Kawasaki disease recurrence with cardiac tamponade

Kardiyak tamponatla rekürrens gösteren Kawasaki hastalığı

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Introduction

Kawasaki disease is an acute febrile illness; characterized by bilateral conjunctival congestion, changes of lips and oral cavity, polymorphous exanthema, changes of peripheral extremities and acute nonpurulent cervical lymphadenopathy. It is an acute vasculitis syndrome of unknown etiology that primarily affects small and medium sized arteries, particularly coronary artery (1). One of the interesting features of the disease is its recurrence.

In this case report, we describe a patient with recurrent Kawasaki disease who was admitted with the signs of cardiac tamponade and was successfully treated with pericardiocentesis and pulse methylprendisolone therapy.

Case report

A six-year-old girl was admitted with two weeks history of high fever, rash, sore throat and cervical mass. Ten days before, she had been admitted to the hospital and diagnosed as lymphadenitis. Parenteral cephazoline treatment had been prescribed. Despite antibiotic therapy, her elevated temperature had continued for two weeks and conjunctivitis, diffuse edema on the hands and feet had been observed on follow-up. She had been referred to our hospital.

On physical examination the following findings were detected: weight: 20 kg, height: 115 cm, temperature: 40 ºC, pulse rate: 130/min, respiratory rate: 34/min, and blood pressure: 90/60 mmHg. Physical examination demonstrated an irritable child with bilateral cervical lymphadenopathy, pharyngeal erythema, bilateral conjunctival congestion, maculopapular rash predominantly on the chest, injected and fissured lips, diffuse arthralgia, edema on the palms and soles.

On laboratory evaluation, the white blood cell count was 21600/mm^3 with 80% polymorhonuclear leukocytes. There were thrombocytosis with platelet count of 950000/mm^3 and normocytic anemia with hemoglobin of 9 gr/dl. The erythrocyte sedimentation rate (ESR) (130 mm/h) and C-reactive protein (CRP) (30 mg/dl) were of very high levels. Repeated blood, urine, stool cultures were negative. Diagnostic tests for acute measles, Epstein-Barr virus, Enterovirus infection and scarlet fever were negative. Renal and liver function tests were normal. The ANA, Anti-DNA, ANCA and RF were negative.

Electrocardiogram (ECG) was normal. Echocardiographic examination revealed 5 mm diffuse aneurysmatic dilatation of left anterior descending coronary artery (LAD) (Fig. 1). Ejection fraction and fractional shortening of the both ventricles were within normal limits.

With all these findings, she was diagnosed as Kawasaki disease. Treatment with intravenous immunoglobulin (IVIG) (2 gr/kg, single dose), aspirin (90 mg/kg/day, in four doses) and dipyridamole (1 mg/kg) were started. Second dose of IVIG (2gr/kg) was given because she was still febrile at the end of 48 hours after first dose. The fever subsided within five hours. Irritability, rash, conjunctival congestion, edema of the palms and soles were completely regressed. Diffuse desquamation from the fingertips was observed. Ten days after second dose of IVIG treatment, ESR was 50 mm/h and CRP was 1,5 mg/dl. She was discharged from the hospital with aspirin (75mg/kg) and dipyridamole (1 mg/kg) treatments. On follow-up, physical examination was completely normal; ESR decreased to 30mm/h, and CRP to 0,5mg/dl. Aspirin dose was gradually decreased to antiaggregant dose (5 mg/kg/day).

One month after discharge from the hospital, the patient was readmitted with the two days history of high fever, severe chest pain, respiratory distress and rash on the trunk. On physical examination patient was dyspneic, ortopneic and tachypneic with the respiratory rate of 46/min. Temperature was 40 ºC. There were tachycardia with the pulse rate of 180/min hypotension and pulses paradoxus. Laboratory evaluation revealed increase in acute phase reactants: White blood cell count: 18400/mm^3, CRP: 15 mg/dl and ESR: 85 mm/h. There was thrombocytosis with the platelet count of 1000000/ mm^3.

On ECG QRS voltages were low in the limb leads and there was ST segment elevation in the leads representing the left ventricle. Echocardiography revealed 20 mm of pericardial effusion compressing right atrium and ventricle in diastole (Fig. 2). There was no any sign of thrombosis or rupture involving the LAD. These clinical and laboratory findings made us to think the recurrence of Kawasaki disease with signs of acute cardiac

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tamponade. To prevent a circulatory collapse, emergent echocardiography-guided pericardiocentesis was undertaken and 300 ml of serohemorrhagic fluid was discharged. Culture of this fluid was negative. For the treatment of reactivation, pulse methylprednisolone therapy (30 mg/kg/day IV for 3 days) was given. Progressive clinical improvement was observed within 24 hours. Fever was subsided and acute phase reactants rapidly decreased. On the 7th day of hospitalization, physical examination was completely normal and acute phase reactants were within normal limits. During the 14 months of follow up, no recurrence was observed and LAD aneurysmatic dilatation has decreased in size to 2 mm.

Discussion

One of the interesting features of the Kawasaki disease is its recurrence, which is reported in the United States of America, in Japan and also in other countries. According to nationwide surveys, the proportions of recurrences among all patient populations are 0.8 % in the Unites States and 3 % in Japan (2,3).

There are two cohort studies that showed risk factors associated with recurrent Kawasaki disease. One is a hospital-based follow up study in the United States of America, which indicates that recurrence is significantly more frequent among Asian patients (2). The other study shows that Kawasaki disease recurred 1.47 times more often in children treated with IVIG than in those without therapy (3). The study done by Nakamura et al shows that cardiac sequel occurs more frequently after a recurrence of Kawasaki disease than after the initial episode (4). Linked data of the initial and second episodes of Kawasaki disease shows that the risk of developing cardiac sequel attributable to recurrent Kawasaki disease is high in those with or without the sequel at the initial episode (5,6).

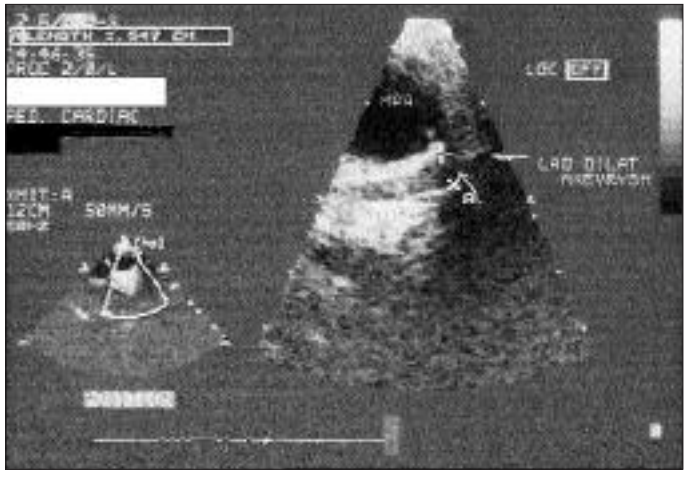

Figure 1. Two-dimensional echocardiography of the left anterior descending artery with diffuse aneurysmatic dilatation

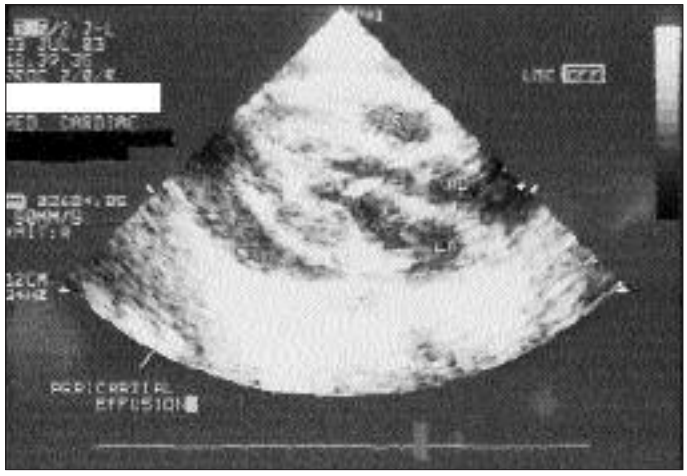

Figure 2. Parasternal long-axis view of the heart with massive pericardial effusion compressing the right atrium and ventricle during diastole

In the presented case, cardiac tamponade occurred after complete recovery, which was achieved with second dose of IVIG. Various alternative diagnoses for the pericardial effusion like bacterial or viral infections, connective tissue disease and drug related polyserositis were excluded. Neither IVIG nor aspirin are known to provoke such pericardial effusion. The absence of even subtle signs of glomerulonephritis, arthritis, and gastrointestinal complaints was incompatible with connective tissue diseases, other forms of vasculitis or serum sickness.

The patient was successfully treated with pulse methylprednisolone and pericardiocentesis. Pulse methylprednisolone was given not only for cardiac tamponade but also given for disease reactivation after the repeated IVIG treatment.

In this IVIG resistant patient, acute life threatening cardiac tamponade occurred as a serious complication of recurrent Kawasaki disease. Pericardial effusion as a complication of Kawasaki disease was reported in 6-24.5% of the patients (7,8). However, cardiac tamponade is very rarely seen and most of them are secondary to the rupture of coronary artery aneurysm (9). In our patient, cardiac tamponade occurred without rupture of aneurysm. Similar to our case, Dahlem reported a child with apparent Kawasaki disease in whom cardiac tamponade developed 14 days after responding to IVIG and aspirin, and who then responded well to three daily doses of intravenous methylprednisolone (10).

The administration of high dose IVIG is observed to reduce both the duration of fever and the incidence of coronary artery aneurysm when given within a few days of the onset of the disease. Nonetheless, approximately 10% of the patients have persistent or recurrent fever despite IVIG (6). These patients are at greatest risk of developing coronary artery aneurysm (11). Repeating IVIG tends to resolve many cases. Pulse steroid therapy should be discussed as a form of treatment for the patients who exhibit resistance to repeated IVIG treatment (11,12). Corticosteroids have a potential to increase the risk of coronary artery aneurysm formation. So, they are not used as a first line treatment modality. Steroid treated subjects can show transient coronary dilatation coincident with this therapy. The authors therefore suggest caution and careful echocardiographic examinations of patients receiving such therapy (11). In our patient, we did not observe increase in the size of previously detected coronary aneurysm.

As a conclusion, in recurrent Kawasaki disease we should be careful for the presence of pericardial effusion and/or cardiac tamponade as a cardiac complication. In the patients with cardiac tamponade, pericardiocentesis combined with pulse steroid therapy may be used as an effective treatment modality.

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