptomatic left ventricular dysfunction to complications of congestive heart failure, thromboembolism and arrhythmias (2). It is associated with arrest of normal embryogenesis of both the endocardium and myocardium (3). Patho-

histological findings of the hypertrabeculated region may reveal myocardial disorganization, presence of fibrosis and scarring of the myocardium, all likely due to an inadequate blood supply (3).

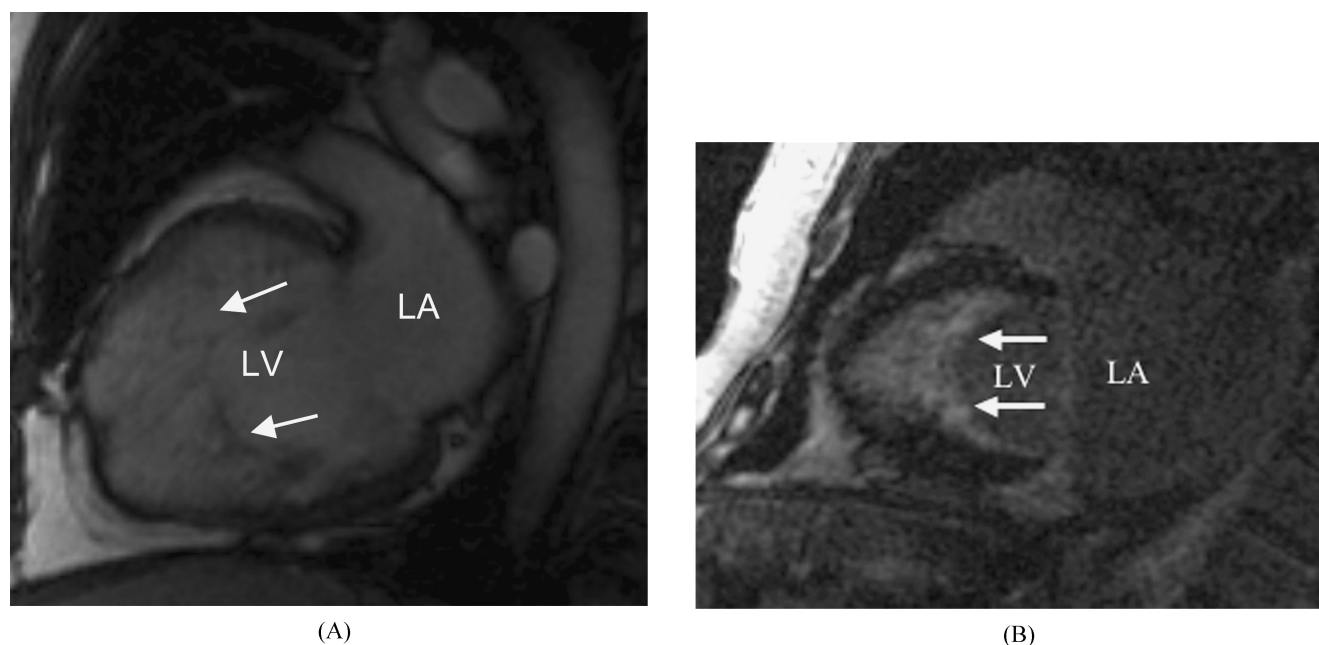


Figure 2. (A) Cine-MRI 2-chamber view in end-diastole demonstrates left ventricular dilatation and prominent trabeculations in the mid and apical regions (arrows). LV, left ventricle; LA, left atrium. (B) Delayed-enhancement MRI 2-chamber view demonstrates hyperenhancement of the trabeculations within the left ventricle, suggesting fibrosis (arrows). LV, left ventricle; LA, left atrium.

The diagnosis of LVNC is best screened by transthoracic echocardiography, demonstrating prominent ventricular trabeculations and deep intertrabecular recesses, left ventricular dilation and reduced systolic function (3, 4). The diagnostic echocardiographic criteria for LVNC include: (1) two-layered wall structure with a compacted thin epicardial band and thicker noncompacted endocardial layer of trabecular meshwork with deep endomyocardial spaces (ratio of noncompacted to compacted thickness >2); (2) localization of the trabeculations in the mid-lateral, apical and mid-inferior segments; and (3) color Doppler flow in deep perfused intertrabecular recesses (3, 4).

Cardiac magnetic resonance imaging (CMR) may provide complementary information in delineating the anatomy due to its high spatial resolution, in particular when noncompacted myocardium is confined to the left ventricular apex or in cases with poor echocardiographic windows (5). A trabecular mass greater than 20% of the total myocardial mass has been recently suggested as a useful index for the diagnosis of LVNC on CMR (6).

The technique of delayed enhancement cardiac magnetic resonance imaging (DE-CMR) has been described in several ischemic and non-ischemic cardiomyopathies to detect irreversible myocardial injury (myocardial necrosis or fibrosis) (7). A review of the literature published between 1966 and 2005 revealed only one previously described case in which CE-CMR was negative in detecting intramyocardial fibrosis in an asymptomatic patient with LVNC (6). To our knowledge, this is the first time that delayed-enhancement MRI is described to allow the visualization of hyper-enhancement within the trabeculations of left ventricular noncompaction, suggesting fibrosis. It is plausible that these areas of fibrosis with hyperenhancement may serve as an important marker of a high-risk patient by serving as a substrate for lethal ventricular arrhythmias as seen in our case.

CONCLUSION

Cardiac MRI may thus serve as an important comprehensive noninvasive tool in the evaluation of patients with left ventricular noncompaction in not only confirming the presence of recesses and hypertrabeculations in the left ventricle through Cine-MRI, but also demonstrating the high risk patient with fibrosis within these trabeculations through Delayed-enhancement imaging.

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