A case of hemolytic uremic syndrome preceded by intussusception

Eun Young Ko, M.D.1, Joo Young Kim, M.D.1, Hye Jin Lee, M.D.1, Hyun Seung Lee, M.D.1, Ji Whan Han, M.D.1, Young Hoon Kim, M.D.1, Jin Tack Kim, M.D.1, Hae Il Cheong, M.D.2, and Pil Sang Jang, M.D.1

Department of Pediatrics1, College of Medicine, The Catholic University of Korea, Seoul, Department of Pediatrics2, Seoul National University Children’s Hospital, Seoul, Korea

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Corresponding author: Pil Sang Jang, M.D. Department of Pediatrics, Uijeongbu St. Mary’s Hospital, 64-1, Gumo-dong, Uijeongbu-si, 480-717, Korea Tel: +82.31-820-3572, Fax: +82.31-821-3108 E-mail: drmagic@catholic.ac.kr

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Introduction

The onset of hemolytic uremic syndrome (HUS) is usually preceded by gastroenteritis, characterized by fever, vomiting, abdominal pain, and diarrhea that is initially watery but then becomes bloody. These prodromal symptoms may mimic acute abdomen, appendicitis, inflammatory bowel disease, and intussusception. Intussusception may be a gastrointestinal (GI) complication during the course of HUS1-2. Although intussusception is one of the differential diagnoses of HUS, HUS may present with intussusception3-7. Here, we report a case of HUS preceded by intussusception in a 17-month-old boy.

Case report

A 17-month-old, previously healthy boy presented at the emergency department with a 2-day history of blood-tinged stools and a 1-day history of worsening irritability. On abdominal examination, no mass was palpable. Intestinal sonogram revealed findings compatible with intussusception. An air enema reduced intussusception successfully without complications. He was observed for 6 hours in the emergency department and discharged without symptoms. The next day, he showed bloody diarrhea and revisited the emergency department with more than 20 episodes of mucoid hematochezia, abdominal pain and low-grade fever. Intestinal sonogram did not identify intussusception and laboratory tests were normal including prothrombin time and partial thromboplastin time. He was admitted to the general ward.
for further treatment. On hospital day (HD) #2, bloody diarrhea and irritability persisted but his vital signs were normal. Laboratory results showed hemoglobin (Hb) 10.6 g/dL, platelets (PLT) 61,000/μL, blood urea nitrogen (BUN) 33.1 mg/dL, serum creatinine (Cr) 1.26 mg/dL, and lactate dehydrogenase (LDH) 3,425 IU/L. On HD #3, he showed edematous eyelids, mild pretibial pitting edema, and slightly decreased urine volume. Laboratory tests showed Hb 9.0 g/dL, PLT 41,000/μL, BUN 40.6 mg/dL, Cr 1.71 mg/dL, and LDH 3,678 IU/L. A peripheral blood smear showed findings compatible with microangiopathic hemolytic anemia. He was diagnosed as HUS but Escherichia coli O157:H7 was not isolated in stool culture. Diet with microangiopathic hemolytic anemia. He was diagnosed as HUS and thrombotic thrombocytopenic purpura are the classical diseases associated with thrombotic microangiopathy (TMA)18. Gianantonio et al.19 reported extrarenal pathology in autopsy cases of the acute stage of HUS. The colonic wall was the most severely affected and, microscopically, hemorrhage and necrosis with mucosal ulceration were significant in the cecum and the ascending colon18. These findings may account for a potential leading point for an ileocolic or colocolic type of intussusception in HUS.

In summary, persistent bloody stools after reduction of intussusception may suggest HUS. Exclusion of recurrence of intussusception, close monitoring of urination and laboratory tests including disseminated intravascular coagulation profile are warranted for early detection of HUS in younger children. To our knowledge, this is the first case report of HUS preceded by intussusception in Korea.

**Discussion**

HUS is the most common cause of acute renal failure in previously healthy infants and young children, and is a substantial cause of acute mortality and morbidity in these patients2. The incidence in children younger than 3 years is higher than that in older children and adolescents8-10. Although HUS is usually a self-limiting disease with spontaneous recovery, a high index of suspicion and early recognition are essential for better outcomes, especially in children aged <3 years. Infection with Shiga/Shiga-like toxin-producing *Escherichia coli* (STEC) O157:H7 precedes >80% of HUS cases in developed countries but other strains, which are more difficult to detect, have also been implicated211. Unlike most O157:H7 isolates, the majority of non-O157:H7 STEC strains cannot be isolated using media such as sorbitol MacConkey agar212. In Korea, O157 is not the most commonly isolated serotype of STEC in patients with HUS and reports of HUS by non-O157 STEC strains are increasing steadily12-15. In cases of HUS with negative cultures, detection of anti O-type specific lipopolysaccharides Ig M antibodies is useful for diagnosing possible causative EHEC16. Although the presentation of our patient was unusual with no related food history and negative stool cultures for HUS, early recognition of HUS and meticulous maintenance of fluid balance seemed to prevent acute exacerbation and dialysis.

Hemorrhagic colitis is the most common and widely recognized GI manifestation of HUS7. Although intussusception is one of the differential diagnoses of hemorrhagic colitis and HUS, cases of HUS preceded by intussusception are rare. Our patient showed a 2-day history of GI symptoms prior to diagnosis of intussusception and others reported mostly a 1-week history of GI and respiratory symptoms. Air enema was successful in this case but all the reported cases underwent laparotomy to resolve intussusception. HUS was diagnosed 4 days after reduction of intussusception in this patient while others needed 1-6 days for final diagnosis19-7. In Korea, there have been no previous reports with a similar history, although one case of HUS preceded by ischemic colitis has been reported7.

In our case, the etiology of HUS preceded by intussusception was not identified. HUS and thrombotic thrombocytopenic purpura are the classical diseases associated with thrombotic microangiopathy (TMA)18. Gianantonio et al.19 reported extrarenal pathology in autopsy cases of the acute stage of HUS. The colonic wall was the most severely affected and, microscopically, hemorrhage and necrosis with mucosal ulceration were significant in the cecum and the ascending colon18. These findings may account for a potential leading point for an ileocolic or colocolic type of intussusception in HUS.

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