Successful Catheter Ablation Against Ventricular Tachycardia Associated with Myotonic Dystrophy

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Abstract

Myotonic dystrophy (MD) is characterized by myotonia and muscular dystrophy and cardiac involvement with tachy-arrhythmia is rarely encountered. We report a case of MD complicated with severe left ventricular hypofunction and incessant ventricular tachycardia (VT) with varying heart rates. The morphology of VT suggested that it originated from the right ventricular outflow tract, and electrophysiological study disclosed that the mechanism of VT was abnormal automaticity. Catheter ablation was performed to treat this VT. The patient had a cardiomyopathy with normal coronary arteries. The specimen of RV biopsy showed moderate hypertrophy, mild fat infiltration and slight fibrosis. These findings are histologically consistent with myotonic dystrophy.

Case Report

Present illness

A 54-year-old woman was admitted to the Department of Neurology, Kyoto Prefectural University of Medicine Hospital on May 22, 1998 for treatment of cerebral infarction. She was referred to a cardiologist because of cardiomegaly in chest X-ray and incessant ventricular tachycardia on electrocardiogram (ECG). She had suffered from myotonic dystrophy characterized by muscle weakness and percussion myotonia for two years. Her cardiovascular history was negative for palpitation, presyncope or chest pain. She had no exertional chest pain or respiratory symptoms.

Physical examination

She was slight and pale. Her height was 155 cm and weight, 40 kg. There was no abnormal jugular venous pulsation. The lungs were clear to percussion and auscultation. The first and second cardiac sound were normal and there were no clicks. A grade 3 systolic regurgitation murmur was audible at the cardiac apex. Her face was myopathic with wasting of the masseters and temporal muscles. There was marked weakness and atrophy of the upper and lower limbs without fasciculation. The grip was myotonic.

Examinations

The chest X-ray showed mild cardiomegaly (cardiothoracic ratio 54%), but no pulmonary venous congestion. The ECG (Fig. 1 left) at rest showed mild PQ prolongation and multiple ventricular premature beats. Episodes of incessant ventricular tachycardia with variable heart rates (HR90–150) were frequently recorded (Fig. 1 right). VT morphology was left bundle branch block and inferior axis. Holter ECG monitoring (Fig. 2 upper) showed total premature ventricular excitations (VEs) were 6,932 (total heart beats 70,660 beats/day). VEs were recorded dominantly on daytime and total count of VT was 42 carneter ablation.
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Figure 1. ECG in the left column was recorded at rest and showed normal sinus rhythm. ECG in the right column was recorded during palpitation attack and showed ventricular tachycardia.

Figure 2. Trends of premature ventricular contractions were shown. Upper panel: recording before ablation, Lower panel: recording after ablation. The number of premature ventricular contractions was markedly decreased after ablation.
Myotonic Dystrophy with VT

Figure 3. The left figure shows coronary arteriogram (upper: right coronary artery, lower: left coronary artery). There were no abnormal findings in coronary angiograms. The right figure shows left ventriculography (upper: end diastole, lower: end systole). Diffuse hypokinesis was demonstrated.

Electrophysiologic study

An electrophysiologic study was performed to examine the mechanism of VT and its origin. During sinus rhythm with a cycle length of 960 msec, AH interval was 120 msec and HV...
interval was 60 msec. Sinus node recovery time was 1,350 msec. Right atrial effective refractory period (ERP), atrioventricular nodal function and right ventricular ERP were within the normal range. Sustained monomorphic VT with a cycle length of 360–500 msec and multifocal premature ventricular contractions spontaneously occurred over and over. VT was not induced by premature ventricular stimulus. QRS morphology of VT was identical to clinical VT with left bundle branch block pattern and inferior axis.

**Interventional treatment**

The morphology of VT suggested that this arrhythmia originated from the right ventricular outflow tract and catheter pacemapping (Fig. 5) was performed in this area. Nearly identical morphology was obtained (8/12) when electrical stimuli were delivered at mid-anterolateral wall of RV outflow (Fig. 6). At this site, the local electrogram during VT preceded the initial deflection of QRS on surface ECG by 40 msec (Fig. 7). During sustained VT, sinus beat or ventricular ectopic beat from another origin could capture the ventricle. Even in this case, the same potential configuration was continuously recorded at a regular rhythm (Fig. 7). This phenomenon suggested the entrance block of propagated excitation to the VT focus, where the ablation catheter tip was located. Entrainment of VT was not possible. The mechanism of VT was presumed automaticity. RF energy was applied to this site during VT. VT was terminated in 8 seconds and converted to sinus rhythm. After 9 times of application of RF energy to this site, spontaneous VT was abolished. Clinical VT could not be induced even with isoproterenol infusion. Holter ECG monitoring showed (Fig. 2 lower) total VEs markedly decreased to 2,784 and VT was not observed. The morphology of residual premature ventricular contraction was different from VT.

**Discussion**

Cardiac involvement in MD is well established. Cardiac conduction disturbances occur in 90% of patients (8). Bundle branch blocks and high-grade atrioventricular block are common in patients with myotonic dystrophy and permanent pacemaker implantation is frequently required. Whereas atrioventricular block and ventricular tachyarrhythmias are considered as the main cause of sudden death in MD patients, reports on VT (3–5) or on sick sinus syndrome (SSS) (9) in association with MD are few. The present case had VT, sinus bradycardia and poor LV function, which are less common, but risky complications of this disease (6, 9–11).

**Pathologic findings**

This patient had a global impairment of LV function with normal coronary arteries. The specimen of RV biopsy showed moderate hypertrophy, mild fat infiltration and slight fibrosis. Other investigators (9, 12) have found that there is increased interstitial myocardial fibrosis, fatty tissue infiltration, myofibrillar degeneration and prominent I-bands in myocardial tissue obtained from patients of myotonic dystrophy. The present findings are histologically consistent with myotonic dystrophy.

Abnormal AV conduction has been well described in MD patients (7, 8, 11, 13). The most common abnormality is HV prolongation, but AH interval may be prolonged and Wenckebach conduction threshold may also be low. EPS of the present case showed mild prolongation of HV interval. Holter ECG monitoring showed that total heart beats and minimal heart rate were decreased. These findings suggested the

**Figure 5.** Catheter position at the successful ablation site. Left 30° right anterior oblique view, Right 60° left anterior oblique view.
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Figure 6. The left column of ECG showed clinical VT and that in the right, paced QRS at the successful ablation site (8/12 matched).

diagnosed of sick sinus syndrome (Rubenstein type 1), although the sinus node recovery time was normal by the overdrive suppression test. These findings suggested that the pathological changes occurred in the sinus node and atrial cardiomyocytes. It was reported (11) that the histopathological changes in SSS patients with MD are the same as those found in SSS without MD.

VT was aggravated by increased sympathetic tone because it was observed dominantly at daytime. VT cycle length was variable from 360 to 500 msec. It could not be induced by premature ventricular stimulus and it had a protected focus with entrance block (Fig. 7). These characteristics suggested that the mechanism of VT was abnormal automaticity.

The increased incidence of sudden death in myotonic dystrophy has been attributed to heart block, ventricular standstill and arrhythmia. The pacemaker is an established method to treat SSS and AV block. However, prevention of tachyarrhythmia was difficult because the use of pharmacological treatment should be limited for patients of poor cardiac function. Therefore in such cases, we should take all therapeutic opinions into consideration, including catheter ablation and implantable cardioverter defibrillator implantation.

References

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Figure 7. Intracardiac ECG. Clinical VT occurred spontaneously (second to sixth QRS). The fourth QRS was a fusion beat. The initial deflection preceded the beginning of QRS during VT by 40 ms.