

Pediatric sigmoid volvulus of an extremely long sigmoid colon with hypoganglionosis: a case report

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Abstract

Sigmoid volvulus is an extremely rare cause of intestinal obstruction in pediatric patients. This condition occurs when a redundant sigmoid loop with a narrow mesenteric base of attachment to the posterior abdominal wall rotates around its mesenteric axis. This situation might result in vascular occlusion and large bowel obstruction. There are only a few predisposing factors of sigmoid volvulus, such as a long-term history of constipation or pseudo-obstruction with an excessive sigmoid colon. Underlying hypoganglionosis can also lead to large bowel obstruction. There have only been two reported cases of hypoganglionosis with sigmoid volvulus, and both were in adults. Sigmoid volvulus usually presents with abdominal pain, nausea, vomiting, constipation and abdominal distension, an absence of stool, or the presence of melenic stool in the rectum. Initial treatment options are non-surgical for stable patients, although surgical management might be necessary. If sigmoid volvulus is not recognized and resolved, it may lead to serious complications and death. Pediatric sigmoid volvulus is frequently the fulminant type, and therefore, a decision about treatment must be prompt. We present an unusual pediatric case of an extremely long sigmoid colon with hypoganglionosis, which twisted and caused obstruction. This condition was resolved with surgical resection.

Keywords

Sigmoid volvulus, hypoganglionosis, pediatrics, surgery, intestinal obstruction, colon

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Introduction

Sigmoid volvulus (SV) is a common cause of intestinal obstruction in adults owing to redundancy of the sigmoid colon loop. A redundant sigmoid loop is rare in children. Among children, several predisposing factors of SV have been identified, such as Hirschsprung's disease, congenital anomalous fixation of the colon, and chronic constipation. The incidence of SV in adults shows a significant geographical distribution in the so-called "volvulus belt" of Africa and the Middle East, which suggests that dietary habits are the probable cause. However, a characteristic geographical distribution of SV is absent in the pediatric population, and thus the cause of SV is unclear.^{1,2} The exact incidence of SV in children is still unknown. There have only been a few case reports and a relatively small case series on SV, with only 93 cases of SV reported worldwide by 2007,^{1,3} and only a few more reported by 2020.⁴ One minimally reported predisposing condition is hypoganglionosis. There have only been two case reports on hypoganglionosis present with SV in adults, but no reports in children.^{5,6}

Abdominal pain and distension are the most common complaints of SV, and spontaneous detorsion can be interpreted as constipation.⁷ Unrecognized and untreated SV can result in hemorrhagic infarction, perforation, septic shock, and death.⁸ We report an unusual pediatric case of an extremely long sigmoid colon with hypoganglionosis, which twisted and caused obstruction.

Case presentation

The reporting of this study conforms to the CARE guidelines.⁹ A 10-year-old girl was admitted to the hospital because of abdominal pain, which started the previous day, and vomiting after every intake of fluid. She had no stool or flatus passage for 2 days. A physical examination showed a distended, meteoristic, tympanic abdomen with left upper quadrant tenderness and high-pitched bowel sounds. Laboratory results showed hypokalemia (2.7 mmol/L), hyponatremia, and hypochloremia. A plain radiograph of the abdomen (Figure 1) showed grossly dilated large bowel loops up to 8 cm, with no obvious cause of

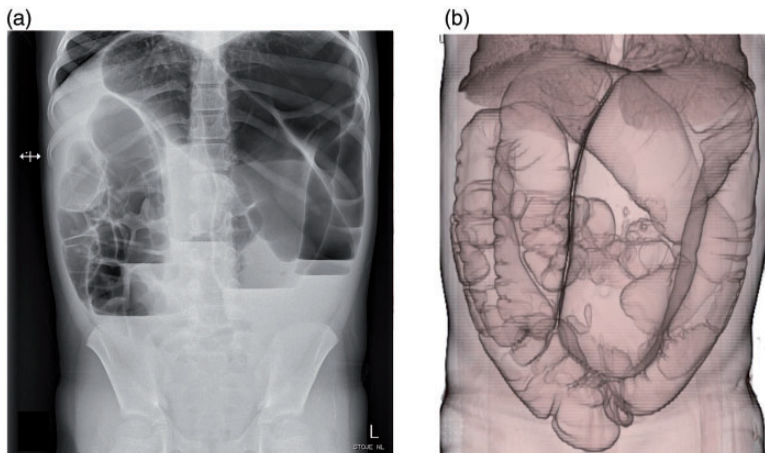


Figure 1. (a) Abdominal X-rays showing ileus with dilated large and small bowels in the upper abdomen with air–fluid levels. There is no evidence of intraperitoneal free air. (b) Computed tomographic three-dimensional reconstruction (MinIP) showing air distribution and the exact degree of colon dilatation

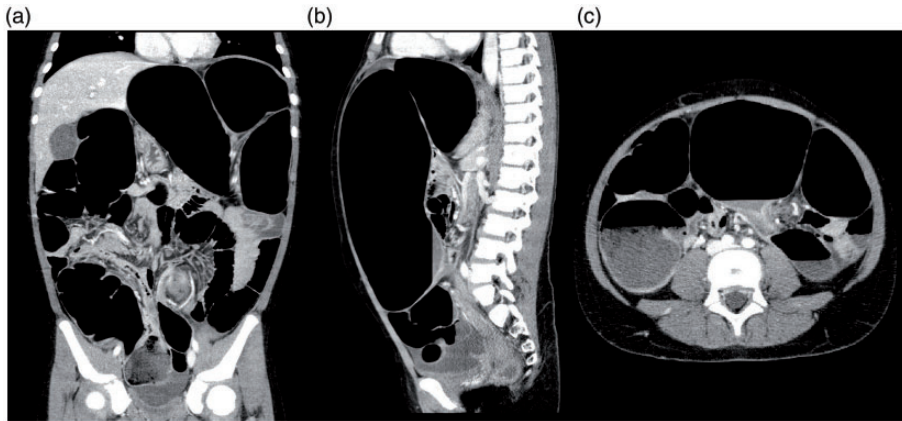


Figure 2. (a) Computed tomographic scan of the abdomen with contrast administration (portal vein phase in the coronal plane) showing “whirlpool sign” in the central part of the abdomen. There is rotation of the mesentery and mesenteric vessels centrally. (b) Computed tomographic scan in the sagittal plane showing an extremely dilated sigmoid colon (9 cm), which lies unusually ventral and high, and the upper part of the sigmoid colon is under the left hemidiaphragm. (c) Computed tomographic scan in the axial plane shows “bird’ beak sign and prestenotic dilatation of the sigmoid colon near the point of mesenteric rotation. Additionally, there are dilated bowels with air–fluid levels, without free air or free fluid intraperitoneally, and no signs of perforation

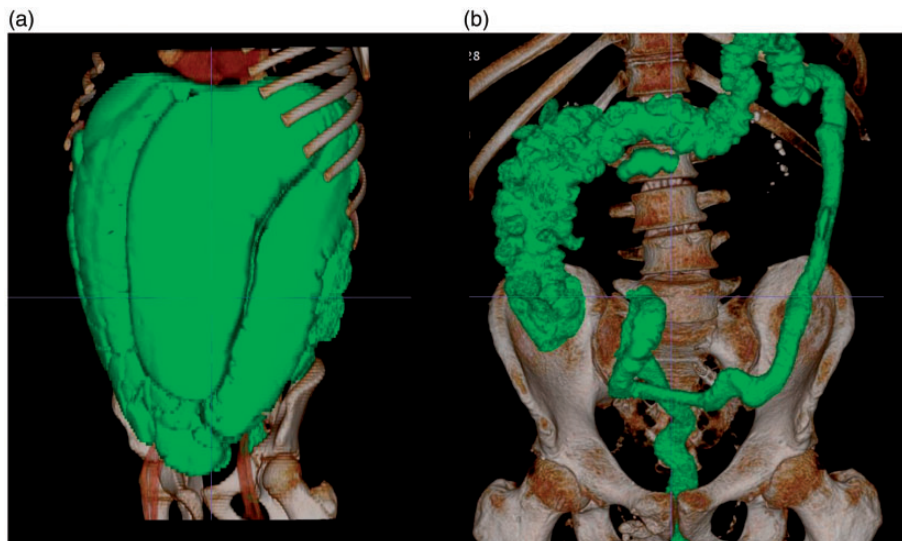


Figure 3. (a) Computed tomographic three-dimensional reconstruction in the paracoronar plane showing typical “coffee-bean” sign in the upper left part of the abdomen. This sign was not able to be seen on previous images because of superposition of dilated loops. (b) Computed tomographic three-dimensional reconstruction of a normal large intestine

obstruction. A barium enema (BE) was attempted, but it was unsuccessful. An abdominal computed tomographic (CT) scan (Figures 2 and 3) revealed a bowel obstruction with swirling of the mesentery, which suggested an SV. An emergency laparotomy was performed, which confirmed torsion of the sigmoid colon. A length of 340 mm of the elongated sigmoid colon was resected, with side-to-side anastomosis because the two intestinal lumens had different diameters. She was discharged without complications. Samples of the resected sigmoid colon were sent for a histological examination and reviewed by two separate examiner teams. This examination showed hypoganglionosis because ganglia were present, but there were fewer ganglia than usual and fewer ganglion cells within them.

Discussion

Hypoganglionosis of the myenteric plexus is a rare innervation disorder associated with a small amount of intestinal ganglion cells and is related to some intestinal pathologies, such as chronic and/or severe acute constipation, pseudo-obstruction, or enterocolitis.^{10,11} Rarely, hypoganglionosis can be present with volvulus.^{5,6} Our patient's histological findings clearly suggested hypoganglionosis, while Hirschsprung's disease was ruled out. She had no history of chronic constipation or any other motility disorder or underlying condition.

During surgery, the redundant sigmoid colon was resected in our patient. A previous large study¹² measured the length of parts of the colon on various CT scans performed on children. This study showed that the mean (standard deviation) length of the sigmoid colon in children aged between 9 and 11 years was 223 ± 75 mm. Our patient's sigmoid colon was 340 mm long, which is extremely long for her age. No underlying cause for such a long sigmoid

colon, other than hypoganglionosis, was identified, and her family reported no previous problems, such as constipation.

The diagnostic approach to children with abdominal pain rarely includes SV as a differential diagnosis. The first imaging modality commonly used to diagnose a suspected SV is plain abdominal radiography, but this technique contributes less to the diagnosis of SV in pediatric patients than in adults. Zeng et al.¹³ showed that plain film diagnosis of SV was made in 17% to 30% of pediatric cases versus 60% to 90% in adults. In our case, a plain radiograph only showed signs of considerable colonic dilatation with no "coffee bean" sign, which has been reported in only 16% to 29% of pediatric patients.¹⁴ Ultrasound has little diagnostic value for SV because of the difficulty of visualization, with impairment by the presence of gas and other intestinal content.

A BE is advisable for showing a spiral twist at the site of the bowel torsion. A roentgenographic appearance of this twist has been referred to as a "bird beak" or "ace of spades" deformity, which is pathognomonic for SV. A BE can also be therapeutic in uncomplicated cases, as shown in a large case series study of successful reduction of 11/14 cases of colonic volvulus.¹⁵ In our case, BE showed some sort of obstruction, but it was inconclusive regarding the cause.

Performing a CT scan on a child raises doubt over the high dose of ionization, especially if there is underlying constipation. However, after an unsuccessful enema and inconclusive ultrasound, we decided to perform a CT scan in our patient, which revealed an SV. Tannouri et al. reported a small series of 11 cases of SV in children and they concluded that CT scanning is a much more sensitive modality compared with plain film (55.6% versus 22.2%).¹⁶

The treatment options of SV in children remain controversial.¹⁴ Non-operative reduction, such as a BE or sigmoidoscopy, can be attempted if the child is stable and shows no signs of perforation or peritonitis. One review showed that SV reduction by a BE was successful in 77% of cases compared with sigmoidoscopy and rectal tube reduction, which have lower success rates in children (47%) and a greater risk of perforation.^{1,14} Alagumuthu et al. showed that non-operative methods of decompression were less successful in children compared with adults (33% vs. 76%).¹⁷ In children, all non-operative modalities for decompression pose a risk of perforation because of a thin-walled colon compared with adults whose sigmoid colon is frequently thickened by mesosigmoiditis.¹ If a non-operative approach is unsuccessful at reducing the volvulus or the patient develops signs of perforation/peritonitis, then immediate surgery is warranted.² We decided to operate because of the greatly dilated colon and fear of perforation in our case. According to Salas et al.,¹⁴ sigmoidectomy with either primary anastomosis or colostomy is the definitive treatment for SV. Recurrence is common when detorsion without resection is performed (operative: 25%, non-operative: 35%).^{14,17,18}

Conclusions

To the best of our knowledge, hypoganglionosis as a redundant sigmoid colon with SV has not been previously reported. However, whether hypoganglionosis caused elongation of the sigmoid colon or overlap of these two conditions (hypoganglionosis and redundant sigmoid colon) resulted in SV remains unclear. Nevertheless, in acute abdominal pain in children, SV should be considered as a possible differential diagnosis because of its rapid progression to a life-threatening situation. An early CT examination is crucial for revealing the

underlying cause of SV. Prompt surgical intervention is indicated owing to the high incidence of bowel gangrene and perforation.

Ethics statement

Approval for the study protocol was not required by our institution because this was a case report. We anonymized all of the patient's details and her parents gave verbal consent for publication.

Declaration of conflicting interest

The authors declare that there is no conflict of interest.

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Author contributions

All of the authors contributed to drafting of the manuscript, reviewed the literature, collected and collated the clinical data, and read and approved the final manuscript.

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