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Atrial Tachycardia Revealing Non Compaction of the Left Ventricle: A Case Report and Literature Review

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Abstract **Case Report**

Non-compaction of the left ventricle is a rare hereditary cardiomyopathy, belonging to a group of so-called "unclassifiable" cardiomyopathies, whose clinical manifestations and prognosis vary from patient to patient. Diagnosis is based on cardiac imaging, represented mainly by transthoracic echocardiography, as well as cardiac magnetic resonance imaging. Transthoracic echocardiographic screening is systematically performed in the family of every patient with this pathology. We report here the case of a 64-year-old man admitted to our department for the management of palpitations in connection with atrial tachycardia on a terrain of non compaction of the left ventricle.

Keywords: Atrial Tachycardia, Palpitations, Non-Compaction of the Left Ventricle.

INTRODUCTION

Left ventricular non-compaction (LVNC) is a rare hereditary cardiomyopathy caused by incomplete myocardial compaction of the left ventricle (LV) during embryonic development. Positive diagnosis is based essentially on transthoracic echocardiography and cardiac magnetic resonance imaging (MRI). Since its first presentation, there have been only a few isolated cases, and small cohorts of patients, especially in children. Although there is new interest and growing recognition of this anomaly, the diagnostic criteria, clinical manifestations and prognosis of this uncommon congenital disease, which is part of the non-stratified cardiomyopathies, are still poorly investigated. The discovery of this pathology requires systematic family screening. Patients with non-compaction of the left ventricle often present with cardiac arrhythmias. These anomalies may include tachycardias or, in some cases, atrial or ventricular fibrillations. We report the case of a patient, who presented with palpitations, and in whom non compaction of the left ventricle was suspected on echocardiography and confirmed on MRI.

CASE PRESENTATION

A 64-year-old patient with modifiables cardiovascular risk factors: well-balanced type 2 diabetes (glycated hemoglobin: 6.3%), well-controlled arterial hypertension on dual therapy, with no particular history. He consulted the cardiology department for several episodes of palpitations with exertional dyspnea (New York Heart Association grade II), which he had experienced for several months. There was no family history of cardiomyopathy.

On clinical examination, he showed no dysmorphia, his heart rate was 140 beats per minute, with a regular rhythm, his blood pressure was 134/78 mm Hg, and he had a holosystolic murmur at the mitral focus, rated 2/6. The rest of the examination was normal.

Electrocardiogram (ECG) revealed a 2/1 atrial tachycardia with mean ventricular rate at 142 beats per minute, Heart axis to the left with left anterior Hemi block (Figure 1).

Transthoracic echocardiography showed the presence of multiple left ventricular trabeculations and deep intertrabecular recesses (Figure 2; 3).

Cardiac magnetic resonance imaging revealed extensive trabeculation of the left ventricle with a ratio of non-compacted area to compacted area greater than 2.3, consistent with non-compaction of the LV with slightly dilated left ventricle, with moderate left ventricular systolic dysfunction, left ventricular ejection fraction at 47%, with absence of left intraventricular thrombus (Figure 4; 5).

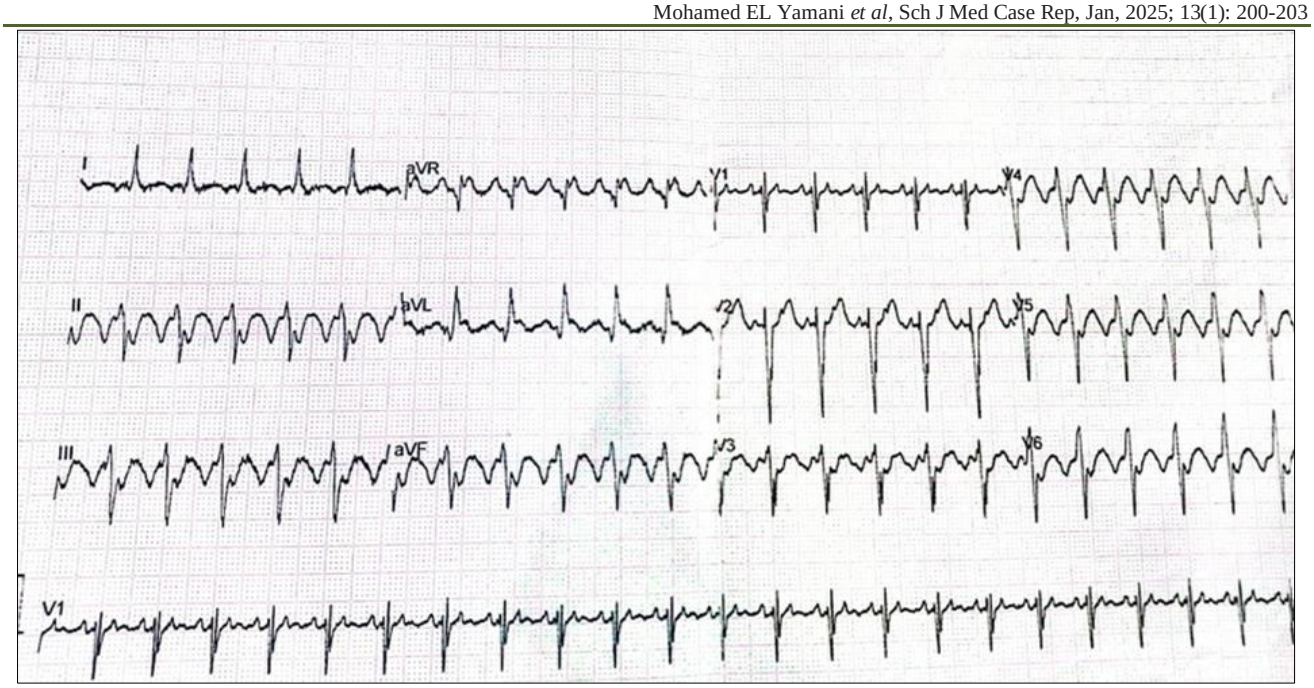

Figure 1: EKG showed 2/1 atrial tachycardia with an average ventricular rate of 142 beats per minute, with left anterior hemi block

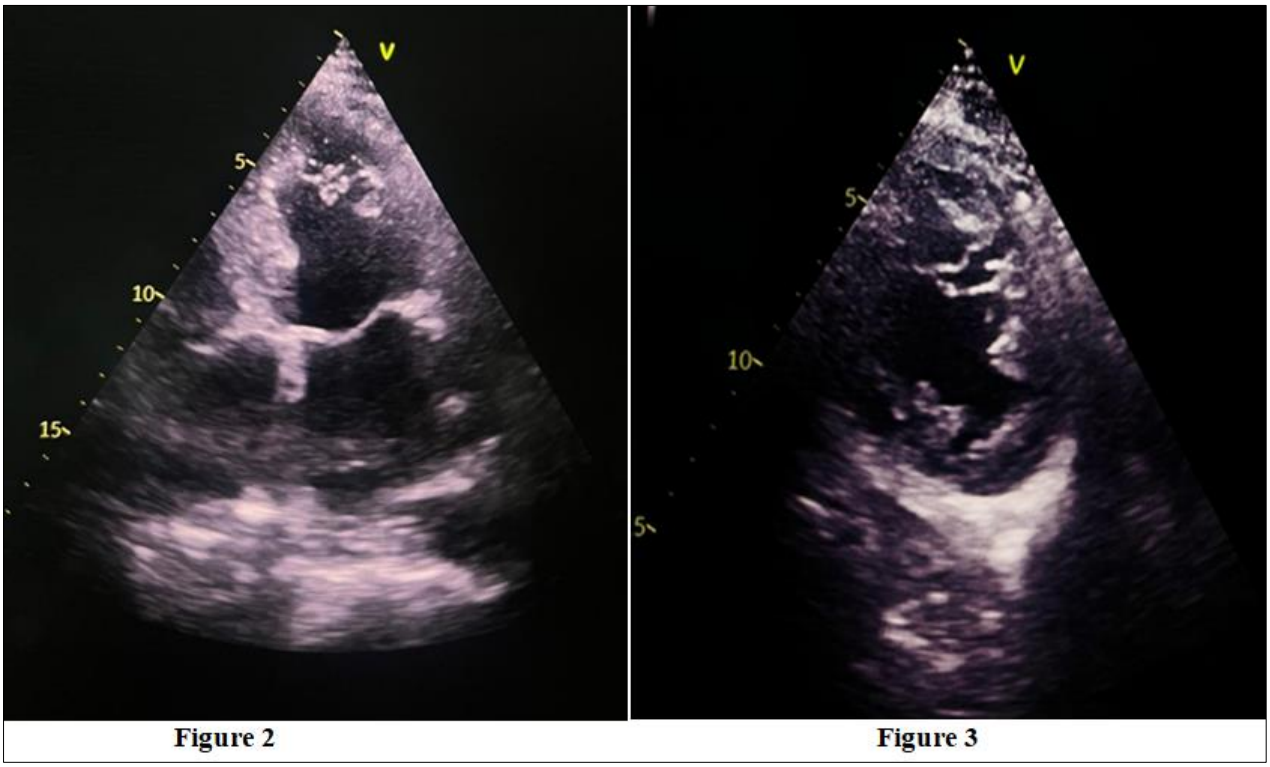

Figure 2 **Figure 3**

Figure 2; 3: four cavities and short-axis echocardiography showing trabeculations and intertrabecular recesses in the left ventricle

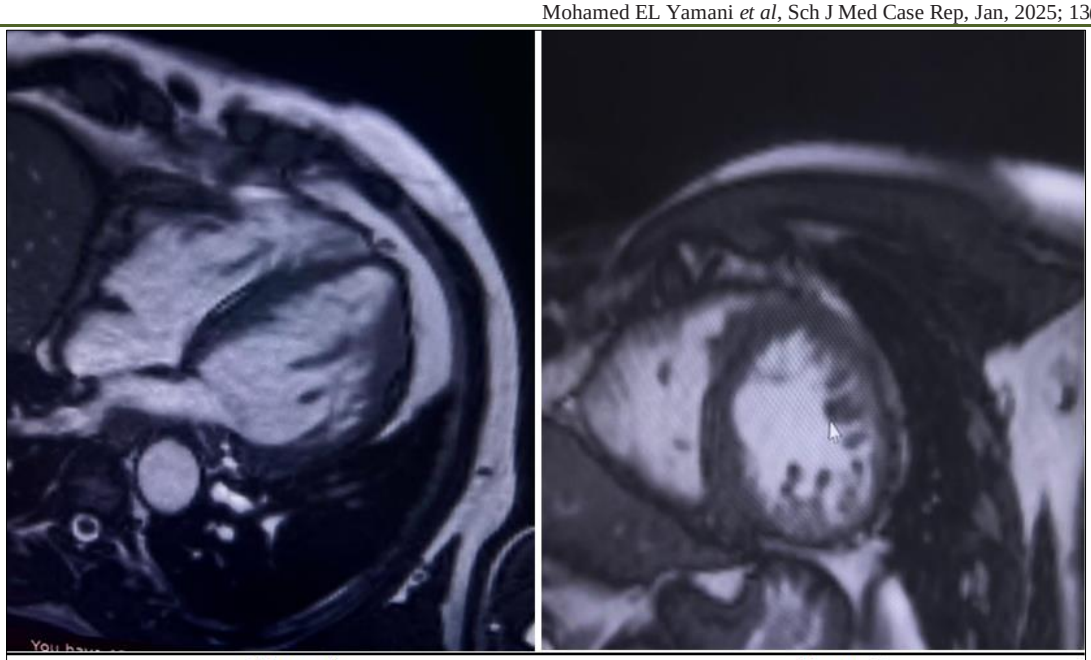

Figure 4 Figure 5

Figure 4; 5: Cardiac MRI showing many trabeculations of the left ventricle, with a ratio of non- compacted area to compacted area greater than 2,3

DISCUSSION

The frequency of LVNC is poorly defined around the world, as it is under-diagnosed and under-recognized. Various studies have revealed a prevalence ranging from 0.014 to 0.45% (Ritter *et al*., 1997) . The disease is inherited, with an autosomal dominant mode (Gebhard *et al*., 2012). In literary reviews, the non compaction of the left ventricle is considered a condition that mainly affects men (Tukker *et al*., 2023).

Non-compaction of the left ventricle results from the cessation of normal myocardial embryogenesis. It is mainly characterized by the formation during embryonic life of many deep myocardial ventricular trabeculations, most often in the apex of the left ventricle (Fennira *et al.*, 2019) . In terms of genetic origin, familial forms linked to mutations of the G4.5 gene on chromosome Xq28 have been described, so systematic family screening is essential when this pathology is discovered(Fennira *et al*., 2019).

The imaging, represented essentially by echocardiography and cardiac magnetic resonance imaging, is the cornerstone for establishing the diagnosis, prognosis and monitoring of any patient with LVNC (Fennira *et al*., 2019) . Because of its ease of access and simplicity, transthoracic echocardiography remains the gold standard for the diagnosis of LVNC. Diagnosis is established using two echocardiographic criteria: Jenni's criteria, based on the existence of a two-level structure (Jenni *et al*., 2001); and Chin's criteria, which emphasize cavity depth in relation to trabeculation height .

These criteria can be used to describe :

- Multiple left ventricular trabeculations;
- Deep intertrabecular recesses;
- Color Doppler flow within the recesses and in communication with the left ventricular cavity;

And classically, the diagnosis of non-compaction is confirmed when the ratio of non-compacted area to compacted area is greater than 2 (in systole). In difficult cases, other echocardiographic techniques can be used for the diagnosis: contrast enhancement, three-dimensional echocardiography, speckle tracking as well as tissue Doppler imaging (Gebhard *et al*., 2012). In the case of our patient, echocardiography alone allowed us to suspect the diagnosis, with the presence of multiple LV trabeculations and deep intertrabecular recesses in the apical and anterolateral wall, confirmed by cardiac MRI. Cardiac MRI is a more effective examination, complementing the informations provided by transthoracic echocardiography to better highlight areas of non-compaction and trabeculations, particularly in doubtful cases and in patients with poor echogenicity (Petersen *et al*., 2005).

At present, in the absence of clear recommendations for the management of LVNC, treatment is based on conventional heart failure therapeutics (Niemann *et al*., 2012). Anticoagulant treatment remains necessary in cases of severe LV dysfunction, supraventricular rhythm disorders and/or the presence of a history of thromboembolism (Stöllberger & Finsterer, 2005). Whereas for the automatic implantable defibrillator, in addition to the standard indications ,some authors recommend it for

patients with a high risk of death, based on certain factors: dyspnea New York Heart Association class III/IV, chronic atrial fibrillation, bundle branch block, sustained ventricular arrhythmias, a dilated left atrium, and a higher LV end-diastolic diameter at initial presentation(Petersen *et al*., 2005). Our patient presented with supraventricular arrhythmia, in addition to ventricular hyperexcitability on holter electrocardiogram, hence the indication for anticoagulation; introduction of Beta-blocker as well as treatment of heart failure with moderate left ventricular ejection fraction and ablation of atrial tachycardia, implantable defibrillator is quite rarely performed as first intention in our social and economic context. Because of the high risk of asymptomatic arrhythmias in LVNC, periodic rhythmic holter screening is essential.

CONCLUSION

LVNC is a cardiomyopathy characterized mainly by the presence of numerous ventricular myocardial trabeculations, localized mainly at the apex of the left ventricle. Imaging techniques, notably echocardiography and cardiac MRI, have improved the diagnosis of this pathology, although standardized criteria are still needed to reduce the risk of over- and under-diagnosis. The existence of cases discovered in adulthood sometimes suggests pathophysiological pathways other than the cessation of embryological phenomes, hence the complexity of diagnosis. It illustrates the challenges and opportunities associated with the management of rare heart diseases, underlining the importance of interdisciplinary collaboration and ongoing research to better meet patients' needs.

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