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The myocarditis as a regular phenomenon in primary noncompact myocardium: clinical and morphological diagnostics, significance and treatment

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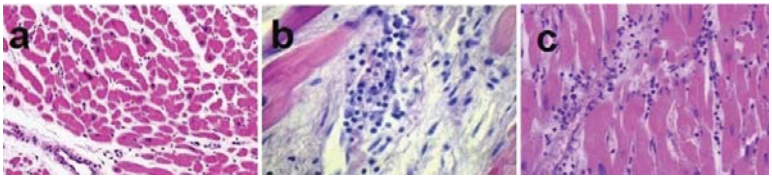
Purpose: To estimate the frequency of myocarditis in adult patients with noncompact myocardium (NCM) of the left ventricle (LV), its influence on the disease, the outcome and results of treatment.
Methods: We included 103 adult patients with NCM (61 males, 45.6±14.9 years). The average EDD LV was 6.0±0.8 cm, LV EF 38.8±14.5%. To diagnose NCM were performed Echo-CG, CT (n=81) and MRI (n=39). We performed NGS sequencing, followed by Sanger sequencing of detected variants (revealed in 9% of patients in the genes MYH7, MyBPC3, LAMP2, DES, DSP, TTN). The examination also included anti-heart antibodies and viruses (PCR) study and morphological study in 19 patients (endomyocardial biopsy in 14, intraoperative biopsy in 1, explanted heart study in 1 and autopsy in 3). The mean follow-up was 12 [2; 32] months.
Results: The myocarditis was diagnosed in 55 (53.4%) patients with NCM: in 19 patients by morphological study (active in 10 and borderline in 9 patients) and in 36 patients on the basis of a complex examination (the relationship with a respiratory infection, a 3–4-fold increase in the anti-heart antibodies, the viral genome in the blood, pericardial effusion, subepicardial LGE). The myocarditis was virus-positive in 17 patients (36.1%). The parvovirus B19, human herpes virus type 6, cytomegalovirus, herpes simplex virus and Epstein-Barr virus were detected in the myocardium in 8 patients (42.1%), and in 14 patients (25.5%) in the blood. Pathogenic genetic variants were revealed in four patients with morphologically verified myocarditis. The rate of myocarditis depended on the clinical form of NCM: 44.4% in arrhythmic form, 12.5% in chronic ischemic form, 57.5% in dilated pattern and 50.0% in association of NCM with other cardiomyopathies. In 10% of all patients, acute myocarditis was the first manifestation of the disease. In 5% the myocardial necrosis was the main manifestation of the myocarditis. The association of NCM with myocarditis led to more severe myocardial dysfunction (NYHA class 2 [1; 3] v 1.75 [0; 2], $p<0.01$, EF 33.8±13.5 v 44.7±13.6%, $p<0.001$), higher rate of the nonsustained ventricular tachycardia (67.3% v 29.3%, $p<0.01$), appropriate shocks of defibrillators (38.9% v 0, $p<0.05$), death (16.4 and 4.2%, OR 5.75, 95% CI 1.21–27.43, $p<0.05$), and heart transplantation (7.3% v 2.1%, $p>0.05$). Only in patients with myocarditis, there was a significant increase in EF (in acute myocarditis from 25.4±7.9 to 38.6±9.5%, $p<0.01$), decrease sizes of the LV and systolic pulmonary artery pressure as a result of basic therapy.
Conclusion: Myocarditis is a regular phenomenon that develops in half of patients with primary NCM. The nature of myocarditis in NCM may be different (primary infectious and immune, secondary in response to genetic/ischemic damage to cardiomyocytes). However, it leads to a significant deterioration of structural and functional parameters, an increase in life-threatening arrhythmias, unfavorable outcomes and requires basic therapy.



Morphological verification of myocarditis