Successful radiofrequency catheter ablation of a premature ventricular contraction triggering ventricular fibrillation in a patient with short QT syndrome

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Introduction

Short QT syndrome (SQTS) is a very rare inherited arrhythmic disease that is associated with sudden cardiac death (SCD) caused by ventricular tachyarrhythmias. Gussak and colleagues[1] reported the first case of SQTS in 2000, and Gaita and colleagues[2] reported family sudden death of patients with SQTS in 2003. If patients experience aborted cardiac arrest (ACA) or ventricular fibrillation (VF), implantation of an implantable cardioverter-defibrillator (ICD) is required to prevent SCD. Some patients experience frequent cardioversions of an ICD and require antiarrhythmic drugs to reduce the VF attacks. Quinidine and sotalol are thought to be effective for preventing VF, but evidence of the effectiveness of those drugs is not sufficient. Some reports showed that premature ventricular contractions (PVCs) triggered VF.[3,4] The occurrence of PVCs will promote random reentries on ventricles with remarkable abbreviated effective refractory periods (ERPs). However, PVCs that trigger VF have not been sufficiently investigated, and it is not clear whether suppression of PVCs can control the occurrence of VF attacks.

We present a patient with SQTS who was resuscitated from cardiac arrest. The patient experienced an ICD discharge and had frequent PVCs and polymorphic ventricular tachycardias (VTs). We performed radiofrequency catheter ablation (RFCA) for the triggered PVCs to control the occurrences of VF.

KEY TEACHING POINTS

- Short QT syndrome (SQTS) is a rare inherited arrhythmic disease and a cause of sudden cardiac death (SCD). Implantable cardioverter-defibrillator (ICD) can prevent SCD in SQTS.
- Quinidine can reduce ICD therapy for ventricular fibrillation (VF) in SQTS. If quinidine fails to reduce multiple ICD therapy, more advanced therapeutic option should be considered.
- Recording of premature ventricular complexes (PVCs) that trigger VF in 12-lead electrocardiography will result in a success of radiofrequency catheter ablation to control VF occurrences in SQTS.
- The mechanism of PVCs triggering VF has not been evaluated in SQTS. Late-phase 3 early afterdepolarizations might be the most likely mechanism of PVCs in SQTS.

Case report

A 50-year-old woman lost consciousness while resting in the morning and recovered from VF by cardiopulmonary resuscitation and electric defibrillation. Just after she was resuscitated, 12-lead electrocardiogram (ECG) showed a short QTc interval (QTc by Bazett formula: 308 ms, heart rate [HR]: 68 beats/min) despite hypopotassemia (2.8 mEq/L) (Figure 1A).

KEYWORDS Late-phase 3 early afterdepolarization; Quinidine; Radiofrequency catheter ablation; Short QT syndrome; Sotalol; Ventricular fibrillation
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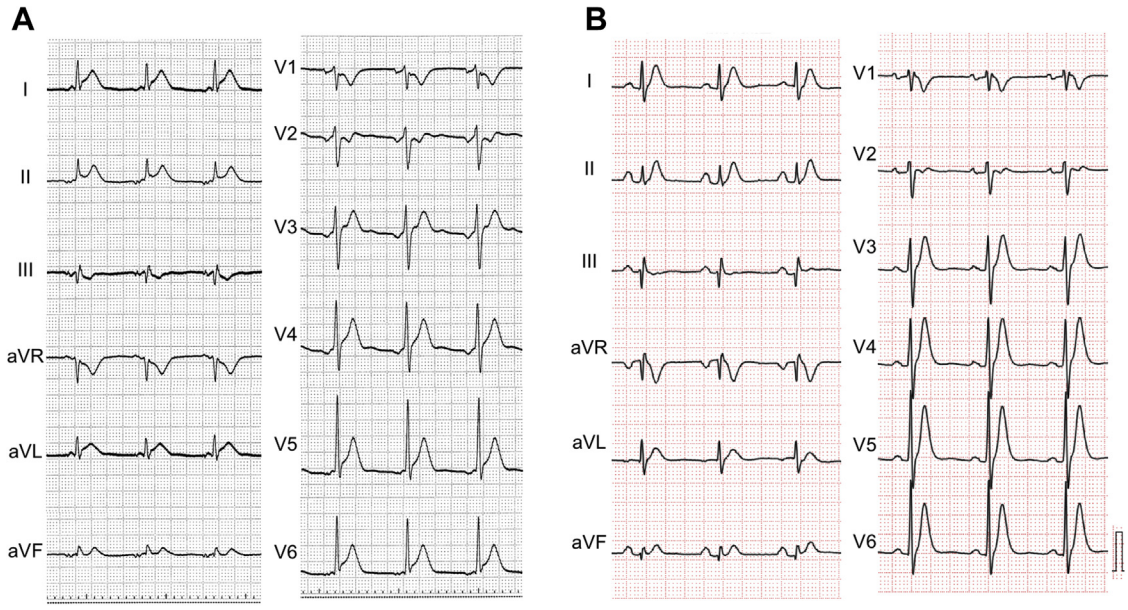

Figure 1 Electrocardiograms (ECGs) of the patient. **A:** Twelve-lead ECG immediately after resuscitation. The QT interval was remarkably short despite hypopotassemia (QTc 308 ms, K 2.8 mEq/L). **B:** Twelve-lead ECG 7 days after resuscitation: the potassium level was in the normal range (K 4.0 mEq/L), but the QT interval was still short (QTc 319 ms).

Two days later, her QTc interval was still short (QTc: 302 ms, HR: 96 beats/min) under hypothermic therapy (body temperature: 34°C) with mild hypopotassemia (3.3 mEq/L). After intensive care, she completely recovered without any brain damage. Various imaging modalities, including echocardiography, coronary angiography, and cardiac magnetic resonance imaging, revealed no organic abnormality in her heart. However, her QT interval was still abbreviated even after normalization of laboratory data (QT: 289 ms, QTc: 319 ms, HR: 75 beats/min) (Figure 1B).

She did not have any symptoms before the experience of the ACA. Her aunt died suddenly at the age of 51 years. Her son and elderly brother also had short QT intervals (QTc: 321 ms, HR: 59 beats/min and QTc: 330 ms, HR: 78 beats/min, respectively), but they did not have any symptoms. An electrophysiological study revealed very short ERPs at the right ventricular apex (150 ms at a pacing cycle length [PCL] of 600 ms and 130 ms at a PCL of 400 ms) and induced VF by 2 extrastimuli with very short coupling intervals (S1/S2/S3 = 400/150/120 ms). Agonists and antagonists of the autonomic nervous system did not influence her QTc interval. Antiarrhythmic drugs (quinidine, disopyramide, nifekalant [a pure I_{Kr} blocker], and pilsicainide [a pure sodium channel blocker]) failed to normalize the QT interval, and sodium channel blockers did not induce Brugada ECG pattern. Although a causative mutation of SQTS was not detected in *KCNH2*, *KCNQ1*, *KCNJ2*, and *SCN5A*, we diagnosed her as having SQTS and implanted an ICD. There were no ventricular arrhythmias during her 1-month hospitalization, and she was discharged without any drugs.

Two months after discharge from the hospital, an appropriate ICD shock terminated a VF event in the early morning. In the intracardiac electrogram at the VF event, a PVC at a coupling interval of 270 ms initiated VF (Figure 2B). Twelve-lead ECG showed the frequent occurrence of monofocal PVCs on the T wave of the previous beat, and they triggered nonsustained polymorphic VTs (Figure 2A). The morphology of the PVC was left bundle branch block configuration with superior axis. The coupling interval of the PVCs was constantly 270 ms and it was coincident with the PVC that triggered VF. The morphology of the second beats of the polymorphic VTs was almost the same as that of the monofocal PVCs, and QRS morphologies of successive beats degenerated and resulted in polymorphic VTs. Oral intake of quinidine sulfate (300 mg/day) and sotalol (80 mg/day) could not normalize QT interval and control the occurrence of PVC and polymorphic VTs. Hence, we decided to perform urgent RFCA targeting the PVCs that were triggering polymorphic VTs. Mechanical stimulation by a catheter manipulation within the right ventricle (RV) easily induced VF, and electrical defibrillation was required to terminate the VF. Accordingly, we could not perform detailed substrate mapping in the RV and performed activation mapping by using a 3.5-mm-tip irrigation catheter (NaviStar, ThermoCool, Biosense Webster, Diamond Bar, CA). A local bipolar electrogram recorded at the inferolateral free wall of the RV showed a prepotential that preceded the onset of the PVC by 30 ms. Pace-mapping at the site showed the same QRS configuration as that of the spontaneous PVCs (Figure 3). There were no abnormal potentials such as fractionated, low voltage, and delayed potentials at the site. Application of RFCAs to the site (RFCA: 30 W, 2 points [57 s/124 s]) and the surrounding regions eliminated all ventricular arrhythmias. After RFCA, she did not experience any arrhythmic events without taking antiarrhythmic drugs during a 17-month follow-up period.

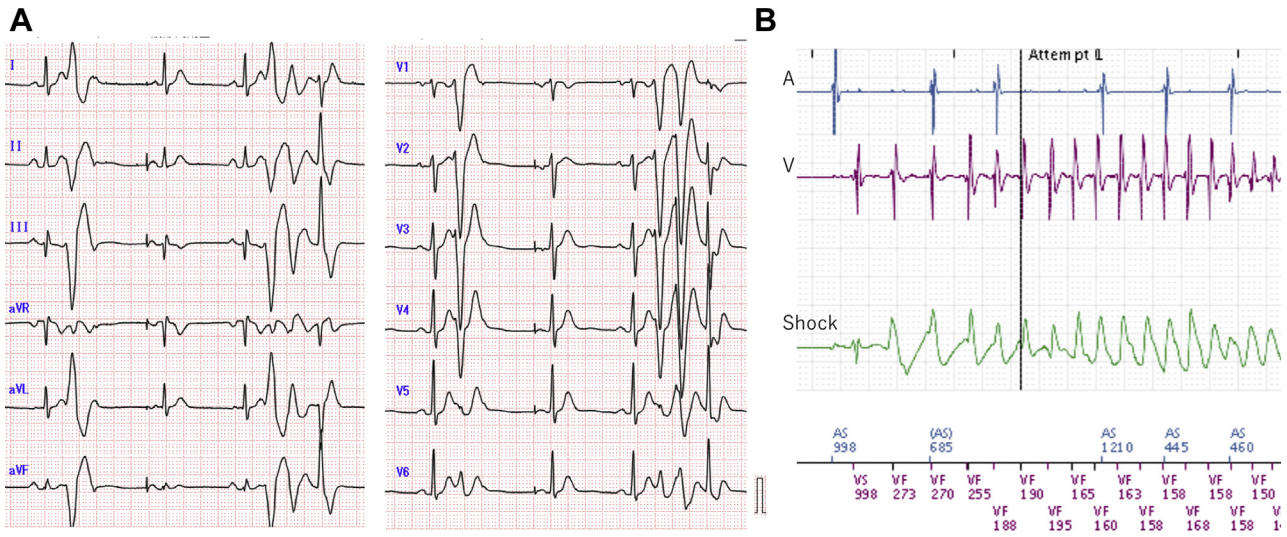

Figure 2 The occurrence of premature ventricular contractions (PVCs) and polymorphic ventricular tachycardia. **A:** Twelve-lead electrocardiogram (ECG) showed that frequent occurrence of monofocal PVCs induced nonsustained polymorphic ventricular tachycardias with a short coupling interval (270 ms). **B:** Intracardiac ECG recorded by an implantable cardioverter-defibrillator at the onset of recurrent ventricular fibrillation episode. The coupling interval of the initial PVC was 273 ms.

Discussion

We presented a female patient with SQTS who experienced ACA and had polymorphic VT and a VF attack after ICD implantation. Treatment with quinidine and sotalol failed to control the ventricular arrhythmias. RFCA targeting the triggered PVCs eliminated the occurrence of polymorphic VTs and VF. To the best of our knowledge, this is the first case report of RFCA for triggered PVCs being able to suppress the occurrence of VF in a patient with SQTS.

Quinidine and sotalol are thought to be effective for preventing VAs in SQTS patients.[5,6] Some studies showed that quinidine was effective for normalizing the QT interval and preventing life-threatening arrhythmic events in patients with SQTS.[7,8] Sotalol is a potassium channel blocker with the ability to prolong QT interval, but there is no clinical evidence regarding the effectiveness of sotalol for SQTS. Some case reports showed that sotalol could not normalize the QT interval and control arrhythmias since the mutant channel lost its affinity to sotalol in SQTS.[9,10] Although appropriate ICD therapy was delivered to terminate VF in the present case, quinidine and sotalol could not suppress the occurrence of ventricular tachyarrhythmias, and intravenous injection of a pure potassium channel blocker (nifekalant) also failed to normalize the QT interval.

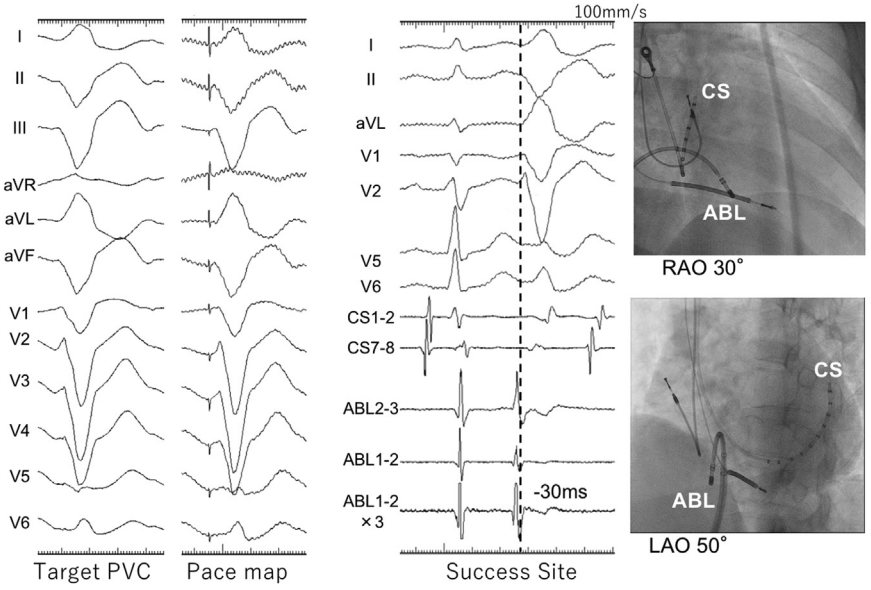

Figure 3 Radiofrequency catheter ablation for triggered premature ventricular contractions (PVCs). A prepotential of the PVC and a good match of the QRS configuration obtained by pace mapping at the inferolateral free wall of the right ventricle. Application of radiofrequency catheter ablation to that site and additional ablation around that site completely eliminated PVCs and ventricular tachycardias. ABL = ablation; CS = coronary sinus; LAO = left anterior oblique; RAO = right anterior oblique.

Unexpectedly, the patient had a frequent occurrence of PVCs that had the same morphology and coupling interval as those of the triggered PVCs at VF onset. We considered that elimination of PVCs would be required to control the polymorphic VTs and VF. Indeed, RFCA successfully abolished the occurrence of ventricular tachyarrhythmia, and the patient was free from arrhythmias without taking any antiarrhythmic drugs. In this case, PVCs were monofocal and detection of triggered PVCs should be a clue to perform successful RFCA.

As for the mechanism of VF in SQTS, the salvos of extrasystoles could promote random reentry on the ventricles with a very short ERP and transmural dispersion of repolarization.[11] However, the mechanism of PVCs triggering VF has not been evaluated. Why do PVCs occur from the ventricle with very short ERPs? Since the PVCs that triggered polymorphic VTs had a constant coupling interval of 270 ms, abnormal automaticity, which has various coupling intervals, would not be a mechanism of PVCs. We also thought that the mechanism of PVC was not a reentry because the local electrogram at the ablation site did not show an abnormally delayed potential. Triggered activity is a potential mechanism of PVCs in SQTS. The occurrence of PVCs on the T wave with a short QT interval also ruled out the possibility of early afterdepolarizations (EADs) at phase 2 and delayed afterdepolarizations. In general, EAD is likely to develop under long QT condition. Late-phase 3 EADs are the most likely mechanism of PVCs in SQTS. Tang and colleagues[12,13] reported that late-phase 3 EADs induced triggered activities and VF via intracellular calcium overload in a short QT rabbit model. The occurrence of late-phase 3 EAD requires autonomic balance changes,[14] and it might have been the reason for the occurrence of VF at night or in the early morning in our patient.

SQTS is an inherited arrhythmic disease, and VF usually occurs in infancy and the ages of 20 to 40 years. The reason for the occurrence of PVCs from only 1 focus at the age of 51 years in the present case is unclear, but any acquired factors would promote the occurrence of arrhythmias. The present case shows that if trigger PVCs are recorded in 12-lead ECG, RFCA can reduce frequent ICD therapy owing to VF in patients with SQTS.

Conclusion

RFCA for PVCs can be a therapeutic option to control ventricular arrhythmic events in SQTS. To our knowledge, this is the first case report of successful RFCA of the PVC that triggered VF in SQTS. Further studies will be required to clarify the efficacy and safety of RFCA in patients with SQTS.

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