

Is Transesophageal Echocardiography May Be Useful Diagnosis of Left Ventricular Noncompaction: A Case Report

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Abstract

The left ventricular non-compaction (LVNC) is a primary cardiomyopathy and is characterized by prominent trabeculations on the luminal surface of the ventricle and deep inter-trabecular recesses (DITR) communicating with ventricular lumen and regional wall motion abnormalities. The diagnosis of LVNC is generally made with transthoracic echocardiography (TTE) and cardiac magnetic resonance imaging (CMRI). The spectrum of clinical features are highly variable, ranging from no symptoms to progressive deterioration in cardiac function which results in congestive heart failure, systemic thromboembolism, life threatening ventricular arrhythmias, and sudden cardiac death (SCD). A case of LVNC with sustained ventricular tachycardia (VT) was presented. An implantable cardiac defibrillator (ICD) was implanted because of the individual's high SCD risk.

Keywords: Left ventricular non-compaction; Implantable cardioverter defibrillator; Transesophageal echocardiography

Case Report

A 50-years-old woman was admitted to cardiology department with chest distress, associated with palpitation. She had a two-month history of mild to moderate progressive dyspnea with a decreased exercise tolerance. Her 48-years-old elder sister suddenly died 6 years ago; however her family had refused autopsy. On physical examination, the patient was confused and her blood pressure was 70/50 mmHg, oxygen saturation 80%. Breath sounds were coarsening on the lung examination. The initial ECG showed sustained, monomorphic VT with a rate of 210 beats per minute. The patient was given a 300 mg bolus of amiodarone intravenously over 20 minutes, which rapidly led to stabilization of the heart rhythm then ECG showed atrial fibrillation with low ventricular response (Figure 1). TTE showed left ventricular (LV) dilation and global hypokinesia (left ventricular end diastolic diameter was 67 mm, ejection fraction was 35%), moderate mitral regurgitation and pulmonary hypertension (estimated systolic pulmonary artery systolic pressure was 50 mmHg). The right ventricle was mildly dilated (end diastolic diameter was 47 mm) and there was no wall motion abnormalities.

Two and three dimensional echocardiography (2DE, 3DE) images showed sparse trabeculations at LV apico-septal region (Figure 2a-2d). Moreover, cardiac MRI showed sparse trabeculations and DITR of the LV (Figure 3). Because the patient had chronic obstructive lung disease, TTE and cardiac MRI showed insufficient images. So, we used

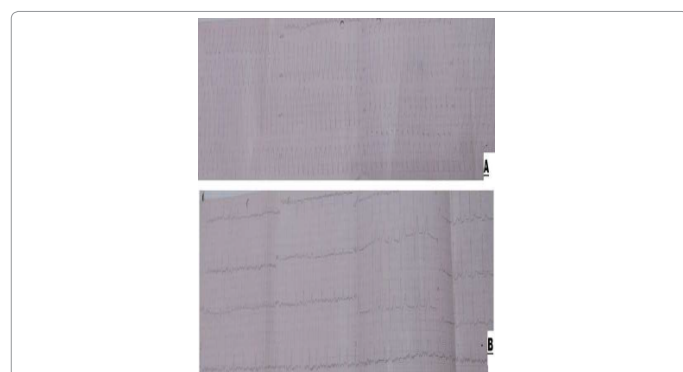


Figure 1: Electrocardiogram at emergency department showing sustained monomorphic ventricular tachycardia (a) and after amiodarone infusion, showing atrial fibrillation with low ventricular response (b).

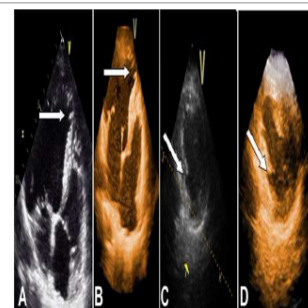


Figure 2: Apical four-chamber views of two (a) and three (b) dimensional echocardiogram, and parasternal short-axis views of two (c) and three (d) dimensional echocardiogram showing sparse trabeculations and recesses of the left ventricle.

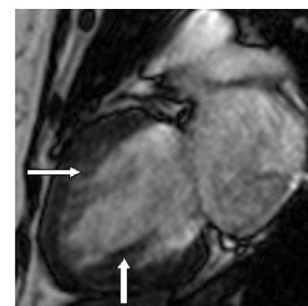


Figure 3: Cardiac magnetic resonance imaging showing sparse trabeculations and recesses of the left ventricle.

a transesophageal echocardiography (TEE), which revealed clearly prominent trabeculations, DITR and color flow imaging demonstrating

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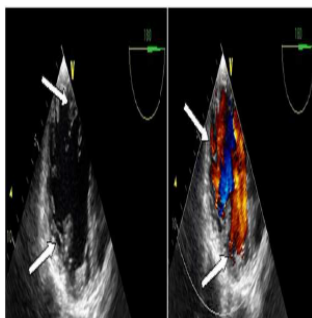


Figure 4: Short-axis view of transesophageal echocardiogram revealing the prominent trabeculations and deep inter-trabecular recesses of the left ventricle, and direct blood flow within the deep inter-trabecular recesses.

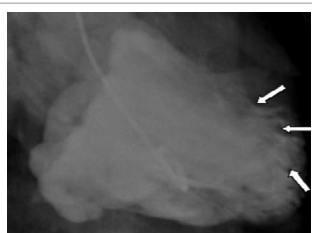


Figure 5: 'Lawn man' image on left ventriculography.

direct blood flow within DITR at the mid-apical, lateral wall and anterior walls of the LV (Figure 4).

Coronary angiography revealed normal coronary arteries. In left ventriculography were seen prominent trabeculations and opaque material which entered in recesses created 'lawn man' image (Figure 5). The patient was given ramipril 5mg/day, aspirin 100 mg/day, carvedilol 12.5 mg/day, and spironolactone 25 mg/day. She was considered for a possible ICD implantation for secondary prevention of SCD. Considering LVNC with decreased left ventricular ejection fraction, sustained ventricular tachycardia episodes and a family history of SCD, the patient was candidate for ICD implantation with class I indication according to the recent guidelines for the management of patients with ventricular arrhythmias and the prevention of SCD. A single-chamber ICD device was implanted. The patient was discharged without complications.

Discussion

The left ventricular non-compaction (LVNC) is characterized by prominent trabeculation on the luminal surface of left ventricle and deep inter-trabecular recesses (DITR), which relation with ventricular lumen and regional wall motion abnormalities [1-3]. Left VNC is myocardial disease with a genetic basis that lead to heart failure, arrhythmia, sudden death (SCD), and thromboembolism [4]. It is true that the diagnosis of this disorder is usually delayed due to difficulty in elucidating the diagnostic findings [4]. Several diagnostic methods have been used in the description and characterization of LVNC containing, but not limited to magnetic resonance imaging (MRI), two-dimensional echocardiography (2DE), contrast-enhanced 2DE [5]. 2DE is by far the most commonly used diagnostic modality. Usually, multiple 2DE studies are needed before the diagnosis is made. The new technic of real-time three-dimensional echocardiography (3DE) could provide an enhanced capability to diagnose and characterize the morphology of complex myocardial disorders like LVNC [6].

Cardiac MRI has good correlation with echocardiogram, which be

useful in patients with poor echocardiographic window and to clarify doubtful echocardiographic abnormalities [7]. In our case, on TTE apical four chamber view there were trabeculations in left ventricle which could be confused with hypertrophy. Also, the left ventricular parasternal short axis TTE view and cardiac MRI had low diagnostic value because of patient's chronic obstructive lung disease. Therefore, transesophageal echocardiography (TEE) was used for diagnosis and short-axis views showed prominent trabeculations and deep recesses within color Doppler flow.

Treatment of LVNC is not specific and is according to its three most important clinical implications; heart failure, cardiac arrhythmias and embolic events. Proper treatment for heart failure is important and is similar to that of other causes [1]. If standard treatment for heart failure is unsuccessful, heart transplantation may be considered as the only therapeutic possibility [8,9]. For patients with arrhythmia, either symptomatic or not, anti-arrhythmic drugs are recommend. Attending to the risk of SCD, ICD may have a role in these patients [10]. Long-term prophylactic anticoagulation is recommended for all patients with LVNC irrespective of the identification of intra-cardiac thrombus [1]. In our case, we decided to perform ICD implantation on the basis of the younger age, the family history of SCD, the left ventricular systolic dysfunction and the documentation of sustained ventricular tachycardia. As a result, in patients with left ventricular dilatation and low ejection fraction should be considered TEE when TTE and CMRI have insufficient images for LVNC.

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