Successful transvenous lead extraction and upgrade to implantable cardioverter-defibrillator in a patient with subtype 2 short QT syndrome with ventricular fibrillation

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Introduction: Short QT syndrome (SQTS) is a hereditary cardiac disease associated with atrial and ventricular arrhythmias leading to syncope and sudden cardiac death. Though there is limited data of SQTS, various device managements are sometimes required in patients with SQTS.

Methods: We presented a case of a 40-year-old male diagnosed with subtype 2 SQTS.

Result: He was admitted to our hospital because of out-of-hospital sudden cardiac arrest and survival. He was diagnosed with bradycardia atrial fibrillation in his childhood, VVI pacemaker was implanted at the 21-year-old because of cardiac arrest for 5 seconds. Genetic testing showed V141M KCNQ1 mutation and electrocardiogram revealed QTc interval was 330 milliseconds. He had a high probability of subtype 2 SQTS according to the diagnostic criteria proposed by Gollob et al. At the 40-year-old, he was found in witnessed out-of-hospital cardiopulmonary arrest. Bystander cardiopulmonary resuscitation was continued and one biphasic defibrillation for VF was performed by an automated external defibrillator. Then return of spontaneous circulation was achieved. Systemic inspections by multiple modalities showed there was no evidence of structural heart disease. We diagnosed with VF associated with subtype 2 SQTS. He had no neurological disability. An implantable cardioverter-defibrillator (ICD) was necessary for the purpose of secondary prevention of VF. Because venography revealed his right subclavian vein including the right ventricular lead was occluded totally, transvenous lead extraction and upgrade from the pacemaker to the ICD were performed at the same time. After transvenous lead extraction using Evolution® RL and dilator sheath, the extracted lead had heavy calcified and fibrous tissue. Finally ICD system was implanted successfully.

Conclusion: In this case, various device management was necessary for bradycardia atrial fibrillation and VF in the patient with subtype 2 SQTS. In my best knowledge, this was a first case report that SQTS with V141M KCNQ1 mutation occurred VF.