

Myocardial Noncompaction: Unusual Presentation – Case Report

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Introduction

Myocardial noncompaction (MNC) is considered a cardiomyopathy classified as congenital, initially described in pediatric patients, but it has recently been detected in adults, whose clinical characteristic is congestive heart failure.¹ Its incidence in adults is 0.05%. Diagnosis on echocardiography is difficult and is done in only 9% of cases. Cardiac magnetic resonance imaging (CMRI) currently corroborates the findings of transthoracic Doppler echocardiography.²

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Assessment of 19-year-old female patient with palpitations and tiredness referred for cardiological evaluation. Cardiac auscultation with no murmurs, blood pressure of 120 x 70 mmHg and heart rate of 76 bpm. Resting electrocardiogram showed sinus rhythm with conduction disturbance of the left branch and HR:94 bpm. The patient had no other comorbidities, reporting physical activity at the gym in the previous month — with functional capacity (NYHA) I.

The assistant physician requested Holter and transthoracic echocardiography (TT-ECHO). Holter showed: irregular sinus rhythm (respiratory sinus arrhythmia) with mean heart rate of 83 bpm, ranging from 51 to 174 bpm. Very frequent ectopic ventricular activity (149 EV/h), polymorphous, isolated, bigeminated, 110 pairs, short three-beat accelerated idioventricular pace and short non-sustained three-beat ventricular tachycardia at 118 bpm:3% of ectopic ventricular beats, two episodes of tachycardia, bigeminy and isolated and paired EV (Figure 1).

Transthoracic echocardiography showed moderate enlargement of the left ventricle (LV) - LV end-diastolic diameter:61 mm and LV end-systolic diameter:48 mm, diffuse hypokinesis with an ejection fraction (LVEF) of 39% and lush prominent ventricular muscle trabeculae in previous segments, side and bottom of the LV, mild tricuspid and mitral regurgitation (Figure 2).

Keywords

Heart Defects, Congenital/complications; Heart Defects, Congenital/diagnosis; Myocardium/pathology; Arrhythmias, Cardiac/complications.

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Because of the findings on transthoracic Doppler echocardiography, CMRI was conducted and revealed: myocardium with lush trabeculations in inferolateral and anterior LV medioapical segments. The relationship between the compacted and the noncompacted portion of the myocardium was greater than 2.3 in these segments. LVEF:38% (Figure 3). The assisting physician initiated drug treatment with beta-blocker and angiotensin-converting enzyme (ACE) inhibitor. The patient is currently asymptomatic and in functional class I (NYHA).

Discussion

It was thought that MNC was rare.³ However, the prevalence among unselected patients referred for TT-ECHO in a tertiary center ranges from 0.014% to 1.26%, and in patients with heart failure it varies from 3% to 4%. There is a genetic linkage in 50% of cases where patients have close relatives with cardiomyopathy. Genetic confirmation or exclusion of MNC is currently a challenge with genetic mutation associated with the disease. Sometimes it cannot be identified and its absence does not exclude MNC.⁴

It was reported that more than 25% of the individuals of the sample without heart disease or symptoms could meet imaging criteria for MNC, as we have recently shown in the Multi-Ethnic Study of Atherosclerosis (MESA).⁵

The prominent myocardial trabeculae and deep intertrabecular recesses are likely to result from trapping at the beginning of the compaction process during embryogenesis and are, therefore, morphological characteristics of the left ventricular noncompaction (MNC).⁶

As shown by Oechslin et al. in the Journal of the American College of Cardiology in 2000, the clinical presentation and the consequences from the MNC range from lack of symptoms to severe systolic dysfunction, heart failure, arrhythmias, thromboembolic events and sudden cardiac death.⁷

Comparison of patients affected with excessive trabeculation, systolic dysfunction of the LV with individuals who have imaging criteria for MNC and preserved LV systolic function is essential for clinical management.

In patients with a clear diagnosis of MNC with compromised LV systolic function, treatment should follow the current guidelines of HF, such as the 2013 recommendations of the American College of Cardiology Foundation and the American Heart Association, which contain a specific analysis of ACE inhibitors, beta-blockers, cardiac resynchronization therapy and implantable cardioverter defibrillators.⁸

Furthermore, patients with MNC with and without atrial fibrillation (AF) are at increased risk for thromboembolic complications. Patients with MNC and AF must receive

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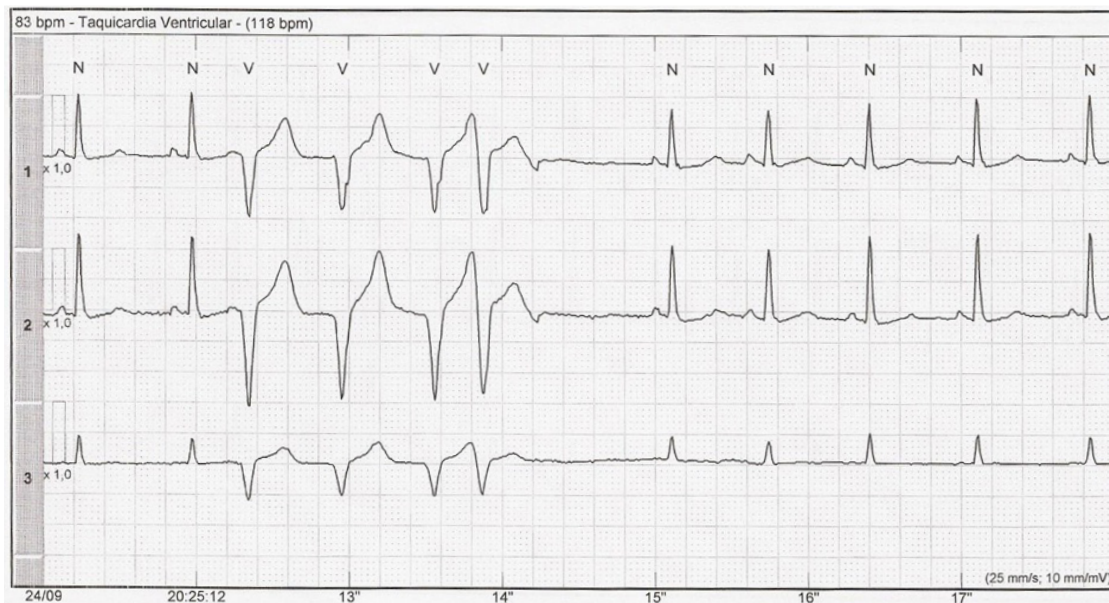


Figure 1 – Holter: ventricular tachycardia.

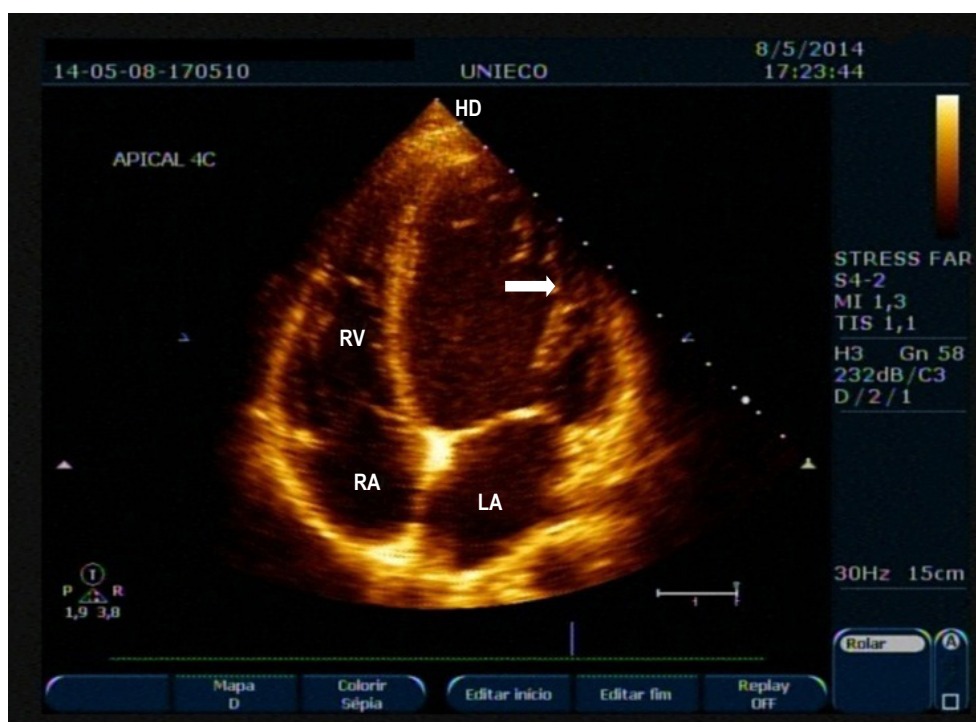


Figure 2 – TT-ECHO: apical window – four chambers observing exuberant trabeculations in LV inferolateral wall (arrow). RV: right ventricle; RA: right atrium; LA: left atrium.

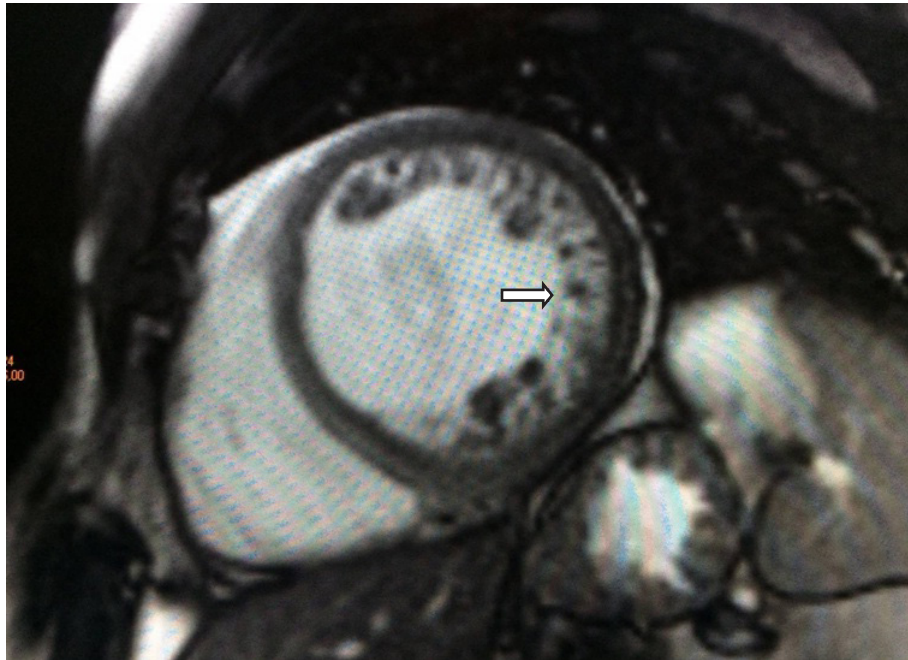


Figure 3 – CMRI: short LV axis observing exuberant trabeculations in anterolateral LV wall (arrow).

anticoagulation according to standard guidelines (if CHA2DS2-VASc score ≥ 1). Anticoagulation should also be considered when they did not meet the resynchronization criteria, especially in patients with ejection fraction $< 40\%$.

According to Ashrith et al., published recently in the Journal of Cardiovascular Magnetic Resonance, patients with established HF and cardiomyopathy with more exuberant LV trabeculations showed a small improvement in LV function after initiation of treatment and remained with more symptoms compared to similar patients, but with less LV trabeculations.⁹

Treatment of patients with MNC, however, has not been established in large cohort or clinical trials, and all recommendations must be justified and considered separately.

In the most common clinical scenario, where we find excessive trabeculations in a patient suspected to have MNC, we must avoid the stereotype patient diagnosed with MNC.¹⁰ MNC patients usually have progressive and gradual worsening of HF (not abrupt as reported in the case).

Individuals with high suspicion of MNC in view of the symptoms or positive family history and preserved systolic function should remain under careful monitoring, especially with cardiac imaging (TT-ECHO and CMRI). In clinical practice, it is also recommended to do a family screening of first-degree relatives.¹¹

Conclusion

In summary, a high degree of LV trabeculae is a common finding in cardiac imaging. In most cases, they do not

involve cardiomyopathy. Those patients with recent onset of arrhythmia and HF (common finding) must have extended investigation, since they may have MNC (unusual finding). MNC diagnostic labeling and proposed follow-up strategies must be done using both the clinical information and cardiac imaging investigation.

Authors' contributions

Research creation and design: Saback NG, Souza ME, Siqueira MHA; Data acquisition: Saback NG, Souza ME, Zeringota AMA, Siqueira MHA; Analysis and interpretation of data: Saback NG, Souza ME, Neiva AA, Cabral ACR, Barrote S, Zeringota AMA, Siqueira MHA; Statistical analysis: Saback NG, Barrote SL, Siqueira MHA; Manuscript drafting: Saback NG, Siqueira MHA; Critical revision of the manuscript as for important intellectual content: Saback NG, Siqueira MHA.

Potential Conflicts of Interest

There are no relevant conflicts of interest.

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Academic Association

This study is not associated with any graduate program.

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