

Septated pericarditis associated with Kawasaki disease: a brief case report

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Kawasaki disease (KD) is primarily the systemic vasculitis of childhood that affects mainly the medium-sized arteries, such as the coronary arteries. KD is the leading cause of acquired heart disease, whereas the incidence of rheumatic fever has declined. The most serious complication is coronary artery involvement. Among the children with KD who developed cardiac complications, pericarditis is a rare complication, with an incidence of 0.07%. We report our experience in a 5.5-year-old child with KD complicated with aneurysm of the left anterior descendant coronary artery and septated pericardial effusion, which has not been reported in the literature. The pericardial effusion disappeared very dramatically with intravenous immunoglobulin (IVIG) therapy. We would like to point out that septated pericardial effusion in cases of KD do not need any further therapy other than IVIG and high-dose acetylsalicylic acid.

Key words: Kawasaki disease, septated pericardial effusion, coronary artery aneurysm, septated pericarditis.

Kawasaki disease (KD) was described in 1967 by Tomisaki Kawasaki¹. It is primarily the systemic vasculitis of childhood that affects mainly the medium-sized arteries, such as the coronary arteries². KD is the leading cause of acquired heart disease whereas the incidence of rheumatic fever has declined³. Although it affects children of all races worldwide, the attack rates are highest in those of Asian ethnicity³. The etiology of KD is still unknown and mostly affects children under five years old, with the peak incidence occurring at 9-11 months of age^{3,4}.

There is no specific diagnostic test available for KD and the diagnosis is based on presence of characteristic clinical findings⁴. The most serious complication of KD is coronary artery involvement. Among the children with KD who developed cardiac complications, pericarditis is a rare complication, with an incidence of 0.07%⁴. We report our experience in a 5.5-year-old child with KD complicated with

aneurysm of coronary arteries and septated pericardial effusion, which was not reported in the literature previously.

Case Report

The patient, who was previously healthy, presented at the age of 5.5 years with fever (>38.5°C), strawberry tongue, arthritis of the knees, cervical lymphadenopathy and macular rash on the lower extremities for the last 13 days. The clinical history revealed previous hospitalization for intravenous antibiotic usage for the treatment of lymphadenitis but no recovery of any symptoms. During his treatment, he developed generalized edema, dyspnea, pyuria and elevation of serum transaminase levels. He had cardiac friction rub in his physical examination, chest X-ray revealed pleural effusion, echocardiography revealed pericardial effusion, and abdominal computerized tomography revealed intra-abdominal effusion.

The patient was hospitalized with the diagnosis of KD. He had desquamation of the skin at the anal region and upper extremities, and levels of serum acute phase reactants including erythrocyte sedimentation rate (62 mm/h) and C-reactive protein (9.01 mg/dl) were high. His new echocardiography and magnetic resonance imaging (MRI) angiography revealed aneurysmal dilatation at left anterior descending coronary artery with a diameter of 4.8 mm and minimal dilatation with diameter of 4 mm at the proximal region of the right coronary artery (Fig. 1). His echocardiography revealed the septated pericardial effusion with approximately 20 mm in diastole at the posterior wall of the left ventricle (Fig. 2). The investigations including viral serologic tests and bacteriologic tests for the etiology of septated pericardial effusions were all negative.

Intravenous immunoglobulin (IVIG) (2 g/kg/total dose) and high-dose (100 mg/kg/day, divided into 4 doses/day) acetylsalicylic acid treatment was applied. Since the acute phase reactants and platelet levels were still high, high-dose acetylsalicylic acid treatment continued for 14 days. The repeated echocardiography on the 14th day revealed that the septated pericardial effusion disappeared but there was no change in the coronary aneurysm. The levels of serum acute phase reactants, including erythrocyte sedimentation rate and C-reactive protein, decreased. The patient was discharged from the hospital with low-dose 3 mg/kg/day acetylsalicylic acid. The echocardiography repeated after one month revealed minimal change in aneurysmal dilatations. At the 6th day of illness, selective coronary angiography revealed fusiform aneurysm with a 4 mm



Fig. 1a. Echocardiography showing the aneurysm with 4.8 diameter at the left anterior descending artery.



Fig. 1b. Fusiform aneurysmal dilatation at the segment 2 level of the left anterior descending artery (MRI).



Fig. 2. Multiple septated (S) pericardial effusion (PE) at the posterior wall of left ventricle (echocardiography).

diameter at the beginning of the left anterior descendant artery. The other branches of left and right coronary arteries were normal in size and appearance.

Discussion

Kawasaki disease is a multi-system disease with unknown etiology and is an acute vasculitis affecting medium-sized arteries, such as coronary arteries⁴. KD mostly affects children under five years, children aged six months to five years old, with the peak incidence occurring at 9-11 months of age^{4,5}. The diagnosis of KD is based on the clinical features including fever $>38.5^{\circ}\text{C}$ for at least five days together with rash, bilateral conjunctival injection, changes in the peripheral extremities, lymphadenopathy, and oral changes⁶. The presence of coronary artery aneurysm (CAA) with three other features is sufficient for the diagnosis.

The most life-threatening and commonest complication of KD is the CAA^{3,8}. If KD is untreated, 25% of patients develop CAA and a small proportion develop myocardial infraction and death⁵. The early diagnosis and treatment of KD are very important to prevent complications, especially cardiac. IVIG (2 g/kg) plus high-dose acetylsalicylic acid (80-100 mg/kg/day in 4 divided doses) combination therapy³ is considered as treatment and should be given preferably within 10 days of KD⁷.

Among the children who developed cardiac complications, coronary artery ectasia (46%), CAA (27%), and myocarditis (20%) are the most commonly seen complications⁴. Pericarditis is a rare complication, with an incidence of 0.07%⁴. A previous study in our center revealed that one of the nine patients with KD had pericardial effusion and myocarditis⁸. In our case, who

fulfilled the diagnostic criteria, complications including CAA, septated pericardial effusions and pericarditis developed. Although CAA did not seem to respond to the IVIG and high-dose acetylsalicylic acid therapy, the septated pericardial effusion did respond to treatment very dramatically. There is no reported case with septated pericarditis associated with KD in the literature, so we want to point out that septated pericardial effusion might be associated with KD and hence, in cases with septated pericardial effusions with undefined etiology, KD should be kept in mind.

In conclusion, septated pericardial effusion may be a sign of KD, in addition to coronary artery involvement, and may not necessitate other treatment modalities.

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