A case of ventricular fibrillation: Brugada syndrome, early repolarization syndrome, or a variant?

ABSTRACT

Idiopathic ventricular fibrillation (VF) is diagnosed after exclusion of structural heart diseases and other known ion channelopathies. A 23-year-old male patient was resuscitated after sudden cardiac arrest due to VF. VF was defibrillated using an automated external defibrillator during transport to our emergency department. Electrocardiograms (ECGs) obtained at the coronary care unit showed prominent J waves in the right precordial V2 lead. The J waves were not followed by ST-segment elevation or T-wave inversion, and thus were distinct from the typical ECG changes in Brugada syndrome. The results of the laboratory and imaging studies were within normal limits. A flecainide provocation test failed to reveal Brugada-type ECG changes. Under the diagnosis of idiopathic VF, a cardioverter-defibrillator was implanted. Follow-up ECGs obtained at the outpatient clinic showed a spontaneous fluctuation of J waves that coincided with the VF episodes, strongly suggesting that the J waves were pathophysiologically linked to the VF events. In this paper, we discuss the diagnostic ambiguity in variants of idiopathic VF and propose an expanded concept of the J-wave syndrome.

Key words: • ventricular fibrillation • early repolarization • J wave

Introduction

Idiopathic ventricular fibrillation (VF) is diagnosed after exclusion of structural heart and electrical ion-channel diseases such as Brugada syndrome (BS), short QT syndrome, or early repolarization syndrome (ERS).1-3 In some survivors of sudden cardiac death (SCD), the diagnosis remains unclear because the electrocardiographic (ECG) features do not satisfy the proposed diagnostic criteria of the known ion-channel diseases.4,5 In this paper, we report the case of an SCD survivor who presented with right precordial J waves that did not satisfy the diagnostic criteria of either BS or ERS. This case demonstrates the need for modification of the current diagnostic criteria for BS or ERS, and the need to propose a new definition of J-wave syndrome.

Case

A 23-year-old male patient was admitted to our emergency department on January 3, 2004 after re—
susception from sudden cardiac arrest due to ventricular fibrillation (VF). The VF was defibrillated using an automated external defibrillator during transport to our hospital. The patient showed severe hypoxic brain damage and laboratory features of rhabdomyolysis, and was managed at the coronary care unit (CCU). ECGs obtained at the CCU showed prominent J waves in the right precordial V2 lead (Figure 1, upper panel). The J waves were not followed by ST-segment elevation or T-wave inversion, and thus were distinct from the typical ECG changes in BS. However, the follow-up ECGs obtained 4 days after the event showed only minor ST-segment elevation in the right precordial lead, and the prominent J waves were not recorded (Figure 2B). The results of the laboratory test for myocardial infarction and imaging studies were within the normal limits. The patient completely recovered normal mental status 20 days after hospital admission. A flecainide provocation test failed to reveal typical Brugada-type ECG changes. Under the diagnosis of IVF, an implantable cardioverter-defibrillator was implanted. During out-patient follow-up, the patient experienced appropriate shocks, and the ECGs obtained within 1 day after the shock revealed a definite right precordial J wave similar to that observed during the pre-event period (Figure 2A and B). Otherwise, the follow-up ECGs obtained at the outpatient clinic in the absence of appropriate shocks displayed only subtle ST-segment elevation (Figure 3, right and left panel) with occasional small J waves (Figure 3, middle panel).

Figure 1. The upper panel shows the ECG obtained on January 4, 2004, the day after the VF episode. A prominent (0.5 mV) J wave is recorded in the right precordial lead V2. The J wave was not followed by ST-segment elevation or T-wave inversion. The follow-up ECG in the lower panel demonstrates only minor ST-segment elevation in V2.
Figure 2. The patient experienced shock delivery and visited our outpatient clinic. Examination of the defibrillator revealed 2 episodes of aborted ventricular fibrillation and 1 episode of ventricular fibrillation that was terminated by the defibrillator shock (A). The ECG recorded at the clinic showed a prominent J wave in the right precordial lead V2 (B).

Figure 3. The J waves were recorded only during the peri-event periods or immediately after the appropriate shock. The other ECGs obtained remote from the episodes demonstrated only minor ST-segment elevation in the right precordial leads (right and left panel) or only a small J wave (middle panel).
Discussion

The present case highlights 2 clinically important observations. First, some variants of BS or ERS were observed, which do not satisfy the diagnostic criteria of either disease. Second, the close temporal relationship of the J waves with the VF events indicates that the J waves were causally related with the VF episodes. Thus, the ERS or BS variant found in the present case should be considered a type of J-wave syndrome.

The present case illustrates the diagnostic ambiguity encountered in actual clinical practice. The ECG changes observed in the peri-event period closely resembled those in BS. However, the current diagnostic criteria of Brugada-type ECG changes require a J-wave amplitude of ≥2 mm and a downsloping ST segment elevation in at least 1 right precordial lead.5 The diagnosis of ERS is based on the J-wave changes present in precordial leads other than the right precordial lead. Therefore, the diagnosis should be idiopathic in the present case. However, the ECG morphology in the patient is similar to the typical patterns in ERS and BS. In the present case, the distribution of the J waves in the right precordial lead and the morphological similarity suggest BS, but the morphology of the J waves (isolated prominent J waves, not followed by ST/T wave changes) suggest a variant of ERS.

Apart from this diagnostic ambiguity, patients with such variant ECGs share common clinical manifestations with patients with BS and ERS. The circadian patterns of the VF episodes cluster at night or early in the morning. The dynamic patterns of the J waves follow bradycardia–dependent augmentation and tachycardia–induced suppression. VF events are responsive to infusion of intravenous isoproterenol or cardiac pacing. This implies that these variant J waves share a common pathophysiological background with the J waves recorded in patients with ERS or BS.

Recently, the term J-wave syndrome was proposed to incorporate BS and ERS because BS and ERS share common electrophysiological mechanisms and clinical manifestations. We propose a broader concept of the so-called J-wave syndrome, and the case introduced in this paper should be included in this new, expanded definition of J-wave syndrome because the J waves played a key role in the initiation of VF but could neither be classified as BS nor ERS. Further investigations are needed to validate our proposal.

References