Sigmoid volvulus: A pediatric case report and review of management

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abstract

Volvulus in the pediatric population commonly occurs in the small bowel associated with malrotation or internal hernia whereas sigmoid volvulus (SV) is extremely rare. Herein we report a case of SV, review the literature and provide a suggested algorithm for diagnosis and management.

Volvulus in the pediatric population commonly occurs in the small bowel associated with malrotation or internal hernia [1]. Sigmoid volvulus (SV), while common in the elderly, is rare in the pediatric population. Patients most frequently present with abdominal pain, abdominal distention and vomiting, symptoms that may easily be misattributed to a more benign condition. Untreated, SV may progress to colonic ischemia and perforation; as these consequences are potentially life-threatening, clinicians should consider SV in the differential for patients presenting with acute or recurrent episodes of abdominal pain and bowel obstruction [2]. This article presents a child diagnosed with SV secondary to functional constipation and reviews the current algorithm for treatment.

1. Case report

A 6 year old, previously healthy boy presented with acute onset of severe abdominal pain after defecation. The bowel movement was normal color and consistency, with no blood or tarry appearance. The patient had one episode of emesis in triage after oral ibuprofen. There were no fevers, chills, or sick contacts. Patient’s parents reported daily bowel movements, but only after coaxing; they reported he voluntarily retained stool. He passed stool at birth. He was neurologically normal and took no medications. There was no family history of bowel disease.

On physical examination, the patient was tachycardic, but all other vital signs were stable. He was writhing in pain while clutching his abdomen. He had abdominal distension without focal tenderness. There was no rebound. Bowel sounds were decreased. There was no hepatosplenomegaly, masses, or hernia. Plain abdominal radiographs showed markedly dilated bowel loops with coffee bean sign (Fig. 1). Ultrasound showed no intussusception. On barium fluoroscopy, beaking of the dilated colon at the level of the mid sigmoid colon was identified and successfully reduced. A rectal tube was placed proximal to the obstruction (Fig. 2). He then underwent full-thickness rectal biopsy. After it returned negative for aganglionosis, ruling out Hirschsprung’s Disease (HD), he underwent laparoscopic sigmoidectomy with primary anastomosis (Fig. 3). He experienced difficulty tolerating oral intake initially, but advanced to a normal diet by post-operative day 5. At the time of discharge, he was experiencing regular bowel movements.

2. Discussion

Sigmoid volvulus occurs when a redundant loop rotates around its elongated mesentery. Obstruction of the intestinal lumen and impairment of vascular perfusion occur when the degree of torsion exceeds 180° and 360°, respectively [3]. In children, the median age of diagnosis is approximately 7–12 years old. There is a 3.5:1 ratio of male to females, however a case series in Turkey showed that 17
out of 19 cases were male [2,4]. It is common in African, Asian, Middle Eastern, Eastern European, and South American countries [5]. In underdeveloped countries, it is often associated with Chagas disease [6-9].

SV is often an overlooked diagnosis and can present in two ways. Most frequently, it presents acutely with a mean duration of symptoms of 1.5 h. Less commonly, patients present with recurrent episodes of abdominal pain [2]. Because spontaneous detorsion is followed by the passing of stool or flatus, it is commonly misdiagnosed as constipation [10]. If untreated, SV can result in hemorrhagic infarction, perforation, septic shock, and death [11].

Abdominal radiographs are often obtained in patients with suspected SV. Unlike in adults, abdominal radiographs are less beneficial in aiding in diagnosis of SV in pediatric patients. Diagnosis by plain films was made in 17–30% of pediatric cases versus 60–90% in adults [12]. On plain radiographs, the most common finding is colonic dilatation. The coffee bean sign is reported in only 16–29% of pediatric patients [2,13]. Barium enema, which serves as a treatment option for SV, can also be of diagnostic value. As in adults, barium enema increases radiographic diagnostic sensitivity in pediatric patients (71–82%) [12].

Treatment for SV remains controversial in children. If the patient is stable, non-operative reduction of the volvulus with barium enema or sigmoidoscopy may first be attempted [2,12]. One review found that SV reduction by barium enema (BE) is successful in 77% of cases [2]. Sigmoidoscopy and rectal tube reduction have lower success rates in children (47%) [2]. BE reduction is preferred over endoscopic detorsion [2,14], although sigmoidoscopy allows for suction and more rapid decompression. All non-operative modalities for decompression carry a risk of perforation. If a non-operative approach is unsuccessful at reducing the volvulus or the patient develops signs of perforation/peritonitis, then immediate surgery is warranted [13,14].

The definitive treatment for SV is sigmoidectomy, either with primary anastomosis or colostomy. Recurrence is common when detorsion without resection is performed (operative 25%, non-operative 35%) [2,4]. Recurrent SV after sigmoidectomy is unreported [15]. Endoscopic sigmoidopexy has been a successful alternative to surgery in high-risk patients, in particular the elderly [16,17]. It has also been successful in non-gangrenous ileosigmoidal knotting pediatric cases, which have a high mortality rate [4,18].

There has been an established association of sigmoid volvulus with HD, thus a full-thickness biopsy is warranted to rule out aganglionosis [2,12,19,20]. Therefore, we propose a modified

Fig. 1. Abdominal x-ray demonstrates coffee bean sign.

Fig. 2. Contrast enema demonstrates a) bird’s beaking at the sigmoid colon, b) a small amount of contrast passing through bird’s beak, c) rectal tube placed past bird’s beak.
algorithm to the one suggested by Salas et al (Fig. 4). Rather than performing sigmoidectomy at the time of rectal biopsy, we suggest that sigmoidectomy be delayed until the results of the biopsy are available. If the biopsy is positive for HD, pull-through surgery is indicated [20]. This may be performed concurrently with resection of any remaining redundant sigmoid colon. This approach avoids a 6–8 week delay for definitive management, as well as a colostomy in patients without bowel compromise.

Fig. 3. Intraoperative photographs demonstrate a) massive dilation of sigmoid, as rectal tube had been removed at the time of the rectal biopsy several days prior, b) externalization of sigmoid colon with transition point at point of volvulus with proximal dilation, c) transition point, d) decompressed colon distal to volvulus, stapled off, next to dilated proximal end, arrow pointing to a healthy, non-dilated bowel.

Fig. 4. Algorithm for management of sigmoid volvulus.
3. Conclusion

Sigmoid volvulus is an uncommon diagnosis in children. However, case series and case reports of SV are becoming more prevalent in the literature. Failure to recognize SV may result in life-threatening complications such as sigmoid gangrene/perforation, peritonitis, sepsis and death [2]. Thus, clinicians should consider SV as a “do not miss diagnosis” in the differential for children presenting with acute or recurrent episodes of abdominal obstruction. A proper diagnostic and treatment algorithm should be followed to optimally treat the growing number of SV cases and avoid long-term sequelae and mortality.

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References