Case study

A RARE SUPRA-VENTRICUALAR ARRTHYMIA REVEALING A LEFT VENTRICULAR NON-COMPACTION CARDIOMYOPATHY IN A YOUNG WOMAN. CASE REPORT.

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 ventricular cavity without evidence of communication to the epicardial coronary artery system. Frequent premature supra ventricular arrythmia as unique finding in LVNC cardiomyopathy are rare manifestation of this disease. We report a case of frequent persistent supraventricular tachycardia as first manifestation of a patient with LVNC cardiomyopathy in a young healthy woman who in spite after radio frequency ablation rest symptomatic. 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

Comment [SV1]: Modify the sentence.

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 has been described as, that during the embryological heart development process the myocardial compaction is disrupted through unknown mechanisms, and the latter that LVNC could be acquired and developed on time [1,3]. Frequent premature supra ventricular arrythmia as unique finding in LVNC cardiomyopathy are rare manifestation of this disease.

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Clinical manifestations and the age at 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 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, contrast TTE has been the diagnostic test of choice for noncompaction [11]. Herein, we describe a case of ~~f~~ supraventricular tachycardia as first manifestation of a patient with LVNC cardiomyopathy in a young healthy woman who in spite after radio-ablation rest symptomatic. 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.

Comment [SV3]: Avoid repeated reference to the patient

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In common with most myocardial disorders, LVNC is often a heritable trait [9], 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 io channel disorders [10].

Case Report

A 44-year-old patient went to the emergency room after an intense sporting activity of running more than 6 km for palpitations associated with chest discomfort which prompted the call for help at the end of the race. On admission, the patient no longer complains of chest pain but persistence of palpitation felt as a 'jump heart beats'. In addition, the patient has a history of hypothyroidism of autoimmune origin under LEVOTHYROX 50ug per day, well followed by her attending physician with a 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 blood pressure value of 119mmHg systolic pressure and 70mmHg diastolic pressure, 99% oxygen saturation in ambient air without no signs of cardiac insufficiency. Cardiac auscultation finds an irregular rhythm with no abnormal heart sounds. On the ECG, showed a supra-ventricular tachycardia probably an atrioventricular nodal re-entry tachycardia (AVNRT) (Figure 1) which could not be reduced by medical therapy, 3 days later patient underwent an electrophysiology study which confirmed an AVNRT type slow-fast reduced by capture and enhancement thus requiring an ablation by standard radio-frequency at the basal part of the inter-atrial septum.

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Comment [SV6]: Check syntactic errors

Comment [SV7]: Correct the error

Comment [SV8]: Please explain this study.

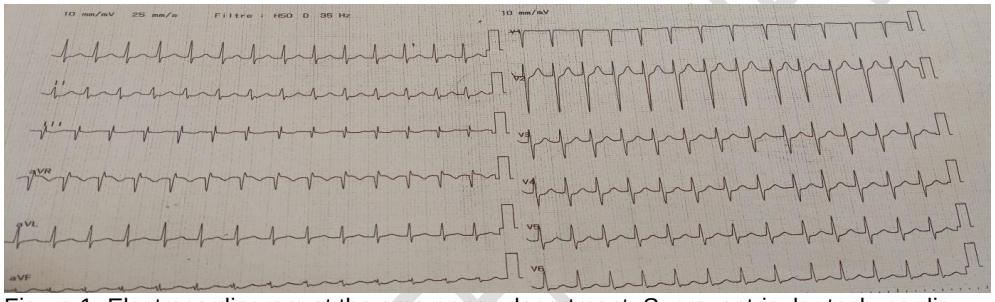


Figure 1: Electrocardiogram at the emergency department: Supraventricular tachycardia probably junctional tachycardia.

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 (Figure 2).

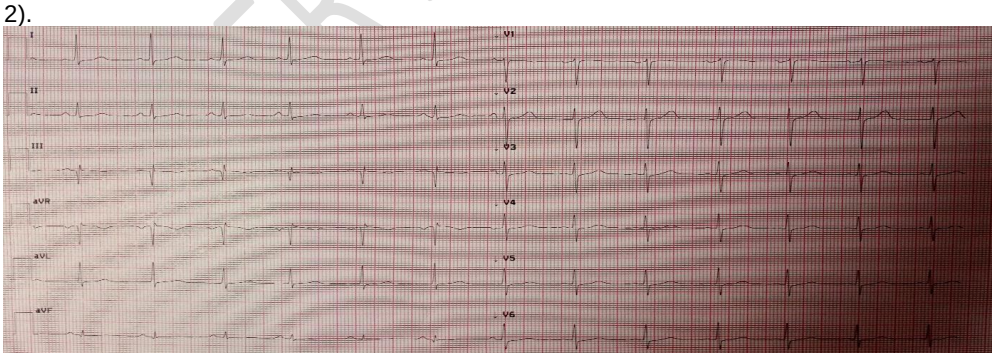
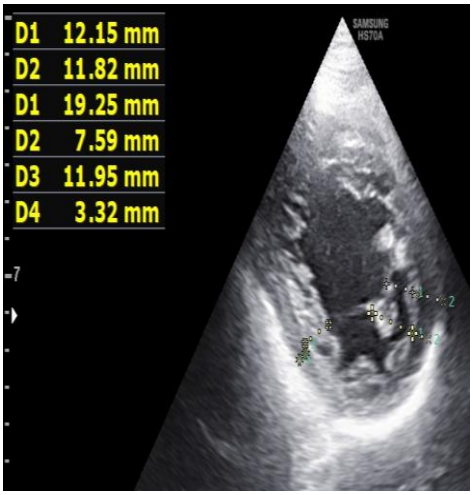


Figure 2: Electrocardiogram post-ablation by radio-frequency: Regular Sinus rhythm.

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 (Figure 3).



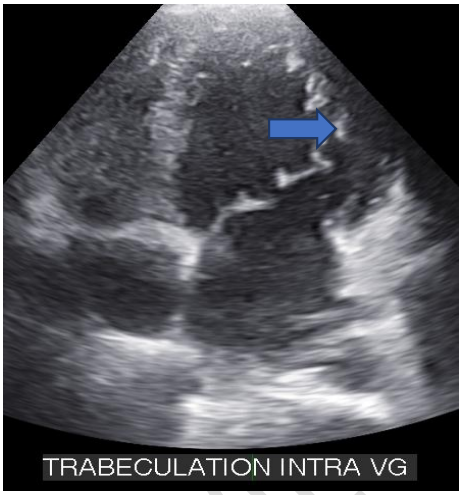


Figure 3-A: Trans thoracic echocardiography; short axes view: shows left ventricular trabeculations predominant at the antero-lateral wall with a ratio of 2.7.

Figure 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.

The systolic function of the left ventricle preserved with an LVEF at 65% without mitro-aortic valve disease with good longitudinal systolic function of the RV without pulmonary hypertension. The biological assessment was unremarkable. A cardiac MRI was carried out in the patient confirmed the presence of a ventricular non compaction (Figure 4).

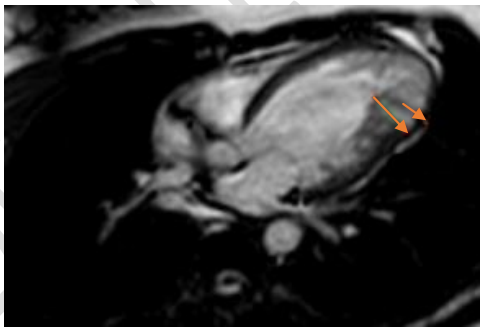


Figure 4 : Cardiac magnetic resonance imaging (MRI) T2 sequence; four-cavity slice horizontal long axe; Hypertrophy of the endocardium with the endocardium/epicardium corresponding to LVNC.

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Our patient was placed on medical therapy after a control 24 hours Holter ECG which confirmed frequent premature ventricular complexes (PVC's) Figure 5.

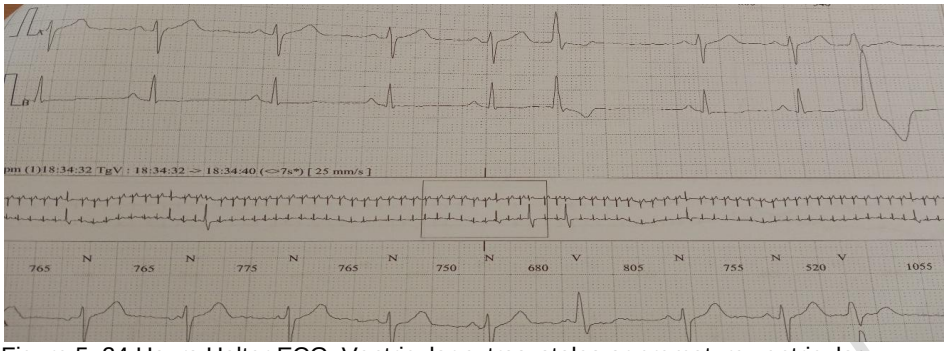

Figure 5: 24 Hours Holter ECG: Ventricular extrasystoles or premature ventricular complexes.

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 hours Holter ECG after 3 months.

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 stablished 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 considered to be 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 – MRI cardiac.

Abnormal electrocardiographic findings related but not specific to LVNC are left or right bundle brunch 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. 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 radio frequency ablation our patient developed premature ventricular rhythm.

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

Until present, there are no specific guidelines for management of LVNC. 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 [18]. In the studies that presented data on asymptomatic patients with preserved systolic function, no thromboembolic events were reported during follow-up [19,20,21,22], and the use of warfarin in LVNC patients with preserved LV systolic function is controversial. In this case report only

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betablocker was prescribed for the patient without 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 absences of repeated ventricular tachycardia.

Conclusion

Supra-ventricular arrythmias are rare manifestations of LVNC. It's important for cardiologists to examine with care the ventricular structures during trans thoracic echocardiography and diagnosis should always be confirmed with a 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 patient with preserved LV systolic function. Holter ECG is a simple accessible tool in the follow-up of patients with documented or non for arrythmias.

ETHICAL APPROVAL

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

CONSENT

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

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