Atypical Kawasaki Disease Presenting as Intestinal Pseudo-obstruction

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Intestinal pseudo-obstruction in atypical Kawasaki disease (KD) is rare. A boy aged 2 years and 6 months presented with a 7-day history of fever, coffee-ground vomit, and abdominal pain. Abdominal radiography and ultrasound showed a dilated duodenum. Peeling of the skin on his fingers and toes developed on hospitalization day 9. Echocardiogram revealed right and left coronary artery dilatation compatible with KD. He was treated with 2 g/kg intravenous immunoglobulin (IVIG), with rapid resolution of fever and relief of abdominal pain. Follow-up abdominal radiography and ultrasound showed improvement of bowel dilatation. This case illustrates that atypical KD can present with intestinal pseudo-obstruction. A high index of suspicion is required for early diagnosis, and prompt treatment with IVIG is recommended. [J Formos Med Assoc 2006; 105(3):252–255]

Key Words: atypical, immunoglobulin, Kawasaki disease, pseudo-obstruction

Patients who do not fulfill the clinical criteria for the diagnosis of Kawasaki disease (KD) but who nevertheless go on to develop coronary abnormalities are described as having incomplete or atypical KD. There is no diagnostic laboratory test that can recognize these atypical cases. Strict adherence to the diagnostic criteria for KD might lead to failure to recognize the disease, with potential sequelae of myocardial infarction or sudden death. Coronary artery aneurysm was more frequent in atypical KD patients (20%) than in KD patients (7%). A high index of suspicion is important for early diagnosis and prompt intravenous immunoglobulin (IVIG) therapy is recommended in atypical KD.

Severe acute abdomen was reported in 4.6% of children with KD, and some even required surgical intervention. Intestinal pseudo-obstruction is an uncommon but important manifestation in 2–3% of KD cases. It may be a prominent early feature of KD, and can occur before the development of more well recognized clinical features. This may confuse the clinical picture and cause delays in diagnosis and treatment. Here, we describe an atypical KD patient presenting with intestinal pseudo-obstruction. To our knowledge, intestinal pseudo-obstruction in atypical KD has not been previously reported.

Case Report

A boy aged 2 years and 6 months presented with a 7-day history of fever, bilious vomiting, and abdominal pain. On examination, he was febrile (39.0°C) with mild dehydration. He had a tender but soft abdomen without muscle guarding. Bile with coffee-ground stain was drained from the nasogastric tube. The remainder of the physical examination was normal. Abdominal radiography and ultrasound showed a dilated duodenum (Figures 1 and 2). Upper gastrointestinal series...
KD presenting as intestinal pseudo-obstruction

dilatation (Figure 3A). Computed tomography of the abdomen showed dilated bowel loops with multiple air-fluid levels consistent with small bowel obstruction (Figure 3B). A repeat complete blood count showed mild anemia (hemoglobin, 9.3 g/dL), WBC of 15,700/mL (segment cells, 34%), and platelet count of 275,000/mL. Serum albumin was 2.0 g/dL. The patient was managed conservatively with nasogastric tube drainage, bowel rest, and intravenous antibiotics. He continued to have abdominal pain and had occasional loose stools.

On the 6th day of hospitalization, complete blood count showed hemoglobin of 9.5 g/dL, WBC of 14,000/mL (segment cells, 49%), platelet count of 608,000/mL, and CRP of 10.5 mg/L.

showed mild stasis of the small intestine. The boy was admitted and managed with intravenous fluids for presumed viral gastroenteritis. None of his family members had fever or signs of infection.

Complete blood count at presentation revealed hemoglobin of 11.1 g/dL and white blood cell count (WBC) of 13,300/mL (segment cells, 36%). C-reactive protein (CRP) was 144.7 mg/L (normal, < 5 mg/L). Platelet count was 162,000/mL. Serum electrolytes were normal. On day 3 of admission, the severity of abdominal distension worsened and was accompanied by bilious vomiting with coffee-ground consistency. Follow-up abdominal radiography showed progression of small bowel obstruction and was accompanied by bilious vomiting with coffee-ground consistency. Follow-up abdominal radiography showed progression of small bowel obstruction.

Figure 1. Abdominal radiography shows that the second and third portions of the duodenum are dilated.

Figure 2. Abdominal ultrasound shows that the second portion of the duodenum is dilated.

Figure 3. On day 3 of hospitalization: (A) abdominal radiograph shows progression of small bowel dilatation, and (B) computed tomography of the abdomen shows dilated bowel loops with multiple air-fluid levels.
Serum albumin also recovered to 3.4 g/dL after 3 days of intravenous albumin supplementation. He continued to complain of abdominal pain and was placed on total parenteral nutrition. Urine and blood cultures were negative.

On day 9 of hospitalization, peeling of the skin on his fingers and toes developed, but there was no cervical lymphadenopathy, rash, conjunctival injection, lip fissure, strawberry tongue or other abnormal physical findings. His platelet count had risen to 1,109,000/mL. In view of the prolonged fever and suspicion of KD, echocardiography was performed, which revealed right (3.7 mm) and left (3.1 mm) coronary artery dilatation consistent with KD. He was treated with 2 g/kg IVIG, which led to rapid resolution of the fever and abdominal pain. Abdominal radiography and ultrasound the next day showed that the bowel dilatation had resolved. He was discharged 3 days later in a stable condition.

The patient was doing well at follow-up 8 weeks after discharge. Serial echocardiograms demonstrated persistent right coronary artery dilatation, with some improvement over the following 6 months.

Discussion

KD is the leading cause of acquired heart disease, and is an acute systemic vasculitis of unknown etiology with a variable incidence. A recent study found that after Japan, Taiwan had the second highest incidence of KD among children < 5 years old in the world. The diagnosis of KD is established by the presence of fever of at least 5 days’ duration, and at least four of the five principal criteria without any other explanation for the illness. Patients with atypical KD are now being identified with increasing frequency, both in Japan and abroad. The incidence of atypical presentations in previous studies was 36.2% in the USA, 15% in Taiwan, and 10% in Japan. The age of patients in these studies ranged from 5 months to 11 years. In Hsieh et al’s study from Taiwan, 31% of children aged < 1 year had atypical KD, as compared with 16% of those aged 1–4 years. The onset of KD in our patient, who was 2.5 years old, was, thus, earlier than usual.

Recognizing cases that do not fully meet the KD diagnostic criteria is a great challenge. It is often a late consideration in patients with longstanding unexplained fever without classical symptoms. Fukushige et al reported that changes in the lips and oral cavity were the most common presentation of atypical KD (96%), followed by bilateral conjunctival infection. Our patient did not have either of these. Our patient developed skin peeling on his fingers and toes on day 9 of admission, a finding consistent with Fukushige et al’s observation that 60% of patients with atypical KD had changes in their peripheral extremities. We emphasize that this symptom is the important diagnostic sign of atypical KD.

Intestinal pseudo-obstruction syndrome has clinical symptomatology of a serious ileus impairment without signs of any mechanical intestinal obstruction. It is characterized by episodes resembling mechanical obstruction in the absence of organic disorder, with abdominal radiography showing prominent gaseous intestinal distension. The pathogenic mechanism of this syndrome is not known. A few KD cases complicated by intestinal pseudo-obstruction have been reported. Miyake et al reported that seven of 310 patients with KD developed pseudo-obstruction. The pathogenesis of pseudo-obstruction in KD has been proposed to be mesenteric artery vasculitis with bowel ischemia and associated dysfunction of the myenteric plexus. To the best of our knowledge, there have been no previous reports of atypical KD with intestinal pseudo-obstruction. Awareness of this atypical form should allow early diagnosis and treatment.

Treatment of patients with KD in the first 10 days of illness with a 2 g/kg dose of IVIG reduces the prevalence of coronary abnormalities from 20% to 2–4%. Although our patient was given IVIG after a prolonged fever of 16 days, the symptoms still improved dramatically. It is important for physicians to be aware of the features of atypical KD.
cal KD in order to facilitate prompt diagnosis and early treatment with IVIG. A high index of suspicion is needed in patients with thrombocytopenia, skin peeling, and prolonged fever without any focus but intestinal pseudo-obstruction.

References