An Autopsied Case of an Elementary School Boy with Sudden Death Four Years after Kawasaki Disease: On the Problem of Present Method of Cardiac Mass Screening of School Children

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An 8 year-old boy died suddenly 4 years after the onset of Kawasaki disease. He was examined by a cardiac mass screening for school children one and a half year before the death, and was evaluated as having no sequela of Kawasaki disease. The autopsy showed coronary arterial aneurysms and obstruction with fresh and old myocardial infarction. One of the problems of the present method for cardiac mass screening for school children is that it is performed only with history taking, physical findings and electrocardiograms. This method is completely insufficient to find out coronary involvement as a sequela of Kawasaki disease. All the children with history of Kawasaki disease should be examined by two dimensional echocardiography, which is the most sensitive and specific noninvasive method to detect the coronary involvement in our experience.

CASE REPORT

Case: 8 year-old boy.

Family History and Personal History
Nothing particular.

Present Illness
He had suffered from Kawasaki disease at 4 years of age. An electrocardiogram and a chest film at that time in a hospital revealed no abnormality. After one month from the onset, he was judged that he had completely recovered from the disease. Electrocardiograms were taken twice during the following 9 months at the hospital, and the findings were almost negative (Fig. 1). The patient was concluded at that time that he had no sequela of Kawasaki disease and further follow up was discontinued.

At 6 years of age, when the patient entered an elementary school, a cardiac mass screening with history taking and electrocardiographic examination was performed. He was checked up for the further examination because of positive history of Kawasaki disease and sinus arrhythmia on an electrocardiogram. The second examination containing physical examination and an electrocardiogram (Fig. 1) revealed nothing particular, and he was concluded as having no abnormality and no necessity for the restriction of exercise.

At 8 years of age, when the patient was the second-year of the elementary school, he ran slowly about 250 meters with his classmates as an exercise training in physical education. Immediately after the running, he was found to fall down on the ground. Paleness and cyanosis of the face and weakened peripheral pulse were noted, and he was transported to a near emer-

Key Words:
Kawasaki disease
Coronary aneurysm
Myocardial infarction
Sudden death
Mass screening

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Fig. 1. Serial electrocardiograms in a patient with Kawasaki disease. The tracings were essentially within normal limits.

Findings of Autopsy
Autopsy was done one and a half hour after his death. Macroscopically, there were 6 aneurysms at the left coronary artery. The largest one was at the bifurcation of the left coronary artery and the size was 1.5 cm in diameter (Fig. 3). The others were much smaller in size, less than 0.5 cm in diameter. Four aneurysms were found at the anterior descending artery and
other one was at the circumflex artery. Cutting the largest aneurysm, a red-brown fresh thrombus was found at the inlet of the left main artery to the aneurysm. The right coronary artery revealed no abnormal findings on macroscopic examination. The surface of the sliced left ventricular wall showed white and dark-brown discoloration in parts (Fig. 4).

Microscopically, at the inlet of the left main coronary artery to the largest aneurysm, the structure of the three layers of the muscular artery was destroyed with remarkable arteriosclerotic changes. Elastic fibers of the media were almost diminished (Fig. 5). The lumen of the artery at the inlet was markedly obstructed. Fresh thrombus was found at the narrowing. Fresh and older organized thrombi were noted at the inner surface of the aneurysm. As to discoloration of the left ventricular wall, white region was piles of fibrosis and was thought to be old infarction. On the other hand, dark-brown areas were bleeding with wavy change, myolytic region and granular degeneration of the myocardial fibers, which was considered as an expression of fresh myocardial infarction (Fig. 4). The right coronary artery revealed significant thickening of the intima, in spite of a normal macroscopic appearance and normal findings of postmortem coronary angiograms.

**DISCUSSION**

Kawasaki disease was first reported in 1967 by Dr. Kawasaki. This is an acute febrile illness mainly prevailing in Japan, and the etiology is yet unknown. The number of the patients is increasing year by year.

Incidence of the school children with a history of Kawasaki disease was as high as 0.2% at the first-year of the elementary school. Kawasaki disease is now becoming one of the biggest problems in the field of health management of school children.

The most important sequela of Kawasaki disease is coronary arterial involvement. Incidence of coronary aneurysm had been reported as 15–20% of the patients. Among 315 patients with a history of Kawasaki disease, to whom coronary cine-angiography were performed without sparse selection in our hospital, incidence of coronary aneurysm was 20.6% (65 cases).

To find out coronary aneurysm noninvasively, sensitivity and specificity of the methods of

Fig. 4. The short axial cut surface of the ventricle showed areas of white and dark brown. White area was due to myocardial fibrosis, indicating presence of an old infarction. Dark brown areas were apparently due to wavy change, myolysis and granular degeneration of the myocardial fiber. These were thought to be related to a fresh myocardial infarction.

Fig. 5. Microscopic findings of the inlet of the main coronary artery to the largest aneurysm. The structure of the three layers of the muscular artery was destroyed with remarkable arteriosclerotic changes. The lumen of the artery at the inlet was obstructed with a fresh thrombus.

examination among 65 cases with coronary aneurysm on angiograms are as follows: history taking; 0.03, 0.25, physical examination; 0.03, 0.25, electrocardiogram; 0.06, 0.44, chest film; 0.03, 0.25, two dimensional echocardiogram; 0.81, 0.93 4 As clearly demonstrated, the most useful noninvasive method is two dimensional echocardiography. Even in this case, aneurysm of the left coronary artery could be detected by two dimensional echocardiography before the autopsy. To prevent unexpected sudden death among the school children with a history of Kawasaki disease and without any apparent positives by "routine" screening methods, two dimensional echocardiogram should be performed to all the children with a history of Kawasaki disease. Even if the history is thought to be doubtful, echocardiographic examination is proposed because it is not time-consuming and harmless.

On the fate of aneurysm, in our study of successive coronary cine-angiographies with an interval of one year, significant number (7 cases among 20 cases) showed progressive stenosis of the coronary artery just at the points of inlet and outlet of the aneurysm and of bifurcation of the coronary arteries. On the reported case, the primary cause of the death is thought to be acute occlusion of the markedly stenosed artery by fresh thrombus at the point of inlet of the aneurysm. Susceptibility of stenosis at these point may be due to turbulence of flow and micro-thrombi.

Although the reported case had had no subjective complaints, such as chest pain, after the "recovery" from Kawasaki disease, autopsy disclosed not only fresh but old myocardial infarction. This may be thought of as having a defective anginal warning system, as recently discussed. 5

Based upon our investigation, among 65 cases with coronary aneurysm, 7 was disclosed as having combined obstructive lesions of the coronary artery by cine-angiogram. Among the cases with coronary arterial obstruction, only one patient had symptoms suggesting an attack
of myocardial infarction. Myocardial imaging by thallium-201 revealed hypoperfusion or perfusion defect in 7 cases out of 16 with obstructive change, and excercise electrocardiogram by treadmill found out ischemic change in one case out of 5.

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