A Case of J Wave Syndrome Due to Severe Hypercalcemia with Ventricular Fibrillation Storm and Successful Treatment of Isoproterenol Infusion

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Summary

A case of J wave syndrome with ventricular fibrillation (VF) storm and severe hypercalcemia due to primary hyperparathyroidism is presented. VF storm subsided with an isoproterenol infusion. Prominent J waves and a Brugada-like electrocardiogram pattern disappeared after parathyroidectomy. Ventricular tachyarrhythmia was not induced during an electrophysiological study. The patient remained asymptomatic up to the 12-month follow-up.

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Key words: Prominent J waves, Brugada syndrome, Hyperparathyroidism

Hypercalcemia is known to be a cause of J wave syndrome;¹⁾ however, the development of ventricular fibrillation (VF) has been reported in only a few cases.²⁻⁴⁾ We experienced a case of J wave syndrome with VF storm due to secondary hypercalcemia. In addition, we demonstrated for the first time that the intravenous isoproterenol (ISP) was effective not only for the prevention of VF but also for the disappearance of the prominent J waves.

Case Report

A 52-year-old man was hospitalized for the examination of fatigue, anorexia, and drowsiness. His family history was unremarkable, with no history of sudden death. He was not taking any medications. On admission, his Glasgow Coma Scale was E4V5M6, but physical examination was unremarkable, except for elevated blood pressure of 158/110 mmHg. There were no abnormalities of the neck, chest, and abdomen. The electrocardiogram (ECG) on admission showed a sinus rhythm of 66 bpm with a type 1 ST elevation in V1 and type 2 ST elevation in V2 (Figure 1A). The chest X-ray was normal. Blood examination showed a high corrected serum calcium level of 18.3 mg/dL (normal range: 8.6-10.2 mg/dL) and a markedly elevated intact parathyroid hormone level of 1250 pg/mL (normal range: 10-65 pg/mL), a serum albumin level of 4.8 g/dL, and normal magnesium (2.1 mg/dL) and potassium (4.5 mmol/L) levels. Renal insufficiency was found, with a serum creatinine of 1.80 mg/dL. Magnetic resonance imaging of the brain and a cardiac echocardiogram did not demonstrate any abnormalities.

After hospitalization, combination therapy of forced saline diuresis (2000 mL/24 hours NaCl 0.9% and furosemide 40 mg/24 hours) and elcatonin 80 units/day was started. Although the serum calcium level decreased only slightly, from 18.3 to 16.3 mg/dL, his symptoms improved. On the fifth day of hospitalization, when the patient talked with his family, he suddenly lost consciousness because of VF (Figure 2). The VF was successfully converted to sinus rhythm with a single direct current (DC) cardioversion of 150 J. He was transferred to the coronary care unit. VF occurred repeatedly, and DC cardioversion was required several times. The ECG during sinus rhythm showed type 1 ST elevation in V1-3 and prominent J waves in V4-6 and the inferior leads (Figure 1B). Thus, J wave syndrome including Brugada syndrome was suspected, and ISP was infused intravenously at 0.02 µg/kg/minute. After the infusion of ISP, the ST elevation and J waves disappeared, and the VF storm subsided (Figure 1C). Then, oral quinidine 300 mg/day and cilostazol 100 mg/day were started. After forced saline diuresis, cinacalcet hydrochloride 50 mg/day, and hemodialysis, the serum calcium level was normalized to 9.4 mg/dL on the ninth day. Parathyroid ultrasonography showed a hypoechoic nodule at the right upper lobe. Parathyroid scintigraphy with Tc-99m MIBI (sestamibi) showed increased activity in the right upper lobe, and this activity persisted on late images. The patient underwent parathyroidectomy, and the histopathological examination confirmed the tumor was a parathyroid adenoma. Electrophysiological study (EPS) was conducted 3 weeks after parathyroidectomy without medication. Neither ventricular tachycardia nor VF could be induced during the EPS with programmed ventricular

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Figure 1. Electrocardiograms (ECG). **A:** On admission, ECG showing a sinus rhythm with a type 1 ST elevation in V1 and a type 2 ST elevation in V2. **B:** ECG after defibrillation of VF shows type 1 ST elevation in V1–3, and prominent J waves in V4–6 and the inferior leads (arrows). **C:** After the infusion of isoproterenol, ST elevation and J waves disappear, and VF storm subsides.

Figure 2. Electrocardiographic monitoring record of ventricular fibrillation.

Figure 3. Electrocardiograms during a pilsicainide provocation test. **A:** Baseline, **B:** after 3 minutes, and **C:** after 5 minutes. Drug-provocation test using intravenous pilsicainide of 1 mg/kg is performed. Although a slight type 1 elevation is observed in V1, the typical Brugada ECG pattern is not induced.

stimulation (PVS) using up to three extra stimuli until S3 to S4 of 180 milliseconds, with pacing cycle lengths of 400 milliseconds from the right ventricular (RV) apex and RV outflow tract. Coronary angiography showed no stenotic lesions. A drug-provocation test using pilsicainide 1 mg/kg intravenously was performed. Although a slight type 1 ST elevation was observed in V1, a typical Brugada-like ECG pattern was not induced (Figure 3).

Because the VF storm was considered to have been caused by reversible hypercalcemia due to primary hyperparathyroidism, a cardioverter-defibrillator (ICD) was not implanted. The patient has remained asymptomatic up to 12-month follow-up.

Discussion

There have been a few case reports on the development of VF with prominent J waves due to hypercalcemia.²⁻⁴⁾ Of the case reports, one case was concomitant with drug-provoked Brugada syndrome.⁴⁾ The present case demonstrated, for the first time, the efficacy of ISP infusion for the secondary J wave syndrome due to hypercalcemia; ISP was effective not only for the prevention of VF but also for the disappearance of the prominent J waves and Brugada-like ECG pattern.

Hypercalcemia was associated with prolongation of both the PR interval and QRS duration, with shortening of the QT interval and J point elevation.⁵⁾ In addition, hypercalcemia has been reported to be a cause of J wave syndrome and acquired Brugada ECG pattern or phenocopies.¹⁾ Concerning arrhythmogenicity, hypercalcemia facilitates re-entry by shortening the action potential, decreasing conduction velocity, and facilitating delayed afterdepolarizations.⁶⁾

ICD implantation is usually recommended for the secondary prevention of VF episodes, but an ICD was not implanted for the following reasons: 1) the patient had no family history of sudden death; 2) after normalization of the serum calcium level, the J waves and Brugada-like ECG pattern disappeared and VF did not recur; 3) aggressive PVS during the EPS did not induce ventricular tachyarrhythmias; and 4) the drug-provocation test using intravenous pilsicainide did not show the typical Brugada ECG pattern. Although late-onset Brugada syndrome could not be denied, it was considered that the present patient exhibited

secondary J wave syndrome due to temporarily severe hypercalcemia. The patient remained asymptomatic during 12-month follow-up, but careful clinical and electrocardiographic follow-up seems mandatory.

Disclosure

Conflicts of interest: None.

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