Ischemic Heart Disease 31 Years After Possible Kawasaki Disease

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We present a 38-year-old woman who had experienced an acute febrile illness lasting more than 1 week at the age of 6, with erythema on the palms and soles following skin desquamation in the subacute phase and skin eruption. Thirty one years later, she experienced acute myocardial infarction and episodes of angina pectoris. She had no coronary risk factors or autoimmune diseases. Coronary angiography revealed an aneurysm of the proximal left coronary artery with occluded lesions at the distal site. The right coronary artery was also occluded at the proximal site. These findings strongly suggest Kawasaki disease should be considered in the differential diagnosis of early-onset ischemic heart disease in young adults.

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Mucocutaneous lymph node syndrome (Kawasaki disease) is an acute febrile illness of unknown etiology which predominantly affects infants and children below the age of 6 years! Kawasaki disease frequently causes cardiovascular angitis, which characteristically forms coronary aneurysms. About half of the coronary aneurysms regress spontaneously, but some become stenotic or obstructed, causing angina pectoris or myocardial infarction. Some reports have indicated that myocardial infarction usually develops within 6 years after the onset of Kawasaki disease. In this paper, we report a patient with acute myocardial infarction (AMI) and subsequent attacks of angina pectoris 31 years after suspected Kawasaki disease.

CASE PRESENTATION

A 38-year-old woman visited our hospital because of chest pain on effort. At the age of 6 in 1961, she was affected by pyrexia which lasted for more than 1 week. Her other symptoms and signs had been erythema on the palms and soles with arthralgia following skin desquamation in the subacute phase, and skin eruption. A diagnosis of rheumatic fever was made at that time. She recovered from the illness without specific therapy. She safely gave birth to two children at 28 and 31 years of age, respectively. She had been doing well until she developed severe chest pain with cold sweating which lasted about 1 h at the age of 32. At the age of 37, she felt a similar chest pain which lasted 3 h and visited a local hospital. A diagnosis of subendocardial AMI was made based on an elevated creatine kinase level (CK: 266 IU/L) and changes in the electrocardiogram (ECG; negative T waves in II, III, aVF), and she was admitted to the hospital. After several months, she was referred to our hospital because of repeated chest pain on effort.

On admission, she showed no symptoms or signs of congestive heart failure. Blood
pressure was 104/62 mmHg with no laterality. Ocular fundi were normal. No vascular bruit was audible. She had no coronary risk factors: no smoking, no diabetes, and her serum level of total cholesterol was 152 mg/dl. A blood test did not demonstrate any obvious inflammatory diseases. The serum CK level was normal. ECG at rest showed normal sinus rhythm and QS waves in V_{1,2} without ST-T changes. Exercise-ECG showed sagging ST depression in I, II, aVL, aVF, V_{3-6} accompanied by chest pain, which was relieved by sublingual nitroglycerin. Stress thallium-201 myocardial perfusion imaging showed a stress-induced transient perfusion defect in the anteroseptal-apical area, and a persistent perfusion defect in the inferior area and part of the septum (Fig 1). Left ventriculography showed hypokinesis of the septum and inferior wall. Coronary angiographic findings were as follows (Fig 2). Fluoroscopy showed calcification of the right (RCA) and left coronary arteries (LCA) along the proximal sites. There was no ostial stenosis. The RCA and LAD were occluded at the proximal sites. The proximal LCA was dilated maximally at the left main trunk (diameter = 6.7 mm), like an aneurysm with an irregular wall. There were no organic lesions of the distal branches, but the main arteries distal to the occluded sites were poorly opacified via the developed collaterals. She has been treated by medicine including anti-anginal agents and aspirin thereafter, and was not operated upon because the distal arteries to be anastomosed were so poorly viewed by contrast medium that we thought these arteries were not suitable for a bypass graft operation. She continues to do well, except for angina attacks on moderate exercise.
DISCUSSION

Patients who develop AMI in young adulthood frequently have some coronary risk factors, such as hypercholesterolemia (especially "familial") or uncontrolled diabetes mellitus, or an underlying disease, such as vasculitis associated with autoimmune diseases. Our patient, however, had no coronary risk factors or inflammatory diseases. It is likely that she was affected by Kawasaki disease at the age of 6, because her three clinical features (pyrexia for more than 5 days, changes in the extremities, and eruption) and the characteristic findings of our coronary angiographic study (coronary aneurysms with occluded lesions) are compatible with Kawasaki disease. These coronary lesions are thought to be the cause of ischemic heart disease in this patient. Since her illness occurred in 1961, 6 years before the first report of Kawasaki disease in 1967, a diagnosis of Kawasaki disease could not have been made. It is possible that Kawasaki disease was sometimes diagnosed as rheumatic fever at that time in Japan.

Kawasaki disease frequently induces coronary aneurysms during the acute phase within 1 month after its onset. Half of the aneurysms regress spontaneously during the following subacute phase, but several percent become stenotic or obstructed, which may cause angina pectoris or myocardial infarction. The natural course of the coronary lesions largely depends on the size of the coronary aneurysms. Aneurysms less than 4 mm in diameter regress spontaneously within a short time, whereas those larger than 8 mm in diameter often become stenotic or obstructed. Since the maximal diameter of the coronary artery in our patient was 6.7 mm, it is likely that the aneurysm was larger in the acute phase but regressed partially, and some lesions were obstructed in the subacute or chronic phase. AMI has been reported to develop several years after the onset of Kawasaki disease, but usually within 1 year. As reported by Kato et al, ischemic heart disease rarely develops more than 20 years after Kawasaki disease.
disease in adult survivors. Our patient also had no history of chest pain until the age of 32, 26 years after the suspected Kawasaki disease. We confirmed that she had ischemic heart disease based on her symptoms, ECG changes, CK levels, thallium-201 myocardial scintigram, left ventriculography and coronary angiography. However, it is unclear why ischemic heart disease appeared after a long symptom-free period in this patient.

In conclusion, Kawasaki disease should be considered as a possible cause of ischemic heart disease in young adulthood, especially when patients have no obvious susceptible factors. It is important for cardiologists to be aware of this sequelae and to carefully examine the medical history of the patient’s childhood.

REFERENCES

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