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A Young Healthy Woman with Supraventricular Tachycardia Reveals an Underlying Left Ventricular Non-compaction Cardiomyopathy: A Rare Case Report

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

LVNC (left ventricular non-compaction) is a rare congenital cardiomyopathy with a reported incidence of 0.05% in adults. It can occur in isolation or affect both ventricles. It's characterized by prominent LV trabeculae and deep intertrabecular recesses which are filled with blood from the

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ventricular cavity without evidence of communication to the epicardial coronary artery system. Frequent premature supra ventricular tachycardia as unique finding in LVNC cardiomyopathy is rare manifestation of this disease. We report a case of a frequent persistent supraventricular tachycardia as first manifestation of a patient with LVNC cardiomyopathy in a young healthy woman who despite radio frequency ablation therapy of the supraventricular tachycardia remains symptomatic. The patient was later placed on medical therapy based on a non-cardio selective beta-blocker with a good clinical outcome without recurrent of supra-ventricular arrythmias.

Keywords: Left ventricular non-compaction; supra-ventricular tachycardia; premature ventricular complexes; beta-blockers.

1. INTRODUCTION

"Isolated LVNC (left ventricular non-compaction) is characterized by prominent LV trabeculae and deep intertrabecular recesses which are filled with blood from the ventricular cavity without evidence of communication to the epicardial coronary artery system. It's an uncommon unclassified cardiomyopathy or genetic myocardial disorder [1,2], which is understood by the explanation that during the embryological heart development process the myocardial compaction is disrupted through unknown mechanisms, and the latter, LVNC could be acquired and developed on time" [1,3]. Frequent premature supra ventricular tachycardia as unique finding in LVNC cardiomyopathy is rare manifestation of this disease.

"Clinical manifestations and the age of symptom onset vary greatly and depend on the associated cardiac abnormalities. Reported complications include systemic thrombo-embolism, ventricular arrhythmias, sudden cardiac death, and progressive heart failure" [4,5,6]. "The diagnosis of LVNC requires demonstration of large and/or numerous ventricular trabeculae, a normal or thin compacted myocardial layer, and the presence of deep intertrabecular recesses that communicate with the left ventricular cavity either by trans thoracic echocardiography (TTE) or cardiac -MRI (Magnetic resonance imaging). However, in spite of the efforts of numerous investigators, there is still no consensus on the imaging criteria for LVNC" [7,8]. Despite this, contrast TTE has been the first diagnostic test of choice for noncompaction [9]. Herein, we describe a case of a rare supraventricular tachycardia as the first symptom manifestation of LVNC cardiomyopathy in a young healthy woman who despite of radio-frequency ablation therapy remains symptomatic. The patient was later placed on medical therapy based on a non-cardio-selective beta-blockers with a good clinical outcome and absence of recurrent supra-ventricular arrythmias.

"In common with most myocardial disorders, LVNC is often a heritable trait [10], and sometimes may be caused by genes implicated in classical cardiomyopathies such as sarcomeric (ACTC1, MYH7, MYBPC3, TNNT2, and TPM1), desmosomal (DSP and PKP2), nuclear envelope protein (LMNA), and z-disk (LDB3), as well as genes implicated in muscular dystrophy, and mitochondrial and ion channel disorders" [11].

2. CASE REPORT

A 44-year-old patient went to the emergency department after an intense sporting activity of running more than 6 km, presented symptoms like palpitations associated with chest discomfort which prompted the call for help at the end of the race. On admission, the patient's symptoms subsided except the persistence of palpitation felt like as a 'jump heart beats'. In addition, the patient has a history of hypothyroidism of autoimmune origin under LEVOTHYROXINE 50ug per day, well followed by her attending physician with final control of normal TSH value without a known family history of heart disease. The clinical examination finds a patient in good general state, hemodynamically and respiratory stable with a systolic blood pressure value of 119mmHg and diastolic blood pressure of 70mmHg, 99% oxygen saturation in ambient air without no signs of heart failure. Cardiac auscultation finds an irregular rhythm with no abnormal heart sounds. The ECG, showed a supra-ventricular tachycardia probably an atrioventricular nodal re-entry tachycardia (AVNRT) (Fig. 1) which could not be reduced by medical therapy, 3 days later, patient underwent an electrophysiology study which was performed by inserting catheters and then wire electrodes in order to measure electrical activity through blood vessels that enter the heart. The latter which confirmed an AVNRT type slow-fast reduced by capture and enhancement requiring thus an ablation therapy by standard radio-

frequency at the basal part of the inter-atrial septum.

Successful ablation at the level of the slow conduction way was carried out, without post-ablation complication or disturbance of nodal conduction and also without disturbance of infra-nodal conduction with HV max at 42ms. The post -ablation ECG was normal (Fig. 2).

The transthoracic echocardiography performed showed a non-dilated left ventricle (LV) with an end-diastolic diameter of 47 mm, site of intra-LV trabeculations and inter-trabecular recesses evoking non-compaction of the LV with a NON-compact LV/compact LV ratio during systole greater than 2 predominant on the antero-lateral wall and the apex of the LV (Fig. 3).

The systolic function of the left ventricle preserved with an LVEF at 60% without mitro-aortic valve disease with the good longitudinal systolic function of the RV without pulmonary hypertension. The biological assessment was unremarkable. A cardiac MRI was carried out on the patient confirming the presence of a ventricular non compaction (Fig. 4 A-B).

Our patient was placed on medical therapy after a control 24-hour Holter ECG which confirmed frequent premature ventricular complexes (PVCs) Fig. 5.

A non-selective cardiac beta-blocker based on her history of autoimmune thyroiditis was used (propranolol) dose at 40mg daily with a good clinical outcome. A regular follow-up was carried out with the repetition of a 24-hour Holter ECG after 3 months.

3. DISCUSSION

"LVNC is a rare congenital cardiomyopathy with a reported incidence of 0.05% in adults. It can occur in isolation or affect both ventricles" [12]. "The diagnosis is usually established by identifying the morphologic diagnostic criteria proposed by Jenni and al. on transthoracic echocardiography [13] and MRI which is the second modality, with an excellent spatial resolution, is the best method because it has not only a diagnostic but also a prognostic role" [14]. "LVNC can be diagnosed with ECG-triggered low-dose CCT (cardiac computed tomography) with a very good correlation of NC:C ratio in TTE and CCT" [15]. Our patient was diagnosed and confirmed with coupled TTE – cardiac MRI.

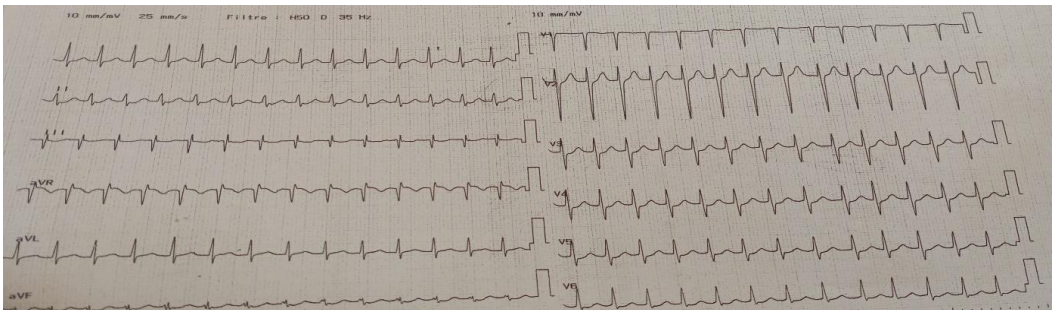

Fig. 1. Electrocardiogram at the emergency department: Supraventricular tachycardia probably junctional tachycardia

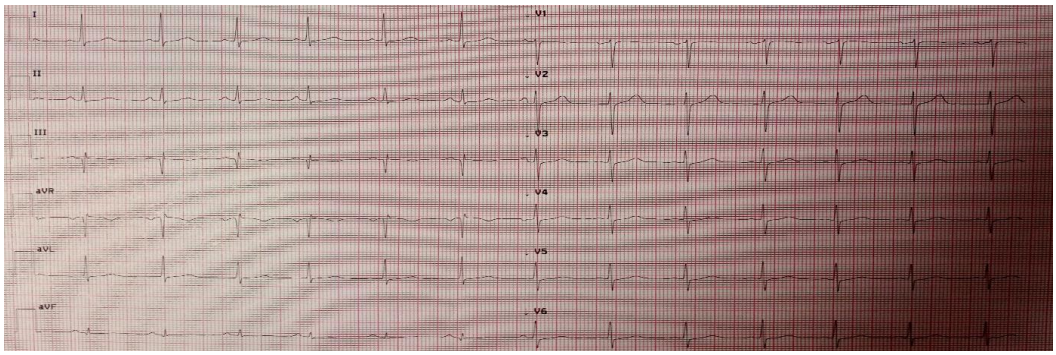

Fig. 2. Electrocardiogram post-ablation by radio-frequency: Regular sinus rhythm

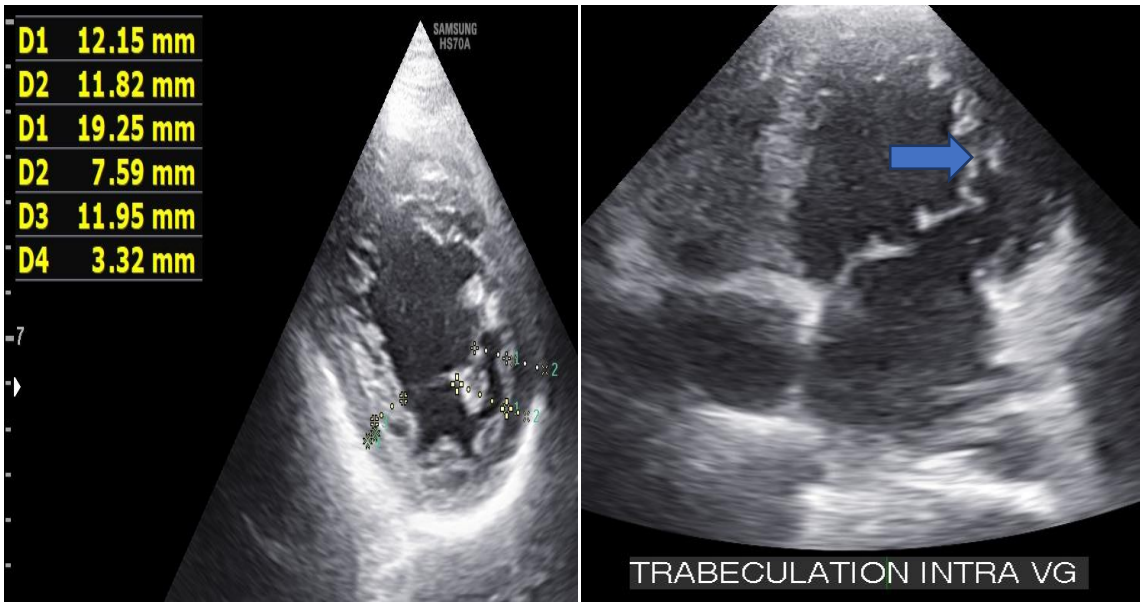

Fig. 3-A. Trans thoracic echocardiography; short axes view: Shows left ventricular trabeculations predominant at the antero-lateral wall with a ratio of 2.7

and

Fig. 3-B. Trans thoracic echocardiography; 4 chambers apical view: Shows left intra ventricular trabeculations predominant at the antero-lateral wall of a non-dilated heart.

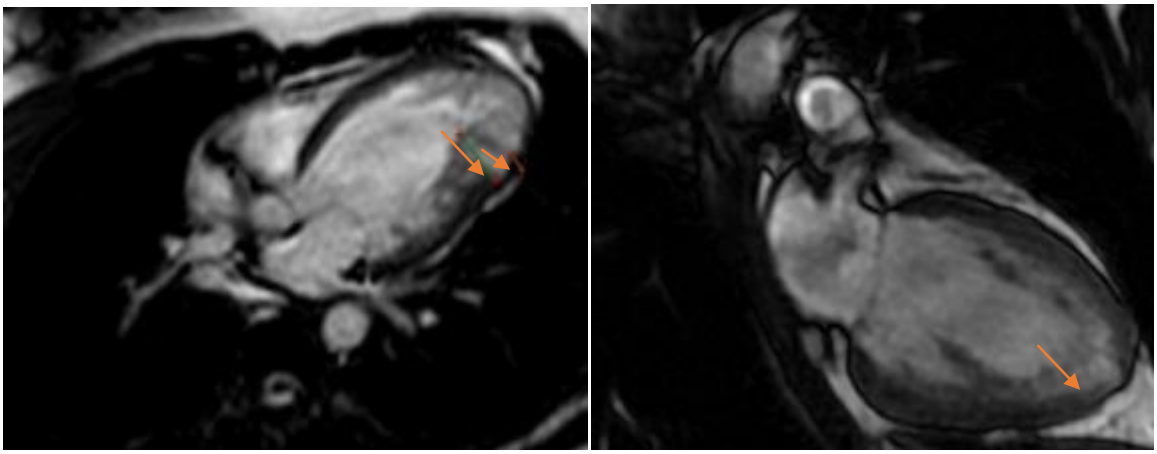

Fig. 4-A. Cardiac magnetic resonance imaging (MRI) T2 sequence; four-cavity slice horizontal long axe; Hypertrophy of the endocardium with the endocardium/epicardium ratio more than 2 corresponding to LVNC without endomyocardial fibrose. The ventricular ejection fraction calculated at 55%

and

Fig. 4-B. Cardiac MRI: Sagittal cross section view: Non dilated cardiomyopathy associated with LVNC (Arrow)

"Abnormal electrocardiographic findings related but not specific to LVNC are left or right bundle branch blocks, fascicular blocks, repolarization abnormalities such as T-Wave inversion and ST-Segment changes high-degree atrioventricular block, AF, atrial flutter, VT and Wolff–Parkinson–White syndrome mainly in children" [1]. In this case report we relate a non-specific and rare rhythmic manifestation of LVNC, type AVNRT which unveiled the diagnosis in a sporty woman.

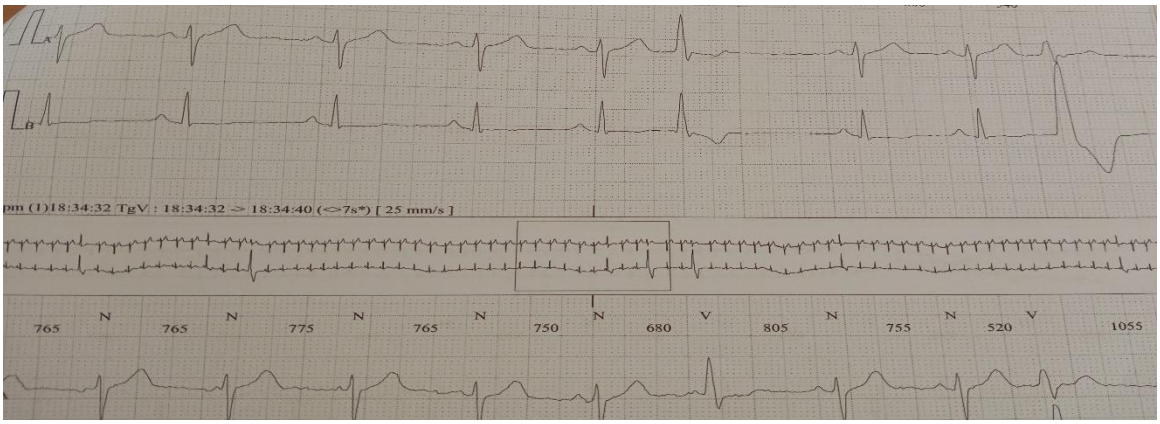

Fig. 5. 24 Hours Holter ECG: Ventricular extrasystoles or premature ventricular complexes

The mechanism of this supra-ventricular rhythm abnormality in LVNC is still unclear compared to ventricular conduction abnormality which is due to ventricular hyper trabecular structure impairing the development of the His-Purkinje system during the embryogenic period [16]. Although, rapid treatment by radiofrequency ablation our patient developed a premature ventricular rhythm.

Complications ranging from heart failure to sudden death due to arrhythmias, and thromboembolic events have been documented in this disease [17]. Our patient quickly benefited from a curative treatment for her AVRNT with a regular 24-hour Holter ECG follow-up.

“Until present, management of patients with LVNC is complicated because prospective studies of large cohorts do not exist” [18]. “However, management should include clinical monitoring for asymptomatic patients with normal LV size and function or clinical guidance according to current therapeutic evidence in symptomatic patients due to LV dysfunction and/or arrythmias. Furthermore, oral anticoagulation is indicated for patients with atrial fibrillation, impaired LV systolic function, history of systemic embolism, or demonstrated intracardiac thrombi” [19]. “In the studies that presented data on asymptomatic patients with preserved systolic function, no thromboembolic events were reported during follow-up [20,21,22,23], and the use of warfarin in LVNC patients with preserved LV systolic function is controversial. Beta-blocker therapy was evaluated in a small, retrospective study of patients with LVNC in which the LV mass was reduced on beta-blocker therapy compared to the patients that were not on betablocker therapy at approximately one-year follow-up” [24]. In this case report, only beta-blocker was prescribed for the patient to treat her frequent PVCs without an oral anticoagulant. It should be noted that regular follow-up is necessary in patients with LVNC especially at the rhythmic level. Since there were only episodes of isolated PVCs in the 24 hours Holter ECG, medical therapy should be considered in absence of repeated ventricular tachycardia.

4. CONCLUSION

Supra-ventricular arrhythmias are rare manifestations of LVNC. It’s important for cardiologists to examine with care the ventricular structures during transthoracic echocardiography and diagnosis should always be confirmed with second performant imagery like cardiac MRI. Radio-frequency ablation therapy is one of the best options for rhythm control. More studies are needed in the future to judge the necessity of anticoagulation therapy in patients with preserved LV systolic function. Holter ECG is a simple accessible tool in the follow-up of patients with documented or non for arrhythmias.

CONSENT

As per international standard or university standard, patient(s) written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

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COMPETING INTERESTS

Authors have declared that no competing interests exist.

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