



Keywords

Early Repolarization Syndrome,
Brugada Syndrome,
Ventricular Tachycardia

Received: August 22, 2015

Revised: September 21, 2015

Accepted: September 22, 2015

Early Repolarisation Syndrome Associated to Brugada Syndrome Like

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Citation

S. Marrakchi, I. Kammoun, S. Kachboura. Early Repolarisation Syndrome Associated to Brugada Syndrome Like. *International Journal of Clinical Medicine Research*. Vol. 2, No. 5, 2015, pp. 61-64.

Abstract

Recent studies have suggested an association between several channelopathy diseases. However, the mechanism remains unclear. The aim of this case was to demonstrate the relation between Brugada Syndrome (BrS) and Early Repolarization Syndrome (ERS). We presented herein a 46-year-old woman with no familial history of sudden death. She was hospitalized for ventricular tachycardia with a lipothymia episode. We recorded in the electrophysiology study a fractionated electrogram in the same time with ventricular depolarization after fascicular electrograms. This fractionated electrograms precede slurring of the terminal portion of the R wave (J wave) and the atrial stimulation with extrastimulus decelerated the fractionated electrograms responsible of Brugada Syndrome type 1 on the ECG. Brugada Syndrome (BrS) and Early Repolarization Syndrome (ERS) suggesting that two syndromes with J wave's disorders have the same mechanism: wave front fragmentation in the anterolateral free wall of the RVOT responsible of ventricular tachycardia (VT).

1. Introduction

Early repolarization, or J - wave, is characterized by elevation of the J - point, which is the junction between the end of the QRS complex and the beginning of the ST - segment, on 12 - lead ECG. Early repolarization is a common ECG finding found in 1-10% of healthy individuals, and for decades has generally been considered benign. However, there is increasing evidence that J - point elevation is associated with an increased risk of ventricular fibrillation and sudden cardiac death. [1]Brugada syndrome (BrS) is characterized by ST - segment elevation in the right precordial leads (V1-3) of the 12 - lead ECG and episodes of ventricular fibrillation in the absence of structural heart abnormalities, although recent studies have identified functional and morphological abnormalities in the right ventricular outflow tract and even in the left ventricle[2].

We report a case presented with early repolarisation syndrome associated to Brugada syndrome

2. Case Report

It is about a 46-year-old woman with no familial history of sudden death. She was hospitalized for ventricular tachycardia with a lipothymia episode, converted back to sinus rhythm twice with an intravenous bolus of 100mg of Lidocaine (Xylocaine*), and electric cardioversion.

The electrocardiogram (ECG) post electrical external choc (CEE) showed a PR interval of 160 ms and with slurring of the terminal portion of the R wave (J wave) in the inferior

chest leads and lateral leads evoqued Early Repolarization Syndrome Fig. 1.

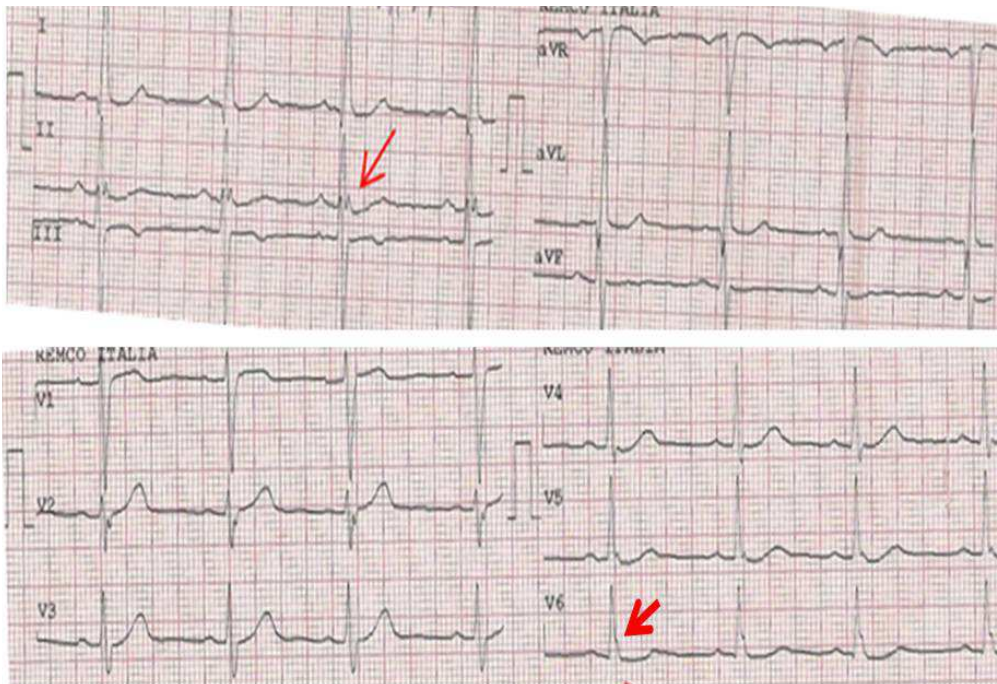


Fig. 1. Sinusalrhythm with slurring of the terminal portion of the R wave (J wave) in the inferior chest leads and lateral leads evoqued Early Repolarization Syndrome.

The cardiac echography showed adilated cardiomyopathy (DCM) with a left ventricular ejection fraction (LVEF) of 45 - 50%.

electrograms in the same time with ventricular depolarization after fascicular electrograms. This fractionated electrograms precede slurring of the terminal portion of the R wave (J wave) by 35 ms (Fig. 2)

The electrophysiological study: We recorded a fractionated

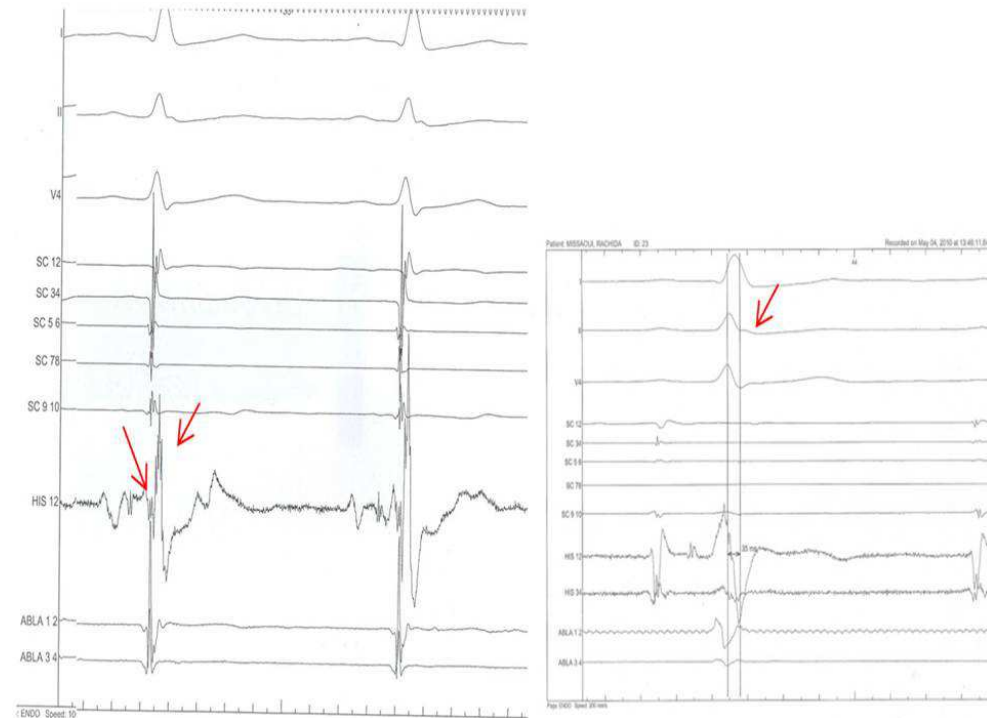


Fig. 2. We recorded a fractionated electrograms in the same time with ventricular depolarization after fascicular electrograms. This fractionated electrograms precede slurring of the terminal portion of the R wave (J wave) by 35 ms.

The atrial stimulation with extrastimulus decelerated the fractionated electrograms responsible of Brugada Syndrome type 1 in the ECG. (Fig. 3)



Fig. 3. The atrial stimulation with extrastimulus decelerated the fractionated electrograms responsible of Brugada Syndrome like type 1 in the ECG.

The ventricular stimulation in this site is responsible of ventricular tachycardia (VT) declenchement. The onset and the arrest were induced by extra - stimuli. The spontaneous arrest of VT is concomitant with two fractionated electrograms. This finding demonstrated Brugada Syndrome may simply reflect a localization RV variant of an extreme form of J point elevation in spectrum of early repolarization disorders

Other electrophysiological study was normal

Her treatment was compound of 200mg of Amiodarone, 50mg of Atenolol, and 100 mg of Captopril per day.

Flecaine test was negative.

Etiologic recherche was negative.

3. Discussion

Early Repolarization (ER), consisting of a J - point elevation, notching or slurring of the terminal portion of the R wave (J wave), and tall/symmetric T wave, is a common finding on the 12 - lead electrocardiogram. In 2007–2008, a high prevalence of ER in patients with idiopathic VF was reported and subsequent studies reinforced the results.[1, 3, 4]

Abnormalities of both depolarization and repolarisation in the right ventricular outflow tract, and heterogeneities of conduction between the endocardium and epicardium have been implicated in the electrographic manifestations of BrS and arrhythmogenesis.[5]

Electrophysiological testing in humans has also identified conduction abnormalities such as prolongation of the His - ventricular (HV) interval and Corrado et al demonstrated that the latest endocardial ventricular electrogram was from the

RVOT[6]. Kanda et al found that patients with readily inducible VF had a longer QRS duration, a higher incidence of late potentials, longer HV intervals and a longer conduction time from the RVOT to the left ventricle compared with non - inducible group[7]. Most of the arrhythmias were induced by extrastimuli applied to the RVOT and the degree of depolarization abnormalities was thus linked to the risk of ventricular arrhythmia. Wavefront fragmentation along anterolateral free wall of the RVOT of functional conduction block, were shown in isochronal maps to initiate polymorphic VT and subsequent degeneration into VF in 5 patients. There were also steep restitution gradients in the RV reflecting repolarization abnormalities in the endocardium that would contribute to the arrhythmic substrate.[8, 9].

At the ECG's patient, the diagnostic of ERS is suggested but this patient shows a ventricular flutter and we don't record ventricular fibrillation. The electrophysiology study shows a Fractionated electrograms in the anterolateral free wall of the RVOT. An extrastimulus ventricular decelerated this fractionated electrograms responsible for a right bloc in ECG brugada type 1. Infact, this fractionated electrogram recorded 35ms before the R' wave in V1 lead at ECG. The findings in this case, although not typical of classical Brugada syndrome or early repolarization syndrome, possess enough pathological features to suggest that there may be a relationship between the two diseases, illustrating the issue of diagnostic classification in the presence of both a typical type 1 ECG and marked structural abnormalities. Infact, the Brugada and early repolarization syndromes share genetic backgrounds. To date, there have been 15 different genes

associated with BrS and 6 associated with early repolarizationsyndrome[10, 11]. Interestingly, all of the 5 causative genes of early repolarization syndrome have been also associated with BrS, indicating the presence of a common pathogenesis in these diseases. Mutations in SCN5A are the most common genetic basis, accounting for 20–30% of patients with BrS, and we have identified mutations in SCN5A in patients with early repolarization syndrome[12]. Mutations in genes encoding the L - type calcium channel, including CACNA1C, CACNB2b, and CACNA2D1, have been associated with both diseases[13, 14].

This case shows, also, that the use of lidocaine might be interesting in converting back to sinus rhythm in use of rapid ventricular tachycardia.

4. Conclusions

Both repolarization and depolarization abnormalities probably contribute to arrhythmogenesis in the Brugada Syndrome. Early Repolarization could cause phase 2 re - entry resulting in closely coupled extrasystoles initiating VT/VF. Conduction delay due to abnormal ion channel kinetics and subtle structural derangements could contribute to this dispersion of repolarization and provide the substrate for the maintenance of ventricular arrhythmia.

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