Isolated Left Ventricular Arrhythmogenic Cardiomyopathy: A Case Report

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Introduction

Left ventricular involvement in the advanced stage of arrhythmogenic cardiomyopathy (AC) is an evolution of this desmosomopathy. Mutation in intercalated discs explains the phenotypic variability. Abnormal trafficking of the intercalated discs at the site of end-to-end contacts between cardiomyocytes and the canonical Wnt/β-catenin and Hippo signaling pathways have been implicated.

In AC, T-wave inversion in lateral leads is the hallmark of left ventricular (LV) involvement. We herein report a case of isolated LV AC in which the initial ECG has typical features: fragmented QRS (fQRS), T-wave inversion (TWI) in lateral leads, ventricular tachycardia from inferobasal LV wall.

Case Report

A 52-year-old Caucasian male presented with a history of non-ischemic retrosternal pain and two previous syncopal episodes. The first syncope occurred at rest while watching TV, and the second one while driving a motorcycle. He had a negative family history for sudden death in young or middle-aged (33 ± 14 years) relatives (≤47 years old).

The standard 12-lead electrocardiogram (S-ECG) showed incomplete right bundle branch block; fragmented QRS (fQRS) in leads III and aVF and lateral TWI (Figure 1).

Figure 2 shows stress exercise tests, at 1-minute recovery, provoked sustained monomorphic ventricular tachycardia (VT) (heart rate 240 bpm) (Figure 2A) followed by salvos of very fast polymorphic VT (330-375 of heart rate) associated with hemodynamic instability (hypotension (80/60 mmHg) and presyncope) (Figure 2B). Luckily the VT was spontaneously reversed back to sinus rhythm.

Signal-averaged electrocardiogram (SAECG) using 25 Hz filters was performed (Figure 3).

Transthoracic echocardiography was normal.

Coronary angiography showed absence of obstructive coronary artery disease and ventriculography revealed LV apical hypokinesis.

Cardiac magnetic resonance imaging showed apical LV fibro-fatty replacement with micro aneurysms and extensive area exclusively in the LV lateral wall, compatible with isolated left dominant AC (LDAC) (Figure 4).

Discussion

In many cases, the definition of epsilon waves/epsilon potentials remains difficult, because some authors consider that these waves may be inside of the QRS complex, manifested as fQRS or QRS notching (1). In AC, fQRS has a high diagnostic value similar to epsilon potentials by highly amplified and modified recording techniques, such as right precordial leads ECG (R-ECG) and Fontaine leads (F-ECG) (2). fQRS refers to the 'slurs or notches' appearing on the R or S waves or if the total QRS complex had ≥ 4 spikes. fQRS can be registered as a normal variant mainly in a senior endurance athlete's heart if it appears randomly in just a few leads. However, fQRS presenting in multiple leads is more likely pathologic. The underlying cause is the regional delay in propagation of ventricular depolarization. fQRS is very prevalent in AC patients when applied to amplified and modified ECG recording techniques, including the use of the Fontaine bipolar precordial leads system (2,3). In practice, nevertheless, most ECGs available from AC patients and family members were obtained by using only the S-ECG recording technique. fQRS is easily recognizable from S-ECG, and they are much more common in AC patients when compared with control subjects. Among them a notch before the end of the R or S wave is characteristic, seen in 51% of AC vs. 26% in controls. In AC, fQRS is often seen in multiple leads (4). Such changes, however, are common in control subjects as well. In the latter, the QRS complex is wider (5). fQRS complex, with various morphologies, has been described as a diagnostic criterion of AC; however, fQRS is also prevalent in other types of cardiomyopathies (both ischemic and non-ischemic) (6).

It is very important to note that the presence of an epsilon wave in patients with AC may be seriously underestimated. Current ECG guidelines set the cutoff at 150 Hz for adolescents and adults. Nonetheless, in routine clinical practice a 40 Hz cutoff frequency is used to reduce muscle noise and improve the appearance of the trace (7).

The diagnosis of AC is generally established by the clinical presentation of right ventricular (RV) disease as well as the other features as represented in the 2010 Task Force Criteria. We do not think that this case meets these criteria. To have a definitive diagnosis we need genetic confirmation and/or histological evidence. LV disease in AC without RV involvement is rare and represents a diagnostic challenge. The average age of these patients was 44 ± 16 years (14-81) (8). Though genetic study is recommended, the chance of obtaining positive mutation result is generally low. Moreover, we do not think it is reasonable to perform a myocardial biopsy to establish the disease.

In this case, the monomorphic VT (240 bpm) had a concordant QRS pattern across precordial leads from V1 to V6. The QRS complexes are entirely positive during wide QRS complex tachycardia. Positive precordial concordance results when ventricular activation originates from the inferobasal LV. Additionally, the propensity toward arrhythmia without ventricular dysfunction helps to differentiate from dilated cardiomyopathy and myocarditis (non-ischemic retrosternal pain) (8). Since VT occurs even in the absence of severe systolic dysfunction, an implantable cardioverter-defibrillator should be indicated.

Conclusion

In the present case, we have three typical ECG features for patients with an under-recognized clinical entity: the isolated LV AC by the presence of fQRS in the inferior leads III and aVF, TWI in the lateral leads, and life-threatening arrhythmias of LV origin. Additionally, magnetic resonance imaging showed epicardial fat and microaneurysms in the LV lateral wall and the apex and sub- epicardial late gadolinium enhancement in the LV lateral wall

Disclosure

The authors do not have any conflict of interest regarding this work.

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Legends to figures

Figure 1 Standard 12-lead ECG (S-ECG) at rest

A. Sinus rhythm, heart rate: 61 bpm, P wave duration: 90 ms, P voltage: 1.8 mV, P axis: +25°, PR interval: 150 ms, QRS with low voltage in the frontal plane, QRS duration: 110 ms, QRS axis: +5°, triphasic pattern in V1 (RSR'), broad final R wave in aVR, incomplete right bundle branch block, ST segment with saddleback appearance in V1-V2, and at least 1 mm remains above the isoelectric line, followed by positive T wave. T-wave inversion in low lateral wall (V5-V6). **Conclusion:** Incomplete right bundle branch block, TWI in lateral leads and fQRS in inferior leads (III and aVF).

B. S-ECG shows bipolar in III and unipolar in aVF with clear fQRS (arrows) inside of the QRS complexes.

F-ECG: Fontaine bipolar precordial leads FI, FII and FIII, respectively.

HR: heart rate; S-ECG: standard ECG

Figure 2

A. Stress exercise tests, at 1-minute recovery: sustained monomorphic VT (rate 240 bpm) with right bundle branch block morphology and positive concordance throughout the chest

leads (positive precordial concordance), highly suggestive of VT with focus in the inferobasal LV. Additionally, fusion beats (F) are observed.

B. After physical effort at 2:47-minute recovery: very fast polymorphic VT (330-375 bpm).

Figure 3 Signal-averaged electrocardiogram (SAECG)

Positive late potential is shown. Filter: 40-250 Hz; Cycle: 288; LPs (late potentials) = 40 μV; QRSd (QRS duration) = 116 ms; RMS (root mean square) voltages: Last 40 ms = 31 μV, SD (standard deviation) of noise = 0.10 μV; LAS (low-amplitude signal) < 40 μV = 40 ms..

Figure 4 Cardiac magnetic resonance imaging findings

Long-axis left ventricular outflow (A) and four chamber (B) bright blood SSFP images remarkable for abnormal amounts of epicardial fat and microaneurysms (arrows) in the left ventricle (LV) lateral wall and the apex. Long-axis images showing midwall and sub-epicardial late gadolinium enhancement in the LV lateral wall (arrows) (C,D). Double inversion recovery left ventricular outflow (E) and four chamber (F) images demonstrating fat infiltration of the LV lateral wall (arrows).