—Case Reports—

# A Case of Kawasaki Disease with Intussusception

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Kawasaki disease (KD) is a systemic vasculitis of unknown cause and is associated with various digestive disorders, although only a few cases of intussusception associated with KD have been reported. We describe a case of intussusception followed by KD in a 3-year-old boy. The patient was admitted to our hospital for evaluation of severe abdominal pain. Because the target sign was seen on ultrasonography, intussusception was diagnosed and hydrostatic reduction was performed. On the second day after admission, he developed a high fever (38°C) and an irregular rash over his whole body. On the fourth day after admission, the high fever continued, and bilateral nonexudative conjunctivitis, erythema of the lips and oral mucosa, strawberry tongue, indurated edema of the dorsa of the hands and feet, and diffuse erythema of the palms and soles appeared, and KD was ultimately diagnosed. He was treated with intravenous immunoglobulin 2 g/kg, aspirin 30 mg/kg/day, and prednisolone 2 mg/kg/day. The high fever and other clinical symptoms resolved immediately after the start of treatment. There was no relapse of KD symptoms after initial treatment, and periungual desquamation was observed on the 10th day after admission. He was discharged on the 15th day, without abnormalities such as coronary dilatation, 3 months after the onset of KD symptoms. Patients with intussusception and KD were older (≥3 years vs <3 years) than those with intussusception alone. In addition, the site of intussusception in KD was mainly colonic rather than ileocolic. If intussusception precedes development of the characteristic clinical symptoms of KD, diagnosis of KD may be delayed. KD should be considered in children older than 3 years with intussusception at a colonic site. (J Nippon Med Sch 2020; 87: 346–349)

**Key words:** intussusception, Kawasaki disease

## Introduction

Kawasaki disease (KD) is a systemic vasculitis of unknown cause. Severe inflammation of coronary arteries results in coronary sequelae such as coronary artery aneurysms and stenosis in about 3% of patients with KD[1,2]. There have been several reports of gastrointestinal manifestations associated with KD, including vomiting, diarrhea, abdominal pain, and abdominal distension[3–5]. However, only a few cases of intussusception associated with KD have been reported[6–14]. We report a case of KD preceded by intussusception and discuss the relevant literature.

## Case

A 3-year-old boy was admitted to our hospital with severe, crampy, progressive abdominal pain with vomiting. Severe abdominal pain continued for 6 hours. On admission, his temperature was 36.9°C, and he gradually became lethargic. A sausage-shaped mass was noted during palpation of the lower right abdomen, and peristalsis was increased. Capillary refill time was <2 seconds. Right cervical lymph node swelling (diameter: 1 cm) with mild tenderness was noted. The patient had no conjunctival congestion, no abnormal findings of the lip or tongue, and no terminal changes of the limbs, such as palmar erythema.

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Table 1 Laboratory findings on admission

| | | | | | | | |
|---|---|---|---|---|---|---|---|
| Blood count | | Biochemistry | | | | Urine | |
| WBC | 25,500 /μL | T-Prot | 7.1 g/dL | ANA | <40 X | protein | (1+) |
| Stab | 3.0 % | Alb | 3.9 g/dL | RF | <3 X | ketone | (2+) |
| Seg | 78.0 % | BUN | 9.9 mg/dL | ASO | 11 IU/mL | occult blood | (-) |
| Lym | 4.0 % | Crnn | 0.20 mg/dL | ASK | 40 X | WBC | (2+) |
| Mono | 14.0 % | Na | 132 mEq/L | Ferritin | 210.9 ng/mL | bacteria | (-) |
| RBC | 442 $\times 10^4$/μL | K | 4.3 mEq/L | IgG | 877 mg/dL | | |
| Hb | 12.3 g/dL | Cl | 101 mEq/L | CH50 | 54 U/mL | | |
| Ht | 35.4 % | Ca | 11.9 mEq/L | C4 | 28.8 mg/dL | | |
| PLT | 48.6 $\times 10^4$/μL | T-Bil | 1.2 mg/dL | C3 | 122 mg/dL | | |
| | | AST | 215 U/L | IL-2R | 2,133 U/mL | | |
| Coagulation | | ALT | 96 U/L | | | | |
| PT | 10.4 sec | LDH | 490 U/L | Adenovirus (CF) | <4 X | | |
| APTT | 23.4 sec | Amy | 123 U/L | CMV-IgM | <10 X | | |
| Fib | 649 mg/dL | BS | 108 mg/dL | CMV-IgG | <10 X | | |
| FDP | <5.0 mg/mL | CRP | 12.38 mg/dL | VCA-IgM | <10 X | | |
| | | | | VCA-IgG | <10 X | | |
| Blood Gas | | ESR | | EBNA | <10 X | | |
| pH | 7.524 | 60 min | 89 mm/h | | | | |
| pCO2 | 27.0 mmHg | | | Pharyngeal culture | negative | | |
| pO2 | 39.4 mmHg | BNP | 299.54 pg/mL | Blood culture | negative | | |
| HCO3 | 22.2 mmol/L | | | Stool culture | negative | | |
| ABE | 0.7 mmol/L | | | | | | |
| Lac | 2.1 mmol/L | | | | | | |

Laboratory examination on admission showed leukocytes 25,500/μL (neutrophils 82.0%), platelet count 486,000/μL, C-reactive protein (CRP) 12.38 mg/dL, serum sodium 132 mEq/L, aspartate aminotransferase (AST) 215 U/L, alanine aminotransferase (ALT) 96 U/L, lactate dehydrogenase (LDH) 490 U/L, albumin 3.9 g/dL, and brain natriuretic peptide (BNP) 299.5 pg/mL. Aseptic leukocytes were noted in urine. The results of tests for Epstein-Barr virus (EBV) and cytomegalovirus (CMV) antibodies, antinuclear antibodies, *Yersinia* bacteria, and rheumatoid factor were negative (**Table 1**).

After admission, the target sign was identified in the ascending colon on abdominal ultrasonography, and blood in the stool and intussusception was diagnosed. Nonsurgical reduction using hydrostatic pressure from an amidotrizoic acid enema was performed. Although no findings indicated a filling defect within the bowel lumen, severe stagnation of contrast agent, with a mild coiled spring sign and meniscus sign, was observed in the transverse and ascending colons. Successful reduction was confirmed by free flow of contrast into the small bowel. After reduction, there was no complaint of abdominal pain, and ultrasonography showed no evidence of relapse.

On the second day after admission, he developed a high fever (38°C) and a rash all over his body. The results of an adenovirus rapid test and streptococcal rapid test were negative. The elevated CRP concentration (18.6 mg/dL) and leukocyte count, high fever, pharyngeal redness, and cervical lymph node swelling indicated a diagnosis of purulent lymphadenitis, which was treated with flomoxef. On the third day, abdominal contrast-enhanced CT was performed to identify intra-abdominal tumors, but no abnormality was found. The high fever continued, and bilateral nonexudative conjunctivitis, erythema of the lips and oral mucosa, strawberry tongue, indurated edema of the dorsa of the hands and feet, and diffuse erythema of the palms and soles were noted on the fourth day after admission. KD was then diagnosed, and he was treated with immunoglobulin 2 g/kg, aspirin 30 mg/kg/day, and prednisone 2 mg/kg/day. After the start of treatment, the high fever resolved immediately, without relapse of KD symptoms. On the 10th day after admission, periungual desquamation was observed, and he was discharged on the 15th day after admission. One and 3 months after the onset of KD, cardiac ultrasonography was performed, and no abnormalities such as coronary dilatation were found.

## Discussion

Although gastrointestinal disorders (diarrhea, vomiting, or abdominal pain; 61%) are common complications in

Table 2 Cases of Kawasaki disease with intussusception

| Age | Sex | Interval from initial symptom (days) to referral | Interval from initial symptom (days) to diagnosis of intussusception | Interval from initial symptom (days) to diagnosis of Kawasaki disease | Site of intussusception | Treatment of intussusception | Treatment of Kawasaki disease | Coronary artery lesions | Reference number |
|---|---|---|---|---|---|---|---|---|---|
| 4 y | F | NA | 2 | 4 | NA | pneumatic reduction | IVIG, ASA, PSL | NA | 6 |
| 3 y | M | 0 | 0 | 6 | colon-colon | pneumatic reduction | IVIG, ASA | (-) | 7 |
| 3 mo | M | 2 | 3 | 8 | colon-colon | surgery | IVIG, ASA | RCA, LMT | 8 |
| 3 y | M | 0 | 0 | 5 | colon-colon, ileum-colon | hydrostatic reduction | IVIG, ASA | (-) | our case |
| 6 y | F | 5 | 9 | 7 | colon-colon | hydrostatic reduction | IVIG, ASA | (-) | 9 |
| 4 y | M | NA | NA | NA | NA | pneumatic reduction | IVIG | NA | 10 |
| 3 y | F | 5 | 8 | 5 | ileum-colon | hydrostatic reduction | ASA, PSL | (-) | 11 |
| 6 mo | M | 2 | 6 | 3 | colon-colon | hydrostatic reduction and surgery | NA | (-) | 12 |
| 5 y | M | 7 | 7 | 7 | ileum-colon | NA | IVIG, ASA | (-) | 13 |
| 4 mo | F | NA | NA | NA | colon-colon | hydrostatic reduction | NA | NA | 14 |

Abbreviations: ASA: aspirin, IVIG: intravenous immunoglobulin, LMT: left main trunk, NA: not applicable, PSL: prednisolone, RCA: right coronary artery

KD[2,3], only a few cases of intussusception have been reported[4–12]. Kato reported that a resected specimen of small intestine from a patient with intussusception and KD showed no aneurysm, no stenosis of mesenteric arteries, and no swelling of mesenteric lymph nodes[12]. Because KD is characterized by generalized vasculitis, we hypothesize that the pathogenesis of intussusception in Kawasaki disease is related to mesenteric small-vessel vasculitis with bowel ischemia and associated myenteric plexus dysfunction[11,13].

Diagnosis and treatment of KD might be delayed if symptoms triggered by organic complications appear before the characteristic clinical features of KD. Therefore, we tried to identify risk factors for intussusception in KD. **Table 2** summarizes all reported cases of intussusception with KD, including our case[15,16]. These reports[15,16] include many patients (n=877[15], n=219[16]) with intussusception and show the actual number of patients. Age at onset (≥3 vs <3 years) was older for patients with both intussusception and KD than for those with KD alone[15]. In addition, the site of intussusception was mainly colonic in children with KD but ileocolic in others (97%)[16].

In conclusion, if intussusception precedes the characteristic clinical symptoms of KD, diagnosis of KD may be delayed. The underlying causes of intussusception in older children (≥3 years) with colonic intussusception should be identified. When KD symptoms are present in children with intussusception, a diagnosis of KD should be considered in those who present with fever.

**Conflict of Interest:** None.

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