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Cardiac resynchronization therapy in a patient with left ventricular non-compaction cardiomyopathy: A case report

Juan Wang, Dasheng Lu, Mingchao Zhang, Yunong Han, Lingfei Yang

ABSTRACT

Introduction: Left ventricular non-compaction cardiomyopathy (LVNC) is a rare and specific type of congenital cardiomyopathy with an incidence rate between 0.05% and 0.24%. Dilated cardiomyopathy (DCM) and ischemic cardiomyopathy have poor prognosis. Cardiac resynchronization therapy (CRT) improves cardiac function and long-term prognosis in patients with heart failure with left bundle branch block, but its efficacy on LVNC is uncertain.

Case Report: This paper reported a case of a 57-year-old male with LVNC who was hospitalized for complaining of palpitations and shortness of breath on exertion and then diagnosed as heart failure. Electrocardiogram revealed a sinus rhythm with a QRS duration of 164 ms, as well as left bundle branch block morphology. Chest X-ray showed marked cardiomegaly and mild pulmonary congestion, and ultrasound echocardiography showed abnormal echo of left ventricular myocardium. The myocardium was cavernous sinus-like, forming sinusoidal cavities in varying sizes, communicating with left ventricular cavity, with the ratio of non-compacted to compacted myocardium of 3:1. The patient featured a large left ventricular end-diastolic diameter of 91 mm, and the function was severely impaired with an ejection fraction (EF) of 31%. After diuretic treatment, improvement of myocardial remodeling, heart strengthening, vasodilation, etc., he recovered gradually and was discharged 20 days later. However, the patient was admitted to hospital again due to shortness of breath within one month without obvious inducement. Considering the poor effect of drug therapy, we recommended that the patient undergo cardiac resynchronization therapy, he recovered and was discharged seven days after surgery. During the 6-month follow-up period, his left ventricle (LV) function and clinical symptoms showed significant and sustained improvement. B-type natriuretic peptide (BNP) levels decreased from 4836.79 to 1253.18 pg/mL. QRS duration (164 ms) on ECG significantly decreased after CRT.

Conclusion: We reported the management of a rare case of congenital LVNC with recurrent heart failure and frequent premature ventricular contractions. Patients with LVNC have clear indications and poor pharmacological response. Early cardiac resynchronization treatment might benefit them.

Keywords: Cardiac resynchronization therapy, Heart failure, Left ventricular non-compaction cardiomyopathy (LVNC), Ventricular arrhythmia

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INTRODUCTION

Left ventricular non-compaction cardiomyopathy (LVNC) is a rare, congenital, unclassified cardiomyopathy characterized by a prominent left ventricular trabecular reticular structure, thin and dense lamina, and deep trabecular depression, which is a morphological description rather than a functional diagnosis [1].

The annualized mortality in LVNC was up to 8%, with heart failure (HF) as the main cause [2]. Cardiac resynchronization therapy (CRT) is a recognized non-pharmacologic treatment for asynchronous HF, which can improve symptoms and quality of life, and reduce morbidity and mortality in patients with HF associated with LV dyssynchrony [3, 4]. However, as far as we know, studies on CRT on LVNC are rare and incomplete, and it is unclear whether patients with LVNC will benefit from CRT.

CASE REPORT

A 57-year-old male patient was admitted to hospital due to asthma after exercise. Physical examination showed a temperature of 36.2°C, pulse of 91 beats/min (bpm), respiration rate of 20 breaths/min, and blood pressure (BP) of 128/65 mmHg (1 mmHg = 0.133 kPa). Auscultation revealed coarse breath sounds in both lungs, with wet rales. The heart was enlarged with apical impulse felt in left, and the heart rate (HR) was 91 bpm with mitral systolic murmur and aortic diastolic murmurs. Laboratory tests suggest BNP of 4836.79 pg/mL (Table 1). Electrocardiogram showed complete left bundle branch block (QRS duration 164 ms) and multi-focal ventricular premature beats (Figure 1A). Chest X-ray showed marked cardiomegaly and mild pulmonary congestion. Transthoracic echocardiography revealed that the myocardium was cavernous sinus-like, forming sinusoidal cavities in varying sizes, communicating with left ventricular cavity, with abnormal echo of left ventricular myocardium (Figure 1C and D). Moderate mitral valve and severe aortic valve insufficiency could also be found by transthoracic echocardiography. The left ventricle was severely dilated (end-diastolic diameter of 91 mm) (Table 1). Diffuse hypokinesia was manifested by a significant decrease in systolic function (LVEF 31%, calculated with the Simpson method) (Figure 1B). During hospitalization, furosemide was given to the patient to reduce cardiac load, sacubitril-valsartan to improve cardiac remodeling, metoprolol tartrate to control ventricular rate, and digoxin to strengthen the heart. The patient's symptoms improved significantly and he was discharged.

However, the patient was readmitted within a month due to worsening shortness of breath with no apparent cause. Cardiac function was significantly worse than before. Laboratory examination showed that BNP was more than 5000 pg/mL (Table 1). Considering the poor effect of regular anti-heart failure drugs, cardiac resynchronization therapy was recommended. On the tenth day after admission, CRT was performed. Cardiac resynchronization therapy was primarily planned to provide dual ventricular pacing through two pacing guides of the cardiac venous system. Three guiding wires were delivered from left axillary vein puncture to deliver electrodes to right ventricle, left ventricle and right atrium respectively. The parameters were as follows: the right ventricular threshold was 0.5 V with an impedance of 550 Ω; the left ventricular threshold was 0.75 V with an impedance of 790 Ω; the right atrial threshold was 1.0 V with an impedance of 450 Ω.

On the third day after CRT, the patient could fall asleep completely on his back. Laboratory tests suggested that levels of BNP were more obviously lowered from more than 5000 pg/mL to 3679.09 pg/mL (Table 1).

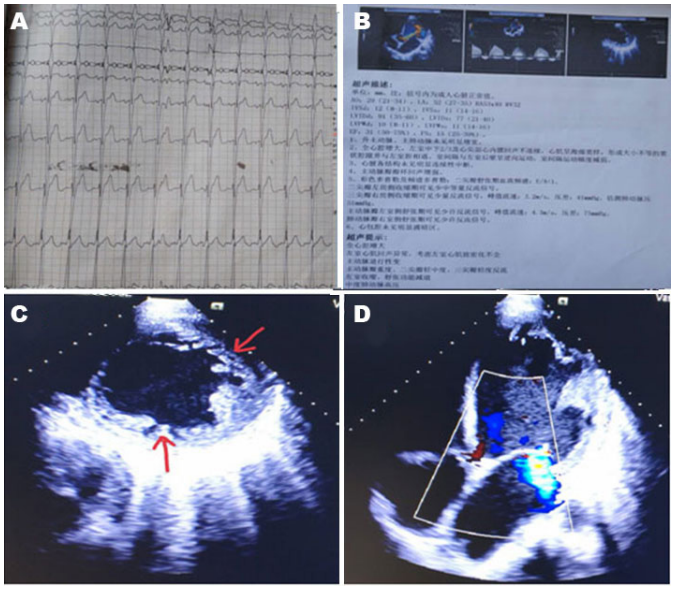

Figure 1: ECG and UCG at first hospitalization. (A) ECG shows wide QRS complex and complete left bundle branch block. (B) UCG revealed abnormal echo of left ventricular myocardium. (C, D) Prominent trabeculations at the apex of the left ventricle are clearly visible. The location in apical segment is typical for LVNC, a ratio of non-compacted/compacted myocardium >2 is diagnostic for LVNC.

Table 1: Laboratory test data

Indicators	First admission	Before discharged	Second admission	2nd-day after-CRT	3 month follow-up	6 month follow-up
BNP (ng/mL)	4836.79	3220.50	>5000	3679.09	2029.80	1253.18
LVIDd (mm)	91		89	87	83	81
LVIDs (mm)	77		76	72	65	61
EF (%)	31		32	36	38.71	41.06
SDI (ms)					32.4	24.2

Abbreviations: BNP: B-type natriuretic peptide, LVIDd: left ventricular end diastolic diameter, LVIDs: left ventricular end systolic diameter, EF (%): ejection fraction, SDI: systolic asynchrony index, CRT: cardiac resynchronization therapy

Electrocardiogram showed that QRS complex width was decreased from 164 to 127 ms (Figure 2A). Seven days after CRT, the patient was discharged, and the drug treatment was continued outside the hospital.

During 3-month follow-up, the symptoms of heart failure were significantly improved (from NYHA III to NYHA II). Electrocardiogram showed that the QRS complex width was decreased from 127 to 117 ms (Figure 2B). Echocardiography suggested that end-diastolic diameter decreased from 91 to 83 mm (Table 1), and the LVEF increased from 31% to 38.71% (Figure 2C). Laboratory examination revealed that the BNP was decreased from 4836.79 to 2029.80 ng/mL (Table 1). Echocardiography showed that the LVEF were increased to 41.06% six months after CRT procedure (Figure 2D).

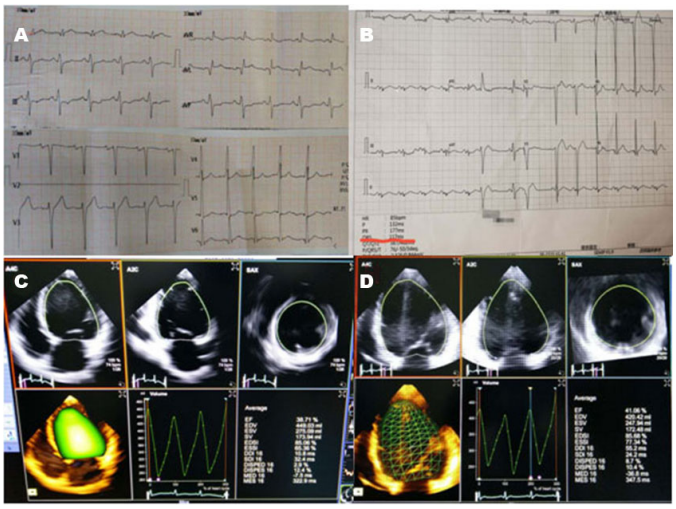

Figure 2: ECG after CRT, three dimensional echocardiography 3-months and 6-months after CRT. (A) ECG shows the QRS complex was narrower than preoperation. (B) 3-months follow-up revealed QRS complex was further narrowed. (C) LV end-diastolic diameter was decreased to 83 mm and LVEF was increased to 38.71%. (D) UCG shows LV end-diastolic diameter was decreased to 81 mm and LVEF was increased to 41.06%.

Meanwhile, the 6-min walking test recorded a distance of more than 450 meters.

DISCUSSION

Left ventricular non-compaction cardiomyopathy (LVNC) is a rare cardiomyopathy with an incidence rate between 0.05% and 0.24% [5], which is characterized by prominent left ventricular (LV) trabeculae and deep intertrabecular recesses. Its typical symptoms are cardiac dysfunction, arrhythmia, and thromboembolism, which are also three fatal risk factors for this disease [6, 7]. The diagnosis of LVNC mainly depends on ultrasound echocardiography (UCG) and clinical manifestations. The characteristics of UCG include: (1) thick lamellar myocardium composed of non-dense and dense layers; (2) prominent trabecular formation; (3) deep trabecular recess. Although cardiac magnetic resonance imaging (CMR) is generally better than echocardiography in identifying non-compacted myocardium [8], it is difficult to determine its sensitivity and specificity due to the lack of recommended gold standards and diagnostic guidelines. In most cases, especially in adult patients, the key factor in the diagnosis decision is not LVNC itself, but the associated LV dilation and/or dysfunction, ventricular involvement, arrhythmia, etc. Ultrasound echocardiography is convenient and economical, and still has irreplaceable advantages. In recent years, there have been few instances of CRT in patients with LVNC being reported.

For this patient, his clinical symptoms and imaging examination were diagnosed as LVNC, the status was New York Heart Association Class III and the effect of drug treatment was poor. Echocardiogram revealed a sinus rhythm with a QRS duration of 164 ms along with a left bundle branch block morphology. Ultrasound echocardiography showed LV dysfunction and left and right ventricular asynchrony, CRT was more effective, so it was wise to choose this method. Cardiac resynchronization therapy is a rapidly evolving treatment option for patients with drug-refractory heart failure. Large clinical trials have demonstrated sustained benefits of CRT in adult patients with moderate-to-severe heart failure (NYHA III or NYHA IV), systolic dysfunction (LVEF ≤35%), and a widened QRS complex (≥120 ms). The results showed that the effect of cardiac synchronization therapy combined with drug therapy was obvious after half a year. During half-a-year follow-up for the patient, combined with ultrasound, ECG and other examinations and the patient's response to pacemaker, the parameters of pacemaker were optimized by program control, so that the function of pacemaker was most beneficial to the patient, playing the role of pacemaker synchronization treatment. The patient's cardiac function was improved from NYHA III to NYHA II, and could be competent for ordinary physical labor. Echocardiography showed that the heart shrank, left ventricle was prominent, ejection fraction increased, and ventricular synchrony gradually improved. In a domestic study, Zhang Bin reported a case of myocardial insufficiency. The patient in the case had a poor response to drug therapy with severe heart failure, the patient's symptoms improved significantly after CRT treatment. Qiu et al. [9]. explored left ventricular (LV) remodeling and mechanical synchronicity before and after CRT in patients with LVNC. The results show that LV ejection fraction increased from 27.6 ± 5.5 to 39.1 ± 7.0% ($p<0.01$) during follow-up, but LV volumes did not change significantly (both were $p>0.05$). Five patients with LVNC (33.3 %) responded to CRT, and all of them were super-responders (reduction in LVESV>30%). In conclusion, CRT improved cardiac function, morphology, and mechanical dyssynchrony in patients with LVNC.

However, this paper is a single case observation and follow-up and the patient was not complicated by other diseases, his own conditions were better, which might

result in well response to treatment, which were also the beneficial factors for the patient in the treatment. Therefore, the evaluation of ventricular synchronization combined with drug therapy for LVNC still needs to increase the sample size and study from all aspects and perspectives.

CONCLUSION

In conclusion, the diagnosis of LVNC is not difficult, but the rate of missed diagnosis is high. The combination of color ultrasound and magnetic resonance imaging (MRI) is used to improve the accuracy of diagnosis. Under the current medical conditions, there has been no specific method to treat the disease. Previous studies have reported that patients with LVNC have poor prognosis, high mortality, and irreversible disease progression, with the main causes for death of heart failure and malignant arrhythmia. Currently, as cardiac resynchronization therapy becomes more widely available and guidelines are revised, more and more patients with heart failure will benefit from it. In this case, it was observed that the surgery for patients with LVNC who meet the indications for CRT could have satisfactory results. Therefore, cardiac synchronization therapy may also be a new treatment direction and method.

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Author Contributions

Juan Wang – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Dasheng Lu – Design of the work, Acquisition of data, Analysis of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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Yunong Han – Acquisition of data, Analysis of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Lingfei Yang – Design of the work, Acquisition of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Guarantor of Submission

The corresponding author is the guarantor of submission.

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Consent Statement

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of written consent is available for review by the Editor this journal.

Conflict of Interest

Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.

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