A CASE OF NON-COMPACTION CARDIOMYOPATHY

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ABSTRACT

The aim of this case report was to present a rare myocardial disease, the so-called isolated non-compaction cardiomyopathy (NCCM). This disorder, a primary genetic cardiomyopathy, attracts increasing attention. A woman with a sudden onset of shortness of breath and isolated NCCM is described. Criteria for diagnosis as well as echocardiographic findings and magnetic resonance imaging of the heart are presented. Although rare, a new option of myocardial disease with various modes of clinical presentation but, most commonly, with acute or chronic heart failure should be taken into consideration. Multiple imaging modalities are available and should be used for proper diagnosis.

Key words: *non-compaction cardiomyopathy, acute heart failure, echocardiography, magnetic resonance imaging, case report*

INTRODUCTION

Pathogenesis of non-compaction cardiomyopathy (NCCM) is a genetically determined disturbance of the myocardial compaction process during fetal endomyocardial morphogenesis. Echocardiography is the diagnostic method of choice. The diagnosis is based on the following echocardiographic criteria: presence of at least four prominent trabeculations and deep intertrabecular recesses, blood flow from the ventricular cavity into the intertrabecular recesses, and a typical bilaminar structure of the affected portion of the left ventricular myocardium. NCCM can also be diagnosed with magnetic resonance imaging (MRI) of the heart. The clinical presentation of NCCM is different and includes heart failure, thromboembolic events, and arrhythmias. Patients with symptomatic NCCM present with adverse prognosis (1).

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CASE REPORT

A female patient was admitted to the hospital for the first time on the occasion of sudden breath shortness and palpitations. Blood pressure was 220/120 mm Hg and antihypertensive medications were administered. The patient had no history of arterial hypertension. However, she reported a similar episode a few weeks ago. Family history showed a sister with arettrtial hypertension. Physical examination at admission revealed a sinus rhythm, heart rate of 110/min, T3 gallop, crepitation in the lower lung fields, and blood pressure of 145/100 mm Hg.

CT of the thorax demonstrated pulmonary congestion with dilated pulmonary arteries: pulmonary trunk - 26 mm; right main branch of pulmonary artery – 16,5 mm and the left main branch of pulmonary artery - 20 mm. There were no signs of pulmonary thromboembolism (Fig. 1).

Electrocardiogram (ECG) displayed a sinus rhythm, intraventricular conduction disturbancies, QRS - 109 msec, and non-specific ST-T changes (Fig. 2).

The following imaging methods were used for diagnostic purposes:

Echocardiography

Echocardiography demonstrated trabecularization of the segments of the left ventricle with a

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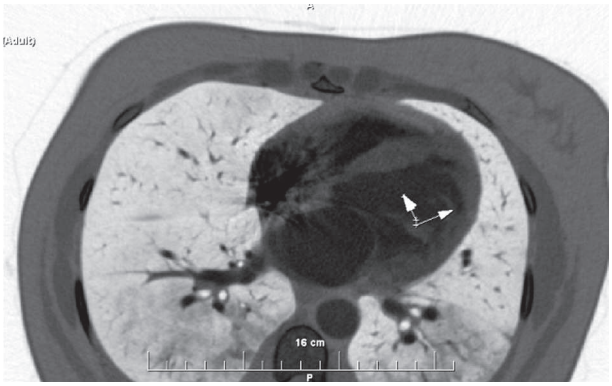

Fig. 1. Patient's CT of the thorax

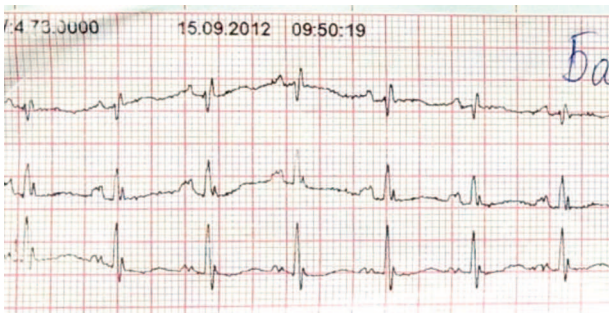

Fig. 2. Patient's ECG

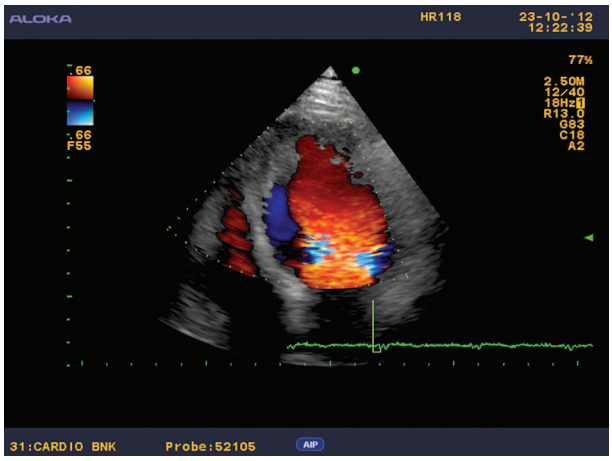

Fig. 3. Patient's echocardiographic images

thin layer of compact epicardial layer and a thicker non-compact endocardial one in a ratio >2,0; multiple lacunas engaging three top segments, three medium and three basal ones with colour blood flow in incisures (Fig. 3). Simpson - EDV/ESV 113 /60 mL; EF 33%, LAA - 18 cm²; mitral regurgitation 2+, tricuspid regurgitation 1+, transmitral flow velocity - E- 0,7 m/s and A - 0,4 m/sec (Fig. 4).

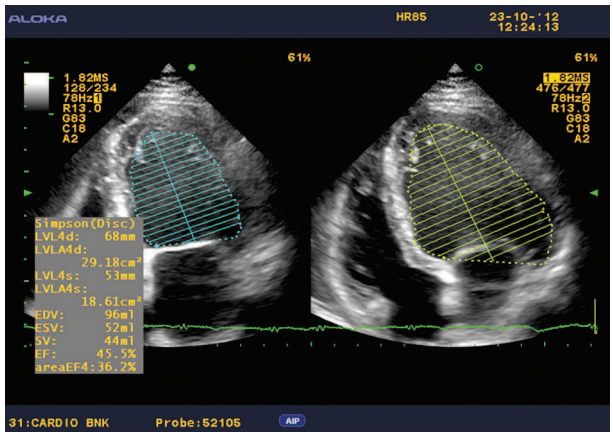

Fig. 4. Simpson echocardiographic patterns

MRI of the heart

MRI displayed a left ventricular hypertrophy, segments with trabecularization, recesses, chords with a thin layer of compact epicardial layer and thicker non-compact endocardial one in a ratio >2,3

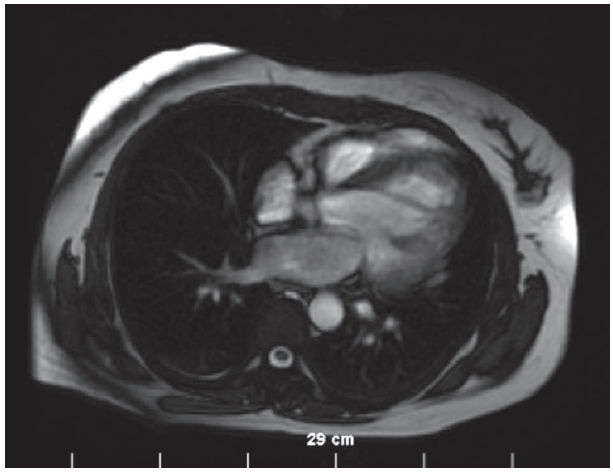

Fig. 5. Patient's MRI findings

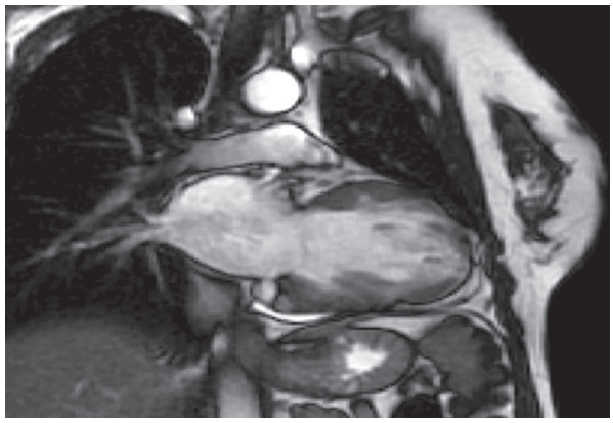

Fig. 6. Patient's MRI features

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engaging the three top segments, three medium and three basal ones (Fig. 5 and Fig. 6).

DISCUSSION

NCCM can arise in all ages and its onset can be either in adults or in children. Heart failure in the acute or chronic setting is the main clinical presentation of NCCM. Thromboembolic complications are common in patients who present with both NCCM and atrial fibrillation. Intraventricular thrombi are also found due to an impairment of left-ventricular compaction in the deep intertrabecular recesses.

Echocardiographic criteria for the diagnosis of isolated NCCM are the following (3,4):

- There are at least four prominent trabecula and deep intertrabecular recesses.
- Blood flow between the cavum of the left ventricle and the recesses is demonstrable with colour Doppler ultrasonography or through the use of ultrasonographic contrast medium.
- The non-compact mural segments have a typical bilaminar structure, and the non-compact subendocardial layer is at least twice as thick as the compact subepicardial layer in systole. Non-compaction is seen mainly at the cardiac apex and in the inferior, central, and lateral portions of the left ventricular wall.
- No other cardiac abnormalities are present.

Table 1 shows the Classification of Cardiomyopathies of the American Heart Association (3).

The differential diagnosis of isolated non-compaction cardiomyopathy includes (5):

- hypertrophic cardiomyopathy
- localized left ventricular hypertrophy
- dilated cardiomyopathy
- endocardial fibroelastosis
- myocarditis and pericarditis
- restrictive cardiomyopathy
- left ventricular thrombi
- aberrant chordae tendineae
- intramyocardial hematoma/abscess, and
- cardiac metastases.

NCCM is myocardial disease due to a disturbance of embryonal endomyocardial morphogenesis. It can be diagnosed either by echocardiography, or by cardiac MRI. Its main clinical manifestations are heart failure, thromboembolic events, and arrhythmias.

The prognosis of advanced NCCM is poor. However, it could be improved if NCCM is diagnosed early by screening of close relatives and appropriate management with standard therapy of heart failure as well as with oral anticoagulation, antiarrhythmic agents and implantation of internal cardioverter defibrillator system to prevent systemic emboli in high-risk patients with impaired left ventricular function (2).

CONCLUSION

Although rare, a new option of myocardial disease with various modes of clinical presentation

Table 1. American Heart Association classification of cardiomyopathies (3)

Primary cardiomyopathies		
Genetic types	Mixed types	Acquired types
hypertrophic cardiomyopathy	dilative cardiomyopathy	infectious/inflammatory (myocarditis)
arrhythmogenic right ventricular cardiomyopathy	restrictive	stress-induced (tako-tsubo)
NCCM		peripartum
glycogen storage diseases		tachycardia-induced
mitochondrial myopathies		children of insulin-dependent diabetic mothers
ion-channel defects		

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