



Cardiac MRI in a Patient with Coincident Left Ventricular Non-Compaction and Hypertrophic Cardiomyopathy

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Received 04 August 2010; Accepted 13 October 2010

Abstract

Left ventricular non-compaction cardiomyopathy is a rare congenital cardiomyopathy that affects both children and adults. Since the clinical manifestations are not sufficient to establish diagnosis, echocardiography is the diagnostic tool that makes it possible to document ventricular non-compaction and establish prognostic factors. We report a 47-year-old woman with a history of dilated cardiomyopathy with unknown etiology. Echocardiography showed mild left ventricular enlargement with severe systolic dysfunction ($EF = 20-25\%$). According to cardiac magnetic resonance imaging findings non-compaction left ventricle with hypertrophic cardiomyopathy was considered, and right ventricular septal biopsy was recommended. Right ventricular endomyocardial biopsy showed moderate hypertrophy of cardiac myocytes with foci of myocytolysis and moderate interstitial fibrosis. No evidence of infiltrative deposition was seen.

J Teh Univ Heart Ctr 2011;6(4):214-216

This paper should be cited as: Alizadeh-Sani Z, Madadi S, Sadeghpour A, Khajali Z, Golnari P, Kiavar M. Cardiac MRI in a Patient with Coincident Left Ventricular Non-Compaction and Hypertrophic Cardiomyopathy. *J Teh Univ Heart Ctr 2011;6(4):214-216.*

Keywords: Magnetic resonance imaging • Cardiomyopathies • Heart defect, congenital

Introduction

Left ventricular non-compaction cardiomyopathy (LVNC), also called spongiform cardiomyopathy, is a rare congenital cardiomyopathy that affects both children and adults.¹ It results from the failure of myocardial development during embryogenesis.^{2, 3} The fact that LVNC has only recently been established as a diagnosis and that it is still unclassified as a cardiomyopathy according to the WHO means that it is not fully understood how common the condition is. Moreover, there have hitherto been not large population studies into the disease and those conducted with small sample sizes have been based primarily upon patients suffering from advanced heart failure. In the largest series

of patients with LVNC, the prevalence was 0.014% of the patients referred to the echocardiography laboratory.⁴ This low number of the reported cases is in consequence of the absence of large population studies; a similar situation occurred with hypertrophic cardiomyopathy (HCM), which was initially considered very rare, but is now thought to occur in one in every 500 people in the population.⁵ It is deserving of note, however, that this has been the subject of intense scrutiny and investigation for over 40 years.⁶ A ratio of non-compacted to compacted myocardium greater than 3 and involvement of three or more segments are indicators of a poor prognosis. Since the clinical manifestations are not sufficient to establish diagnosis, echocardiography is the diagnostic tool that makes it possible to document ventricular

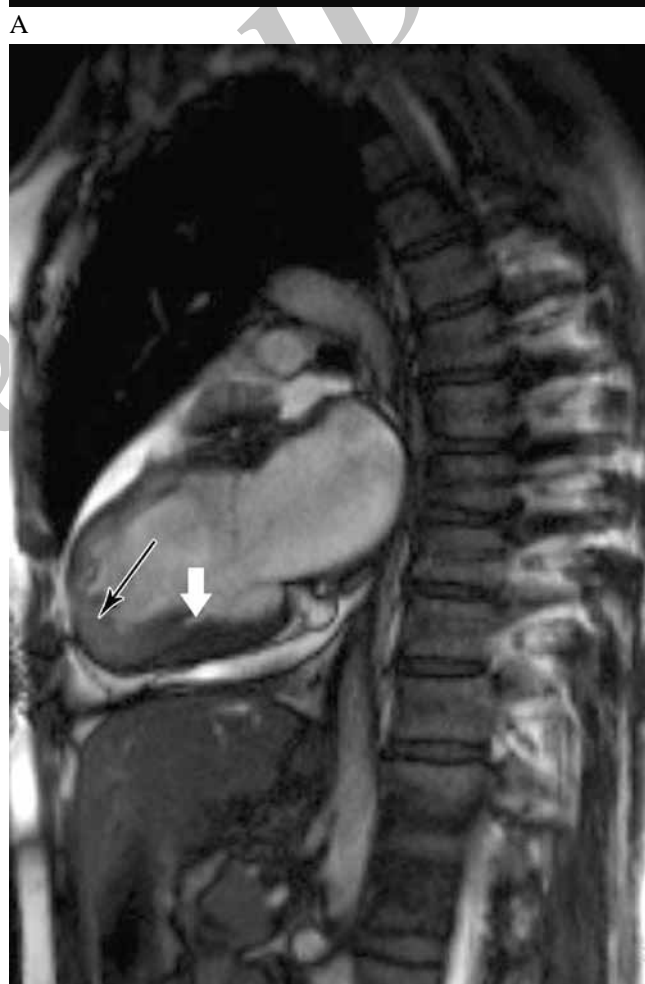
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non-compaction and establish prognostic factors.²

Case Report

A 47-year-old woman was referred to our hospital with a history of dilated cardiomyopathy (DCM), diagnosed by angiography, with unknown etiology. The patient complained of a prolonged history of weakness and palpitation. Her two children had died because of cardiomyopathy. In physical examination, S3 was heard in heart auscultation, with no inspiratory crackles and no peripheral edema. Electrocardiogram showed a normal sinus rhythm, with left-axis deviation, right bundle branch block, and first-degree atrioventricular block. Echocardiography showed a mild left ventricular (LV) enlargement with severe systolic dysfunction (EF = 20 - 25%), grade I LV diastolic dysfunction, normal right ventricular (RV) size with moderate RV dysfunction, hypertrabeculated LV apex (highly suspicion of LVNC), mild left atrial enlargement, mild mitral regurgitation (MR), moderate tricuspid regurgitation (TR), mild pulmonary insufficiency, mild to moderate pulmonary arterial hypertension (systolic pulmonary arterial pressure = 40 - 45 mmHg), plethoric inferior vena cava, and small pericardial effusion. A cardiac magnetic resonance imaging (CMR) study was ordered for risk stratification purposes. CMR was done with 1.5 T Siemens® Avanto® using protocols for functional and flow study with T1, retro IPAT, magnitude, and phase sensitive techniques on different planes, two chambers, three chambers, and four chambers. Short-axis views and MR angiography with Gadolinium revealed biatrial enlargement, obvious MR and TR, mild LV enlargement with moderately reduced systolic function (EF = 37%), localized mid lateral and basal inferior segmental LV hypertrophy (thickness = 15 mm) with bilayered appearance of the RV free wall and LV anterior, and basal lateral and apical segments with a diastolic ratio of non-compacted to compacted myocardial wall of 4 - 5 (Figure 1). In the late enhancement images, a diffuse circumferential mid myocardial stripe of scar at the anterior, septal, and anteroseptal segments and also at the junction of the RV inferior and septal walls was found (Figure 2). According to the CMR findings, biventricular LVNC with HCM was considered and RV septal biopsy was recommended. Tissue Doppler echocardiography was done to evaluate the suitability of cardiac resynchronization therapy (CRT), and it revealed that the patient was an ideal case for CRT (regarding significant delay in the inferolateral segments). Due to frequent arrhythmia, an implantable cardioverter-defibrillator CRT (ICD - CRT) was implanted. The RV endomyocardial biopsy showed moderate hypertrophy of myocytes with foci of myocytolysis and moderate interstitial fibrosis. No evidence of infiltrative deposition was seen.



B

Figure 1. Cardiac magnetic resonance imaging view of four-chamber (A) and two-chamber (B) images with steady-state free precession (SSFP) sequences showed hypertrophy of the mid lateral and basal inferior segments (thick white arrows) with a maximum thickness of 15 mm and presence of a non-compaction process involving mainly the left ventricular basal lateral and apical segments together with right ventricular free wall segments (thin black arrows)

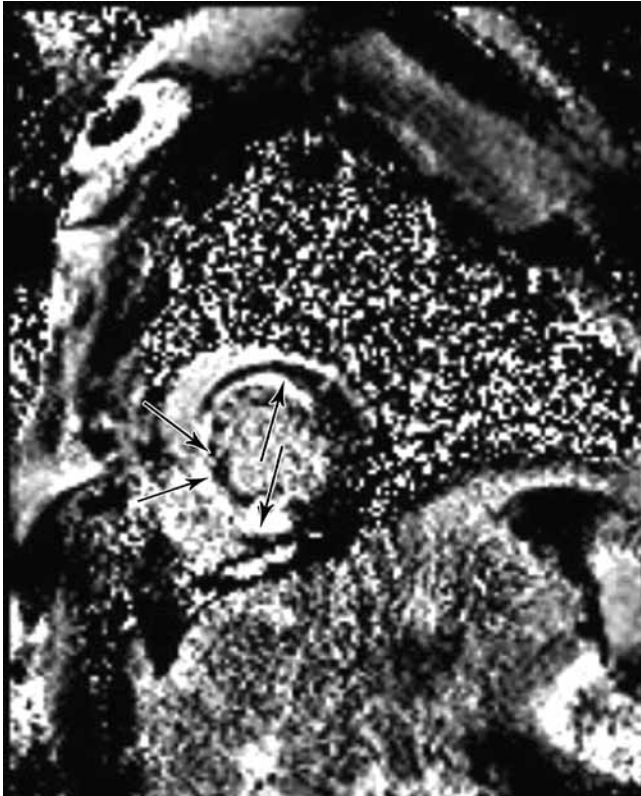


Figure 2. Cardiac magnetic resonance imaging delayed contrast enhancement sequences showed mid myocardial enhancement at the level of anterior, inferior, and septal segments (arrows)

Discussion

Our CMR study demonstrated the presence of definite signs of LVHCM with both ventricular non-compaction cardiomyopathy accompanied by noticeable intramyocardial fibrosis and reduced ventricular function. Anecdotal cases of LVNC being reported in the relatives of patients known to have HCM⁷ and detecting the criteria of HCM in 2 out of 73 patients with an echocardiographic diagnosis of LVNC⁸ show an association between these two. A rationale for this association seems to lie in the finding that particular gene mutations known to cause familial HCM have been also described in families with LVNC.⁹ Independently of which nomenclature is used for the cardiac disease, either apical HCM or LV NC, the clinical phenotypes in the affected individuals appear to be similar in their genetically programmed morphological abnormalities and characterized by a unique high penetrance.¹⁰ Hence, controversy still exists over whether LVNC is a discrete disease entity or not.¹¹

A distinguishing feature of the present case is the presentation of LVNC involving the LV, with an extremely thin compacted portion at this level, whereas most of the described HCM-LVNC dual pathology have been reported in cases with apical HCM.¹² In dilated cardiomyopathy,

the LV walls become thin and paper like, whereas in the patient presented herein localized non-apical myocardial hypertrophy with hypertrabeculation in some other segments was seen.

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