CASE REPORTS

A Case of Familial Ventricular Tachycardia

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Ventricular tachycardia was noted in 4 members of the same family. One showed diffuse hypokinesis of the left ventricle by echocardiography and left ventriculography, 2 showed progressive left ventricular dysfunction and 1 showed regional perfusion defects of the left ventricle shown by thallium scintigraphy. One patient was diagnosed as dilated cardiomyopathy. Although no definitive cause of the left ventricular disturbance was identified in the 3 other patients, they may have all been dilated cardiomyopathy.

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Familial ventricular tachycardia has mainly been reported in patients with hypertrophic cardiomyopathy; long QT syndrome; and arrhythmogenic right ventricular dysplasia. We report here familial ventricular tachycardia that was not associated with any of these diseases.

CASE REPORTS

Case 1 (proband)
A 55-year-old man experienced chest discomfort, palpitation and dizziness while on the telephone on February 4, 1989. At the nearest hospital, his ECG showed ventricular tachycardia at a rate of 235/min. (Fig. 1a), which was restored to sinus rhythm by an intravenous injection of 50 mg of lidocaine. He was referred to our hospital for further examination and treatment. On admission, his blood pressure was 116/82 mmHg and his heart rate was 78/min and regular. No abnormalities were revealed by chest X-ray, ECG, echocardiography, or thallium scintigraphy. In cardiac catheterization, hemodynamic measurements and coronary angiography were normal, but left ventriculography demonstrated a regional wall-motion abnormality (Fig. 2). The left ventricular end-diastolic volume and end-systolic volume were 77 ml/mm² and 31 ml/mm² respectively, and the left ventricular ejection fraction was 62%. He has since been taking 450 mg of mexiletine, 450 mg of disopyramide, and 40 mg of metoprolol. Echocardiography in July 1993 showed an increase in the left ventricular diastolic dimension to 56 mm (from 46 mm in February 1991) and a reduced left ventricular ejection fraction of 41% (Teichholz method).

Case 2 (An elder brother of the proband)
A 50-year-old man was admitted to our hospital for close examination of chest discomfort and palpitations on September 6, 1979. He first felt palpitation in January 1978. On admission, he had a blood pressure of 106/60 mmHg and a regular heart rate of 60/min. ECG, chest X-ray and echocardiography revealed no abnormalities. In the hospital, episodes of ventricular tachy-

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Cardiac occurred frequently (Fig. 1b). Thallium scintigraphy revealed hypoperfusion in the posterolateral area of the left ventricle. Cardiac catheterization showed no abnormalities in hemodynamic measurement, left ventriculography or coronary angiography. In April 1993, he underwent echocardiography, which showed no abnormality. However, his ECG showed a complete left bundle branch block.

Case 3 (A cousin of the proband)

A 49-year old man was taken to a local hospital by ambulance because of syncope while playing golf on July 26, 1989. ECG revealed ventricular tachycardia at 200/min, which was restored to a sinus rhythm by an intravenous injection of 50 mg of lidocaine. He was referred to our hospital for close examination in August 1989. In 1982, ECG abnormalities were noted in a medical examination. In 1984, he was diagnosed as having dilated cardiomyopathy, since there was no cause for the reduced left ventricular wall motion seen in echocardiography, cardiac catheterization and thallium scintigraphy. Fig. 1c shows an ECG of his ventricular tachycardia.

Case 4 (A cousin of the proband)

A 61-year-old man died of congestive heart failure on December 3, 1989. Beginning in 1975, he had occasionally experienced palpitations. In April 1976, he went to an emergency hospital because of palpitation. ECG revealed ventricular tachycardia. The ventricular tachycardia was restored to sinus rhythm by an intravenous injection of procainamide. In November 1977, he was admitted to our hospital for close examination. On admission, ECG revealed sinus bradycardia of 52/min, an electric axis of +60D, QRS interval of 120 ms and a QS pattern in the V1 lead. An ECG of the ventricular tachycardia is shown in Fig. 1d. No abnormalities were found in cardiac catheterization including coronary angiography, left ventriculography and hemodynamic measurements. However, cardiac thallium scintigraphy showed a cold spot in the inferoposterior wall. At the time, his ventricular tachycardia was thought believed to be idiopathic. He was treated with 600 mg of disopyramide daily. In 1985, echocardiography revealed that left ventricular wall motion was markedly reduced, although previous coronary angiography had denied ischemia. In 1989, he died of congestive heart failure.

Table I showed shows the clinical characteristics of the 4 cases.

Fig. 3 shows the family tree. Two other members of this family died suddenly. The younger brother of case 4 showed reduced left ventricular wall motion and left ventricular dilatation by echocardiography, and posterolateral hypop erfusion by cardiac thallium scintigraphy. He was diagnosed as dilated cardiomyopathy because his left ventricular wall motion was diffusely hypokinetic with
no anginal episodes. His father had died suddenly at the age of 47 without any previous serious disease.

**DISCUSSION**

Most reported cases of familial ventricular tachycardia are associated with hypertrophic cardiomyopathy, long QT syndrome or arrhythmogenic right ventricular dysplasia. Cardiac involvement of palmoplantar keratosis\(^6\) or neuromuscular disease\(^7\) is rarely reported. There have been a few reported families in which there were no definitive causes of ventricular tachycardia\(^8,9\) Familial ventricular tachycardia based on dilated cardiomyopathy is also rare.

Case 3 fulfilled the criteria of dilated cardiomyopathy proposed by Goodwin: ie, a decrease in ejection or fraction fractional shortening and enlargement of left ventricular volume\(^10\). Case 1 had left ventricular wall

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**TABLE I CLINICAL CHARACTERISTICS OF THE 4 CASES**

<table>
<thead>
<tr>
<th>ECG</th>
<th>Rate (min.)</th>
<th>VT</th>
<th>Heart disorders in early period</th>
<th>Treatment</th>
<th>Heart disorders in late period (US)</th>
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<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Disopyramide 450 mg</td>
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<td></td>
<td></td>
<td></td>
<td>Mexiletine 450 mg</td>
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<td></td>
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<td></td>
<td></td>
<td>Metoprolol 40 mg</td>
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<tr>
<td>Case 1 NSR 235 R -150° - + -</td>
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<tr>
<td>Case 2 NSR 150 R +120° + - -</td>
<td>Quinidine 1500 mg</td>
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<tr>
<td>Case 3 cLBBB 214 R +160° - + +</td>
<td>Mexiletine 300 mg</td>
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<tr>
<td>Case 4 cLBBB 52/min. 167 R -60° + - -</td>
<td>Disopyramide 450 mg → Catheter ablation</td>
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<tr>
<td></td>
<td>Metoprolol 40 mg</td>
<td>+</td>
<td></td>
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<tr>
<td></td>
<td>Disopyramide 600 mg</td>
<td>+</td>
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motion abnormalities by left ventriculography. After two years had passed, his left ventricular volume had progressively increased by echocardiography. In case 4, wall motion abnormalities were not initially detected by cardiac catheterization, but left ventricular wall motion became diffusely hypokinetic. The cause of ventricular tachycardia in cases 1 and 3 is thought to be a natural history of dilated cardiomyopathy. In a previous report, 10 patients came to medical attention because of a symptomatic arrhythmia, and received echocardiography and cardiac catheterization. No overt abnormalities were recognized. However, 7 of these patients subsequently developed dilated cardiomyopathy and 3 developed restrictive cardiomyopathy. Heart disease sometimes appears with time in idiopathic ventricular tachycardia. In the early stage of dilated cardiomyopathy, some cases are either normal or show a slight decrease in the left ventricular function. Case 2 showed focal perfusion defects by thallium scintigraphy. Goodwin's criteria for dilated cardiomyopathy did not apply to case 2, who did not show cardiac dysfunction for 15 years since his first ventricular tachycardia. However, he had a focal perfusion defect that may have represented left ventricular dysfunction. In addition, another family member did show dilated cardiomyopathy and two showed sudden death. Therefore, in this family, the cause of ventricular tachycardia may have been dilated cardiomyopathy.

Dilated cardiomyopathy is a multifactorial disease which involves environmental factors. An association of has been reported between dilated cardiomyopathy and viral myocarditis. Dilated cardiomyopathy is found with a familial occurrence of from 2 to 16%, and the mode of inheritance is said to be autosomal recessive or X-linked. Thus, this condition could be associated with various individual immune responses that are genetically regulated. Although this family may have a gene for dilated cardiomyopathy, no obvious inheritance pattern was found. However, environmental factors may be important in dilated cardiomyopathy, and the gene responsible for dilated cardiomyopathy may have been weak in this family.

We believe that this familial ventricular tachycardia may have been caused by dilated cardiomyopathy for two reasons: 1) 3 of the 4 cases had dilated cardiomyopathy and the other member had a left ventricular disorder. In addition, this family contained 2 sudden death victims, including a dilated cardiomyopathy patient; and 2) common causes of familial ventricular tachycardia, such as hypertrophic cardiomyopathy, long QT syndrome, and arrhythmogenic right ventricular dysplasia, were not found in this family.

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