

Case report

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Treatment of left ventricular non-compaction with cardiac resynchronization therapy

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Background

Left ventricular non-compaction (LVNC) is a genetic cardiomyopathy often familial and autosomal dominant. It is characterized by morphological abnormalities affecting the left ventricular myocardium with prominent trabeculations of the inner surface of the ventricle, often extending deep into the ventricular wall. There are no pathognomonic histological findings with normal myocytes being interspersed with areas of fibrosis. Both familial and sporadic forms of non-compaction have been described, the prevalence being estimated to be between 0.01% and 0.27%.¹ The diagnosis of LVNC calls for multimodal imaging. Echocardiography, being the most widely available cardiac imaging modality, may raise the initial suspicion of LVNC and can also provide physiological data. See Table 1. Cardiac Magnetic Resonance Imaging (CMR) offers detailed visualization of the extent of non-compaction and supplemental morphological information.⁴ It should be noted that the current criteria for this condition may result in over diagnosis.

A correct diagnosis is important both for subsequent treatment and also to enable appropriate genetic counselling and familial screening. It is suggested that first degree relatives should be screened. CMR is the best imaging modality

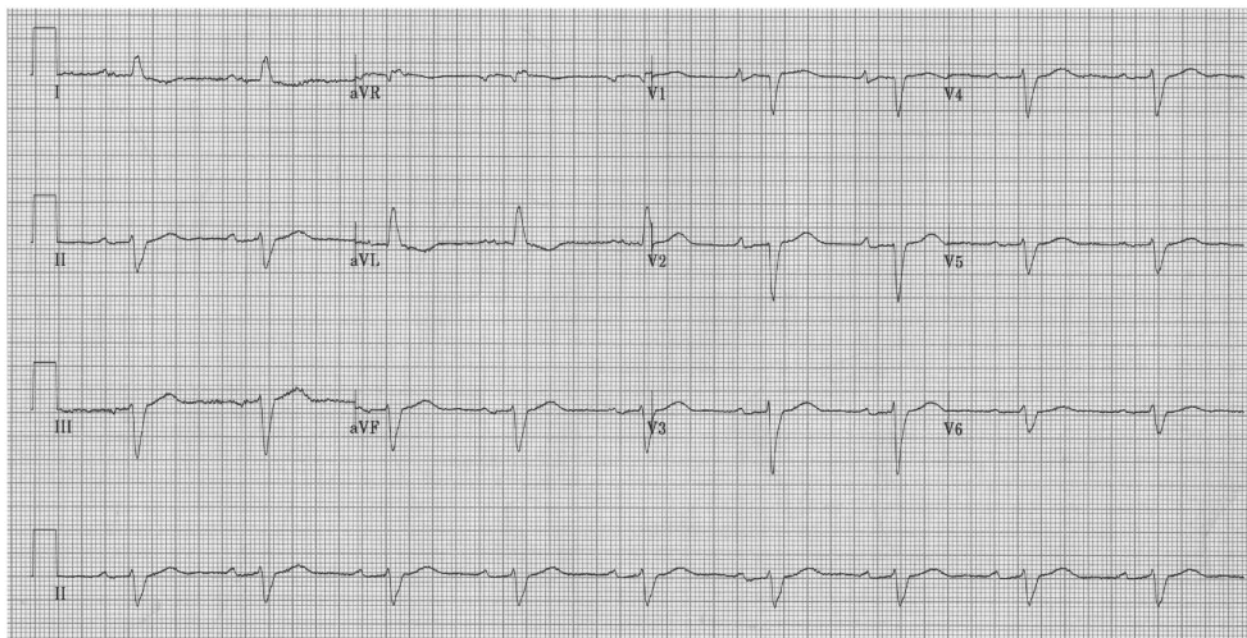
currently, though other techniques in the future may be able to improve on diagnostic accuracy. LVNC is associated with LV systolic dysfunction, due to subendocardial hypoperfusion and microcirculation dysfunction, and at a lesser extent to diastolic dysfunction, ventricular arrhythmias, sudden cardiac death and systemic embolism.

There is no specific treatment for LVNC at present, with the mainstay of therapy being appropriate medication for heart failure and anticoagulation for thromboembolism. The increased risk of systemic cardiac embolism with LVNC has been reported in retrospective studies, which is thought to be due to the formation of thrombi within the inter-trabecular recesses.¹ As LVNC is also associated with a significant prevalence of atrial fibrillation (as in our case), the source of emboli may be either the left atrial appendage or the inter-trabecular recesses. Patients with LVNC sustain an increased propensity to ventricular arrhythmias and sudden cardiac death. After risk stratification, implantable cardioverting defibrillator (ICD) implantation should be considered in isolation or in combination with Cardiac Resynchronisation Therapy (CRT).

CRT has specific pre-implantation criteria.^{5,6} However, there is a lack of evidence with regards to the specific role of CRT in the management of LVNC and CRT. Limited single institution case

Table 1 Criteria for the diagnosis of isolated left ventricular non-compaction

Echocardiography ²	Cardiovascular magnetic resonance ³
Presence of multiple echocardiographic trabeculations, particularly in the apex and free wall of the left ventricle	Diastolic steady-state free precession cine frames to determine the ratio of thickness of the trabecular and compact layers.
Multiple deep intertrabecular recesses communicating with the ventricular cavity, as demonstrated by colour Doppler imaging	The diastolic ratio of 2.3 shows high diagnostic accuracy for distinguishing pathological LVNC from the degrees of non-compaction observed in healthy, dilated and hypertrophied hearts.
A two-layered structure of the endomyocardium with a ratio of end-systolic non-compacted endocardial layer to compacted layer of >2.0 in adults	
Absence of other congenital or acquired heart disease, particularly those causing left ventricular outflow obstruction	

**Figure 1.** The 12-lead resting electrocardiogram (ECG) showing normal sinus rhythm and a broad QRS duration of 146 ms with a left bundle branch block (LBBB) morphology and a normal QT interval.

reports and series show improvement in functional class and LV systolic function post CRT implantation.^{7,8} Our case concurs with these findings, and draws attention to the need for inclusion of LVNC in larger CRT trials. Our case is also unique in that it presents all of the associated imaging.

Case presentation

A 56-year-old Caucasian female was transferred as an inpatient to our institution, for the management of her heart failure and cardiogenic shock. Her past medical history included the diagnosis of a stromal uterine cancer at the age of 40 years, which was

treated with hysterectomy, bilateral salpingo-oophorectomy and pelvic radiotherapy. She had a relapse with a solitary pulmonary metastasis 3 years later for which she received anthracycline-based neoadjuvant chemotherapy (6 cycles adriamycin and ifosfamide) followed by a right lung lobectomy. She has been in remission since. Four months prior to her current admission, the patient presented to her local hospital with an ischaemic middle cerebral artery stroke. This was treated with Tenecteplase resulting in almost complete neurological recovery, the residual deficit being a mild right-sided hemiparesis.

As part of her investigations, she had a transthoracic echocardiogram (TTE), which demonstrated

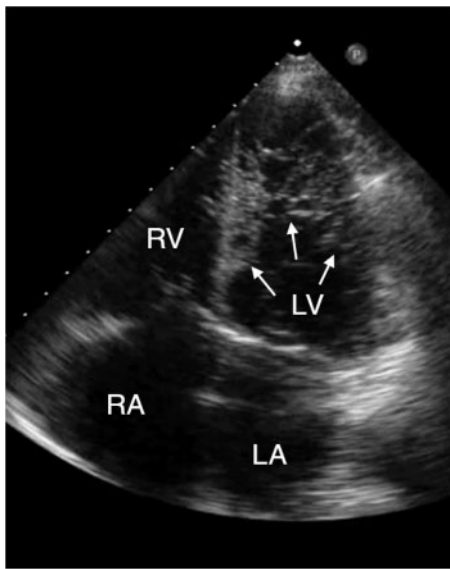


Figure 2. Transthoracic echocardiogram confirmed severe biventricular impairment (LV end diastolic dimension 5.9 cm, end systolic dimension 5.0 cm LVEF 15% and right ventricular dilatation; inlet dimensions 4.3 cm, mid cavity 4.8 cm.) Additionally the LV was heavily trabeculated (white arrows), particularly at the lateral, anterior and apical segments, suggestive of LVNC.

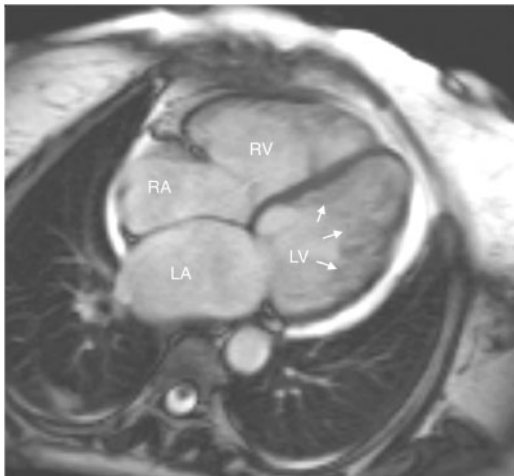


Figure 3. A CMR scan with Gd contrast was performed that was also highly suggestive of LV non-compaction (arrows) in the setting of severe biventricular systolic dysfunction and dilatation.

severe biventricular impairment with a left ventricular ejection fraction (LVEF) of 15% (Biplane Simpson's methodology). A probable left ventricular (LV) mural thrombus was also visible for which she was anticoagulated with warfarin.

Additionally, standard heart failure pharmacotherapy (beta blockade and an ACE-inhibitor) were commenced. However, dosages were limited by



Figure 4. CMR scan: early Gd enhancement demonstrated a left atrial appendage thrombus (white arrow).

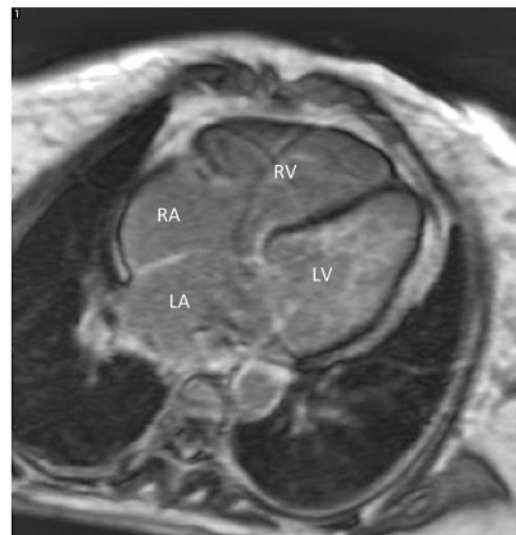


Figure 5. CMR scan: delayed phase Gd enhancement showed diffuse uptake in the non-compacted layers.

symptomatic hypotension. Following discharge she had two subsequent admissions to her local hospital with decompensated heart failure and paroxysmal rapid atrial fibrillation.

On the third admission, she presented with oliguria, hypotension and pulmonary oedema culminating in cardiogenic shock requiring inotropic support and concomitant intravenous loop diuretic therapy. She was transferred to our institution, a tertiary cardiac centre, for advanced heart failure management.

On admission to our hospital she was hypotensive with clinical evidence of biventricular impairment requiring ongoing intravenous loop diuretic therapy. Her 12 lead electrocardiogram (ECG) showed sinus

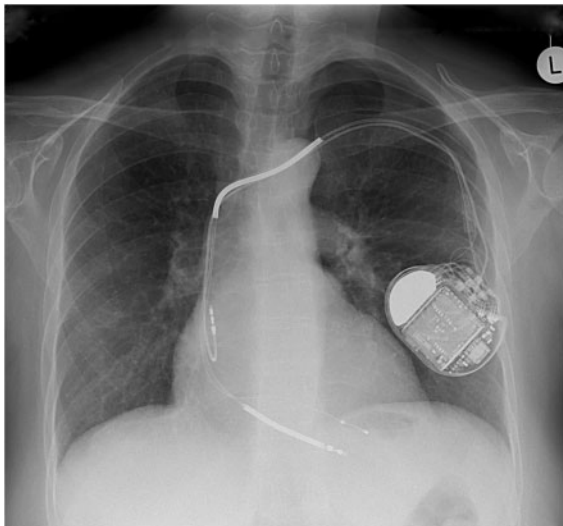


Figure 6. Posteroanterior Chest radiograph demonstrating cardiac resynchronization therapy and defibrillator in situ.

rhythm and a broad QRS duration of 146 ms with a left bundle branch block (LBBB) morphology (Figure 1). B-Type natriuretic peptide (BNP) was elevated at 386 pmol/l (upper limit of normal being 4 pmol/l). A repeat transthoracic echocardiogram confirmed severe biventricular impairment (LV end diastolic dimension 5.9 cm, end systolic dimension 5.0 cm LVEF 15% and right ventricular dilatation; inlet dimensions 4.3 cm, mid cavity 4.8 cm). Additionally the LV was heavily trabeculated, particularly at the lateral, anterior and apical segments, suggestive of left ventricular non-compaction (Figure 2). A cardiac magnetic resonance (CMR) scan with Gadolinium (Gd) contrast was performed, which was also highly suggestive of LV non-compaction in the setting of severe biventricular systolic dysfunction and dilatation. Early Gd enhancement demonstrated a left atrial appendage thrombus and the delayed phase showed diffuse uptake in the non-compacted layers (See Figures 3–5).

Despite maximally tolerated pharmacological therapy for heart failure, she remained in New York Heart Association (NYHA) functional class III. Hence, she fulfilled current guidelines for the implantation of cardiac resynchronisation therapy (LVEF <35%, QRS duration >120 ms, Optimal Tolerated Medical Therapy) (Figure 6),⁵ which was performed. Device implantation led to marked improvement in the patient's symptomatic status (NYHA functional class II) and facilitated uptitration of the patient's heart failure medication following discharge.

One month post discharge, repeat TTE showed improvement in LVEF and LV dimensions (LVEF, 40%; left ventricular end diastolic diameter (LVEDD), 5.2 cm; left ventricular end systolic diameter (LVESD), 4.1 cm) and the BNP level had reduced to 13 pmol/l. This correlated with further recovery in her clinical status with a resolution of her heart failure symptoms (NYHA class I) and reduction in oral loop diuretic requirements.

Conclusions

This case of LVNC highlights the potential cardiac and extracardiac complications of this genetic cardiomyopathic process. It illustrates the need for multimodal cardiac imaging. The case demonstrates the complex management of this condition with particular focus on anticoagulation and heart failure management including cardiac resynchronization therapy.

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Conflict of interest: None declared.

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