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CASE REPORT
Rare diseases of the heart

The 51-year-old patient with a non-compaction cardiomiopathy and multi-vessel coronary artery disease (RCD code: III-5A.1)

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Abstract

Left ventricular non-compaction (LVNC) or "spongy myocardium", is a rare form of a primary genetic cardiomyopathy considered to be the result of abnormal intrauterine arrest of the myocardial compaction process. Left ventricular noncompaction belongs to the group of unclassified cardiomiopathies by the ESC Position Statement. It is is a rare disorder characterised by a thin, compacted epicardial layer and an extensive non-compacted endocardial layer, with prominent trabeculation and deep recesses that communicate with the left ventricular cavity but not with the coronary circulation. Clinical presentation of patients with LVNC is highly variable – it ranges from completely asymptomatic patients who are accidentally diagnosed (e.g. during familial screening) to patients in need of heart transplantation. Most commonly patients present with symptoms from the spectrum of the classic triad of LVNC complications: heart failure, arrhythmias and systemic thromboembolic events. We describe a case of accidental discovery of isolated left ventricular non-compaction in a patient with an acute coronary syndrome. JRCD 2015; 2 (4): 123–126

Key words: spongy myocardium, congenital heart defects, acute coronary syndrome, bradycardia

Case presentation

51-year-old patient was referred to our Departament for cardiological evaluation with suspicion of an acute coronary syndrome. Patient had a history of hypertension but he did not take any medications. Before hospitalization he had never had echocardiography. At admission to our hospital the patient was haemodynamically stable. His heart rate (HR) was 90 beats per minute (bpm) and the blood pressure (BP) – 158/95 mm Hg. Laboratory tests showed increased levels of myocardial necrosis markers, abnormalities in lipid profile, normal renal function parameters, abnormal liver function parameters, normal fasting glucose level. ECG showed sinus rhythm, 60 bpm, normal axis, Q waves in leads II, III, aVF, negative T wave in leads II, III, aVF, V5, V6, elevated J points in leads V2, V3. Coronary angiography unveiled multi-vessel coronary artery disease (CAD): chronic ostial occlusion of right coronary artery, long 60–70% stenosis of anterior descending branch (LAD) (distal to the diagonal artery), chronic occlusion of diagonal artery (Dg) and occlusion of circumflex artery (Cx). The patient was consulted with the cardiac surgeon and was qualified for the coronary angioplasty of the Cx in the first stage of treatment and coronary artery bypass graft (CABG) in the second stage. The patient underwent immediately percutaneous coronary intervention (PCI) and stenting of the circumflex artery (the drug-eluting stent, DES). Carotid ultrasound did not reveal any hemodynamically significant abnormalities. A transthoracic echocardiogram (TTE) revealed both atria enlargement, slightly reduced ejection fraction of left ventricle (EF 49%), segmental wall motion abnormalities: hypokinesis of basal and mid-segment of the inferior, posterior and anterior wall, hypokinesis of basal segment of the septum and antero-lateral wall, combined aortic valve disease: mild stenosis and moderate regurgitation, moderate mitral valve regurgitation, the quotient of maximal compact wall to the total wall thickness was 0,38 (≤0,5), visible (in one im-

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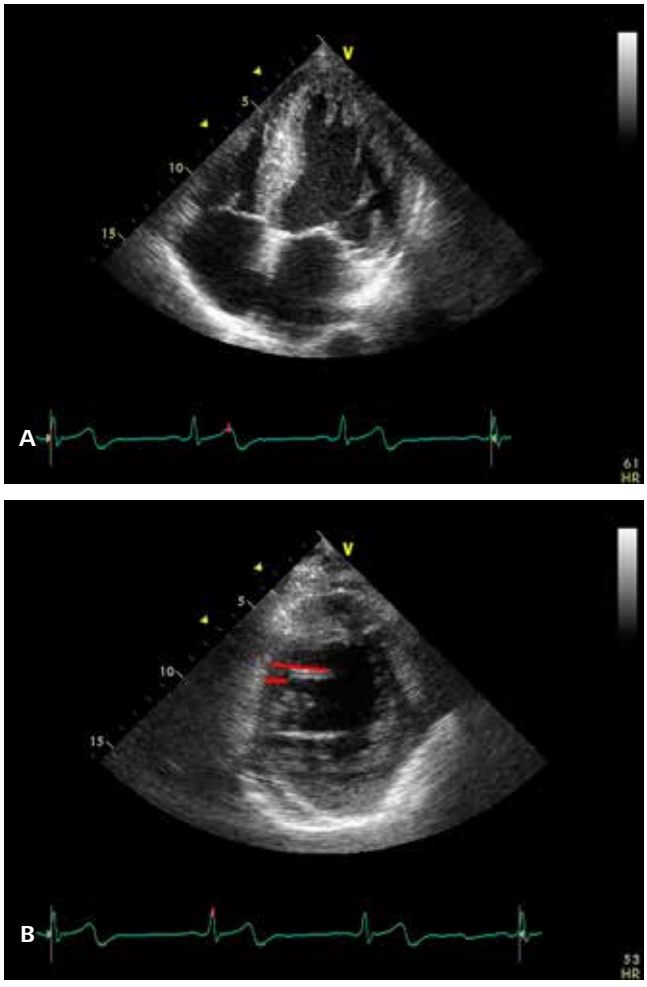

Figure 1. Transthoracic echocardiography demonstrating intertrabecular recesses and trabeculations in the left ventricle. The flow in the trabeculations can be verified by the color Doppler method. The quotient of maximal compact wall to the total wall thickness was 0,38 (≤0,5)

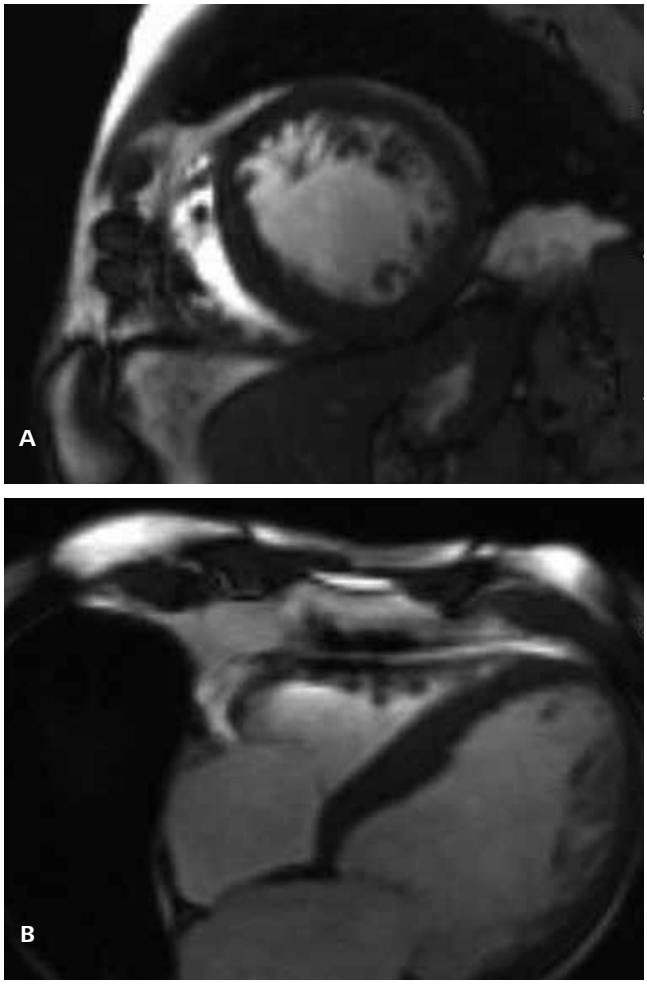

Figure 2. Cardiac magnetic resonance. Increased trabeculation of the apical segments of the anterior and lateral wall and the cardiac apex

age) more than 3 prominent trabeculae at end-diastole that moves synchronously with the compacted myocardium (Figure 1). Due to suspicion of the left ventricular non-compaction (LVNC) in the transthoracic echocardiography (two criteria), the cardiac magnetic resonance (CMR) was performed. CMR scans revealed significant enlargement of the left ventricle with signs of ischemic myocardial injuries (the basal segments of the inferior, posterior and lateral wall and mid segment of lateral wall). It showed also myocardial thickening of the basal part of the front wall (up to approx. 15 mm) and front region of the interventricular septum (up to approx. 16 mm), the middle part of the interventricular septum (up to approx. 17 mm). Increased trabeculation of the apical segments of the anterior and lateral wall and the cardiac apex was confirmed (Figure 2).

During hospitalization 24-hour monitoring Holter was also performed. It registered sinus rhythm of 54 bpm (max. 71 bpm, min. 41 bpm), decrease heart rate, occasional ventricular and supraventricular ectopic beats, one episode of supraventricular tachycardia, multiple episodes of sinus bradycardia with a minimum ventricular action of 28 bps, 10 pauses (≥2 sec.) with the longest pause of 2406 ms. The patient had no history of syncope. He was consulted with an electrocardiologist and his medications were modified according to the recommendations (low dose beta-blocker during the day and theophylline and atropine at night). Control Holter ECG showed similar findings to the previous one. Patient's medications were modified – the dose of beta-blocker was reduced, the dose of atropine was increased and theophylline was discontinued. Because there was no improvement in control ECG Holter monitoring the patient was qualified for pacemaker implantation. During the hospitalization pharmacological therapy including angiotensin converting enzyme inhibitor (ACEi), beta-blocker, aldosterone receptor blocker, statin, acetylosalicylic acid (ASA), clopidogrel and atropine.

Discussion

Left ventricular noncompaction can be diagnosed at any age. The real LVNC incidence is unknown, but based only on echocar-

Table 1. Proposed diagnostics criteria of left ventricular non-compaction

Criterion	Chin et al. [3]	Jenni et al. [7]	Stöllberger et al. [8]	Petersen et al. [9]	Jacquier et al. [10]
imaging technique	Echocardiography	Echocardiography	Echocardiography	CMRI	CMRI
phase cardiac cycle	end-diastole	end-systole	end-systole and end-diastole	Diastole	End-diastole
views	Short axis	Short axis	any	Long axis	Short axis
criteria	Excessive prominent trabeculations with deep intertrabecular recesses. maximal compacted wall/total wall thickness≤0,5	maximal noncompacted / compacted wall >2,0 Intertrabecular spaces filled by direct blood flow	>3 prominent trabeculae visible in 1 image plane at end-diastole that moves synchronously with the compacted myocardium.	maximal noncompacted / compacted wall >2,3	Trabecular mass /total LV mass >20%

diographic studies, reported prevalence is between 0.014 and 1.3% in the general population [5].

In clinical practice the diagnosis of LVNC is usually attained by using non-invasive imaging studies – TTE and CMR [6]. TTE despite its limitations (e.g. operator skills, poor acoustic window, high interobserver and intraobserver variability) but because of its widespread availability, ease of interpretation and low cost, is a good screening test and still remains the front-line imaging modality. At present, there is no consensus on the echocardiographic criteria for diagnosing LVNC but there are 3 commonly used diagnostic criteria: Chin et al. [4], the Jenni et al. [7] and Stöllberger et al. [8]. CMR have been shown to have very high specificity and so it is used to confirm the diagnosis. As with TTE, there are no gold standard criteria but clinically the Petersen et al. [9] and Jacquier et al. [10] criteria are usually used. Both TTE and CMR criteria are based on the ratio of the thickness of the non-compacted layer to that of the compacted layer (Table 1).

Arrhythmias are the second most common (after heart failure) manifestation of LVNC and according to available data are the number one cause of mortality in these patents [11]. In adults most frequent arrhythmias comprise of sustained or non-sustained ventricular tachycardia (VT), atrial fibrillation (AF), QT prolongation and atrioventricular (AV) blocks. In children most frequent are Wolff–Parkinson–White (WPW) syndrome, AV block, VT and bradycardia [12]. Ventricular tachyarrhythmias are reported in 38–47% adult patients and in 13–18% of those who die suddenly [13], which supports the hypothesis that noncompacted myocardium is a highly arrhythmogenic substrate. Episodes of sinus bradycardia as seen in our patient are a rare phenomenon in adult patients with LVNC. It may be a first manifestation of LVNC in this patient but it may as well be secondary to chronic total occlusion of his right coronary artery.

Much controversy surrounds the association between LVNC and atherosclerotic coronary artery disease. Our patient is an example of coexistence of severe CAD and LVNC. At present the question whether the combination of CAD and LVNC is purely coincidental or there is genetic link remains open to further studies, especially those in the rapidly developing field of gene mutation research. An even more important question from the clinical perspective is whether coexistence of CAD influences the prognosis of patients with LVNC. One retrospective study showed that incidence of major cardiac events was similar in LVNC patients with or without CAD but definitely more research is needed [14].

Due to variable phenotypic expressions of patients with LVNC the outcomes are more closely related to the disease severity than to the diagnosis. Since there is no treatment for the primary disease, therapy is directed at the classic triad of LVNC complications and is based on the corresponding international guidelines. Special consideration require primary prevention of cardiac arrhythmias and thromboembolic events.

According to available data sudden cardiac death due to arrhythmias is the number one cause of mortality in patients with LVNC. Currently there are no clinically useful tools for risk stratification

Randomized control trials will be needed in the near future to determine the most appropriate method of treatment. However, given low incidence of the disease and the controversy regarding the diagnosis, case reports may remain the only guidelines for quite a while.

Conclusions

LVNC is a rare cardiomyopathy, which has serious consequences and remains the subject of numerous clinical studies. Cardiac manifestations of LVNC comprise heart failure, thromboembolism, and arrhythmias. LVNC may be associated with coronary artery disease. The complex pathophysiology and the lack of targeted therapy makes this disease needs an accurate estimation of clinical adverse outcomes, and determine appropriate management to treat LVNC.

References

1. Freedom RM, Yoo S-J, Perrin D, et al. The morphological spectrum of ventricular noncompaction. Cardiol Young. 2005; 15: 345–364.
2. Elliott P, Andersson B, Arbustini E, et al. Classification of the cardiomyopathies: a position statement from the European Society Of Cardiology Working Group on Myocardial and Pericardial Diseases. Eur Heart J 2008; 29: 270–276.
3. Chin TK, Perloff JK, Williams RG, et al. Isolated noncompaction of left ventricular myocardium. A study of eight cases. Circulation 1990; 82: 507–513.
4. Trąbka-Zawicki A, Gackowski A, Andres M, et al. Accidentally discovered non-compaction cardiomyopathy in patient with initial diagnosis of acute coronary syndrome. JRCD 2013; 1: 17–22.

5. Ritter M, Oechslin E, Sutsch G, et al. Isolated noncompaction of the myocardium in adults Mayo Clin Proc 1997; 72: 26 –31.
6. Ikeda U, Minamisawa M, Koyama J. Isolated left ventricular non-compaction cardiomyopathy in adults. J Cardiol. 2015; 65: 91–97.
7. Jenni R, Oechslin E, Schneider J, et al. Echocardiographic and pathoanatomical characteristics of isolated left ventricular non-compaction: a step towards classification as a distinct cardiomyo-pathy. Heart 2001; 86: 666–671.
8. Stöllberger C, Finsterer J, Blazek G. Left ventricular hypertrabeculation/noncompaction and association with additional cardiac abnormalities and neuromuscular disorders. Am J Cardiol 2002; 90: 899–902.
9. Petersen SE, Selvanayagam JB, Wiesmann F, et al. Left ventricular non-compaction: insights from cardiovascular magnetic resonance imaging. J Am Coll Cardiol 2005; 46: 101–105.
10. Jacquier A, Thuny F, Jop B, et al. Measurement of trabeculated left ventricular mass using cardiac magnetic resonance imaging in the diagnosis of left ventricular non-compaction. Eur Heart J 2010; 31: 1098–1104.
11. Brignole M, Auricchio A, Barón Esquivias G, et al. 2013 ESC guidelines on cardiac pacing and cardiac resynchronization therapy. Eur Heart J. 2013; 34: 2281–2329.
12. Stöllberger C, Finsterer J. Arrhythmias and left ventricular hypertrabeculation /noncompaction. Curr Pharm Des. 2010; 16: 2880–2894.
13. Oechslin EN, Attenhofer Jost CH, Rojas JR, et al. Long-term follow-up of 34 adults with isolated left ventricular noncompaction: a distinct cardiomyopathy with poor prognosis. J Am Coll Cardiol. 2000; 36: 493–500.
14. Gao XJ, Kang LM, Zhang J, et al. Incidence of coronary artery disease and outcome of patients with left ventricular noncompaction. Zhonghua Xin Xue Guan Bing Za Zhi. 2011; 39: 725–729.