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Clinical and morphologic aspects of noncompact left ventricular cardiomyopathy in the pediatric patient: a clinical case report

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Left ventricular noncompaction cardiomyopathy (LVNC) is a rare genetic cardiomyopathy. The disease has a wide range of clinical manifestations, from asymptomatic conditions to serious cardiovascular complications, making its diagnosis and treatment difficult.

The **aim** of this study was to demonstrate the clinical and morphologic aspects of noncompact LVNC in a child and to analyze the current principles of diagnosis of this disease.

We present a **clinical case** of LVNC (spongy) in a child aged 3 years 9 months. Postmortem examination of the child's body was performed in the Pathology Department of the MNPE «Regional Clinical Hospital of the Ivano-Frankivsk Regional Council». Histological sections of the heart were stained with hematoxylin and eosin and Van Gieson's picrofuchsin. Microscopic examination and photography of samples were performed using an Olympus CX23 light-optical microscope with an LC30 digital CMOS camera and cell Sens Entry software. The presented clinical case allowed us to identify the main pathomorphologic criteria of LVNC: hypertrabecular «spongy» myocardium, hypertrophy of muscle fibers, pleomorphism of cardiac myocytes, endocardial fibrosis, and small-focal myocardial sclerosis.

Despite the lack of standardized methods for diagnosing LVNC, ultrasound, echocardiography, magnetic resonance, and cardiac computed tomography are traditional and widely used. Genetic evaluation of cardiomyopathies is an important clinical priority. Genetic testing and phenotyping will allow appropriate treatment of specific LVNC targets, thereby improving survival, reducing morbidity, and improving quality of life.

The research was carried out in accordance with the principles of the Declaration of Helsinki. The informed consent of the patient was obtained for conducting the studies.

No conflict of interests was declared by the authors.

Keywords: noncompact cardiomyopathy of the left ventricle, clinic, diagnosis, pathomorphology.

Клініко-морфологічні аспекти некомпактної кардіоміопатії лівого шлуночка у дитини: клінічний випадок

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Некомпактна кардіоміопатія лівого шлуночка (Left ventricular noncompaction cardiomyopathy, LVNC) – це рідкісна генетична кардіоміопатія. Захворювання має широкий спектр клінічних проявів, від безсимптомних станів до серйозних серцево-судинних ускладнень, що утруднює його діагностику та лікування.

Мета: продемонструвати клініко-морфологічні аспекти LVNC у дитини та проаналізувати сучасні принципи діагностики цього захворювання.

Представлений **клінічний випадок** LVNC (губчастої) у дитини віком 3 роки 9 місяців. Посмертна діагностика тіла дитини проведена на базі патологоанатомічного відділення КНП «Обласна клінічна лікарня Івано-Франківської обласної ради». Гістологічні зрізи серця забарвлювали гематоксиліном та еозином, пікрофуксином за Ван Гізона. Мікроскопічне дослідження та фотографування зразків проводили із використанням світлооптичного мікроскопу Olympus CX23 з цифровою CMOS камерою LC30 та програмним забезпе-

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ченням cell Sens Entry. Представлений клінічний випадок дозволив виділити основні патоморфологічні критерії LVNC: гіпертрабекулярний «губчастий» міокард, гіпертрофія м'язових волокон, плеоморфізм ядер кардіоміоцитів, ендокардіальний фіброз, дрібно-вогнищевий міокардіосклероз.

Попри відсутність стандартизованих методів діагностики LVNC традиційними та основними є ультразвукове дослідження, ехокардіографія, магнітно-резонансна та комп'ютерна томографія серця. Генетична оцінка кардіоміопатії є ключовим клінічним пріоритетом. Генетичне тестування та визначення фенотипу дозволяють проводити відповідне лікування конкретних мішеней LVNC, що сприятиме покращенню виживаності, зниженню захворюваності і підвищенню якості життя.

Дослідження виконано відповідно до принципів Гельсінської декларації. На проведення досліджень отримано інформовану згоду батьків дитини.

Автор заявляє про відсутність конфлікту інтересів.

Ключові слова: некомпактна кардіоміопатія лівого шлуночка, клініка, діагностика, патоморфологія.

Introduction

Cardiomyopathies (CMPs) are a group of diseases characterized by myocardial pathology, in which structural or functional abnormalities occur that are not caused by coronary heart disease, hypertension, valvular disease, or congenital heart disease. This definition applies to both children and adults and makes no a priori assumptions about etiology (which may be familial/genetic or acquired) or myocardial pathology. Working Group of Experts on the Treatment of Cardiomyopathies of the European Society of Cardiology (2023) has developed guidelines for the management of patients with cardiomyopathy. These guidelines aim to provide recommendations on the specifics of diagnosis and highlight aspects of therapy based on the current evidence base [9,14]. According to this guideline, the following phenotypes of cardiomyopathies are distinguished:

Dilated cardiomyopathy – defined as the presence of left ventricular dilatation and global or regional systolic dysfunction not explained by abnormal stress conditions (e.g., hypertension, valvular heart disease, coronary artery disease) or coronary artery disease alone.

Hypertrophic cardiomyopathy is defined as the presence of increased left ventricular wall thickness (with or without right ventricular hypertrophy) or mass not explained by abnormal stress conditions alone.

Restrictive cardiomyopathy – defined as restrictive pathophysiology of the left and/or right ventricle with normal or reduced diastolic volume (of one or both ventricles), normal or reduced systolic volume, and normal ventricular wall thickness;

Arrhythmogenic right ventricular cardiomyopathy – presence of predominantly dilated and/or dysfunctional right ventricle with histological lesions and/or electrocardiographic abnormalities according to published criteria.

Non-dilated cardiomyopathy (systolic dysfunction with minimal dilation, mitochondrial diseases, fibroelastosis).

LVNC is a newly classified cardiomyopathy and probably the most controversial without clinical guidelines. The American Heart Association has classified LVNC as

a separate primary cardiomyopathy with a genetic etiology [13]. However, it is considered an unclassified cardiomyopathy by the Expert Working Group (ESC) and the World Health Organization. According to the ESC (2023), LVNC is not a cardiomyopathy, but rather a phenotypic feature that may occur either alone or in combination with other developmental abnormalities, ventricular hypertrophy, dilatation, and/or left ventricular systolic dysfunction. The term «left ventricular noncompaction» is used to describe a ventricular phenotype characterized by prominent left ventricular trabeculations and deep intertrabecular depressions. Left ventricular (LV) noncompaction is often a familial trait and is associated with variants in a number of genes, including those encoding sarcomeric, Z-disc, cytoskeletal, and nuclear envelope proteins. Considering the absence of morphometric evidence of ventricular compaction in humans, the ESC (2023) recommends using the term «hypertrabeculation» rather than LVNC, especially when the phenomenon is transient or clearly begins in adults [9].

The left ventricular hypertrabeculation phenotype is caused by two different processes:

1. Genetic mutations that cause excessive development of the trabecular layer of the myocardium in relation to the compacted layer. Examples of monogenic inheritance of the phenotype are common, but the association with CMP and other phenotypes suggests that the genetic architecture of LVNC is much more complex. Genetic background may be important in the development of both noncompacted and compacted myocardium [1]. Recent data suggest that isolated LVNC is a distinct phenotype that may not be pathological [2]. Studies show that LVNC has a genetic cause for cardiomyopathies, channelopathies, and congenital heart defects [12,15,18,26]. Particularly in the pediatric population, LVNC has been identified with systemic diseases such as Barth syndrome, chromosome 1p36 deletion syndrome, and CMP, which are the result of neuromuscular, metabolic, or mitochondrial diseases [3,4,10]. Genetic factors play a more critical role in children than in adults, and severe LVNC usually occurs in childhood.

2. Physiological conditions such as pregnancy, athletic training, and hemoglobinopathies in response to conditions of mechanical overload lead to increased trabeculation in the left ventricle. These diseases are observed in adults and are often reversible. It may also be a variant of the norm in some people [5,20].

LVNC is characterized by a specific morphological appearance of the left ventricle («spongy myocardium»). The leak is localized mainly in the apical part of the left ventricular chamber with deep intertrabecular depressions (sinusoids) communicating with the ventricular cavity and is the result of arrest of normal embryogenesis [17,22,24]. Because of fetal developmental abnormalities in LVNC, the myocardium is divided into two layers:

1) a thick, noncompact, spongy layer of myocardium formed by trabeculae;

2) a thin layer of normal homogeneous myocardium with normal contractility.

Due to the low prevalence of the disease and the limited data available in the literature, there is no consensus on the definition, diagnostic criteria, pathogenesis, treatment, or reasonable prognosis of LVNC.

Aim: demonstration of clinical and morphologic aspects of noncompact left ventricular cardiomyopathy in a child, and to analyze the current principles of diagnosis of this disease.

We present a clinical case of LVNC (spongy) in a child aged 3 years 9 months. Postmortem examination of the child's body was performed in the Pathology Department of the MNPE «Regional Clinical Hospital of the Ivano-Frankivsk Regional Council», observing the requirements of biological safety. Heart sections were fixed in 10% buffered formalin for 28 hours, followed by conventional paraffin embedding. The histological sections were stained with hematoxylin and eosin and picrofuchsin by Van Gieson. Microscopic examination and photography of the samples were performed using an Olympus CX23 light-optical microscope with an LC30 digital CMOS camera and cell Sens Entry software.

The parents signed an informed consent to use the results of the study under the condition of confidentiality of the person in the reporting of the article. The study was conducted according to the provisions of the Declaration of Helsinki of the World Medical Association «Ethical Principles for Medical Research Involving Human Subjects». The design of the study with the information on the safety of research is part of the complex research work of the Department of Pathological Anatomy, «Improvement of efficiency of morphological diagnosis of diseases in adults and children with comorbid pathology» (state registration number 0121U110770), approved by the Ethics Committee of the Ivano-Frankivsk National Medical University.

Clinical case

The boy R. was born on 01/26/17 at full term from the second pregnancy. He was admitted to the Ivano-Frankivsk Regional Children's Clinical Hospital (IFR CCH) on 11/20/2020 with complaints of lethargy, general weakness, refusal to eat, decreased physical activity, nausea, and vomiting. Diagnosis at hospital admission: Functional gastric dyspepsia. Noncompact left ventricular myocarditis. From the medical history, it is known that the boy has been ill since June 2019, when he suffered from bilateral community-acquired pneumonia complicated by toxic syndrome and exudative pleurisy, after which changes in the heart were detected. In March 2020, he was treated in the Endocrinology Department of IFR CCH for impaired glucose tolerance, seizure syndrome on the background of hypoglycemia. He was treated under the supervision of a pediatric cardiologist. 2 weeks prior to hospitalization, the child's temperature rose to 38.2°C, followed by vomiting, refusal to eat, and lethargy.

The general condition of the child on admission was serious due to toxic syndrome and manifestations of chronic heart failure. On 26.11.20 at 08:30, the child's condition suddenly deteriorated, and he was taken to the Department of Anesthesiology and Intensive Care, and resuscitation measures were initiated. Resuscitation measures were ineffective; they were stopped at 09.40, and biological death was diagnosed. He was admitted to the hospital from 20/11/2020 to 26/11/2020.

Results of clinical and laboratory tests for 20-25.11.2020: Complete blood count: red blood cells (RBC) – $(4.71-5.53) \times 10^{12}/L$; hemoglobin – 119–130 g/L; erythrocyte sedimentation rate – 3–7 mm/hour; white blood cells (WBC) – $(7.5-13.2) \times 10^9/L$; eosinophils – 3–6%; young 1%: rod-shaped – 2–4%; segmented – 11–29%; lymphocytes – 78–53%; monocytes – 6–8%; platelets – 98–101%; hematocrit – 35.5–41.3 g/L.

Blood biochemistry: total protein – 55.5 g/L; urea – 1.8 mmol/L; creatinine – 27.0; total bilirubin – 15.29, direct – 2.10, indirect – 13.19 $\mu\text{mol}/L$; potassium – 4.88; sodium – 118.3; calcium – 2.20; chloride – 93.0; alanine aminotransferase – 28 units/L; aspartate aminotransferase – 30 units/L, sugar – 4.3 mmol/L.

Blood test for antibodies to COVID-19 dated 11/23/20, SARS-CoV-2 IgM – 0.096 (negative).

General urinalysis: protein – 0.033 g/L; WBC: 2–1–2 / High Power Field (HPF); RBC – 5–6 / HPF, epithelium – 3–4 / HPF, acetone negative.

Coagulogram: prothrombin time – 11.2 s, prothrombin index – 135.6%; International Normalized Ratio – 0.86, Activated Partial Thromboplastin Time – 4.3 s, fibrinogen – 2.66 g/L.

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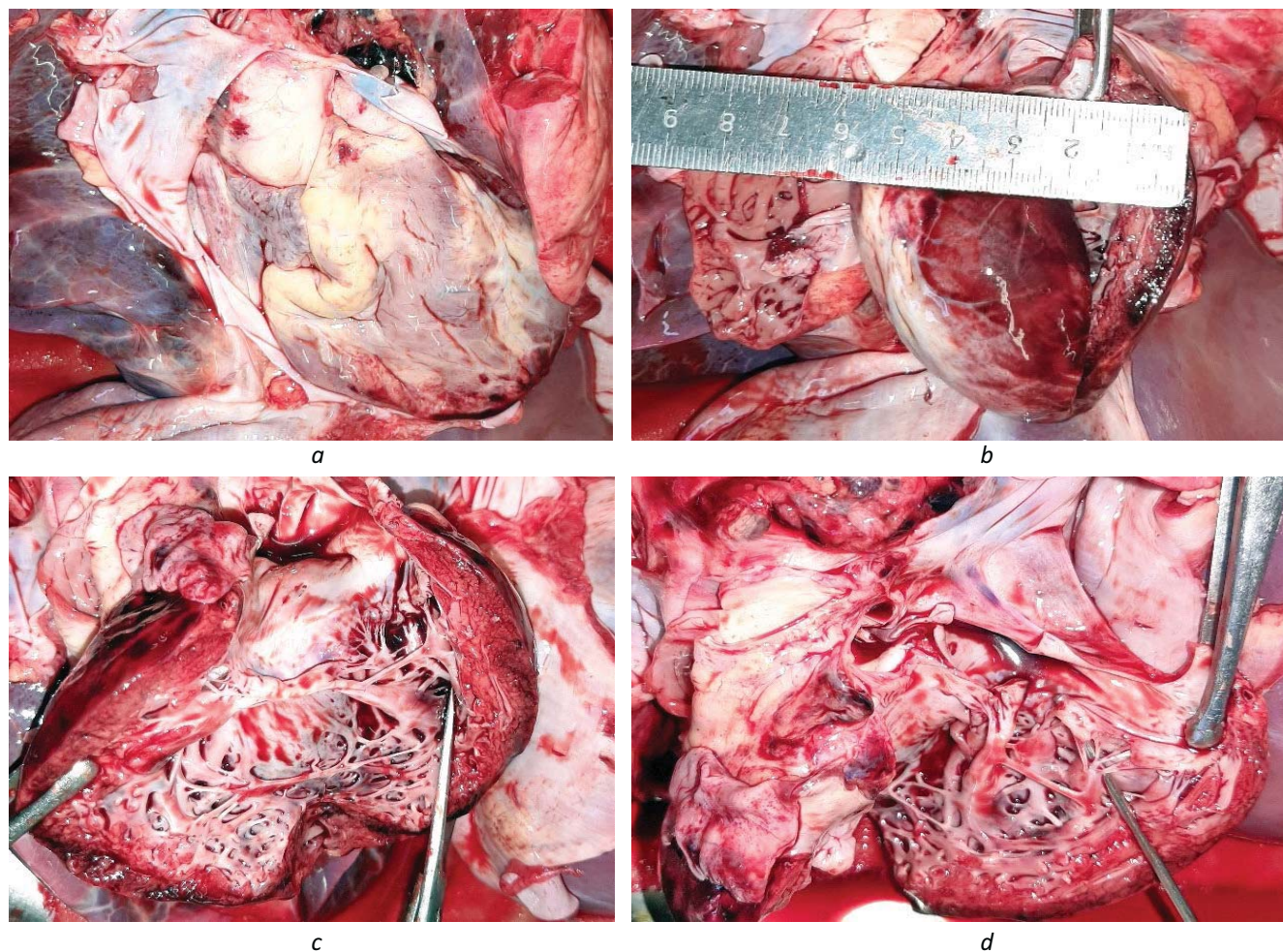


Fig. 1. Macroscopic changes in the heart: a, b – irregular oval heart, increased size; c, d – spongy myocardium, marked trabecularity of the left ventricular myocardium

Glycemic profile: glucose – 4.3 mmol/L – 6.5 mmol/L – 3.7 mmol/L – 3.7 mmol/L – 3.3 mmol/L.

Cardiac ultrasound (11.11.2020): left ventricle enlarged, left ventricular myocardium not compact. Myocardial contractility is reduced. Cardiomyopathy.

Electrocardiogram (ECG) (11/10/2020): Sinus rhythm is correct. Heart rate is 120 beats per minute. electrical axis of the heart (EAH) is deviated to the left. Expressed repolarization abnormalities.

Final clinical diagnosis (11/21/20): I. Underlying disease: Noncompact cardiomyopathy (myocarditis in June 2019) with left ventricular systolic dysfunction. Congestive heart failure of the third functional class. Chronic heart failure of the second stage. Complications: Acute left ventricular heart failure. Relative mitral valve insufficiency. III. Concomitant diseases: Adrenal insufficiency? Ataxic syndrome. Cognitive impairment. Functional dyspepsia of the gastrointestinal tract.

A pathological diagnosis was made based on the results of the autopsy:

Underlying Disease: I. Noncompact (spongy) cardiomyopathy with endomyocardial fibrosis of the left ventricle and sclerosis of the mitral valve leaflets. II. Right subtotal and left posterior bacterial bronchopneumonia (culture from 27.11.20, No.1528, lung tissue was massively contaminated with *St. aureus*, moderately with *Str. pyogenes*).

Complications: Edema and cerebral hemorrhage. Meningoencephalitis (bacterial culture from 27.11.20 No.1527 brain tissue isolated single *St. aureus*). Pulmonary edema and dystelectasis. Left-sided fibrinous pleural effusion with formation of fibrous casts. Interstitial pneumosclerosis with signs of pulmonary hypertension. Interstitial myocarditis, cardiogenic shock. Erosive gastropathy. Thrombo-hemorrhagic syndrome with multiple small epicardial hemorrhages, with focal hemorrhages in the lungs, kidneys, adrenal glands, thymus, stomach, serous membranes. Venous hemorrhage and parenchymal dystrophy of internal organs. Accidental thymic transformation stage IV–V.

Associated Diseases: Adrenal hypoplasia. Funnel chest. Cephalic hypospadias.

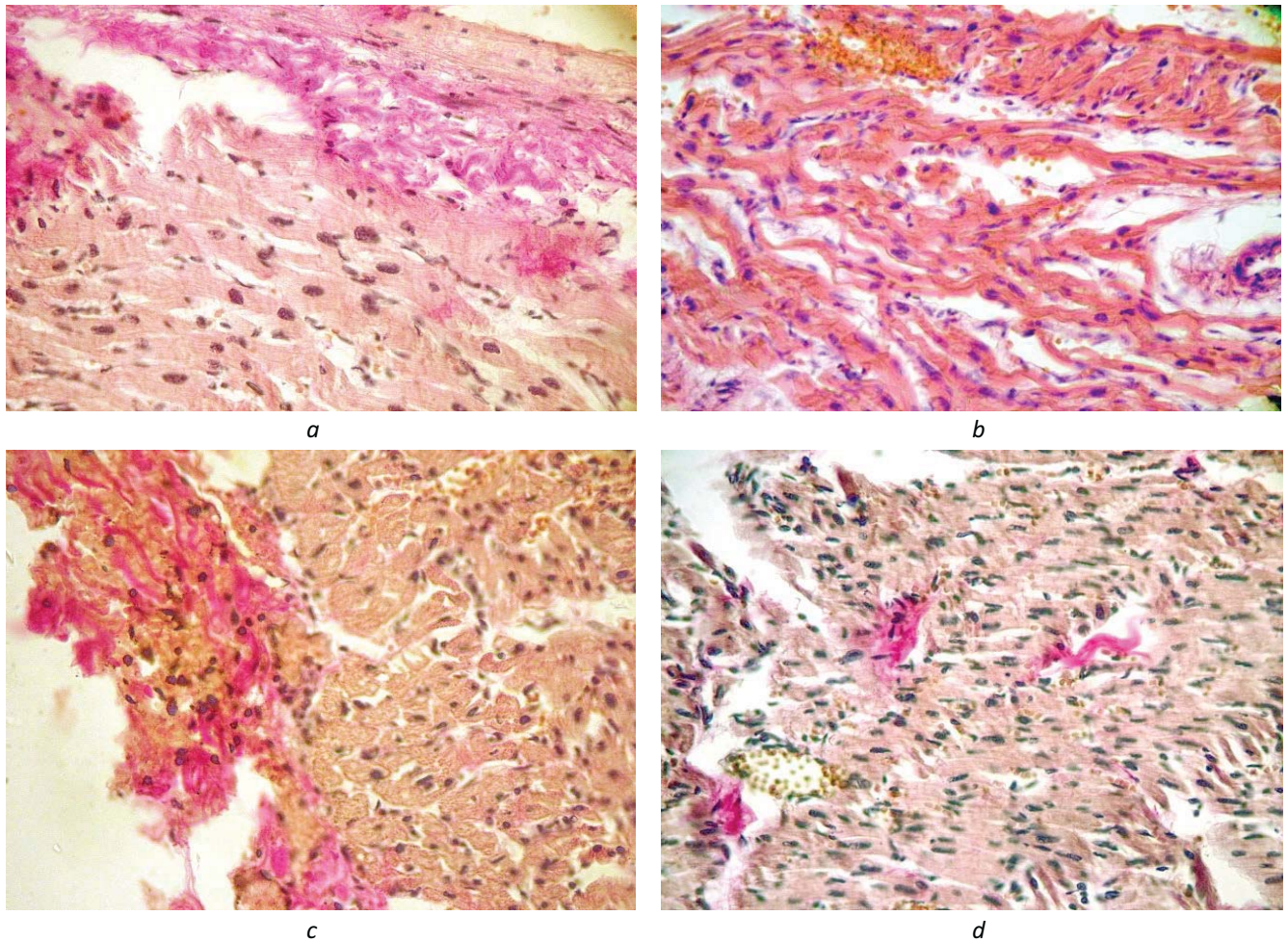


Fig. 2. Histologic changes in the heart: a – hypertrophy of cardiomyocytes with pleomorphic nuclei, foci of sclerosis; b – fragmentation, vacuolization of cardiomyocytes; c – thickened endocardium due to connective tissue proliferation, d – small-focal myocardiosclerosis. Note: b – with hematoxylin and eosin, a, c, d – with Van Gieson's picrofuchsin. Staining: a, b, c, d – $\times 10$

Noncompact (spongy) cardiomyopathy with left ventricular endomyocardial fibrosis and mitral valve leaflet sclerosis was characterized by peculiar macroscopic changes (Fig. 1). The heart was irregularly oval, enlarged $8.0 \times 7.5 \times 5.0$ cm, weighing 73 g. The right atrium and ventricle were slightly dilated and empty. The left atrium was slightly dilated. The left ventricular cavity is dilated in the transverse dimension, the apex is rounded, and the wall is thickened to an average of 1.2–1.5 cm. There is a marked trabecularity of the left ventricular myocardium, which is barely demarcated from the normal layer. The myocardium is elastic, spongy, and its upper part is accompanied by a crunch. Areas of sclerosis appear as whitish streaks. The subendocardial layers of the myocardium and papillary muscles are particularly dense. The parietal endocardium is thickened up to 1–2 mm, compacted in the upper half, and grayish-white («pearly») in color. Mitral valve leaflets are thickened and deformed.

Uneven hypertrophy of muscle fibers throughout the myocardium and papillary muscles was observed on histologic examination (Fig. 2). Fibers are arranged in dif-

ferent directions, subepicardially in severe angiomatosis. Muscle fibers are fragmented and vacuolated, edematous, with loss of transverse striation, with severe diffuse cardiosclerosis and scattered petrification. The endocardium is severely thickened by the growth of thick layers of elastic and collagen fibers. The coronary vessels are unevenly filled with blood, the inner lining is thickened, corrugated, the lumen is narrowed, and there is perivascular sclerosis. Focal subendocardial and perivascular lymphohistiocytic infiltration. The influx of vessels is obliterated and surrounded by a thick layer of elastic tissue.

Primary cardiomyopathies in children are a clinically and genetically heterogeneous group of rare heart muscle diseases. They can cause significant morbidity and mortality in children. In the pediatric age group, the number of reported cases of LVNC has increased over the past few decades, perhaps not due to an actual increase in cases, but rather as a result of improved accuracy of imaging techniques [19]. The clinical presentation of LVNC in children is complex. LVNC may be clinically asymptomatic or pres-

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ent with a variety of symptoms, including chest pain, dyspnea, and palpitations; however, three main clinical symptoms require urgent attention. The most common and important one is heart failure, which is associated with most other clinical symptoms and threatening complications such as thromboembolism and arrhythmias. In addition, these patients often have neuromuscular disease. They may experience fatigue [8], muscle pain, and elevated creatine kinase levels [25]. To diagnose cardiomyopathy in the presence of LVNC, the following tests are usually performed: evaluation of medical and family history, cardiac ultrasound, electrocardiography (ECG), echocardiography with functional assessment of LV size and function, Holter ECG (24 hours), cardiopulmonary exercise testing (CET), cardiac magnetic resonance imaging (CMR), and genetic testing [6,12,23]. The diagnosis of LVNC may be based on various criteria of echocardiography, magnetic resonance, and cardiac computed tomography, none of which are currently standardized to establish an accurate diagnosis. This further complicates the diagnosis of LVNC, which can manifest as other forms of cardiomyopathy, each with specific clinical features that may influence treatment strategies. Echocardiography is traditionally used to diagnose noncompacted left ventricles, and adding contrast significantly increases the sensitivity of this modality. However, cardiac MRI is now considered the method of choice for LVNC. There are no generally accepted echocardiographic criteria for noncompact left ventricle [22].

There are not enough data to establish guidelines for the management of LVNC. The treatment of LVNC is discussed in terms of anticoagulation, electrophysiological studies to prevent arrhythmias, and prevention of heart failure [7,16,24]. The genetic architecture of LVNC reveals both sporadic and familial patterns, with a significant proportion of individuals having a family history suggesting a genetic predisposition. The inheritance pattern is predominantly autosomal dominant, although X-linked and autosomal recessive forms have been documented [4]. Recent studies highlight the multifaceted genetic basis of LVNC, implicating pathways involved in cardiomyocyte differentiation, myocardial energy, and cellular architecture [21]. In 2020, Keiichi Hirono and colleagues first focused on genotype-phenotype correlation in a large pediatric cohort of LVNC patients with ion channel gene variants identified by next-generation sequencing, demonstrating a strong correlation of ion channel gene variants with arrhythmia phenotypes [11]. Genetic assessment of cardiomyopathies is an important and rapidly emerging clinical priority, as high-throughput sequencing is now feasible for clinical trials and routine interventions can improve survival, reduce morbidity, and improve quality of life. Moreover, specific interventions can be guided by genetic analysis. A systematic

approach is recommended: a comprehensive family history, expert phenotypic evaluation of the proband and at-risk family members to confirm the diagnosis and guide the selection and interpretation of genetic testing, referral to expert centers as needed, genetic testing with pre- and post-test genetic counseling, and specific guidelines for drug and device therapy as indicated. Evaluation of infants and children requires special expertise [10]. Despite decades of clinical observation and study of the relevant cardiac developmental processes, the molecular mechanisms underlying LVNC remain unknown. The likely multifactorial nature of LVNC development complicates the search for an underlying causal mechanism. A recent study showed that nearly 40% of children and 54% of adults with LVNC had no identifiable genetic mutations or family history of cardiomyopathy, suggesting that non-genetic factors such as abnormal exercise conditions may also play a role [21]. Among the morphological changes in the heart, the most important are hypertrabecular «spongy» myocardium, hypertrophy of muscle fibers, pleomorphism of cardiomyocytes nuclei, endocardial fibrosis, and small-focal myocardial sclerosis.

There are different opinions regarding how endocardial fibrosis occurs. It may be caused by an immune response, the result of local blood flow abnormalities, or caused by hemodynamic and mechanical factors around the trabecular myocardium [11]. There are isolated reports of myocarditis and focal cardiomyocyte necrosis in LVNC [13]. In our case, the interstitial myocarditis is obviously due to a concomitant severe bacterial pneumonia. Therefore, it would not be appropriate to distinguish it as a pathomorphologic criterion of LVNC.

For a better understanding of the nature of LVNC, a larger number of cases with pathologic examination is needed.

Conclusions

1. The presented clinical case allows us to identify the main pathomorphologic criteria of LVNC: hypertrabecular «spongy» myocardium, hypertrophy of muscle fibers, pleomorphism of cardiomyocytes nucleus, endocardial fibrosis, and small-focal myocardial sclerosis.

2. Despite the lack of standardized methods for diagnosing LVNC, the traditional and main ones are ultrasound, echocardiography, magnetic resonance, and computed tomography of the heart.

3. Genetic evaluation of cardiomyopathies is an important clinical priority. Genetic testing and phenotyping will allow appropriate treatment of specific LVNC targets, which will improve survival, reduce morbidity, and improve quality of life.

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