

Aborted Sudden Cardiac Death as a Presentation of Isolated Non-Compaction Cardiomyopathy

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Left ventricular non-compaction cardiomyopathy (LVNC) is a rare disorder characterized by a thick myocardial wall with two distinct layers consisting of compacted and noncompacted myocardium. The major clinical manifestations of LVNC have been described as heart failure, cardiac arrhythmias, and cardioembolic events. In this report we present a case of a young woman with LVNC who presented with aborted sudden cardiac death. This is the second case of LVNC reported in Puerto Rico, but the first presenting with this complication. Recent advances in the field of cardiology allow the identification and diagnosis of this disease; thus, preventive and treatment strategies could be established for this potentially life-threatening condition. LVNC has to be considered in young patients presenting with sudden cardiac death. [P R Health Sci J 2011;30:84-86]

Key words: Non compaction cardiomyopathy, Aborted sudden cardiac death, Ventricular tachycardia

Left ventricular non-compaction cardiomyopathy (LVNC) is a rare disorder classified by the American Heart Association as a primary genetic cardiomyopathy (1). It is characterized by a thick myocardial wall with two distinct layers consisting of compacted and noncompacted myocardium, also described as loose interwoven meshwork with prominent trabeculae protruding to the main cavity. Arrhythmias are common in patients with ventricular noncompaction. Ventricular tachyarrhythmias have been reported in as many as 47% of patients with sudden cardiac death accounting for half of the fatalities in this group (2).

During early development, the myocardium is a loose network of interwoven fibers separated by deep recesses that link the myocardium with the main cavity, and blood is supplied to the myocardium through this spaces. Gradual regression of myocardial sinusoids normally occurs between weeks 5 to 8 of embryonic life, when the ventricular myocardium undergoes gradual compaction mediated by tissue-specific growth factors (3). LVNC is an uncommon finding thought to be caused by an intrauterine arrest of normal cardiac embryogenesis (4). We report the case of a 21 year-old woman whose first manifestation of LVNC was syncope followed by aborted sudden cardiac death (SCD).

Case Report

A 21 year-old woman without history of any previous systemic illnesses presented with three syncopal episodes 30 days prior to our first evaluation. These episodes were associated with palpitations, shortness of breath and dizziness followed by loss of consciousness. The worst of these episodes occurred while

being at work in an emergency room. Upon evaluation by the medical staff she was unresponsive to verbal or painful stimuli. Vital signs were undetectable. A monomorphic ventricular tachycardia observed on cardiac monitor was terminated by immediate cardioversion as recommended in advanced cardiac life support guidelines. Successful reanimation was established. She was transferred to the intensive coronary care unit of the Puerto Rico and Caribbean Cardiovascular Center for further evaluation and therapy.

Cardiovascular evaluation was remarkable for increased sinus rate, normal S1 and S2, and no gallops or murmurs. The 12-lead-electrocardiogram was essentially unremarkable (Figure 1). A transthoracic echocardiogram demonstrated a marked globally hypokinetic left ventricle (LV) with systolic dysfunction (40% estimated ejection fraction). Concentric LV hypertrophy with prominent trabeculations protruding toward the cavity were also noted. Color Doppler and intravenous contrast-enhanced echocardiography demonstrated blood flow in between the trabeculae, compatible with LVNC (Figure 2). Cardiac catheterization with coronary angiography showed normal coronary anatomy. The diagnosis of isolated LVNC was established.

After initial reanimation, the patient was started on standard heart failure therapy as recommended by the American

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College of Cardiology, including a beta-receptor blocker and an angiotensin-converting enzyme inhibitor. An implantable cardioverter-defibrillator (ICD) was implanted during the hospitalization. Following the event she had multiple episodes of non-sustained ventricular tachycardia which gradually subsided. She was discharged after seven days. At 3-month follow up the patient did not present any further arrhythmogenic episodes, with no device therapy required.

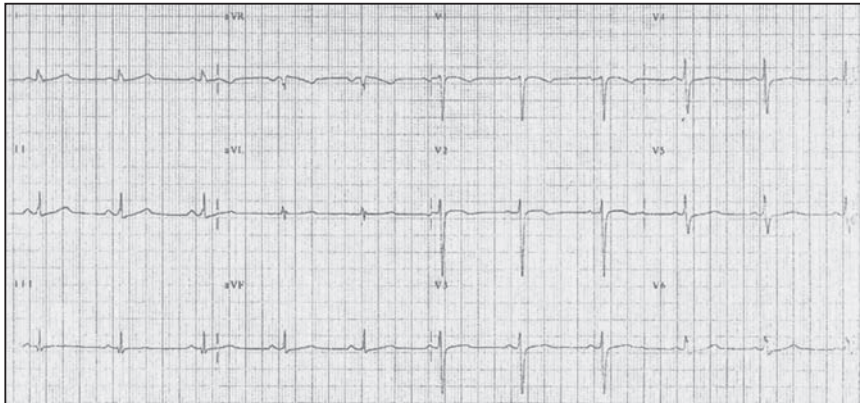


Figure 1. 12 lead electrocardiogram shows non-specific ST changes

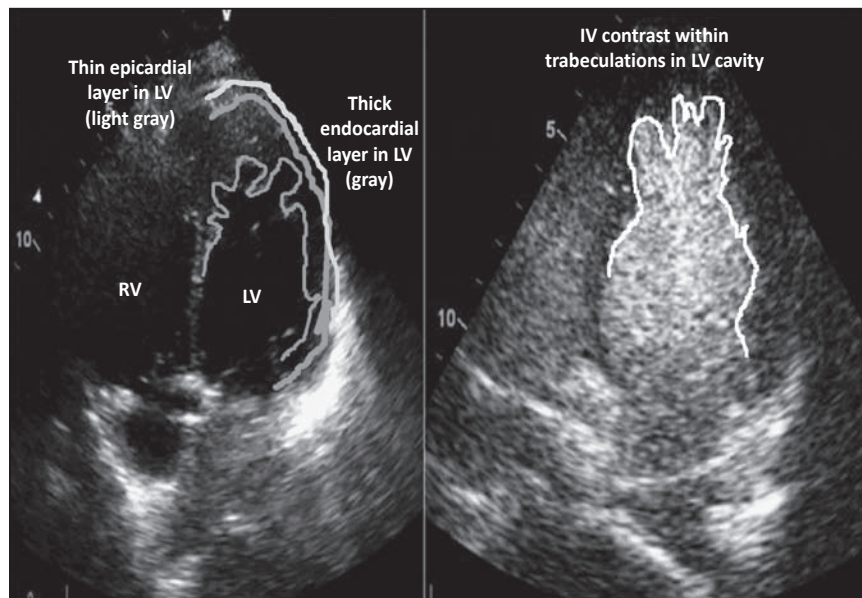


Figure 2. Left. Apical four chamber view of a transthoracic echocardiogram shows concentric LV hypertrophy with prominent trabeculations protruding toward the left ventricle cavity. Right. Intravenous contrast-enhanced echocardiography demonstrates blood flow in between the trabeculae (LV: left ventricle; RV: right ventricle; IV: intravenous).

Discussion

Sudden cardiac death refers to a sudden loss of cardiac activity followed by rapid hemodynamic collapse. Immediate action must be undertaken for a patient to survive this catastrophic event.

Structural heart disease has been determined to be the underlying cause in most patients (5). Our initial clinical evaluation was focused on determining if this was the case for our patient.

The diagnosis of LVNC was established by 2-dimensional and color Doppler echocardiography. Although this imaging modality has been used both to establish the diagnosis and for follow-up, no universally accepted definition for diagnosis has been accepted. Jenni and his group proposed the following criteria in patients

with isolated disease: a thickened left ventricular wall consisting of two layers (a thin compacted epicardial layer and a markedly thickened endocardial layer with numerous prominent trabeculations and deep recesses with maximum ratio of noncompacted to compacted above 2:1), color Doppler evidence of flow within the deep intertrabecular recesses, and a prominent trabecular meshwork in the LV apex or midventricular segments of inferolateral wall (6). All three of these criteria were present in our patient. The presence of all three compaction criteria is rare for diseases other than LVNC (6, 7). This condition must be distinguished from other established clinical entities presenting prominent trabeculations including: apical hypertrophic cardiomyopathy, dilated cardiomyopathy, arrhythmogenic right ventricular dysplasia and endocardial fibroelastosis.

The major clinical manifestations of LVNC are heart failure, cardiac arrhythmias, and cardioembolic events (2). Our patient suffered a very dramatic and potentially fatal complication of this rare disorder. Previous syncopal episodes were most likely related to other ventricular arrhythmia events. Arrhythmias are common in patients with ventricular noncompaction ranging from asymptomatic supraventricular to fatal ventricular events (8).

Available information regarding the appropriate therapy for this condition is limited but focuses on the 3 major clinical manifestations. Carvedilol, a non-selective beta blocker, has been shown to improve left ventricular function as well as both metabolic and adrenergic abnormalities in isolated LVNC (9). Our patient was treated with carvedilol and an angiotensin-converting enzyme inhibitor.

Our patient was a candidate for an implantable cardioverter defibrillator (ICD) according to standard indications. Since she survived a sustained ventricular tachycardia with SCD an ICD was implanted for secondary prevention. A small study published in 2008 showed that ICD therapy was effective for primary and secondary prevention of SCD in this group of patients. Potentially life-threatening ventricular tachyarrhythmias were appropriately aborted, either by anti-tachycardia pacing or ICD shock, after a median follow-up of 36 months in patients with implanted devices for secondary prevention (10, 11).

In conclusion, we present a case of a young woman with LVNC, a rare cardiomyopathy that presented with aborted sudden cardiac death. This is the second case of LVNC presenting at our institution (12), but the first presenting with aborted SCD. This etiology must be considered by practicing physicians in young patients evaluated for syncope and/or after surviving sudden cardiac death events. The prevalence of this condition for Hispanic populations has not been described. One of our goals is to promote awareness on diagnostic criteria for LVNC and provide the framework for further research. Improvement in imaging studies will lead to earlier identification of patients and provide adequate prevention of potentially fatal events like the one described above. Specific management guidelines for treatment should be compiled.

Resumen

Miocardopatía no compacta (MNC) es una entidad clínica muy rara, caracterizada por la presencia de paredes miocárdicas engrosadas compuestas de dos capas distintivas, una compacta y otra no compacta. Entre sus manifestaciones más comunes se encuentra el fallo cardíaco, eventos cardioembólicos y arritmias. Reportamos el caso de una joven con MNC cuyas primeras manifestaciones de esta condición fueron síncope seguida por muerte cardíaca súbita que fue abortada. Este es el segundo caso que es reportado en nuestra institución, pero el primero con esta presentación. Los avances en la cardiología permiten la identificación correcta de esta enfermedad. Esto permite establecer las estrategias de prevención y tratamiento

de consecuencias potencialmente mortales para esta condición. Esta condición debe considerarse como posible etiología en pacientes que desarrollan muerte cardíaca súbita.

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