

Intestinal Pseudoobstruction in Kawasaki Disease

Jonathan D. Akikusa, MBBS*[†]; Ronald M. Laxer, MD*^{‡§}; and Jeremy N. Friedman, MB, ChB^{†¶}

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.

The individual clinical features of Kawasaki disease (KD) are protean and may appear only transiently over what is sometimes a prolonged period of illness. This may obscure their true significance, particularly when a less common manifestation suggestive of an alternate diagnosis dominates the clinical picture. Abdominal symptoms, including those of bowel obstruction, may be a prominent early feature of KD and can occur before the development of more well recognized clinical features.^{1,2} Failure to appreciate this may lead to a delay in diagnosis and treatment, with potential consequences for the development of cardiac sequelae. We present the case of a 3½-year-old boy admitted to hospital with fever and what initially was felt to be gastroenteritis. He subsequently developed paralytic ileus and was managed with a view to a possible surgical cause for his symptoms. At various times during his admission, he demonstrated the typical features of KD, which were transient and not recognized as such because of the perception that he had a primarily “abdominal” disease. Clinical suspicions were raised when he developed desquamation on the 14th day of his illness and confirmed when an echocardiogram revealed coronary artery abnormalities.

From the Divisions of *Rheumatology and ¶Pediatric Medicine, Departments of †Pediatrics and §Medicine, University of Toronto, Toronto, ON, Canada.

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Address correspondence to J.N. Friedman, MB, ChB, Hospital For Sick Children, 555 University Ave, Toronto, ON, Canada M5G 1X8. E-mail: jeremy.friedman@sickkids.ca

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CASE REPORT

A 3½-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.

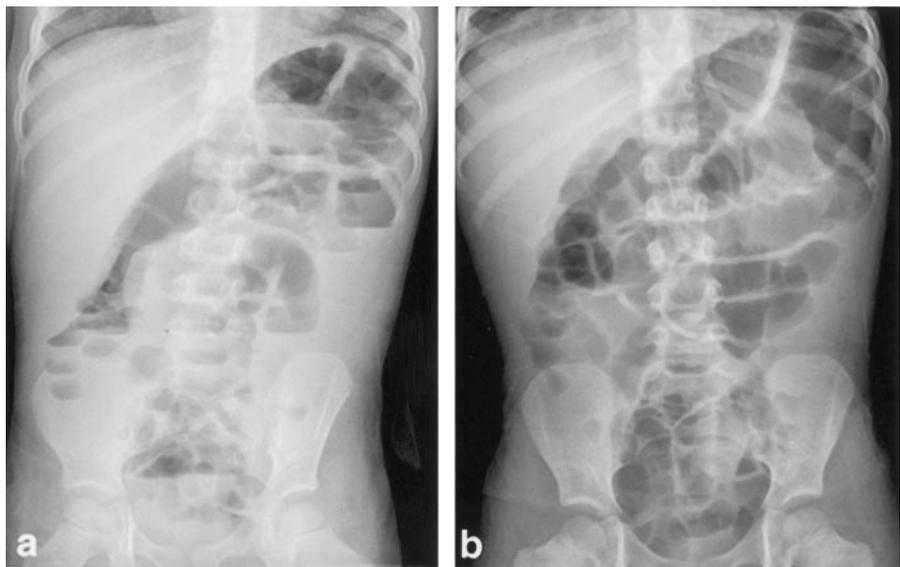
A complete blood count at presentation revealed a hemoglobin of 116 g/L, white cell count of $17 \times 10^9/L$ (normal: $5.0\text{--}12.0 \times 10^9/L$) with neutrophilia, and a platelet count of $317 \times 10^9/L$. Serum electrolytes were normal. An abdominal radiograph showed air-filled loops of 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 bilious 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 ($1.53 \times 10^9/L$; normal: $0.0\text{--}0.01 \times 10^9/L$), with significant lymphopenia ($0.47 \times 10^9/L$; normal: $2.0\text{--}8.0 \times 10^9/L$). 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 $17.69 \times 10^9/L$, respec-

Fig. 1. Erect (a) and supine (b) abdominal radiographs demonstrate small bowel obstruction with multiple air-fluid levels visible on the erect film and multiple air-filled loops of bowel on the supine film.



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 sequela of KD, from 25% to ~3%.^{3,4} 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).⁵ 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.^{6–9} 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,^{7,8,10} 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.¹¹

Surgical complications in KD are well recognized. Complications reported in the literature include bowel infarction,¹² bowel obstruction related to ischemic strictures,^{13–16} focal colitis,¹⁷ and peripheral gangrene.^{12,13,18,19} Intestinal pseudoobstruction has been reported to occur in up to 2% to 3% of children with KD.^{20,21} 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,^{13–16} vomiting in pseudoobstruction tends to be seen early.^{2,20,22}

The pathogenesis of pseudoobstruction in KD is felt to relate to mesenteric artery vasculitis with bowel ischemia and associated dysfunction of the

myenteric plexus^{20,22}; 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 rest^{21,22} to intravenous steroids.²⁰ In 2 cases, IVIG was administered at the time of bowel symptoms (1 after a 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.

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 al¹ 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, but was found to have normal coronary arteries at postmortem, and the other recovered with no treatment and was said to be "normal" 4 years later. Finally, Fang et al² report on 1 child who developed coronary artery abnormalities after presenting with predominant symptoms of fever, abdominal distension, and vomiting. It is unclear whether these outcomes reflect the delay that may occur in recognizing and treating these children or whether, as has been suggested by some,^{1,20} it is a marker of a more significant vasculitis in those who develop this complication.

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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