Discrepancy between Inducibility of Ventricular Tachycardia and Activity of Cardiac Sarcoidosis —Requirement of Defibrillator Implantation for the Inactive Stage of Cardiac Sarcoidosis—

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Abstract

Monomorphic ventricular tachycardia (VT) developed in two patients with cardiac sarcoidosis. Before treatment with prednisolone, technetium or gallium scintigram revealed abnormal accumulation in the heart and bilateral hilar lymph nodes, but programmed electrical stimulation failed to induce VT in either case. Prednisolone was administered and the abnormal accumulation of the scintigrams disappeared. However, VT became reproducibly inducible, and in one of the patients, transient entrainment was demonstrated in clinical VT morphology. Defibrillators were implanted in both patients. Some VTs associated with cardiac sarcoidosis are due to reentry, and inducibility of VT is not associated with the activity of cardiac sarcoidosis. Even though steroid therapy suppresses the activity of cardiac sarcoidosis, defibrillator implantation is necessary to prevent a possible arrhythmic event during the follow-up.

Case Report

Case 1

The patient was a 26-year-old man having sustained monomorphic VT. In August 2000, he complained of palpitation and VT at the rate of 210 bpm as recorded on the twelve-lead electrocardiogram (ECG). He was admitted to Nagaoka Red Cross Hospital. Hematological and serological examinations showed no abnormalities except for a slightly higher serum ACE of 25 IU/l. His twelve-lead ECG showed normal sinus rhythm and complete right bundle branch block. Chest X-p showed mild cardiomegaly and swelling of bilateral hilar lymph nodes. Two-dimensional echocardiogram demonstrated that the basal wall of the left ventricle was thin and wall motion at that site was akinetic. Color-Doppler study did not show prominent valvular regurgitation. Results of coronary angiogram were normal, and ejection fraction calculated from the left ventriculogram was 45%. During electrophysiologic (EP) study, AH and HV intervals were 120 and 80 ms, respectively at the basic cycle length of 830 ms. When single premature stimulation at progressively shorter coupling intervals of 10 ms was applied to the right atrium, the AH interval was progressively prolonged to 240 ms until the pacing site reached the effective refractory period. Atrio-ventricular conduction showed Wenckebach block at the pacing cycle length of 316 ms. To study the inducibility of VT, programmed electrical stimulation consisted of 1–3 extrastimuli and rapid incremental pacing up to 286 ms was attempted at 2 sites of the right ventricle and one site of the left ventricle before and after using isoproterenol, but no VT was induced (Fig. 1). He was referred to our hospital for further therapy but VT became inducible by programmed extrastimulation after treatment with prednisolone by which the abnormal accumulation of technetium and gallium scintigraphy was resolved in the heart.

Key words: ICD, steroid therapy, electrophysiologic study

Introduction

Ventricular tachyarrhythmia (VT) sometimes occurs in patients with cardiac sarcoidosis and can be a cause of sudden cardiac death (1–3). However, it has not been well clarified whether steroid therapy which suppresses the activity of cardiac sarcoidosis can prevent the pacing-induced and spontaneous development of VT. In the two present patients with spontaneous monomorphic VT associated with cardiac sarcoidosis, no VT was induced during the active phase of cardiac sarcoidosis but VT became inducible by programmed extrastimulation after treatment with prednisolone by which the abnormal accumulation of technetium and gallium scintigraphy was resolved in the heart.

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Figure 1. Programmed electrical stimulation. Panel A shows the results before treatment with prednisolone. No ventricular tachyarrhythmia was induced until the end of the whole protocol. Panel B was obtained from the electrophysiological study performed during maintenance treatment with prednisolone. Polymorphic ventricular tachycardia was reproducibly induced at 1–2 extrastimulus from the site of the outflow tract of the right ventricle. S₁S₁: basic stimulation. S₁S₂: coupling interval of first extrastimulus. S₂S₃: coupling interval between first and second extrastimulus. S₃S₄: coupling interval between second and third extrastimulus. HBE: His-electrogram recording area, RVA: apex of the right ventricle.

Case 2
The patient was a 49-year-old man having sustained monomorphic VT. Since November 1998, he had complained of faintness and palpitation, and VT at the rate of 250 bpm was documented on his ECG. Treatment with dl-sotalol (160 mg/day) was effective in preventing spontaneous VT attack, and this drug has been administered since April 1999. He was referred to our hospital for further treatment. On admission to our hospital, hematological and serological examinations showed no abnormalities except for a high serum ACE value (58.0 IU/l). The twelve-lead ECG showed normal sinus rhythm and complete right bundle branch block. Chest X-p showed mild cardi-
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omegaly and swelling of bilateral hilar lymph nodes. The two-dimensional echocardiogram demonstrated diffuse hypokinesia of the left ventricle and the basal wall of the left ventricle was thin. Color-Doppler study did not show any prominent valvular regurgitation. Results of his coronary angiogram were normal and the ejection fraction calculated from the left ventriculogram was 48%. Technetium scintigram revealed abnormal accumulation in the heart and noncaseating granulomatosis was demonstrated by transbronchial lung biopsy. From these findings, cardiac sarcoidosis was diagnosed.

EP study was performed under treatment with dl-sotalol because spontaneous VT had not recurred after dl-sotalol therapy. At the basic cycle length of 860 ms, AH and HV intervals were 120 and 55 ms, respectively. When single premature stimulation was attempted from the right atrium, atrio-ventricular conduction showed AH block at the coupling interval of 400 ms, and Wenckebach block was observed during the pacing cycle length of 428 ms. Programmed electrical stimulation was attempted as in case 1 but no VT was induced (Fig. 3). During sinus rhythm, endocardial mapping was performed, but no abnormal local electrogram was recorded from either of the ventricles. Prednisolone (40 mg/day) was started for cardiac sarcoidosis, but sustained VT with two different QRS morphologies recurred although the abnormal technetium accumulation in the heart was eliminated by prednisolone therapy. A temporary pacing lead was inserted to the apex of the right ventricle, and EP study was performed using the lead. These two VTs were reproducibly induced by ventricular extrastimulus, and constant and progressive fusion were demonstrated in one of the VT morphologies (4, 5). Thus the mechanism of the VT was considered to be reentry with an excitable gap (5, 6). The spontaneous recurrence of VT became infrequent and finally disappeared during maintenance treatment with prednisolone (20 mg/day). A defibrillator device was implanted in August 1999 but monomorphic VT was still inducible by programmed electrical stimulation from the device (Fig. 3).

Discussion

Spontaneous VT which develops in patients with cardiac sarcoidosis is considered to be a manifestation of disease activities or inflammation (1–3). Abnormal automaticity or reentry seems to be the mechanism of the VT, but electrophysiological characteristics of VT associated with cardiac sarcoidosis have not yet been clarified. In the present two patients, monomorphic sustained VT developed during the active phase of cardiac sarcoidosis, and EP study was performed both at the active and then later inactive period. Although no VT was induced at the first EP study, VTs showing clinical and non-clinical QRS morphologies were reproducibly induced by pro-

Figure 2. $^{67}$Ga scintigraphy before and after treatment with prednisolone. Abnormal accumulation in bilateral hilar lymphnodes and in the heart successfully disappeared after treatment with prednisolone.
Figure 3. Programmed electrical stimulation. Panel A showed the results before treatment with prednisolone. No ventricular tachyarrhythmia was induced during the whole induction protocol of ventricular tachycardia. Panel B illustrated the polymorphic ventricular tachycardia which was induced by double extrastimulus during maintenance treatment with prednisolone. See details in the text. Abbreviations as in Fig. 1.
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To prevent the recurrence of possible arrhythmic events and sudden cardiac death.

References