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KAWASAKI Disease: A Rare Case Report with Severe Abdominal Pain

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Abstract

Kawasaki disease is an acute febrile systemic disease with unknown etiology that often affects children younger than 5 years of age. Common symptoms are fever, noninfectious conjunctivitis, inflammation of the mucous membranes and macular skin complications. Severe abdominal pain is a non-common symptom of the disease. Abdominal pain can be reduced with medical and conservative treatment. The patient is a 4-year-old female who referred with fever and severe abdominal pain. She underwent an unnecessary appendectomy due to lack of timely diagnosis. With careful attention to the uncommon symptoms of the disease and early administration of intravenous immunoglobulin and control of coronary aneurysm, the child was discharged with good condition.

Keywords: Kawasaki, Abdominal pain, Rare symptom.
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Introduction

Kawasaki disease is an inflammatory disease that causes blood vessel involvement throughout the body. It is an acute febrile illness with systemic vasculitis that often affects infants and children. The disease was first reported in Japan in 1961. The disease is sporadic or with limited epidemics that occur with an incidence of 7 per 10,000 children under the age of 5 years.[1] The average age of the onset of the disease is 5 and it is rarely seen after the age of 11. The ratio of male to female is 5 to 1.[1-2] The etiology of the disease is unclear and has posed toxical, allergic and immunologic causes. Kawasaki disease is probably the result of the general stimulation of inflammatory response with increased cytokine production, which causes endothelial destruction. Severe thrombocytosis can result in the formation of thrombosis in damaged endothelium.[2-3] The main clinical manifestations of the disease include fever, macular and mild rash, pharyngitis, and mucosal involvement, cervical adenitis and conjunctivitis.[3] Other common manifestations in the early stages of disease are pneumonitis, tinnitus, meningitis, photophobia, uveitis, diarrhea, urinary duct inflammation, arthritis and/or arthralgia.[4] Relatively uncommon clinical findings include fluid accumulation in the pleura, abdominal colic, gall bladder hydrops, jaundice, and tonsils exudate. The worst case manifestations are myocarditis and coronary vasculitis. Coronary aneurysm often occurs at the onset of a disease or appears in the early second week.[5] Risk factors for the incidence of coronary aneurysms include male sex, age below 18 months, Japanese race, and acute febrile and toxical progressive or early clinical myocarditis.[4-5] In laboratory examinations, the number of white blood cells (WBC), platelets, and erythrocyte sediment rate (ESR) increased, and occasionally there is a significant neutropenia. Other findings include increased relative liver enzymes, normocytic and hypocorum anemia, pyuria and increase in the relative number of WBC especially lymphocytes in the cerebrospinal fluid.[1-3] Electrocardiography helps in the initial assessment of ventricular dysfunction, ischemia, or arrhythmia.[4]

Two-dimensional echocardiography is the most sensitive method of showing coronary vasculitis and formation of aneurysm which should be administered upon diagnosis.[4-5] Treatment includes administration of aspirin in anti-inflammatory doses and administration of intravenous immunoglobulin. Often there is a significant response to fever, systemic symptoms, and general symptoms.[5] Administration of glucocorticoid is prohibited because it increases the incidence of coronary aneurysms.[6]

Patient report

The patient is a 4-year-old girl due to severe fever which began alternately a few days before the visit and continued from the previous night with abdominal pain, referring to Bahar Hospital of Shahroud University of Medical Sciences. She was admitted to the children's ward. The patient was the first-born in family. The delivery type was caesarian section without a history of illness or a particular problem during pregnancy and childbirth. The condition began with fever for about 4 days before admission, which was treated with Acetaminophen at home. Due to the continuation and intensification of fever and abdominal pain, she was admitted in the pediatric ward of Bahar Hospital.

Vital signs were examined as the following: BP: 90/50mmHg, PR: 115/min, RR: 20/min, Tem: 38.8 °C. Dry mucus and anorexia was reported. She complained of abdominal pain, especially around the navel. During examination, she was conscious but restless. In the head and neck, there was no problem and heart and lung auscultation was normal. In the abdominal examination, tenderness, especially around the navel and hypogastric activity without organomegaly were reported. Initial tests were requested for the patient whose results are presented in table (1). In the ultrasound examination of the entire abdominal and pelvic cavity, no specific and abnormal problem was reported. The patient was treated with ceftriaxone for a possible diagnosis of urinary tract infection due to clinical symptoms.

Due to the intensification of abdominal pain and vomiting exacerbation after 24 hours, after surgical consultation for acute abdomen, she was transferred to the surgery ward. According to a surgeon's examination and a negative toshe rectal (TR) in terms of suspicion of invagination, and lack of pathologic findings in ultrasound for invagination, the patient was diagnosed with acute abdomen. Therefore, she underwent an appendectomy surgery and 48 hours later she was discharged from the hospital with a normal condition. (It should be noted that the result of the pathological examination of the appendix mass was normal). Seven days after surgery, the patient was brought back to the hospital with severe abdominal pain and fairly severe fever.

In the new examination, the patient was restless and had a dry mouth. The vital signs were as follows: BP: 85/55 mmHg, PR: 125/min, RR: 24/min, Tem: 39.2°C. She had a weight of 16.5 kg and a height of 105 cm. In facial examination, they found bilateral conjunctivitis, dead lips, and an extremely red tongue with prominent bands. Also, three prominent lymph nodes measuring 3×4 cm in the anterior lymphatic chain of the neck were detected on both sides. No specific problem was found in the examination of the chest and abdomen, except for sensitivity and tenderness around the site of the surgery. There was also a mild hand and foot swelling, especially from the wrists to fingers. The results of paraclinical studies are shown in Table (1). In the ultrasound of the entire abdominal and pelvic cavity, mesenteric lymphadenopathy in the ascending colon region and a brief free liquid were detected in the ileocecal area. The urinary system was normal and hepatomegaly and splenomegaly were not observed but the gallbladder had a brief inflammation. At the same time, chest X-rays and electrocardiograms were reported normal. On the third day of admission, systematic macular degeneration damages throughout the body and mild flaking around the nails and the perineum were also identified (Figures 1 to 5).

Table 1. Tests performed at two times referrals to Bahar Hospital of Shahroud

Testing	Primitive	Secondary
CBC:		
Hb(g/dl)	10.7	10.1
HCT (%)	32	30
WBC	10900	12800
PMN (%)	88	75
Lymph (%)	8	21
PLT	435000	504000
PT (Second)	14	15
PTT (Second)	41	43
ESR (1 hour)	63	87
CRP	++	++++
BUN (mg/dl)	12	23
Cr (mg/dl)	0.5	0.9
Blood sugar (mg/dl)	68	59
Na (meqi/dl)	138	134
K (meqi/dl)	3.9	3.5
Alk-P (IU)	503	515
ALT (IU)	63	58
AST (IU)	59	55
Total Bilirubin (mg/dl)	2.6	2.5
Direct Bilirubin (mg/dl)	0.7	0.8
Wright	Negative	Negative
Widal	Negative	Negative
Urine Analysis	Normal	Normal
Urine Culture	Negative	Negative

Figure 1. Lips dryness and scaling

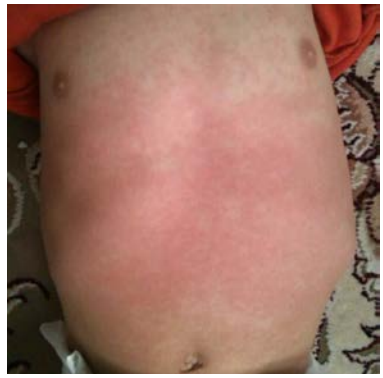

Figure 2. Macular redness of the entire skin of the abdomen and chest

Figure 3. Redness and flaking of the around the nails

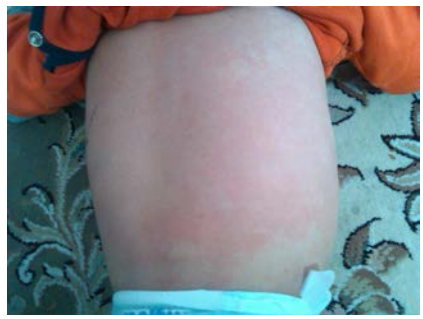

Figure 4. Redness and flaking of the extremity

Figure 5. Bilateral conjunctivitis

Due to clinical signs and laboratory findings, patient with Kawasaki disease diagnosis was treated with intravenous immunoglobulin at a dose of 2 g/Kg. After administration of intravenous immunoglobulin within 24 hours the fever subsided and systemic symptoms such as abdominal pain and flatulence were reduced. Also, aspirin administration was started at a dose of 100 mg/Kg. At the same time, echocardiography was observed, a coronary aneurysm of 3-4 mm in diameter. The patient was discharged with a good general condition, three days after the administration of immunoglobulin and an aspirin dose of 5 mg/Kg. In subsequent reviews and echocardiography 45 days after discharge, the status of coronary arteries was normal and the child had no further conditions.

Discussion

Kawasaki disease does not have a specific diagnostic test and requires a set of clinical criteria for its diagnosis which include a fever in all cases lasting for 5 days or more which can reach more than 40 degrees Celsius. The fever does not subside after antibiotic therapy and gives the anti-fever medication a relative response. The changes in the lips and oral cavity occurred in 90% of cases and includes dryness, redness and lips splits, strawberry tongue and redness of mucous membranes, non-purulent and bilateral conjunctivitis seen in 85% of patients lasting for a few weeks. Polymorphological rash occurs in 80% of patients, which is commonly seen in the trunk with a sign of vasculitis of small blood vessels and perivasculitis of the dermis and subcutaneous tissues. Rash is associated with fever in the acute phase of the illness with fever and then it gradually disappears. Itching often occurs, but vesicles and purpura are not seen. Extremity changes in 70% of patients include erythema and edema, which occurs within few days after the onset of the disease. In the recovery phase, skin scaling begins around the nails. Scaling and edema can occur from anywhere including the perinea zone, which is a valuable diagnostic symptom. Acute and non-puerperal enlargement of the neck lymph nodes is there in 70% of patients, and will improve in the last period of fever phase. For the diagnosis of Kawasaki, four of these criteria plus coronary aneurysm in echocardiography should be met.[1-5] In addition to the diagnostic criteria for the disease, Clinical and paraclinical findings including irritability and lethargy, anemia, leukocytosis, an increase in ESR and a brief increase in liver enzymes can help diagnose Kawasaki's disease. Abdominal pain in these patients is a low prevalence and seen only in 10-15% of patients where intensity of the pain is mild to moderate in all parts of the abdomen, especially around the navel and resolves within two to three days. Significant therapeutic response to intravenous immunoglobulin was also confirmed. The major complication of this patient was abdominal pain and lymphadenopathy around the mesentery. There are reports that Kawasaki disease can be associated with liver-digestive disorders. Biochemical liver tests are abnormal in 31% of cases and 15% of patients suffer from hepatomegaly. Sprained liver is self-limiting.[7] The patient also had abdominal pain, feeling of nausea and vomiting at the time of admission. Three cases of acute abdomen in Kawasaki disease have been reported due to severe inflammation of the lymph nodes in the abdomen.[8] In the patient being introduced, medical treatment was successful and did not occur in a complication requiring surgery or other intervention. Abdominal pain may be related to the shape of the gallbladder in this patient. Failure to evacuate the gallbladder after a meal can be explained for abdominal pain and vomiting, especially after eating, and it seems that there was a disorder in the patient that caused abdominal pain and digestive problems.[9-10] Interestingly, tenderness and severe abdominal pain around the navel was an initial appearance of Kawasaki disease which caused the practitioners who initially examined the patient to suggest the possibility of acute appendicitis, and placed the patient under appendectomy. However, more precise clinical symptoms and diagnosis of Kawasaki disease and timely administration of intravenous immunoglobulin, perhaps, in addition to improving the fever and abdominal pain could prevent unnecessary surgery.[11-12] This report should be accompanied by other unorganized reports, paying attention to less common symptoms and complications of the disease, such as abdominal pain along with more efficient and faster diagnosis of the disease and its complications.

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Conflict of Interest

The authors declare that they have no conflict of interest.

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