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Kawasaki disease in adolescence

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Summary

Kawasaki disease is a vasculitis causing morbidity and mortality due to coronary artery involvement which is more frequent during infancy and early childhood. Since the disease is rare in older children and adolescents, the diagnosis may be delayed and the risk of coronary artery involvement may increase. We herein present a case of Kawasaki disease diagnosed in an adolescent patient, involving the coronary arteries and the lungs. *(Turk Arch Ped 2013; 48: 169-172)*

Key words: Adolescent Kawasaki disease, coronary artery dilatation, pulmonary involvement

Introduction

Kawasaki disease is an acute vasculitis which mostly affects children below the age of 5 and which may involve small and medium arteries (1,2). Although late complications related with coronary artery involvement occur in the adulthood and adolescence, cases diagnosed with acute Kawasaki disease are rare and ignoring this disease in this age group may lead to missing of the diagnosis (3). In untreated patients, severe cardiac complications may develop. Therefore, it is important to perform echocardiographic evaluation in terms of coronary involvement, when compatible clinical findings are observed together with fever at all ages. Here, we present an adolescent patient diagnosed with Kawasaki disease with predominant lung involvement.

Case

A 17-year-old female patient presented with an axillary fever of 39 ºC which started 12 days ago and a lump in the right side of the neck which appeared 10 days ago and did not respond to antibiotic treatment. On pysical examination, the axillary fever was found to be 39°C, the sclerae were subicteric, a hyperemic and adherent lymphadenomegaly with a size of 5x5 cm was found in the right cervical region and hepatosplenomegaly was present. Laboratory findings were as follows: anemia (Hb:10.6 g/dL), thrombocytopenia (85000/mm^3), 78% polymorphonuclear leukocytes on peripheral smear, increased erythrocyte sedimantation rate (52 mm/h; n=0-10 mm/sa) and increased C- reactive protein (37.7 mg/L; n=0-5 mg/L), increased transaminases (AST:160 U/L, ALT: 95 U/L, GGT: 219 U/L, ALP: 190 U/L, LDH: 401 U/L), direct hyperbilirubinemia (4,9 mg/dL) and hypoalbuminemia (2.7 g/dL). Blood and urine cultures were sterile. Normal throat flora was found on throat culture. Epstein-Barr virus, cytomegalovirus, Parvovirus B19, anti nuclear antibody (ANA), anti ds DNA, c-ANCA, p-ANCA were found to be negative and postero-anterior lung graphy was found to be normal. On the first day of hospitalization, maculopapular rash which started from the abdomen and extended to the arms and legs and turned pale when pressure was applied and non-purulent conjunctivitis were observed. Echocardiography (EKO) which was performed in terms of "incomplete" Kawasaki disease was found to be normal. On the third day of hospitalization, peeling of the skin on the tips of fingers, cough and dyspnea developed. Respiratory sounds were decreased in the middle and lower areas of both lungs and expirium was prolonged. A diagnosis of interstitial pneumonia was made with an extensive ground glass appearance and bilateral pleural fluid (Picture 1) on thoracic computarized tomography (CT). Viral serologic tests for Legionella Pneumophilia, Chlamidia Pneumonia, Mycoplasma Pneumonia, influenza H1N1 which were ordered to find the etiology of pneumonia were found to be negative. Piperacillin,

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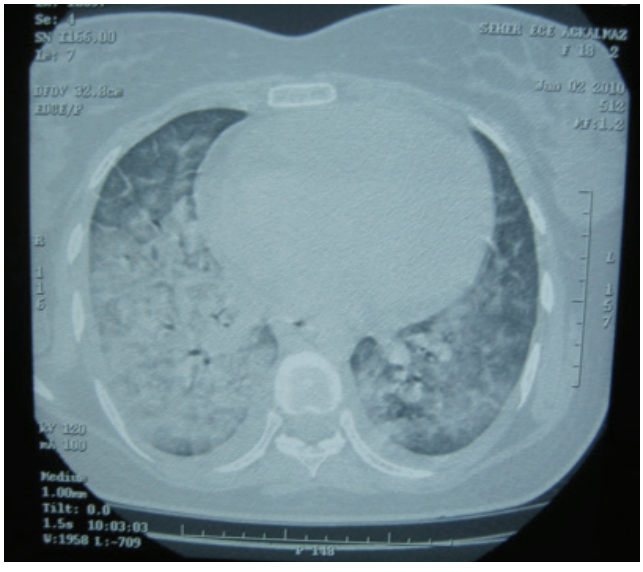
tazobactam and tamiflu treatment was started. When fever persisted and new findings compatible with Kawasaki disease appeared, echocardiography was repeated and extensive dilatation in the coronary arteries (left coronary artery diameter: 4 mm, right coronary artery diameter: 4 mm, LAD (left anterior descending artery): 5 mm) and minimal pericardial fluid (4 mm) were found (Picture 2a,2b,2c). Intravenous gammaglobulin (IVIG) 2 g/kg and acetylsalicylic acid (ASA) 80 mg/kg/gün (3.5 g) treatment was administered. After IVIG, fever and respiratory complaints recovered rapidly starting from the third day. The coronary dilatations returned to normal and the fluid was reduced in the second week. Acetylsalicylic acid treatment was completed to 8 weeks at the antiaggregant dose. Hair loss and 'beau's lines' on the nails were observed in the convalescence period. No complication occured during the one-year follow-up period. The patient was continued to be followed up without any finding.

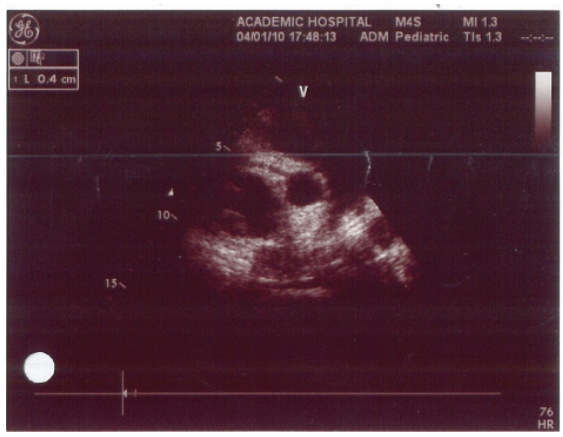
Discussion

Kawasaki disease is the most common cause of acquired heart diseases in the childhood following acute rheumatic fever. Coronary artery aneurism or dilatation which lead to myocardial infarction, sudden death and ischemic heart disease may develop in 15-25% of the untreated children (1,2).

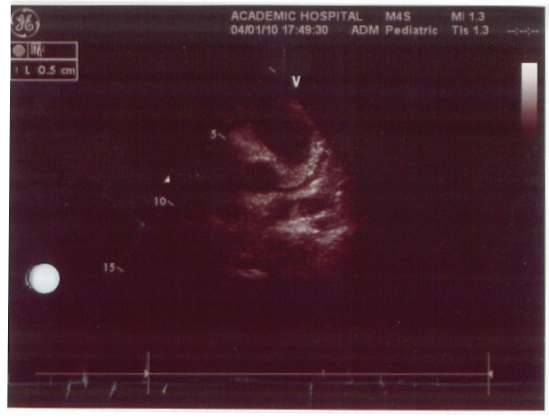
Since there is no specific test for the diagnosis of Kawasaki disease, the diagnosis is based on clinical findings. At least four out of the findings including extremity changes, polymorphous exanthem, bilateral conjunctival congestion, changes in the lips and oral cavity and non-



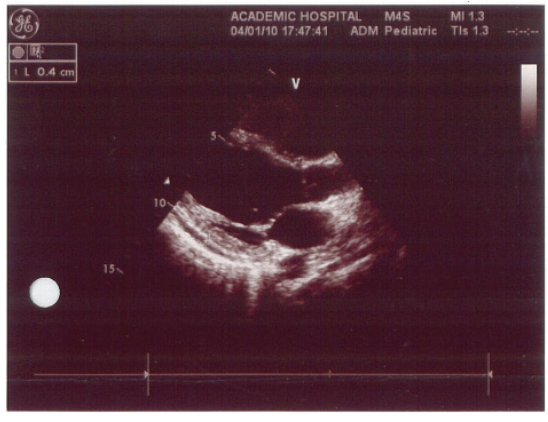
Picture 1. Diffuse interstitial pneumonic infiltration is observed in both lung areas on computarized tomography of the chest.



Picture 2a. Dilated right coronary artery is observed in the parasternal short axis image on two-dimensional echocardiographic examination



Picture 2b. Dilated left coronary artery is observed in the parasternal short axis image on two-dimensional echocardiographic examination



Picture 2c. Pericardial effusion is observed posterior to the left ventricle in the parasternal long-axis image on two-dimensional echocardiographic examination

suppurative cervical lymphadenopathy with a diameter of at least 1.5 cm accompanying fever which lasts at least for 5 days are sufficient for the diagnosis (2). Kawasaki disease may be confused with sepsis, toxic schock syndrome (streptococcal, staphylococcal), scarlet fever, enterovirus, adenovirus, measles, parvovirus, Epstein-Barr virus, cytomegalovirus, mycoplasma, rickettsia, leptospira infections, drug allergy, Stevens-Johnson's syndrome and juvenile rheumatoid arthritis (1,2). In typical patients, investigation directed to all these diagnoses may not be needed, but infectious causes had to be excluded in our patient, since she presented at an older age and atypical findings including interstitial pneumonia in addition to typical findings including lymphadenopathy, polymorphous exanthem, bilateral conjunctival congestion and peeling of the skin on the tips of fingers. In the differential diagnosis, toxic schock syndrome was considered primarily, but presence of coronary artery dilatation, lack of improvement of interstitial pneumonaia despite efficient antibiotic treatment and rapid response to IVIG treatment removed us from this diagnosis.

Although hematological abnormalities including thrombocytosis, leukocytosis and anemia are reported frequently in Kawasaki disease, thrombocytopenia is observed rarely (2,4). Hara et al. (4) reported in their study that thrombocytopenia developed in 10 (2%) of 486 patients with a diagnosis of Kawasaki disease a mean period of 6.8±2.2 days after the disease onset, the number of platalets returned to normal in 1-2 weeks and this was related with clotting-mediated platalet consumption. In our patient, thrombocytopenia found at the time of presentation improved after the second week of treatment.

Although Kawasaki disease can be observed at any age, 85% of the patients are younger than 5 years of age. Kawasaki disease is a rare disease for adults. In the literature, 57 adult Kawasaki patients have been published as case presentations in 46 years. Although the ages of the patients range between 18 and 68 years, 72% are below the age of 30 (3,5). The patient we presented was an adolescent female patient.

Clinical findings are different in adults compared to children. In the study performed by Stockheim al. (3), clinical findings including fever, conjunctivitis, pharyngitis, polymorphous exanthem and peeling of the skin were observed with an equal rate in adults and children, whereas findings including coronary artery aneurism, thrombocytosis, meningitis were reported to be found more frequently in children and findings including cervical LAP, hepatitis and arthralgia were reported to be found more frequently in adults. Adult Kawasaki patients frequently have an "incomplete" course. However, there are also patients who fulfill all the criteria of Kawasaki disease and who have severe findings as our patient (5).

Although lung involvement in Kawasaki disease is a rare clinical finding, interstitial pneumonia is found in 30-90% of the patients at autopsy (6,7). Findings including nasal discharge, cough, dyspnea, tachypnea and interstitial penumonia do usually not respond to antibiotic treatment and are mostly described in infants younger than 6 months of age in contrast to our patient (6,8). In the study performed by Umezawa et al. (9), abnormal lung graphy findings were found in 14.7% of 129 patients who were diagnosed with Kawasaki disease, reticulogranular appearance was observed in 89.5%, peribronchial narrowing was observed in 21.1%, pleural fluid was observed in 15.8%, atelectasia was observed in 10.5% and air trapping was observed in 5.3%. Interstitial pneumonia which was found in our patient did not respond to antibotic treatment either.

The established treatment for Kawasaki disease is composed of 2 g/kg IVIG and 80-100 mg/kg ASA in the acute phase (1,10). In the subacute and convalescent phases, ASA is used at the antithrombotic dose (3-5 mg/kg/day). If there is no coronary abnormality on echocardiography, ASA may be discontinued 6-8 weeks after the disease onset. ASA is continued indefinitely in patients with coronary abnormality. In resistant patients, a second dose of IVIG or methylprednisolone treatment may be administered (11). In our patient, fever dropped on the second day of IVIG and ASA treatment and pneumonia findings returned completely to normal on the fifth day. In the acute phase, regression occurs in approximately 50% of the dilatations or aneurisms observed. However, regression usually does not occur in aneurisms of 8 mm or larger and these tend to cause complications (11,12). In our patient, coronary artery dilatation improved in the second week of treatment.

In Kawasaki patients, the degree of risk is determined according to the findings of echocardiography which is performed when acute phase reactants return to normal; patients with transient coronary dilatation or improved aneurism are evaluated to have a risk level of 2 as in our patient (12,13). Annual pediatric cardiology examination is appropriate. In patients who want to participate in competetive sports, combined stress test may be applied with stress echocardiography or myocardial perfusion scintigraphy. It was planned to perform myocardial perfusion scintigraphy in our patient.

Conclusively, Kawasaki disease can occur in the adulthood and different clinical pictures like lung involvement may be observed in addition to the classical findings of the disease. To prevent cardiac complications because of delay in the diagnosis and treatment it is important to perform echocardiographic evaluation at any age, when compatible clinical findings are observed together with fever.

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