CLINICAL ARRHYTHMIA
Case Report

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Early Repolarization Pattern and Idiopatic Sustained Monomorphic Ventricular Tachycardia: an Infrequent Combination

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ABSTRACT

Malignant early repolarization as cause of sudden death has taken on great importance in recent years. It has been described as an entity capable of producing ventricular arrhythmic events, causing from episodes of syncope to sudden cardiac death. Ventricular fibrillation is the typical arrhythmia in these patients, with no clear relationship to date with idiopathic monomorphic ventricular tachycardia. Electrocardiographic markers related to the development of arrhythmic events in early repolarization syndrome have been described. They seem not only related to the development of ventricular fibrillation, but also to sustained monomorphic ventricular tachycardia, as is the case described in the article.

KEYWORDS: Sudden cardiac death; Ventricular tachycardia; Electrocardiogram; Cardiac arrhythmias.

INTRODUCTION

Early repolarization (ER) syndrome is a known cause of sudden cardiac death. It is characterized by alterations in the baseline electrocardiogram (ECG) and the development of ventricular arrhythmic events, specifically ventricular fibrillation (VF), with no clear relationship to date with idiopathic monomorphic ventricular tachycardia. Electrocardiographic risk markers have been identified for the development of arrhythmic events in this disease.

CASE REPORT

A 36-year-old man with a family history of sudden death, mother died at age 50, and with a history of rapid palpitations episodes accompanied by syncope. Presented to emergency department for a new episode of palpitations.

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Cardiac monitoring was performed upon arrival, observing a monomorphic, regular and wide QRS tachycardia. Due to the presence of arterial hypotension, synchronized electrical cardioversion was performed with 200 joules, with return to sinus rhythm, without achieving a record of the tachycardia.

The resting ECG showed the presence of an ER pattern in inferolateral leads (DII-DIII-AVF and D1-AVL-V5-V6), with the presence of *slurring* morphology in inferior leads and *notching* morphology in lateral leads, in addition to the presence of a descending ST segment in inferior leads, and isoelectric ST segment in lateral leads. Also, the presence of a negative T wave in inferior leads and biphasic T wave in V4 to V6 leads was observed (Fig. 1).

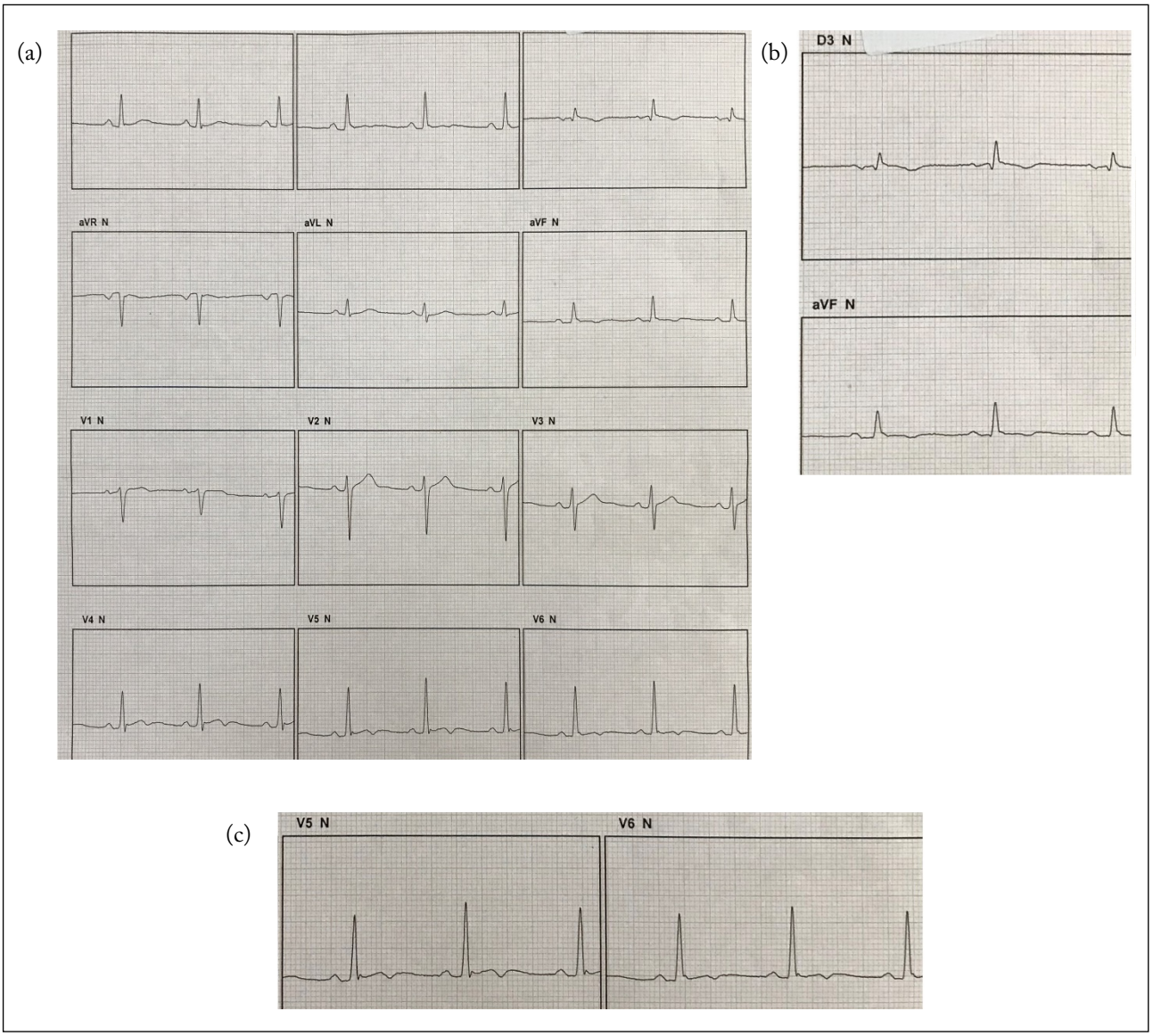

Figure 1. Baseline electrocardiogram. (a) 12-lead electrocardiogram. (b) Inferior leads with slurring morphology and descending ST segment. (c) Lateral leads with notching morphology and isoelectric ST segment.

Metabolic disturbances were ruled out. Transthoracic echocardiogram ruled out macro structural heart abnormalities.

In order to rule out structural heart disease (SHD), cardiac fibrosis, and as an aid to define the arrhythmia mechanism, a cardiac magnetic resonance (CMR) was performed. It showed a structurally normal heart, and absence of evidence of late enhancement or inflammatory tissue.

An electrophysiological study (EPS) was indicated.

The baseline conduction intervals were normal (PA = 35 ms, AH = 53 ms and HV = 41 ms). The atrial stimulation protocol did not induce any sustained supraventricular tachycardia. Subsequently, the ventricular pacing protocol was carried out from the right ventricle (RV) apex, with up to two extra stimuli and with induction of sustained monomorphic ventricular tachycardia (SMVT), with atrioventricular dissociation (Fig. 2a), a cycle length of 350 ms and a QRS morphology compatible with origin in the apex of the left ventricle (Fig. 2b) and with reversal to sinus rhythm by ventricular *burst* stimulation. The same stimulation protocol was carried out from the RV outflow tract, and a pleomorphic ventricular tachycardia (VT) with organization in non-sustained monomorphic ventricular tachycardia (NSMVT) was induced with the same morphology, but with a cycle length of 330 ms (Fig. 3a). When sinus rhythm returned, an increase in the J wave was evidenced in inferolateral leads (Fig. 3b). An implantable cardioverter defibrillator (ICD) was implanted as secondary prevention of sudden death.

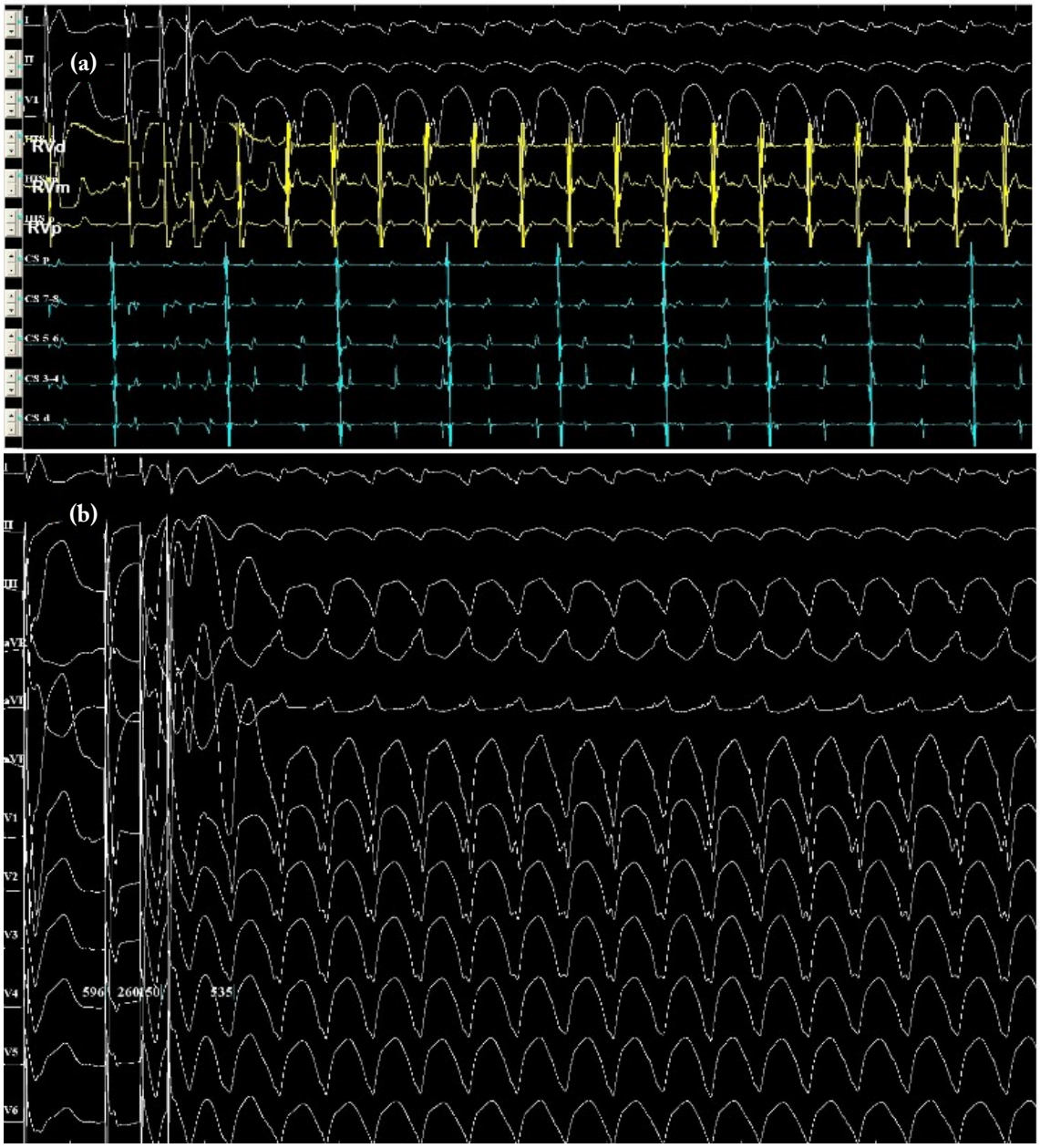

Figure 2. (a) Programmed ventricular stimulation with induction of sustained monomorphic ventricular tachycardia, with atrioventricular dissociation. (b) QRS morphology on 12-lead electrocardiogram.

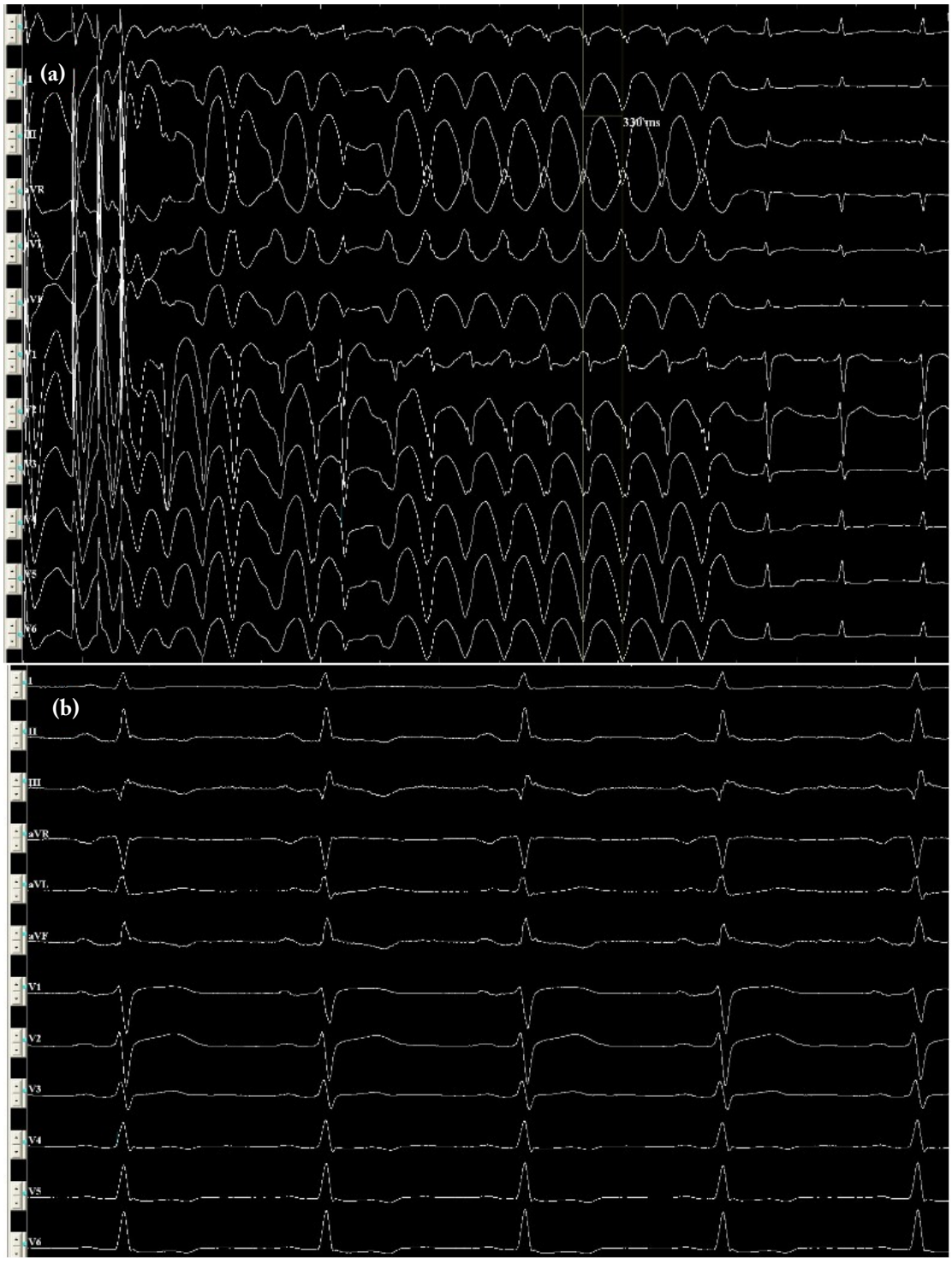

Figure 3. (a) Pleomorphic ventricular tachycardia with organization in non-sustained monomorphic ventricular tachycardia with same QRS morphology. (b) Increase in the amplitude of the J wave.

DISCUSSION

This case describes a patient without SHD, syncope and a SMVT induced during EPS, with a basal ECG compatible with type 2 ER pattern (Fig. 1), which, as described by Antzelevitch et al.[1], corresponds to the presence of J point elevation in inferolateral leads.

The diagnostic criteria for ER pattern are: elevation of the J point greater than or equal to 0.1 mV in at least two leads, terminal part of the QRS with notching or slurring morphology, and QRS duration less than 120 ms[2]. In the case described, in addition to the criteria mentioned, the presence of a descending ST segment in inferior leads (Fig. 1b) and isoelectric ST segment in lateral leads is also observed (Fig. 1c). As Tikkanen et al.[3] demonstrated, this finding is related to the higher risk of death from arrhythmic causes, unlike those in which the ST segment is ascending, in which long-term mortality does not generally differ from the population[4]. However, these ECG markers have been related to the development of VF, although, and as the case described shows, there is probably also a relationship with the development of monomorphic VT (Fig. 2). Voskoboinik et al.[5] have recently described the presentation of ten patients with ER syndrome (ER Sd.), in which three of them presented SMVT at some point. However, in all three cases polymorphic VT or VF was also observed.

Recent evidence shows, through high density electrogram mapping, a wide spectrum of heterogeneous substrates, which may be a consequence of late depolarization or caused by abnormalities in the ER: the first one due to microstructural changes not visible on CMR, and in the second due to a voltage gradient across the ventricular wall, in which case the Purkinje system has an important triggering role[6]. In both cases, the resulting substrate could be the cause of SMVT with a focal mechanism. Current evidence shows ablation as a therapeutic option with favorable results[6].

Another interesting finding of this case is the increase in the amplitude of the J wave, at the time of the induction of the SMVT (Fig. 3b). Haïssaguerre et al.[7] have observed that the amplitude of the J wave increases more than doubles moments before the VF episode.

A differential diagnosis, especially in young and male patients, is myocarditis. Oka et al.[8] demonstrated that, during the acute phase of myocarditis, an ECG pattern of ER may appear in inferolateral leads, which is transient and reversible, and is also not related to the development of malignant ventricular arrhythmias. However, the requested CMR, ruled out myocarditis, showed absence of late enhancement or inflammatory tissue.

Due to all the discussed, it was decided to implant an ICD as secondary prevention of sudden death.

CONCLUSION

ER Sd. has heterogeneous forms of presentation. This case describes an ER Sd. with a type II ECG pattern, and with ECG risk markers for the development of malignant ventricular arrhythmias, as is the case of a descending ST segment in inferior leads and isoelectric ST segment in lateral leads.

Although the typical arrhythmia is VF, this case shows an atypical presentation with the presence of SMVT, in a patient without SHD.

Based on the current evidence available, the mechanism of ventricular arrhythmia could be due to microstructural changes not visible on CMR or to a voltage gradient across the ventricular wall, in which case the Purkinje system plays an important role.

FUNDING

Not applicable.

DATA AVAILABILITY STATEMENT

All dataset were generated or analyzed in the current study.

AUTHORS' CONTRIBUTION

Conceptualization: Pinos J, Leiria TLL, Kruse ML, Lima GG; **Formal Analysis:** Pinos J, Leiria TLL, Kruse ML, Lima GG; **Methodology, Writing – original draft:** Pinos J, Leiria TLL, Kruse ML, Lima GG; **Writing – review & editing:** Pinos J, Leiria TLL, Kruse ML, Lima GG.

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