Acute Colitis Resembling Ulcerative Colitis in the Hemolytic–Uremic Syndrome

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The clinical picture of ulcerative colitis is mimicked by numerous conditions; therefore, an accurate diagnosis is often dependent on the exclusion of other causes of acute colonic disease. Recently, we saw a 10-year-old boy who had proctoscopic and radiographic findings resembling ulcerative colitis. Further evaluation demonstrated that he had hemolytic–uremic syndrome. For the pediatric age group, hemolytic–uremic syndrome should be included in the differential diagnosis of acute ulcerative lesions of the colon.

CASE REPORT

A 10-year-old black male presented to the pediatric clinic of Letterman Army Medical Center with a 4-day history of periumbilical abdominal pain, intermittent vomiting, and diarrhea. Two days before admission, small amounts of blood appeared with each evacuation. At the time of admission, the patient was having 20 stools per day. He had no history of previous gastrointestinal disease, growth retardation, food intolerance, or other major illnesses. The family history was negative for gastrointestinal disease.

Physical examination revealed a well-nourished child in no acute distress. He was alert and cooperative, with a pulse of 140 beats/min; blood pressure of 104/72 mm Hg; temperature, 37.1° C (98.8° F); weight, 28.6 kg (63 lb); height, 134.6 cm (53 in.); (height and weight 50th percentile for age). His abdomen was soft with mild periumbilical tenderness to deep palpation. The liver and spleen were not palpable. Bowel sounds were normal. Rectal examination was normal except for the presence of 4 plus guaiac-positive stool. Proctoscopic examination to 8 cm (3rd hospital day) revealed marked edema, erythema, and friability with a strongly positive swab test.

Laboratory data disclosed an hematocrit of 45%; white blood cell (WBC) count, 20,800/mm^3; 62% neutrophils; 12% lymphocytes; 8% monocytes; 18% bands. Platelet estimate was normal on admission; sodium, 130 mEq/liter; potassium, 3.8 mEq/liter; blood urea nitrogen (BUN), 30 mg/100 ml; and urinalysis, 2 plus protein with a normal sediment. Hemoglobin electrophoresis revealed sickle trait. Stool cultures grew no pathogens; urine and blood cultures were sterile. Stools for ova and parasites were repeatedly negative. Barium enema examination (Figure 1) showed multiple tiny mucosal ulcerations throughout the colon with "thumb printing" in the cecum and ascending colon. An intravenous pyelogram (IVP), chest roentgenogram, and abdominal plain films were normal.

The patient was initially treated with intravenous fluids and bed rest. Following barium enema and proctoscopic examinations, corticosteroids and ampicillin were added to the regimen. His stool frequency decreased to 3 per day but during a 48-hr period his hematocrit dropped to 22%. At this time, the BUN was 35 mg/100 ml, and the platelet count dropped to 15,000/mm^3. The peripheral smear revealed numerous red cell fragments and burr cells. The lactic dehydrogenase (LDH) was 1100 mU/ml; serum glutamic oxaloacetic transaminase (SGOT) was 80 mU/ml; bilirubin, 1.8 mg/100 ml; fibrinogen, 255 mg/100 ml, prothrombin time was 13 sec (control 12 sec); partial thromboplastic time, 23 sec (control 34 sec); reticulocyte count was 9%; Coombs' direct and indirect tests were negative. Fibrin split products were present by the protamine method. The hemolytic–uremic syndrome was diagnosed. His BUN rose to a high of 84 mg/100 ml. A repeat urinalysis revealed 4 plus protein, 4–6 red blood cells and 3–5 white blood cells per high power field. Red cell casts were also seen. The initial high dose of steroids (10 mg methylprednisolone every 8 hr) was tapered over 10 days. Within the following week, his bowel function and all laboratory values returned to normal. Repeat proctoscopic examination still revealed moderate edema and some erythema, but a negative sigmoidoscopic swab test. Two months after discharge the sigmoidoscopic examination was normal with a normal rectal biopsy.

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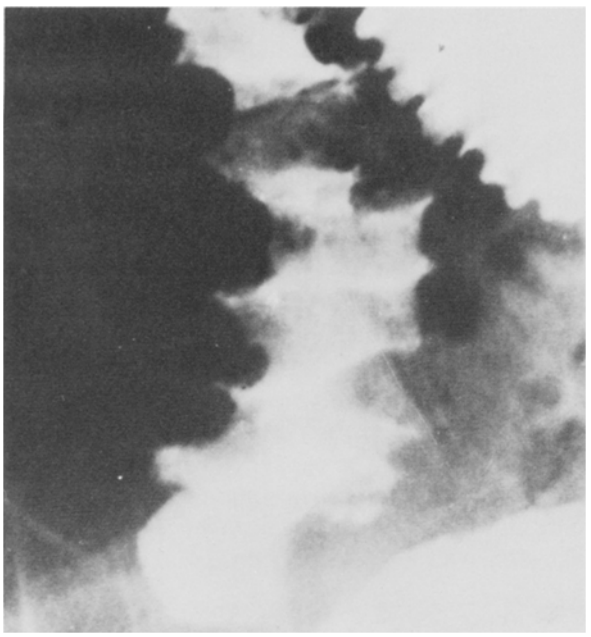

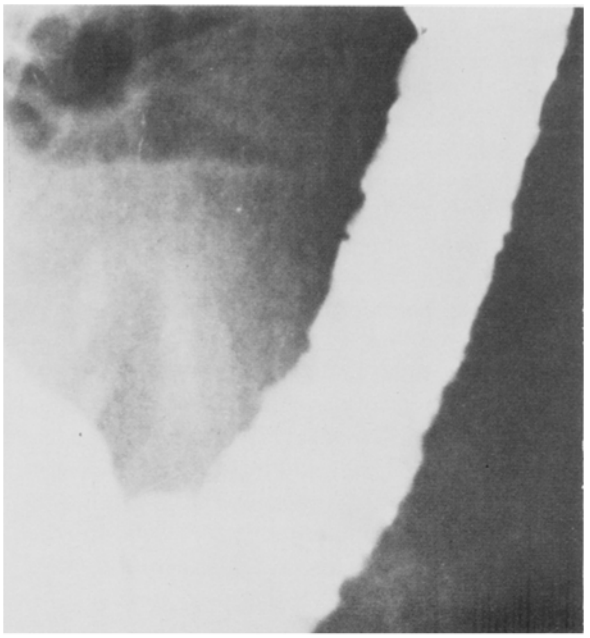

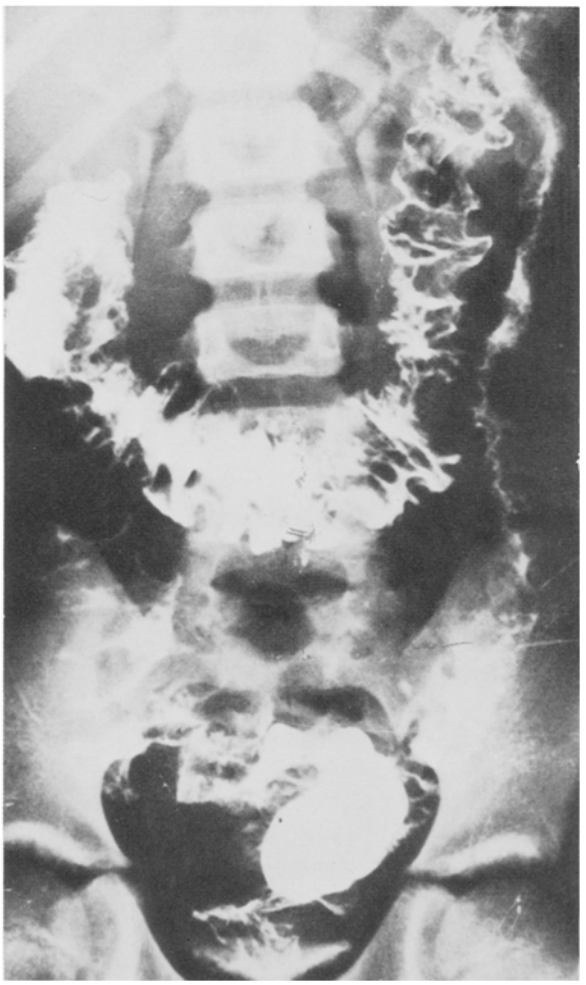

Fig 1. (A, above left) Cecum showing "thumb printing" indicative of submucosal edema. (B, above right) Ascending colon showing fine mucosa ulcerations. (C, below right) Post evacuation film showing majority of edema to be on right side.

DISCUSSION

The hemolytic–uremic syndrome has become a well-defined clinical entity since it was first described in 1955 (1). For unknown reasons, a state of diffuse intravascular coagulation develops in affected children; microangiopathic hemolytic anemia, thrombocytopenia, and renal failure result. Other organ systems may also be involved. Neurological disturbances are common and range from minor disturbances in mentation to seizures or coma.

Gastrointestinal symptoms appear to be common, particularly in the prodromal period. Liberman (2) reported that 34 of 39 patients had gastroenteritis of varying severity from 3 weeks to several days before admission. Usually the diarrhea is only slight to moderate and may or may not be associated with blood streaking of the stool. More severe gastrointestinal bleeding manifested by hematemesis and melena has also been described (3). Berman (4) reported five cases of hemolytic–uremic syndrome which had initially been diagnosed as acute ulcerative colitis on the basis of presenting clinical symptoms and proctoscopic findings. Berman's paper (4) is the only publication to our knowledge that emphasizes the confusion between hemolytic–uremic syndrome and ulcerative colitis. The

proctoscopic findings in that report coincide with those in our case.

Gianantonio et al (3) in Argentina have considerable experience with the hemolytic–uremic syndrome which appears to be endemic in that country. They emphasize on the basis of autopsy material that, except for the kidney, the colon shows the greatest involvement. Extensive areas of hemorrhage and necrosis with mucosal ulceration are seen in the colon; the ulceration is most severe in the cecum and ascending colon.

Because a number of patients with hemolytic–uremic syndrome present with diarrhea associated with gross blood, inaccurate diagnosis can easily lead to improper therapeutic measures. Vigorous hydration leading to fluid overload is often a problem before the accompanying renal failure is recognized. The use of corticosteroids, as in our case, is questionable. In fact, if the pathogenesis of the hemolytic–uremic syndrome is related to diffuse intravascular coagulation, corticosteroids might be contraindicated since they are known to potentiate the generalized Shwartzman phenomenon in animals. Steroids are not part of the standard regimen for hemolytic–uremic syndrome at present. If, as we suspect, the type of colonic disease we are describing is common in the hemolytic–uremic syndrome, it suggests that the colonic disease rarely leads to perforation and needs only supportive therapy.

If there is significant blood in the stool, the diagnosis of intussusception will occasionally be considered early in the course of the hemolytic–uremic syndrome. This diagnosis had been considered in the present case and led to the performance of a barium enema before consultation with the gastroenterologists and before the proctoscopic examination. Despite the impressive radiographs we are able to publish, it is our feeling that barium enema is rarely indicated and usually contraindicated in acute colonic disease, particularly if ulcerative colitis is a consideration. A proctoscopic examination, which infants and children tolerate well, will provide the necessary information for treatment in the acute stage.

The treatment for hemolytic–uremic syndrome is symptomatic and primarily dependent on the extent of renal disease. Peritoneal dialysis appears to have lowered the mortality rate to 7% of cases (5). The use of heparin, which should be useful from the theoretical viewpoint, remains a controversial issue (5).

When presented with a case of what appears to be acute ulcerative colitis, hemolytic–uremic syndrome should be considered in the differential diagnosis, particularly in children under 12 years of age. Examination of the peripheral smear revealing typical findings of microangiopathic hemolytic anemia, thrombocytopenia, and a rising BUN will lead to the diagnosis and early appropriate therapy. However, serial evaluations may be necessary because the colonic manifestations may precede the more typical features of the hemolytic–uremic syndrome.

The ultimate prognosis for the bowel disease associated with the hemolytic–uremic syndrome appears to be complete recovery; there are no reports of long-term colonic sequelae. Although most of the surviving children recover completely from the renal complications, some will have persistent renal disease as well as hypertension.

SUMMARY

The case report of a 10-year-old boy, admitted to the hospital after he had experienced 4 days of periumbilical abdominal pain, intermittent vomiting, and diarrhea, is presented. He had proctoscopic and radiologic findings resembling ulcerative colitis. However, further analysis of laboratory data suggested hemolytic–uremic syndrome. Since the patient in the pediatric age group presents with a clinical picture mimicking ulcerative colitis, this hemolytic–uremic syndrome should be included in the differential diagnosis. Examination of a peripheral smear revealing typical findings of microangiopathic, hemolytic anemia, thrombocytopenia, and a rising blood urea nitrogen value will lead to the diagnosis of hemolytic–uremic syndrome and early appropriate therapy.

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