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CASE REPORT | INFLAMMATORY BOWEL DISEASE

Obstructing Sigmoid Volvulus: An Unusual Complication in a Pediatric Patient With Ulcerative Colitis

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ABSTRACT

Acute colonic dilation in pediatric patients with ulcerative colitis (UC) raises a concern for toxic megacolon, but other rare conditions such as sigmoid volvulus may present in a similar manner. We report a rare case of a teenager with UC without prior surgery who developed an obstructing sigmoid volvulus managed with endoscopic detorsion and decompression. Colonic inflammation in patients with UC may result in a volvulus in the absence of other predisposing factors and should be considered in the differential diagnosis of patients with UC who present with obstructive symptoms with an atypical presentation.

KEYWORDS: pediatric ulcerative colitis; sigmoid volvulus; colonic dilation; pediatric IBD; volvulus; colonic distension

INTRODUCTION

Causes of colonic obstruction in children with inflammatory bowel disease (IBD) include toxic megacolon in UC, which occurs less frequently compared with adults; stricture(s) in Crohn's disease; and adhesions in patients with prior abdominal surgery.[1,2] In the setting of IBD, other causes of bowel obstruction such as volvulus may be overlooked. Colonic volvulus most commonly occurs in older individuals with chronic constipation, prior abdominal surgeries, or bowel dysmotility and may occur less frequently in children with similar comorbidities. UC is not believed to be a risk factor of colonic volvulus. We present an unusual case of an adolescent with UC without prior surgery who developed an obstructing sigmoid volvulus managed with endoscopic detorsion and decompression.

CASE REPORT

A 16-year-old adolescent girl with UC managed with infliximab infusions (6 mg/kg every 8 weeks) presented with abdominal pain, hematochezia, and diarrhea for 2 weeks. She had mildly elevated fecal calprotectin—210 mg/kg (normal: $<$ 50 mg/kg), positive *Clostridium difficile* toxin polymerase chain reaction with negative enzyme immunoassay, and otherwise unremarkable laboratory test results. She failed to respond to outpatient therapies for 7 days including oral vancomycin for suspected *C. difficile* infection and oral corticosteroids for a suspected IBD flare. Magnetic resonance enterography (MRE) was performed as an outpatient to evaluate the extent of disease given the failure of outpatient therapy. MRE demonstrated a normal terminal ileum and a dilated proximal colon raising concern for toxic megacolon vs distal colonic stricture, leading to hospital admission after study for further management (Figure 1). However, clinically the patient did not have any symptoms of bowel obstruction or abdominal distension at the time of MRE.

At admission, the patient stopped passing stools; however, she was passing flatus without abdominal distension, pain, or vomiting. Kidney ureter bladder x-ray (KUB) showed a dilated large bowel in the left upper quadrant with a maximum diameter of 8.3 cm. The surgery service was consulted to rule out toxic megacolon and deferred acute surgical intervention. She was managed conservatively with nil-per-oral, nasogastric tube decompression and underwent serial x-rays and abdominal examinations. In addition, she received intravenous corticosteroids to empirically manage an IBD flare. On day 3 of admission, she developed obstipation with a KUB showing the transverse colon with a maximum diameter of 10.2 cm (Figure 2).

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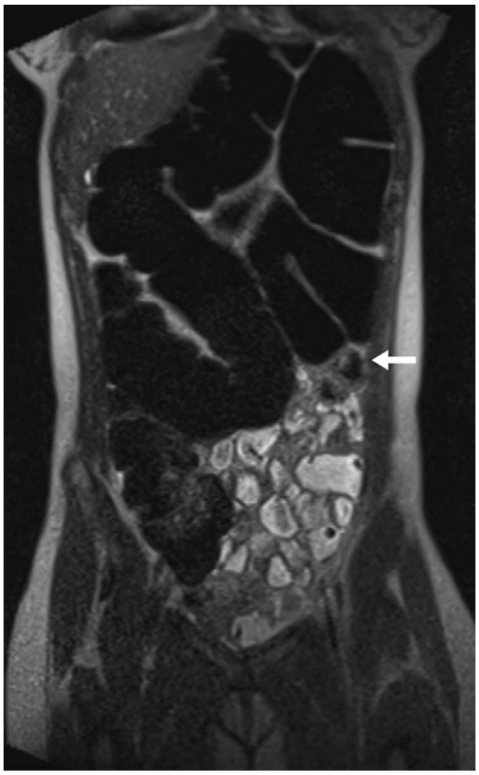

Figure 1. MRE in coronal view showing rectosigmoid inflammation (arrow) with markedly dilated proximal colon concerning for a distal stricture or toxic megacolon.

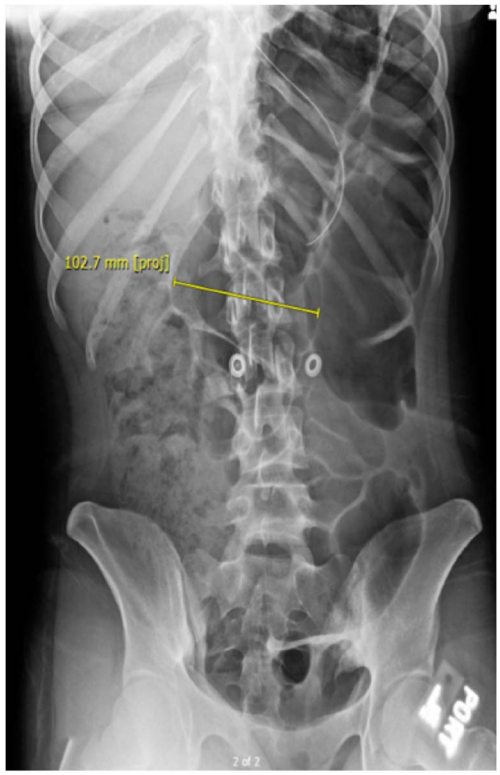

Figure 2. Kidney ureter bladder x-ray (KUB) on day 3 of nil-per-oral (NPO) showing a maximum transverse colon diameter of 10.2 cm in the transverse colon.

She subsequently underwent a colonoscopy to evaluate her disease activity and possibly the etiology of her obstruction. This demonstrated torsion and near-complete obstruction of the sigmoid colon with a pale mucosal appearance consistent with a volvulus (Figures 3 and 4). The obstructed area was able to be traversed and detorsed using a neonatal upper endoscope (outer diameter 6 mm) with subsequent spontaneous passage of flatus consistent with reduction of the volvulus. The mucosa proximal to the obstructed area showed marked pallor with decreased vascularity extending up to the splenic flexure with a normal-appearing transverse colon. There was no evidence of stenosis with withdrawal of the endoscope. Repeat stool *C. difficile* toxin polymerase chain reaction was negative. Before discharge the following day, the patient was readily able to pass stools and tolerated a regular diet. Follow-up colonoscopy 1 month later showed mild-to-moderate left-sided colitis from the rectum to the splenic flexure (Figures 5 and 6). Her infliximab dose was increased to 10 mg/kg every 6 weeks with improved therapeutic drug levels. Azathioprine was added to optimize her regimen. She is under clinical remission without recurrence of volvulus at 12-month follow-up, suggesting that inflammation due to UC likely caused her volvulus.

DISCUSSION

Only 100 cases of volvulus have been reported in children to date with a mean age of presentation of approximately 7 years.[3–6] The sigmoid colon is most commonly involved, followed by the right colon.[7] Predisposing factors include underlying neuromuscular disorders (cerebral palsy, myopathy, and Prader-Willi syndrome), late-onset Hirschsprung disease, and infections such as roundworm infestation or Chagas disease in tropical areas.[4,6–8] The absence of these risk factors in our patient suggests that

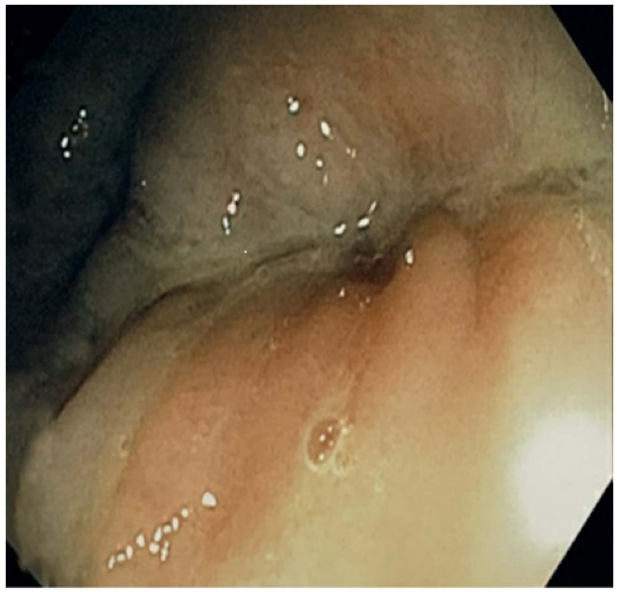

Figure 3. Colonoscopy images of the sigmoid colon (30 cm from anal verge) just distal to the volvulus with the characteristic whirl pattern and scattered exudates. Note the pale appearance of the mucosa.

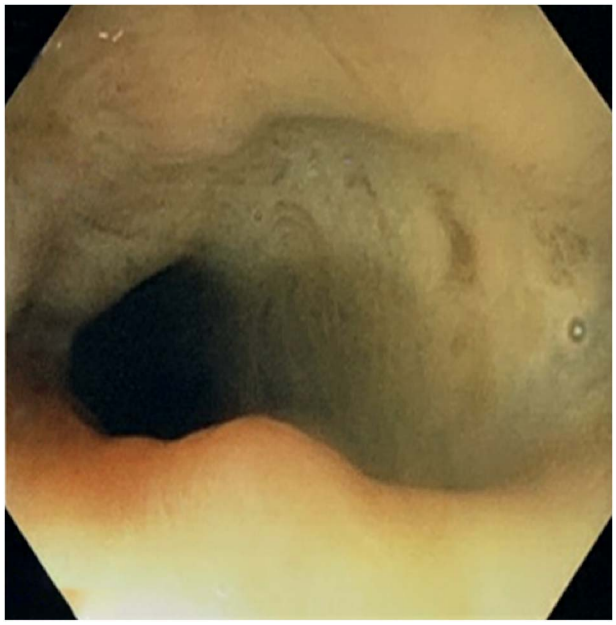

Figure 4. Mid-descending colon, proximal to the area of the sigmoid volvulus, with very pale mucosa and decreased vascularity. This endoscopic appearance extended to the level of splenic flexure.

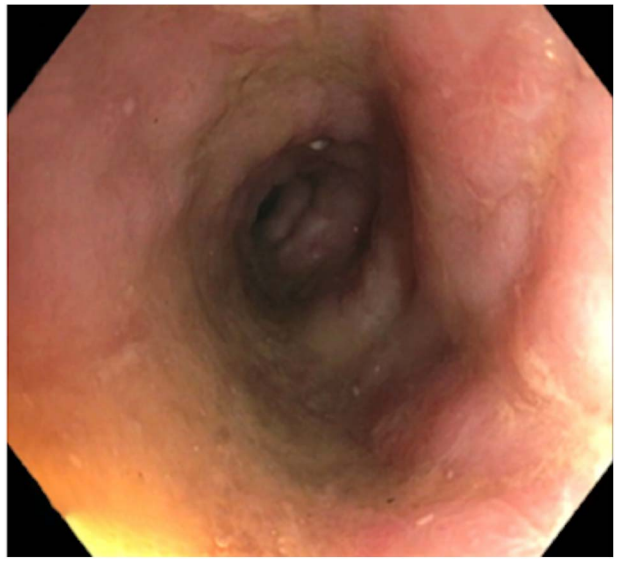

Figure 5. Follow-up colonoscopy 1 month later demonstrated left-sided colitis characterized by decreased vascularity, congestion, erythema, friability, and scattered aphthous ulcerations extending from the anus to the splenic flexure with normal mucosal appearance proximally; this figure demonstrates mucosal changes in the sigmoid colon.

inflammation due to UC and potentially *C. difficile* infection was likely the precipitating cause of her volvulus.

Colonic volvulus is rare in IBD, with only 2 cases reported to date in adults with UC involving the sigmoid colon and cecum, respectively.[7,9] No cases have been reported in children with IBD. The pathophysiology of volvulus in patients with IBD is not fully elucidated; we postulate that abnormal fixation of an inflamed colon to surrounding viscera might be a predisposing factor. Given this hypothesis, our patient should be followed to monitor for the development of transmural disease in the future.

Colonic volvulus typically presents with abdominal pain, vomiting, and abdominal distension, although volvulus can mimic either constipation or infectious colitis in children.[4,5] Our patient was relatively asymptomatic, and there was a greater suspicion for inflammation due to UC as the cause of her obstruction rather than a mechanical cause.

Initial imaging in children with suspected volvulus includes a KUB, which typically shows a dilated colon with inverted-U sign, which was seen in our patient; however, air in the rectum argued against a volvulus.[4] A beak sign on barium enema and "whirl" sign of the mesentery on an abdominal computed tomography scan are also used to confirm volvulus.[10] Our patient had undergone an MRE to evaluate colonic inflammation and the extent of the disease, which demonstrated rectosigmoid thickening and unexpected proximal dilatation of the colon. However, an MRE would not be the initial imaging modality of choice for either toxic megacolon or volvulus. MRE is the preferred modality in pediatric patients with IBD because of the ability to evaluate the small bowel as a substantial fraction of children with IBD have Crohn's disease compared with adults, even if they are initially presenting with isolated colonic involvement.[11] In addition, MRE is preferred over computed tomography because of a higher safety profile with no radiation exposure if it can be obtained in the same time frame in children with IBD who require a high volume of radiologic studies over their lifetime.[12,13]

In a nontoxic patient with colonic dilation, colonoscopy may aid in the diagnosis of volvulus before surgery, especially in

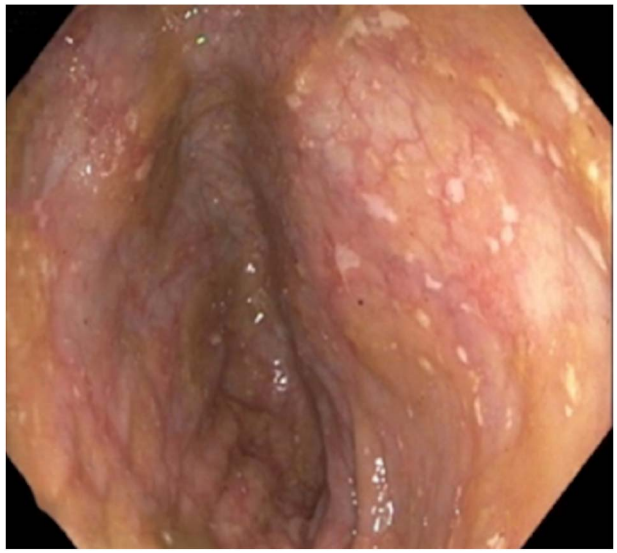

Figure 6. Mucosal changes in the descending colon characterized by scattered aphthous ulcerations and erythema at the time of follow-up colonoscopy 1 month later.

patients with IBD.[14] Volvulus was discovered in our patient at colonoscopy with a typical whirl sign (converging mucosa) at the rectosigmoid junction with immediate passage of gas on traversing this transition point.[8] The examination demonstrated very pale colonic mucosa extending to the splenic flexure, suggesting decreased perfusion of this segment of the colon.

If sigmoid volvulus is diagnosed on imaging, emergent endoscopic detorsion is the preferred method for initial reduction (if feasible), followed by rectal tube decompression, which was not required in our patient. The success rate of endoscopic detorsion is less in children compared with adults, with a comparable recurrence rate of sigmoid volvulus (50%) in both groups.[5,8,15] Our patient is doing well at 12 months after volvulus without surgery or recurrent volvulus, likely due to optimizing her UC therapy.

Physicians should have an increased index of suspicion for volvulus in patients with IBD without prior surgery presenting with evidence of bowel dilation or obstruction with atypical features on examination or imaging. We postulate that underlying inflammation may predispose to volvulus. This condition may mimic toxic megacolon and should be considered in the differential diagnosis. Timely involvement of a multidisciplinary team including a surgeon can optimize the clinical outcomes.

DISCLOSURES

Author contributions: Conception: A. Mittal and M. Kay. Data collection: A. Mittal. Drafting of the manuscript: A. Mittal and M. Kay. Critical revision of the manuscript for important intellectual content: all authors. Final approval of the manuscript: all authors. M. Kay is the article guarantor.

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Informed consent was obtained for this case report.

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