Two Adults Requiring Implantable Defibrillators Because of Ventricular Tachycardia and Left Ventricular Dysfunction Caused by Presumed Kawasaki Disease

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There is an adult patient population in Japan with undiagnosed coronary artery lesions caused by Kawasaki disease (KD) occurring before 1967, the time at which KD was first described. Two adult patients presented with a low left ventricular (LV) ejection fraction and ventricular tachycardia (VT) caused by presumed KD. A 43-year-old man with rapid VT had a history of an acute febrile illness with desquamation of the fingertips at the age of 10 months. Coronary angiography (CAG) showed segmental stenosis of the right coronary artery (RCA) and occlusion of the left anterior descending artery with a giant aneurysm. The other patient was a 48-year-old man with a history of ischemic cardiomyopathy diagnosed after a previous myocardial infarction when he was 32 years old. He had segmental stenosis of the RCA on CAG. Non-sustained VT with transient unconsciousness was observed during 24-h Holter electrocardiography. Rapid VT with syncope was induced in both patients in the electrophysiologic studies and an implantable defibrillator was required to prevent sudden death. Physicians must be aware that VT can occur in older patients with LV dysfunction many years after KD. (Circ J 2005; 69: 870–874)

Key Words: Coronary artery disease; Implantable defibrillator; Kawasaki disease; Left ventricular dysfunction; Non-sustained ventricular tachycardia

Kawasaki disease (KD) is an acute febrile infantile disease, first described in 1967; but there is an adult patient population in Japan with a history of acute KD and cardiac sequelae occurring before 1967. In most of these patients, the coronary artery lesions caused by KD were first recognized after an acute myocardial infarction or at autopsy for sudden death.\(^2\)\(^-\)\(^8\) We present 2 adult patients with ventricular tachycardia (VT) and left ventricular (LV) dysfunction caused by coronary artery lesions after presumed KD.

**Case Reports**

**Case 1**

In 1993 a 34-year-old man visited hospital because of headache. A 12-lead electrocardiography (ECG) revealed an abnormal Q wave in lead III and a QS pattern in V1 and V2. Coronary angiography (CAG) showed segmental stenosis of the right coronary artery (RCA) and occlusion of the left anterior descending artery (LAD) with calcification of a giant aneurysm. At the age of 10 months, he had had an episode of unexplained fever lasting 1 month with desquamation of the palms and fingertips. He was diagnosed in 1993 as having coronary artery lesions caused by KD. Multifocal premature ventricular contractions (PVC) were frequently observed in on 24-h Holter ECG. Furthermore, he had a low LV ejection fraction (LVEF). Beta-blocker was prescribed. At the age of 43 years, he visited another hospital because of fever associated with a common cold. An ECG revealed wide QRS tachycardia at a rate of 198 beats/min (Fig 1), as well as left axis deviation and right bundle-branch block. He was restored to normal sinus rhythm by direct conversion and was referred to us. Body length and body weight were 169 cm and 74 kg, respectively; blood pressure was 130/80 mmHg; total cholesterol was 228 mg/dl. At cardiac catheterization, the LV end-diastolic volume (LVEDV) and LVEF were 97 ml/m\(^2\) and 41%, respectively. The CAG findings were similar to the previous imaging (Fig 2). Electron beam computed tomography showed the occlusion of the LAD with calcification of a giant aneurysm. He underwent coronary artery bypass grafting to the RCA and LAD. Amiodarone was prescribed. During electrophysiologic studies (EPS), 2 clinical and 2 non-clinical episodes of VT were induced in the left postero-septal wall of the left ventricle and a diastolic potential was recorded at the site. Radiofrequency catheter ablation was successful for 3 of the 4 foci. However, it was impossible to ablate the focus inducing rapid VT with syncope, so an implantable defibrillator (ICD) was inserted.

**Case 2**

In 1987, a 32-year-old man visited hospital because of general malaise. He was diagnosed as having ischemic cardiomyopathy after a previous myocardial infarction (MI). There was segmental stenosis of the right coronary artery on CAG but an almost normal left coronary artery. Multifocal PVC and couplets were detected on 24-h Holter ECG. Non-sustained VT with transient unconsciousness was observed during 24-h Holter electrocardiography. Rapid VT with syncope was induced in both patients in the electrophysiologic studies and an implantable defibrillator was required to prevent sudden death. Physicians must be aware that VT can occur in older patients with LV dysfunction many years after KD. (Circ J 2005; 69: 870–874)

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Fig 1. 12-lead ECG of wide QRS tachycardia of 198 beats/min. The ECG shows the left axis deviation and right bundle-branch block.

Fig 2. Case 1: Coronary angiogram shows segmental stenosis of the right coronary artery (Left). (Middle) The left anterior descending artery filled via collateral arteries from the right coronary artery (arrow shows a giant calcified aneurysm of the left anterior descending artery). (Right) The left anterior descending artery is occluded.

Fig 3. Case 2: Coronary angiogram shows segmental stenosis of the right coronary artery (Left) and an almost normal left anterior descending artery (Right).
sensation in his chest immediately after getting up during the night. Non-sustained VT (NSVT) including a 19-beat run was observed during 24-h Holter ECG and he was referred to us. Body length and body weight were 163 cm and 73 kg, respectively; blood pressure was 120/60 mmHg; total cholesterol was 196 mg/dl while taking 3-hydroxy-3-methyl-coenzyme. A reductase inhibitor. He had given up smoking when 32 years old. ECG revealed an abnormal Q wave in lead III, poor progression of the r wave in V1 and V2, and flat T waves in V5 and V6. An episode of acute KD was unknown. LVEDV and LVEF were 147 ml/m² and 32%, respectively, on left ventriculography. The CAG findings were almost the same previously (Fig 3). A perfusion defect in the inferior wall of the left ventricle was found on ⁹⁹ᵐTc-methoxy-isobutyl isonitrile myocardial imaging at rest (Fig 4). The histologic findings of a biopsy of the right ventricle did not correspond to that of any cardiomyopathy. Beta-blocker was prescribed. At EPS, monomorphic NSVT at a rate of 240 beats/min with left axis deviation and polymorphic VT of several beats run were induced by stimulation at the outflow tract of the right ventricle. They stopped spontaneously, and he had presyncope at the time. Furthermore, polymorphic VT was induced by stimulation at the apex of the right ventricle, and it evolved into rapid VT (Fig 5). He collapsed, but was restored to normal sinus rhythm by direct conversion. He had no abnormal potentials at the apex of the right ventricle in sinus rhythm; however, ICD implantation is planned to prevent sudden death.

Discussion

KD is an acute febrile disease affecting children, mainly those less than 5 years of age.¹² Currently in Japan, approximately 6,000–8,000 patients develop KD each year. Its cause remains unknown, but it is a systemic vasculitis involving medium-sized vessels. Diagnosis is based on the major clinical features of acute KD, which include fever of at least 5 days duration, bilateral conjunctival injection, an erythematous reaction involving the lips and oral cavity,
VT Caused by KD

polymorphous exanthema, cervical lymphadenopathy, erythema of the palms and soles and/or firm induration of the hands or feet in the early phases and desquamation of the fingers and toes in the post-inflammation period.1,9 All these symptoms are self-limiting and not all occur in every patient. In addition, the severity of the symptoms varies and for these reasons the diagnosis of KD can be difficult. Acute systemic arteritis particularly affects the coronary arteries. In the 1970s, it was considered that approximately 20% of acute KD patients had cardiac sequela immediately after the acute illness;10,11 however, it was difficult to diagnose both acute KD and the development of coronary artery lesions at that time. Most patients with coronary artery lesions caused by KD are asymptomatic until acute myocardial infarction or sudden death occurs and there are probably many asymptomatic adult patients with coronary arterial lesions caused by KD who remain undiagnosed. The present 2 patients had coronary artery lesions and a low LVEF diagnosed when they were in their 30s. Both had segmental stenosis of the RCA, which is often found in patients with a history of KD and is considered typical of the lesions caused by the disease. It implies the development of several new small vessels, reflecting recanalization after coronary artery occlusion.11,12 Because approximately two-thirds of patients with segmental stenosis or complete coronary occlusion are asymptomatic, coronary artery occlusion cannot be diagnosed without CAG12 and consequently, LV dysfunction after MI can exist unrecognized for many years in such patients. As a result, they are asymptomatic for many years after the episode of undiagnosed acute KD. The 2 patients described here developed VT when in their 40s and it was most likely secondary to myocardial damage after a previous MI. We believe that VT develops with age, many years after the previous MI.

Giant aneurysms in the proximal portion of the coronary arteries are a characteristic coronary artery lesion caused by KD and they often develop late calcification on the outer surface. Case 1 had an occluded and calcified giant aneurysm of the LAD. Stenotic lesions and calcification involving the same segments in which the coronary aneurysms develop occur during the acute phase of KD. Furthermore, affected segments and almost normal segments may be found in the 1 patient.14

These 2 patients were born in the 1950s. The mother of Case 1 remembered the symptoms of his acute illness in infancy, which were consistent with acute KD. Any history of an acute illness in childhood was unknown in the other patient. However, we suspect that the characteristic coronary artery lesions and ischemic cardiomyopathy occurring in middle-age of individual without high risk of atherosclerosis signify a KD etiology. Adults with ischemic cardiomyopathy, severely depressed LV function, and asymptomatic NSVT are at significant risk for future arrhythmic events. Sudden death occasionally occurs in their 20s in patients with LV dysfunction and NSVT after KD.15,16 In such cases there was probably an undiagnosed MI soon after the onset of KD and they then remained asymptomatic prior to their sudden death. We suspect that fatal arrhythmias in such patients are a late complication of MI1,13,18,19 which might occur earlier in KD patients with a low LVEF than in adults with LV dysfunction caused by atherosclerosis.

Treatment for KD patients with long-standing LV dysfunction after previous MI becomes more essential as they get older. An EPS and then antiarrhythmic treatment will be required to prevent sudden death. If critical VT is detected, catheter ablation or implantable cardioverter-defibrillator should be considered20,21 because patients with severe coronary artery lesions caused by KD are likely to have cardiac events earlier than the normal population.

Conclusion

There is a population of adults in Japan with undiagnosed coronary arterial lesions caused by KD. They can be recognized by an episode of ventricular arrhythmia and a low LVEF, as well as by acute MI or sudden death. Detection and treatment of such KD patients is essential to prevent premature death.

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References

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