Case Report

Brugada syndrome associated with J waves in multiple leads and "pseudo-epsilon" wiggle waves in lateral leads: Possible conduction delay in J-wave syndrome

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1. Introduction

A large number of recent reports suggest that early repolarization (ER) and J waves are associated with the occurrence of ventricular fibrillation (VF) [1–4]. In particular, the presence of J waves in global leads, including the inferolateral leads, is thought to increase the probability of VF [3]. Moreover, the prominence of J waves occasionally precedes the patient's degeneration into VF [2]. Here, we present a case of Brugada syndrome with dramatic alteration of J waves and the appearance of "pseudo-epsilon" wiggle waves in the lateral leads, which has not yet been reported. The significance of J wave variability and the presence of pseudo-epsilon wiggle waves in VF are discussed in this report.

2. Case report

A 36-year-old man was hospitalized following a first syncpe attack while awake at night. No history of sudden death was reported in his family. On arrival, his consciousness was clear and his vital signs were stable. An electrocardiogram (ECG) showed sinus rhythm with ST-segment elevations and J waves in leads V1 through V4 (Fig. 1A). In particular, in leads V1 and V2, the ST-segment elevation revealed coved-type Brugada syndrome. Slurring at the terminal portion of the R wave was also apparent in the inferior leads. The QT interval was 380 ms at a heart rate of 72 bpm. Regrettably, we could not determine whether delayed waves were present at the terminal portion of the QRS-complexes in leads V5 and V6, because the record was not in perfect condition. Laboratory data, including electrolytes, showed no abnormalities. Almost 1 h later, he experienced syncpe recurrence. During the attack, VF was documented on the monitoring ECG (Fig. 2). A short-coupled ventricular premature beat with a coupling interval of 320 ms initiated ventricular tachycardia followed by VF. Just before the occurrence of VF, the monitoring ECG showed neither a prominent J wave nor ST-segment elevation. Electrical defibrillation was immediately performed, leading to termination of VF. After defibrillation, the ECG showed atrial fibrillation (AF) with a rapid ventricular response (Fig. 3A). The amplitude of the J waves (sharp, positive deflections at the terminal portion of the QRS-complexes) in lead V2 changed from beat to beat, although significant ST-segment elevation was not observed in any other lead. Notably, minimal jagged deflections
mimicking epsilon waves (pseudo-epsilon wiggle waves) were documented at the terminal portion of the QRS-complexes in leads V5 and V6. The patient was carefully observed in an intensive care unit. During admission, no VF episodes occurred. After spontaneous termination of AF, the ECG showed sinus bradycardia with a heart rate of 48 bpm. Nonspecific ST-segment elevations were seen in the inferior leads and in leads V1 through V6. In contrast, the amplitude of the J wave in lead V2 was attenuated, and the pseudo-epsilon wiggle waves were not recorded in leads V5 and V6. Late potential detected by signal-averaged ECG was positive, with a filtered QRS duration of 148 ms, root mean square voltage of the terminal 40 ms of filtered QRS-complex of 5.3 \( \mu \)V, and a duration of low-amplitude signal (less than 40 \( \mu \)V) in the terminal filtered QRS-complex of 55 ms. Echocardiography revealed normal cardiac function without any structural abnormalities, including right ventricular abnormalities suggestive of arrhythmogenic right ventricular cardiomyopathy. Angiography showed normal coronary arteries. Pilsicainide challenge test revealed mild enhancement of J-point elevation (up to 1.5 mm) in lead V2 without any changes in the other leads (Fig. 4). VF could not be induced with burst pacing or triple ventricular extrastimulation (even after pilsicainide infusion). An implantable cardioverter-defibrillator (ICD) was implanted in the patient. After the ICD implantation, there were several episodes of VF (always at night), eliminated by appropriate shocks. Then, the ECG showed prominent J waves, although the pseudo-epsilon wiggle waves were not detected in the lateral leads. For reduction of VF events, medicinal therapy was attempted. After oral administration of quinidine at an initial dose of 600 mg/d, VF did not occur for a period of 1 year, and the ECG showed a normal pattern.
3. Discussion

The main finding of this case report was possible conduction delay in a patient with frequent VFs associated with multiple J waves. In a multicenter study, Haïssaguerre et al. reported that an increased prevalence of ER was observed among patients with a history of VF [1]. Moreover, Antzelevitch et al. described ER/J-wave syndrome and proposed 3 subtypes of the disorder [3]. According to their definitions, type 3 of the J-wave syndrome shows a global ER pattern in the inferior, lateral, and right precordial leads and includes a specific type of Brugada syndrome. In fact, with regard to our case, the morphology of the J waves in leads V1 and V2 (coved-type ST-segment elevation) and its response to pilsicainide were compatible with Brugada syndrome. Therefore, we viewed our patient as having overlapping Brugada syndrome and J-wave syndrome (i.e., type 3 J-wave syndrome) because of the widespread appearance of J waves, although the definition of J-wave syndrome remains controversial [5,6].

In the present case, the ER/J-wave inscribed various forms on ECG recordings. Morphologies of ER/J-waves detected in this case consisted of 3 types: positive deflection, slurring, and nonspecific ST-segment elevation. Quinidine, a sodium-channel blocker that also inhibits the transient outward current (Ito), eliminated these ERs and restored a normal ECG. Thus, the prominence of J waves was likely to be associated with an increase in Ito, which might lead to phase 2 re-entry [3]. Notably, just after the second VF attack, beat-to-beat alteration of the J wave was clearly recorded in lead V2. The appearance of a similar fluctuation in the J wave followed by an electrical storm has been previously reported by Haïssaguerre et al. [1] and Letsas et al. [7]. In our case, as Fig. 3A shows, J waves were temporarily aligned with pseudo-epsilon wiggle waves. Moreover, J waves (i.e., second R waves) were augmented following short coupling intervals (beats 1 and 4). These findings suggest that the J wave, which forms an early phase in ST-segment elevation, is likely to belong in the depolarization period. Therefore, the variability of J-wave amplitude might be interpreted as depolarization heterogeneity, triggering an increased vulnerability to VF.

The mechanism underlying Brugada syndrome and J-wave syndrome still remains controversial [8,9]. The prevailing view is the repolarization disorder theory, although the depolarization disorder theory is also acceptable. Recently, Letsas et al. indicated that delayed waves, such as epsilon waves, were detected in the right precordial leads in patients with Brugada syndrome, demonstrating the possibility of right ventricular conduction delay in Brugada syndrome [10]. However, there have been no reports describing these delayed waves in other leads among patients with Brugada syndrome or/and J-wave syndrome. In our case, minimal jagged deflections resembling epsilon waves were clearly visible at the terminal portion of the QRS-complexes in leads V5 and V6 (Fig. 3B). Under re-administration of quinidine (200 mg/d), the patient has now been free from VF attacks for more than 15 months, although J waves and pseudo-epsilon wiggle waves are occasionally recorded on the patient’s ECG.

(Fig. 1B). Therefore, the dose of quinidine was tapered. After the confirmation that VF would not occur with a quinidine dose of 200 mg/d, oral administration of quinidine was discontinued. However, 1 month later, VF relapsed. Just after the recurrence of VF, the ECG showed almost the same morphology as at the patient’s initial visit. In addition, both the pseudo-epsilon wiggle waves and the prominent J waves were clearly visible at the terminal portion of the QRS-complexes in leads V5 and V6 (Fig. 3B). Under re-administration of quinidine (200 mg/d), the patient has now been free from VF attacks for more than 15 months, although J waves and pseudo-epsilon wiggle waves are occasionally recorded on the patient’s ECG.

Fig. 3. Pseudo-epsilon wiggle waves in leads V5 and V6. (A) Immediately after direct current shock in an emergency room, (B) Immediately after relapse of ventricular fibrillation (VF), a beat-to-beat alteration of J waves was seen in lead V2 just after direct current shock for VF (dotted arrows). Pseudo-epsilon wiggle waves were simultaneously documented at the terminal portion of the QRS-complexes in leads V5 and V6 (solid arrows). These small wiggle waves were temporally aligned with the J waves. Immediately after relapse of VF, prominent J waves and pseudo-epsilon wiggle waves reappeared. Moreover, the near coved-type ST-segment elevation in lead V2 clearly outlasted the pseudo-epsilon waves in leads V5 and V6.

Fig. 4. Pilsicainide challenge test. After the injection of pilsicainide (60 mg), the amplitude of the J wave in lead V2 increased by 1.5 mm (solid arrows) without any changes in the other leads.
of quinidine at a dose of 300–600 mg/d, the wiggle waves were never visible. Given that these small deflections were primarily recorded just after VF episodes, both the pseudo-epsilon wiggle waves and the J waves may be related to the occurrence of VF. On the basis of a single-case report, we cannot suggest the potential clinical implication of these pseudo-epsilon wiggle waves. However, we would like to propose that these wiggle waves and J waves share a common etiology and that these pseudo-epsilon waves be regarded as a type of J wave. If so, J waves may be related to delayed conduction, which is associated with enhanced VF vulnerability. Moreover, Fig. 3 may give us a clue as to the mechanism of ST-segment elevation in Brugada-type ECG. In Fig. 3B, the near coved-type ST-segment elevation in lead V2 clearly outlasted the pseudo-epsilon waves in leads V5 and V6. Therefore, the latter phase of ST-segment elevation was thought to reflect repolarization, and it is likely that the near coved-type ST-segment elevation in lead V2 consists of both depolarization and repolarization components. Interestingly, Abe et al. recently described the nocturnal enhancement of depolarization abnormalities in idiopathic VF with J waves by detecting late potentials [11]. It is likely that both depolarization and repolarization abnormalities exist in J-wave syndrome [11] and Brugada syndrome [12,13]. Further investigations are recommended.

Conflict of interest

None.

References