Intestinal Pseudoobstruction in Kawasaki Disease

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ABSTRACT. Intestinal pseudoobstruction is an uncommon but important manifestation of Kawasaki disease. Its occurrence at the onset or during the course of the disease may confuse the clinical picture and cause delay in diagnosis and treatment. This delay may be responsible for the high rate of coronary artery abnormalities that have been reported in patients with this complication. We suggest that Kawasaki disease be considered in the differential diagnosis of any child presenting with intestinal pseudoobstruction and fever without definable cause. Pediatrics 2004;113:e504–e506. URL: http://www.pediatrics.org/cgi/content/full/113/5/e504; pseudoobstruction, Kawasaki disease.

ABBREVIATIONS. KD, Kawasaki disease; IVIG, intravenous immunoglobulin.

CASE REPORT

A 31⁄2-year-old African Canadian boy presented with a 2-day history of fever, vomiting, diarrhea, and abdominal pain. On examination he was febrile (39.1°C tympanic) and mildly dehydrated. He had a tender but soft abdomen with no guarding. The remainder of the physical examination was normal.

A complete blood count at presentation revealed a hemoglobin of 116 g/L, white cell count of 17 × 10⁹/L (normal: 5.0–12.0 × 10⁹/L) with neutrophilia, and a platelet count of 317 × 10⁹/L. Serum electrolytes were normal. An abdominal radiograph showed air-filled loops of small bowel consistent with either a small bowel obstruction or paralytic ileus. An ultrasound of the abdomen showed multiple fluid-filled loops of bowel in the right lower quadrant suggestive of enteritis with no evidence of appendicitis. The boy was admitted and managed with intravenous fluids for presumed viral gastroenteritis.

Over the next 2 days the child continued to be febrile and have watery stools. He was noted to have a transient maculopapular rash on his trunk, felt to be consistent with a viral exanthem. On day 3 of admission, he developed abdominal distension with vomiting. A repeat abdominal radiograph at this time showed increased air-fluid levels within the bowel consistent with a small bowel obstruction (Fig 1). Computed tomography of the abdomen was not suggestive of underlying appendicitis. A repeat complete blood count showed the development of mild anemia (hemoglobin: 96 g/L) with continued mild neutrophilia and elevated band count (1.53 × 10⁹/L; normal: 0.0–0.01 × 10⁹/L), with significant lymphopenia (0.47 × 10⁹/L; normal: 2.0–8.0 × 10⁹/L). Stool examination was negative for viruses on 2 occasions. The patient was managed conservatively with nasogastric tube drainage, bowel rest, and intravenous antibiotics. He continued to complain of abdominal pain and to have occasional loose stools.

On the fifth day of admission, he was noted to have swollen, red lips with some mild, nonpurulent conjunctivitis. By day 6, his diarrhea had settled; however, he continued to be febrile and have significant volumes of nasogastric tube drainage, and as such he was placed on total parenteral nutrition.

Over the next 5 days his abdominal symptoms settled, and he tolerated a gradual increase in oral intake. He continued to have intermittent axillary temperatures up to 37.9°C. Investigations revealed continued mild anemia associated with increasing leukocytosis and neutrophilia (white cell count and neutrophil counts: 25.5 and 17.69 × 10⁹/L, respect-
tively) and new-onset thrombocytosis with a platelet count of 474 $\times 10^9$/L. His serum albumin, previously normal, had fallen to 25 g/L, although his liver-function tests remained normal. An erythrocyte sedimentation rate performed around this time was 120 mm/hour. Cultures of urine and blood were negative. Intravenous antibiotics were continued.

On day 12 of admission, he developed peeling of his fingers but did not have cervical lymphadenopathy or other abnormal physical findings. His platelet count had risen to 908 $\times 10^9$/L.

In view of his prolonged fever, the observed peripheral changes, history of rash, mucous membrane changes, and conjunctivitis at various times during his admission, and markers of increasing inflammation on laboratory testing, the diagnosis of KD was entertained. An echocardiogram revealed right and left coronary artery ectasia with a small aneurysm of the proximal right coronary artery. He was treated with 2 g/kg of intravenous immunoglobulin (IVIG) and high-dose aspirin, with rapid resolution of fever. He was discharged from the hospital 3 days later.

On follow-up at 4 weeks and 6 months, the patient was well. Serial echocardiograms at these visits demonstrated persistent right coronary artery dilatation, with some improvement over time. His other coronary arteries were normal.

**DISCUSSION**

KD is one of the most common vasculitides in childhood and the most common cause of acquired heart disease in children in the developed world. Prompt recognition and early treatment with IVIG and aspirin reduces the incidence of coronary artery aneurysms, the most important long-term sequel of KD, from 25% to $\sim$3%. In the absence of a specific test, the diagnosis of KD is a clinical one based on the recognition of a characteristic set of symptoms and signs. The KD case definition of the Centers for Disease Control and Prevention requires at least 5 days of fever “without other more reasonable explanation” and at least 4 of the following: 1) bilateral conjunctival injection; 2) mucous membrane changes with injected or fissured lips, injected pharynx, or “strawberry” tongue; 3) extremity changes with erythema of the palms or soles, edema of the hands or feet, or generalized or periungual desquamation; 4) rash; and 5) cervical lymphadenopathy (with 1 node $>$1.5 cm). It is recognized, however, that not all children with KD, diagnosed on the basis of development of virtually pathognomonic coronary artery lesions, meet these criteria. The concept of “atypical” or “incomplete” KD has been invoked to describe children who do not meet traditional criteria but either are considered to be at risk of developing or have developed coronary artery lesions. It is these children and those with typical KD who meet criteria but in whom other less common manifestations dominate the clinical picture, as in the present case, in whom the challenge of prompt recognition lies. For the pediatrician, this may constitute a considerable part of their experience with the disease. Between 10% and 45% of children may present with atypical or incomplete disease, and more than half, while meeting criteria, may have a “predominant feature” such as concurrent tonsillitis or otitis media that serves to obscure the diagnosis and delay definitive therapy.

Surgical complications in KD are well recognized. Complications reported in the literature include bowel infarction, bowel obstruction related to ischemic strictures, focal colitis, and peripheral gangrene. Intestinal pseudo-obstruction has been reported to occur in up to 2% to 3% of children with KD. A clue to the development of this complication, as demonstrated in the present case, is the presence of persistent vomiting during the course of the illness, which is uncommon in KD. In contrast to those who develop mechanical bowel obstruction secondary to ischemic strictures, in which vomiting generally appears 2 to 4 weeks after the acute illness, vomiting in pseudo-obstruction tends to be seen early.

The pathogenesis of pseudo-obstruction in KD is felt to relate to mesenteric artery vasculitis with bowel ischemia and associated dysfunction of the...
myenteric plexus20,22; however, in those cases in which mesenteric vessel imaging has been performed, no abnormalities have been found,20 suggesting probable small vessel involvement.

The treatment of intestinal pseudoobstruction in KD in reported cases has varied from simple bowel rest21,22 to intravenous steroids.20 In 2 cases, IVIG. In the present case, the intestinal symptoms had resolved with conservative management by the time the child was recognized as having KD and given IVIG.

Of some concern are the reported outcomes of children with pseudoobstruction in KD. In the report by Miyake et al.,20 7 of 310 patients with KD developed pseudoobstruction, of whom 5 were found to have coronary artery disease. Similarly Zulian et al.1 found a coronary artery aneurysm rate of 50% in their report of 10 children with KD presenting with a surgical abdomen, of whom 3 were felt to have intestinal pseudoobstruction. Of the 2 cases reported by Franken et al.,22 1 child died while undergoing an exploratory laparotomy, and in both there seems to have been a subsequent rapid resolution of symptoms.1,2 In the present case, the intestinal symptoms had resolved with conservative management by the time the child was recognized as having KD and given IVIG.

KD can be associated with a number of complications that potentially may obscure the true diagnosis and delay therapy with IVIG. We suggest that KD be considered in the differential diagnosis of any child presenting with intestinal pseudoobstruction and fever without definable cause.

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