SEMINAR ZA LEKARE U PRAKSI
SEMINAR FOR PHISICIANS

Faculty of Medicine, University of Belgrade, Belgrade[1]
Cardiovascular Insitute Dedinje, Belgrade[2]

Seminar za lekare u praksi
Seminar for phisicians
UDK 616.124-007-07
UDK 616.12-008.331.1:616.24
DOI: 10.2298/MPNS1112597N

A MISINTERPRETATION OF THE LEFT VENTRICULAR NON-COMPACTION-ADULT PATIENT WITH PRIMARY PULMONARY HYPERTENSION

PROBLEMI U DIJAGNOSTIKOVANJU NEKOMPAKTNE LEVE KOMORE – ODRASLI PACIJENT SA UDRUŽENOM PRIMARNOM PLUĆNOM HIPERTENZIJOM

Aleksandra NIKOLIĆ[1,2], Ljiljana JOVOVIĆ[2], Velibor RISTIĆ[2], Dejan NIKOLIĆ[2] and Lazar ANGELKOV[2]

Summary – Non-compaction of the left ventricle is a rare cardiac malformation, defined as a primary cardiomyopathy caused by genetic malformations. Although the pathogenesis of this cardiomyopathy is unknown, there are two possible hypotheses (congenital and acquired) which lead to arrest in intrauterine endomyocardial morphogenesis. We are presenting a case of a 60-year-old woman, with a history of bradyarrhythmia, syncope and cyanosis. Two-dimensional echocardiography showed the thickened myocardium with prominent trabeculations and deep intertrabecular recesses in the two thirds of the apical part of left ventricle walls. The right side cavity was enlarged with hypertrophied wall. Tricuspid regurgitation was moderate. Systolic pressure in the right ventricle was 70mmHg. Catheterization of the right heart showed high pressure in the pulmonary artery. According to publications, this is a very rare case with the presence of possible primary pulmonary hypertension and non-compaction of the left ventricle.

Key words: Hypertension, Pulmonary; Heart Ventricles + abnormalities; Adult; Diagnosis; Cardiomyopathies; Heart Defects, Congenital

Introduction

Non-compaction of the left ventricle was a rare and unclassified congenital cardiac malformation until the year of 2006 when new Contemporary Definitions and Classification of the Cardiomyopathies from the American Heart Association Scientific Statement referred to it as a primary cardiomyopathy with genetic cause [1, 2]. Although the pathogenesis of this cardiomyopathy is unknown, there are two possible hypotheses (congenital and acquired) which lead to an arrest in intrauterine endomyocardial morphogenesis.

The first hypothesis resulted from the arrest of an intrauterine compaction of the myocardial fibers in the absence of any other structural heart diseases. The mechanism of the second hypothesis was less convincing than the first one and it was described on the basis of higher hemodynamic demands in some situations (left ventricular pressure or volume overload), which could change the endomyocardium [3]. It is extremely important to recognize this condition because of its high mortality and morbidity due to progressive heart failure, ventricular arrhythmias and thromboembolic events [3].

Case report

A 60-year-old female was admitted to our Institute complaining of slow heart rate and cyanosis on minimal effort.

History

For almost five years (since 2006) she had been complaining of irregular slow heart rate, chest discomfort, worsening dyspnea and cyanosis on minimal effort. In 1993, she was diagnosed by echocardiography to have hypertrophic non-obstructive cardiomyopathy. From that time, the patient was treated by beta blockers, angiotensin-converting enzyme (ACE) inhibitors and diuretics due to hypertension. In May 2006, she had a transient ischemic attack with no neurological consequences according to computed tomography (CT) scan. She was hospitalized three times during 2007 due to hypotension and irregular fast heart rate (atrial fibrillation), after amiodarone had been added to the therapy. The family history was negative. She had two normal pregnancies.

The physical exam did not reveal any pathological findings except for peripheral cyanosis. The patient had a normal body habitus. The laboratory findings were normal, except for low partial pressure of oxygen in arterial blood, which was less than 80mmHg. The electrocardiogram (ECG) showed a sinus bradycardia, signs of right ventricular hypertrophy and strain, with the incomplete right bundle branch block. Iatrogenically-induced bradycardia developed after the administration of amiodarone and beta blockers.

24 h Holter monitoring revealed sinus absolute bradycardia (the average frequency was 43/min,

Corresponding author: Asist. dr Aleksandra Nikolić, Institut za kardiovaskularne bolesti Dedinje, 11000 Beograd, Milana Tepića 1a, E-mail: nikolicdrsasa@gmail.com

Abbreviations
LVNC – left ventricular non-compaction

from 30/min to 53/min during exercise) with 159 pauses (2 seconds or more) and lower escape rhythm. Twelve hundred premature ventricular beats with two different morphologies were detected and two episodes of supraventricular tachycardias (up to 12 complexes).

Transthoracic echocardiographic (TTE) study described a moderately reduced left ventricle systolic function (ejection fraction was 40%), mild mitral regurgitation, thickened myocardium with prominent trabeculations and deep intertrabecular recesses in the two thirds of the apical left ventricle **(Figure 1)**. Transmitral flow had a restrictive pattern. The right cavity was enlarged with hypertrophied walls and with normal morphology. Tricuspid regurgitation was moderate and systolic pressure in the right ventricle was 70 mmHg. The pulmonary artery had normal dimension with mild regurgitation.

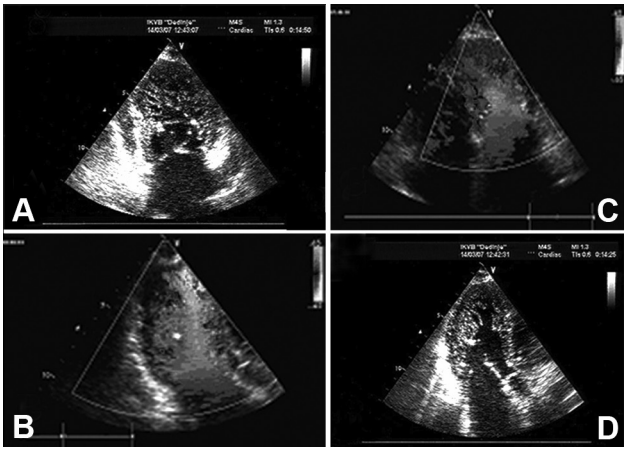

Fig. 1. (A, D) Transthoracic two-dimensional echocardiography shows apical two and three chambers view where hypertrabeculation in the apical two thirds of all walls can be seen. (B, C) Color Doppler confirmed that the deep intertrabecular space is in the connection with the left ventricular cavity.
Slika 1. *(A, D) Transtorakalna 2D ehokardiografija pokazuje da su distalne dve trećine leve komore ispunjene trabekulama, posmatrano iz apikalnog dvo i tro-šupljinskog preseka. (B, C) Kolor doplerom se uočava da krv ulazi u duboke intertrabekularne prostore.*

Coronary angiography showed neither significant stenosis nor abnormalities of the coronary arteries, but we found TIMI 2 flow throughout the coronary system. Catheterization of the right heart showed high pressure in the pulmonary artery pulmonary artery systolic pressure (PASP) 101 mmHg, right ventricle 104 mmHg and pulmonary capillary wedge pressure (PCWP) 6 mmHg **(Figure 2)**.

Chest X-ray showed the enlargement of the right ventricle and enlarged pulmonary arteries shadow.

To confirm the presence of possible primary pulmonary hypertension and exclude other possible diagnoses we performed pulmonary function tests, blood tests for HIV, autoimmune and liver diseases. Doppler exa-

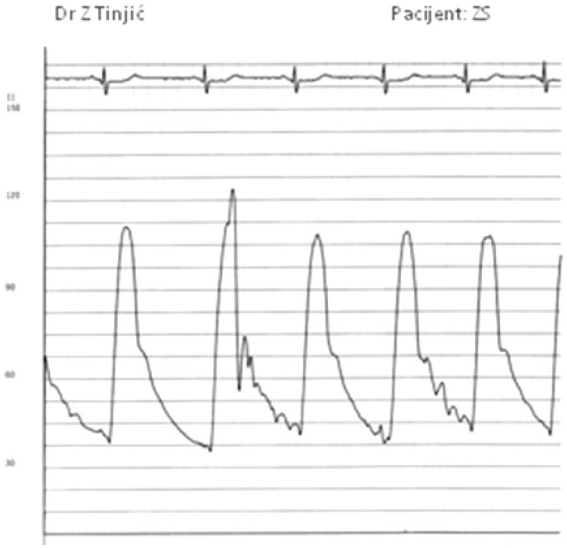

Fig. 2. Catheterization of the right heart showed high pressure in the pulmonary artery (systolic 101 mmHg, diastolic 32 mmHg and mean pressure was 55 mmHg)
Slika 2. *Kateterizacijom desnog srca su dobijeni visoki pritisci u plućnoj arteriji (sistolni 101 mmHg, dijastolni 32 mmHg i srednji pritisak 55 mmHg)*

mination of deep veins was done to exclude chronic thromboembolic pulmonary hypertension according to the current guidelines for pulmonary hypertension. The results of all performed tests were normal.

We did an electrophysiological study with successful ablation of tachycardia near the coronary sinus ostium using an electro-anatomical mapping system.

After the primary ablation of atrial tachycardia in 2010, the patient again had attacks of atrial fibrillation that could not be adequately stabilized by medication therapy, after which it was decided to repeat the ablation. From clinical point of view, the patient complained of intense fatigue and limitation in normal activities but she was assessed as the New York Heart Association (NYHA) functional class II. The patient was admitted to our Institute in September 2010, where the ablation of atrial fibrillation was repeated; however, since atrial fibrillation could not be terminated and the third degree atrioventricular block and left ventricular ejection fraction, estimated to be 30%, developed, we decided to embed a multi-site pacemaker.

When the previous therapy (calcium channel blockers, diuretics) was modified with anticoagulants, her cardiac status became satisfactory. Unfortunately, the patient died several days after discharge due to non cardiac causes.

Discussion

The prevalence of LVNC has been reported to be from 0.05% to 0.24% in the largest adult series and 0.14% in the largest pediatric series, but the true prevalence is thought to be higher [3]. The longest follow-up period of patients with LVNC was 24 years. They dem-

onstrated that some patients could have a favorable prognosis [4]. The new studies in this field found associations among patients with LVNC and patients with other pathologies (like neuromuscular) [5]. The latest study with the largest number of patients [238] found that non-compaction alone did not seem to be a risk factor for malignant supraventricular or ventricular arrhythmias [6]. The most available technique for diagnosis and follow-up of patients with LVNC is the echocardiography. During primary echocardiographic examinations, LVNC was diagnosed in approximately less than 50% of patients [7]. The most frequent misinterpretation has been described as apical or localized hypertrophic cardiomyopathy, dilated cardiomyopathy, endocardial fibroelastosis, endomyocardial fibrosis, restrictive cardiomyopathy, myocarditis, the left ventricular masses or thrombus [8].

The diagnosis of the right ventricular non-compaction by echocardiography is not attempted anymore because of the heavily trabeculated right ventricular apex and the difficult differentiation between normal variants and pathologic patterns. The right ventricular non-compaction may accompany the left ventricular non-compaction in less than 50% of patients [9].

Misdiagnoses of patients with LVNC together with wrong therapy of these patients might deteriorate their condition (hypotension, bradycardia and syncopa).

According to the available data, this paper describes a very rare association between LVNC and possible primary pulmonary hypertension. The pitfalls in the diagnosis of LVNC are present, but, on the other hand, they could encourage doctors to venture into new investigations in this field and try to give the true diagnosis to the patients with LVNC with advanced imaging techniques.

Conclusion

Individual reports could disclose the etiology of left ventricular non-compaction. This is important for further investigations and identification of the patients, who could have complications resulting from wrong diagnoses and treatment. Making the true diagnosis of left ventricular non-compaction together with adequate medication strategy could change bad prognosis in these patients.

References

1. American Heart Association. Scientific statement: contemporary definitions and classification of the cardiomyopathies. Circulation. 2006;113:1807-16.

2. Richardson P, McKenna W, Bristow M, et al. Report of the 1995 World Health Organization/International Society and Federation of Cardiology task force on the definition and classification of cardiomyopathies. Circulation 1995;93:841-84.

3. Stöllberger C, Finsterer J. Left ventricular hypertrabeculation/noncompaction. J Am Soc Echocardiogr 2004;17(1):91-100.

4. Salemi VM, Rochitte CA, et al. Long-term survival of a patient with isolated noncompaction of the ventricular myocardium. J Am Soc Echocardiogr. 2006;19(3):354.

5. Stöllberger C, Finsterer J. Blazek G. Isolated left ventricular abnormal trabeculation: follow-up and association with neuromuscular disorders. Can J Cardiol 2001;17:163-8.

6. Fazio G, Corrado G, Zachara E, et al. Ventricular tachycardia in non-compaction of left ventricle: is this a frequent complication? Pacing Clin Electrophysiol 2007;4:544-6.

7. Frischnecht BS, Attenhofer Jost CH, Oeschlin EN, Seifert B, Hoigne P, Roos M, et al. Validation of noncompaction criteria in dilated cardiomyopathy, and valvular and hypertensive heart disease. J Am Soc Echocardiogr 2005;18(8):865-72.

8. Stöllberger C, Finsterer J. Pitfalls in the diagnosis of left ventricular hypertrabeculation/non-compaction. Postgrad Med J 2006; 82:679-83.

9. Weiford BC, Subbarao VD, Mulhern KM. Noncompaction of the ventricular myocardium. Circulation 2004;109:2965-71.

Sažetak

Uvod

Nedovoljno formiran miokard (LVNC – left ventricular non-compaction) do pre nekoliko godina pripadao je neklasifikovanim kardiomiopatijama. Nove preporuke (Contemporary Definitions and Classification of the Cardiomyopathies) svrstavaju ga u primarne kardiomiopatije koje su prouzrokovane genetskim defektom, koji nastaje kao posledica prekida normalne intrauterine endomiokardne morfogeneze. Iako je patogeneza ovog procesa i dalje nepoznata, u opciji su dve hipoteze kao mogući razlozi prestanka normalnog intrauterinog razvoja – to su urođena i stečena.

Prikaz slučaja

Kroz prikaz slučaja želeli smo da predstavimo pacijenta sa primarnom plućnom hipertenzijom i lako redukovanom sistolnom srčanom funkcijom koja je prouzrokovana neprepoznatom nekompaktnom levom komorom. Pacijenta je kroz niz godina pratila dijagnoza – neopstruktivna hipertrofična kardiomiopatija, što je dovelo do jatrogenog pogoršanja opšteg statusa. Nekompaktna leva komora je manifestovana kroz tranzitorni ishemijski atak i aritmiju. Analizom publikovanih radova u dostupnim bazama podataka, do sada nije objavljen rad koji govori o udruženosti izolovane forme nekompaktne komore i primarne plućne hipertenzije.

Zaključak

Pojedini prikazi slučajeva mogu da otkriju etiologiju nekompaktne leve komore, što je važno za dalja istraživanja i identifikaciju pacijenata, koji bi mogli da imaju komplikacije nastale zbog pogrešnih dijagnoza i lečenja. Postavljanje tačne dijagnoze LVNC zajedno sa adekvatnom strategijom lečenja lekovima mogli bi da promene lošu prognozu kod tih pacijenata.

Key words: *Plućna hipertenzija; Srčane komore + abnormalnosti; Odrasli; Dijagnoza; Kardiomiopatije; Urođene srčane mane*

Rad je primljen 20. VII 2010.
Prihvaćen za štampu 1. IV 2011.
BIBLID.0025-8105:(2011):LXIV:11-12:597-599.