Ventricular Fibrillation Refractory to ICD Therapy

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Abstract

A 14-year-old boy was admitted for the evaluation of recurrent syncope. His ECG on admission revealed a sinus rhythm with an undetermined QRS axis, T wave inversion at leads V3, V4 and abnormal q at leads I, aVL, V5 and V6. However, no underlying disease could be detected by any morphological examination. Programmed ventricular stimulation also induced no ventricular tachycardia or fibrillation (VF). Only signal-averaged ECG showed ventricular late potential and the cause of syncope was not clarified. As his brother with a similar ECG had died suddenly, he was prophylactically treated with an ICD. However, 14 months later he died suddenly after playing a video game. The ICD recorded VF, which was not converted despite 6 cardioversion attempts by the ICD. Progression of myocardial damages and/or elevation of defibrillation threshold may have been the cause of unsuccessful cardioversion.

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Key words: ventricular fibrillation, ICD, sudden cardiac death

Case Report

The patient was a 14-year-old boy who had a history of recurrent syncope after vigorous exercise since he was 6 years old. ECG revealed a sinus rhythm with an undetermined QRS axis, T wave inversion at leads V3, V4 and abnormal q at leads I, aVL, V5 and V6 (Fig. 1A). Electroencephalogram also showed minor abnormalities and syncope was considered to be due to epilepsy or ventricular arrhythmias. He was treated with the combination of sodium valproate and propranolol by a local pediatrician. His elder brother, at the age of 9 years, with a similar ECG died suddenly after exercise, and the pathological examination of autopsy showed non-specific findings. Thereafter, he was admitted to our pediatric department for further evaluation of syncope. Stress ECG, ambulatory ECG showed no serious arrhythmias, and an obvious underlying heart disease was not detected by echocardiography, magnetic resonance image, cardiac scintigraphy, or coronary angiography with both right and left ventriculography, and endocardial biopsy specimen obtained from the right ventricular apex. ECG recordings of his parents and his residual elder brother were within normal range.

Although he was followed with these same medications, the syncope relapsed at age 13 years and he was transferred to our department for further evaluation. Electrophysiological study showed normal sinus node and atrioventricular function. Ventricular arrhythmias were not induced by programmed ventricular stimulation, which was performed both by incremental ventricular pacing up to 200 beats per minute and single and double ventricular extrastimulation (at basic cycle length of 500 ms) up to 200 ms of second extrastimulation, at both right ventricular apex and outflow tract. Ventricular late potential examined by signal-averaged ECG was the only positive finding (Fig. 1B). From those results and family history, he was judged to have a high risk for sudden cardiac death. After obtaining informed consent from him and his parents, he was treated with an ICD concomitant with atenolol at a dose of 25 mg daily. The screw-in lead

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YASUDA et al (Medtronic 6943–58, Minneapolis, USA) was fixed to endocardium of right ventricular apex, and the ICD (Medtronic 7229Cx) was placed in the left precordial pocket. Pacing threshold and R wave sensing were acceptable. T wave shock-induced VF was not converted into sinus rhythm by internal shock with 20 joule (J). The superior vena cava electrode was not available at that time, and by changing the defibrillation polarity, successful defibrillation by ICD with

Figure 1. A) 12-leads ECG showed a sinus rhythm with an undetermined QRS axis, T wave inversion at leads V3, V4, and abnormal q at leads I, V1, V3, and V6. B) Three parameters on signal-averaged ECG, including RMS 40 (13.8 μV), f-QRS (132 ms), LAS (40 ms) showed all abnormal value, in which implied positive ventricular late potential. C) The ICD recorded a sinus tachycardia at a rate of 150/min with premature ventricular contractions followed by polymorphic ventricular tachycardia. It was transformed into ventricular fibrillation, which was not converted into sinus rhythm despite 6 cardioversion attempts by maximum (30J) energy.
20J was obtained twice. ICD was set to the maximum energy (30J) before discharge and he was followed every 2 months at our outpatient clinic. No syncope or serious arrhythmia was documented, and pacing threshold showed no rise during follow-up periods. However, 14 months after the ICD implantation, he collapsed suddenly after playing a video game and could not be saved. In the ICD memory, polymorphic ventricular tachycardia transformed into VF was recorded and it was not converted into sinus rhythm despite 6 cardioversion attempts by maximum (30J) energy (Fig. 1C).

Discussion

Sudden cardiac death in young people is reported in patients with congenital heart disease, cardiomyopathy, long QT syndrome, catecholaminergic polymorphic ventricular tachycardias, or Brugada syndrome (1–4). In the present case, no underlying disease could be detected by any morphological examination in spite of positive ventricular late potential and abnormal ECG. Shimizu et al (5) reported that a certain hypertrophic cardiomyopathy shows only an abnormal Q wave in their early teens, and wall hypertrophy becomes noticeable only in the late teens. From these findings and family history, our case could be a certain hereditary cardiomyopathy, but genetic analysis was not yet performed because of his mother’s refusal. As he was judged to have a high risk for sudden cardiac death, he was implanted with an ICD. However, the ICD could not effectively terminate VF and save his life. Progression of myocardial damage and/or elevation of defibrillation threshold may have been the cause of unsuccessful cardioversion. In the present case, it should have been considered to add a superior vena cava electrode or to select dual-coil lead, and to measure defibrillation threshold regularly even at the outpatient clinic. Moreover, the manufacture of a new ICD with a high energy cardioversion therapy of over 30J might be suggested.

References