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Hirschsprung's Disease-Related Giant Sigmoid Volvulus Complicated by Refractory Hypertension in an Elderly Man

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

BCEG 1 **Shaohan Wu***
BEF 2 **Xiaofang Sun***
BCD 3 **Yawei Yu**
AG 1 **Yiyu Shen**

1 Department of General Surgery, The Second Affiliated Hospital of Jiaying University, Jiaying, Zhejiang, P.R. China
2 Department of Dermatology, The Second Affiliated Hospital of Jiaying University, Jiaying, Zhejiang, P.R. China
3 Department of Clinical Pathology, The Second Affiliated Hospital of Jiaying University, Jiaying, Zhejiang, P.R. China

* These authors contributed equally to this work

Corresponding Author: Yiyu Shen, e-mail: doctorSYY01@163.com

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Patient: Male, 82
Final Diagnosis: Hirschsprung's disease-related sigmoid volvulus complicated with refractory hypertension
Symptoms: Constipation • moderate abdominal pain and progressive abdominal distension • hypertension
Medication: Antihypertension medication
Clinical Procedure: CT scan • Hartmann's procedure
Specialty: General Surgery

Objective: Rare disease





Background: Sigmoid volvulus (SV) is a life-threatening condition occasionally seen in adults. Adult Hirschsprung's disease (HD)-related SV is rarely complicated by difficult-to-control hypertension. In this report we present the case of an elderly man with a rare constellation of HD, SV, and refractory hypertension.

Case Report: An 82-year-old man had long-term constipation, moderate abdominal pain, and progressive abdominal distension. A CT scan revealed the typical "coffee bean sign". Blood pressure was abnormal high. Subsequently, the patient's condition deteriorated. Therefore, he underwent a Hartmann's procedure. A giant and redundant sigmoid colon (length more than 60 cm, maximal diameter about 15 cm) was demonstrated to be the cause of SV during the process of surgery. Moreover, abdominal compartment syndrome caused by SV resulted in his high and refractory blood pressure (BP). Postoperative pathological results revealed HD in his sigmoid colon.

Conclusions: SV is rarely combined with conditions like refractory hypertension or HD among the elderly. Clinical features of SV typically present with long-term constipation, severe abdominal pain, and progressive abdominal distension. The "coffee bean sign" could be observed in imaging examinations. It is important to note that the management of SV is to relieve the obstruction and prevent recurrence, no matter which therapy is used in elderly patients with Hirschsprung's disease.

MeSH Keywords: Antihypertensive Agents • Hirschsprung Disease • Sigmoid Diseases

Full-text PDF: <https://www.amjcaserep.com/abstract/index/idArt/908389>

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Background

Sigmoid volvulus (SV) is a life-threatening condition that results from the twisting of the sigmoid colon on its mesenteric axis. It is the third leading causes of large bowel obstruction, after cancer and diverticulitis, in adults [1,2]. SV accounts for 4% of total cases of large bowel obstruction in the United States [3], and is much higher in Africa, Asia, and India [1,2]. The main cause of SV is a long and redundant sigmoid colon with an elongated mesentery, which is prone to rotating on itself [4,5]. Hirschsprung's disease (HD or HSCR) [6,7] is an uncommon cause of those redundant SVs in elderly patients. The incidence of HD in the elderly is less than 1/1 million [8], and very few cases have been reported in the English literature.

SV is an abdominal emergency issue which is seen more commonly in the elderly, but it can occur in young adults, pregnant women, and children [9–11]. Clinical symptoms vary with disease progression. However, it has 3 typical presentations: constipation, severe abdominal pain, and progressive abdominal distension. SV is not difficult to diagnose among these elderly patients according to their symptoms and auxiliary examinations, but it may be difficult to deal with because older adults usually have other difficult complications or diseases. Herein, we described a case of an 82-year-old man with the triad of symptoms. A computed tomography (CT) scan revealed the typical “coffee bean sign”, and the patient had very high blood pressure (BP). This elderly patient was postoperatively diagnosed with HD.

Case Report

An 82-year-old man was admitted to our department with a 3-day history of abdominal distention, pain, obstipation, and constipation. He had a 25-year history of hypertension and an old myocardial infarction. He regularly took antihypertension medication and his systolic BP was always reasonably controlled at 130–145 mm Hg. Physical examination of his abdomen disclosed obvious abdominal distention, tympanitic percussion, decreased bowel sounds, and a prominent loop in his abdomen. However, there were no signs suggesting perforation or necrosis of his bowel. His BP was 181/109 mmHg, HR was 86 bpm, BT was 36.5°C, and SpO₂ was 83.7%. Blood tests revealed that WBC was 8.0×10⁹/ml and Hs-CRP was 3.96 mg/ml. A CT scan of the abdomen depicted a dilated sigmoid colon with the classic “coffee bean sign” (Figure 1A, 1B) pre-operatively. Thus, these findings helped us to quickly make the diagnosis of SV in this patient.

He was afebrile and hemodynamically stable, and there was no evidence of peritonitis. In consideration of his medical history, at that moment, his BP was relatively high (181/109

mmHg). Therefore, he was first given flexible nasal intestinal decompression tube implantation to keep gastrointestinal decompression, somatostatin to inhibit the secretion of digestive juices, fasting treatment, and antihypertension medication. Unfortunately, the situation of the patient was not improved significantly. By 24 h later, the abdominal distention, and pain were more severe than before, and he had peritonitis symptoms of tenderness, rigidity, and rebound tenderness. Meanwhile, the BP was up to 210/116 mmHg despite using antihypertension medications. The BT was 38.5°C. In consideration of all the facts mentioned above, this patient was immediately taken to the operating room.

A midline laparotomy was used to explore, exteriorize, and resect the redundant sigmoid colon and mesosigmoid. Surprisingly, during the emergency laparotomy procedure, the high BP quickly declined to 143/79 mmHg. Additionally, we found a markedly dilated sigmoid colon (length more than 60 cm, maximal diameter about 15 cm), and a redundant mesentery of the sigmoid colon (Figure 2A, 2C). The bowel seemed nonviable. His sigmoid colon had obvious ischemia and necrosis 14 cm above the anus. Next, the patient underwent a Hartmann's procedure (HP). Sigmoidectomy, proximal colostomy, and closure of the distal colon were performed to treat this severe SV (Figure 2B). The resected sigmoid was sent for histopathology analysis.

Postoperatively, the patient recovered well. His BP returned to 136/75mmHg. A CT scan of the abdomen illustrated no evidence of leakage and intestinal obstruction. The histopathology results of the resected sigmoid colon revealed inflammatory reaction with necrosis and hemorrhage (Figure 3A). Especially, immunohistochemical staining, with S-100 (Soluble protein 100) [12] which used to confirm the HD, indicated occasional ganglion cells and neuronal hypogenesis (small ganglia, Figure 3B). Hyperplasia of nerve bundles was huge and wave-shaped in the distal spastic intestine (Figure 3C). The immunohistochemical results revealed HD of his sigmoid colon. The patient was discharged uneventfully on postoperative day 11. This patient was followed-up for more than 6 months and reported a dramatic improvement in his bowel function and quality of life.

Discussion

SV typically occurred in the elderly, especially in frail patients or individuals with dementia or a psychiatric illness [13]. By 2020, over 23% of China's population is expected to be over 65 years [14]; therefore, doctors and nurses in China need to know more about SV. SV can cause adult large bowel obstruction, with a 33%–80% mortality rate in patients with intestinal perforation and necrosis. However, the mortality rate is



Figure 1. (A, B) Computerized tomography scan demonstrated the giant sigmoid volvulus and coffee bean sign, as well as the distended sigmoid compartments with central double walls.

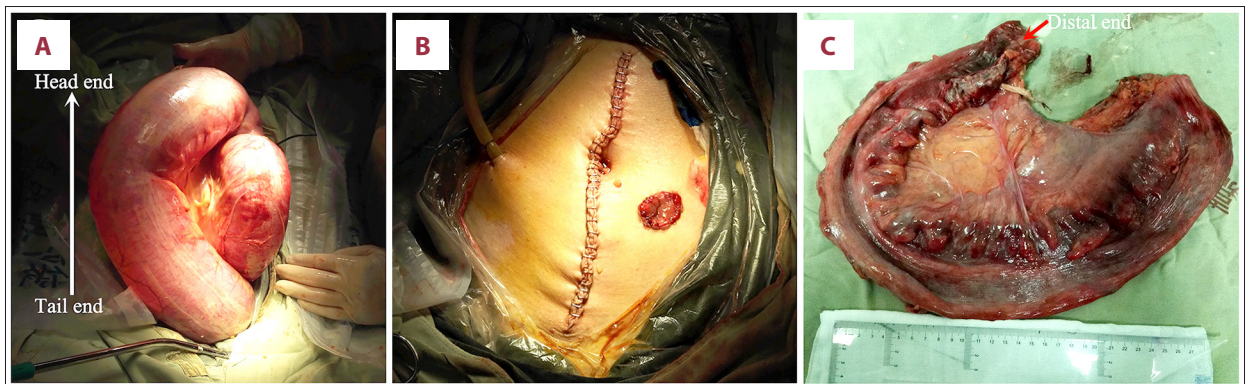


Figure 2. (A) The patient underwent a Hartmann's procedure (HP) with midline laparotomy. (B) The proximal colostomy and closure of distal colon was performed to treat this severe SV. (C) A giant and redundant sigmoid colon was found during the process of surgery, length more than 60 cm, maximal diameter about 15 cm.

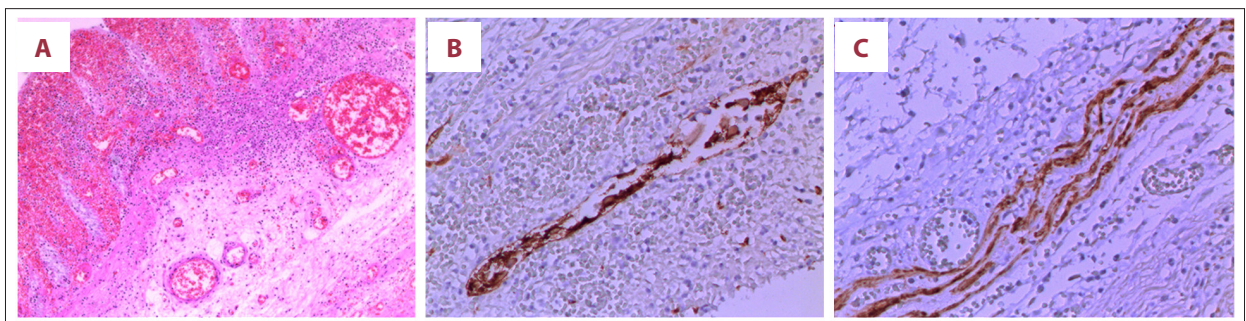


Figure 3. Immunohistochemical staining of resected sigmoid showed inflammatory reaction and Hirschsprung's disease. (A) Hematoxylin and eosin (HE) staining. (B) One occasional ganglion cell with S-100 (soluble protein 100) staining. (C) Hypertrophied and wave-shaped nerve bundles with S-100.

relatively lower in cases without ischemia (0–7%), so early diagnosis and treatment are essential [2,15,16]. Anatomically, the mobile and redundant sigmoid colon, and bowel adhesions associated to abdominal surgery, have been reported as major factors of SV [17]. Although the complaints vary, abdominal distention, pain, and constipation are more common in SV. However, some patients with SV have difficult-to-control BP. In fact, our patient's BP was always controlled and relatively stable because he took antihypertension medication and consulted the doctor on a regular basis, in spite of his 25-year history of hypertension. When he was admitted in our department, his BP was already 181/109 mmHg, so a cardiology consultation was immediately done to guide the use of antihypertensive drugs. Unfortunately, there was no drop in his difficult-to-control hypertension despite using nitroglycerin injection, irbesartan, hydrochlorothiazide tablets, and isosorbide dinitrate tablets. In contrast, along with the progression of abdominal distention, his BP was up to 210/116 mmHg. But it gradually went to 143/79 mmHg after his abdomen was opened. Thus, we thought the abdominal compartment syndrome (ACS) caused by SV was mainly induced by intra-abdominal hypertension and the difficult-to-control high BP. Additionally, his anxieties and fears may also have result in high BP. However, the relationship between SV and severe hypertension is unclear and worth further exploration.

With acute onset of SV, the major symptoms are usually constipation, severe abdominal pain, and progressive abdominal distention, but these situations might be reduced in chronic SV [16]. Diagnosis can be achieved according to clinical features, as well as radiologic and endoscopic findings. Most cases can be diagnosed only with X-ray imaging showing a dilated sigmoid loop known as “coffee bean sign”. Actually, it has much higher sensitivity on CT scan [18,19]. In this report, the patient had the “coffee bean sign” (Figure 1A, 1B), consistent with SV (Figure 2). Notably, hematoxylin and eosin (HE) staining showed edema and inflammatory cell infiltration in the sigmoid (Figure 3A). Immunohistochemical staining of sigmoid specimens showed intestinal neuronal dysplasia and hypertrophied nerve bundles in the myenteric and submucosal of distal narrow zone of colon (Figure 3B, 3C). These observations demonstrated that this patient had Hirschsprung's disease (HD or HSCR). HD is a congenital disorder caused by sparse ganglion cells or intestinal neuronal dysplasia or nerve fibers dysplasia in both submucosal and myenteric plexuses and the transitional segment of colon during development [6,7]. Those disorders lead to the functional obstruction manifesting as severe chronic constipation and colorectal distention, which can result in SV [20,21]. HD mostly causes SV in infants and children and is rare in elderly patients. A few of these patients live to old age, even if they were diagnosed with a giant colon. HD might be misdiagnosed or missed due to the chronic constipation, which can cause acquired giant colon. Moreover, after

surgical operation, the diseased colon may not conduct immunohistochemical staining-related HD in adult patients. In fact, further considering the patient's medical history in this case, he also had decades of constipation and had received multiple conservative treatments. Surgery is the definitive treatment method for adult HD [22,23].

There is no unified therapeutic guideline for the treatment of SV. Dealing with the intestinal volvulus relies on relieving the obstruction and preventing recurrence [24]. Generally speaking, SV treatment is classified into nonsurgical and surgical treatment. Non-operative therapy, such as decompression through a nasogastric tube or colonoscopy, can convert an emergency procedure in an elective procedure. In our case, decompression through colonoscopy was not used due to the 3-day pain history, which was considered an absolute contraindication for decompression through colonoscopy. If there was ischemic colon necrosis, decompression through colonoscopy might lead to bowel perforation during the process. Urgent surgery is recommended if there was evidence of bowel ischemia or necrosis. The findings during the operation, as well as the preference of the surgeon, dictate the surgical alternatives. A prospective, randomized study indicated that sigmoid resection, done either as a HP when a necrotic colon is discovered at laparotomy or as one-stage resection with primary anastomosis in the setting of a viable colon, had the lowest rate of recurrence [25]. In the present report, this huge dilated sigmoid colon with a redundant mesentery was nonviable because of chronic SV. His sigmoid colon had obvious ischemia and necrosis 14 cm from the anus. Considering this and the patient's complex situation, in our case, we chosen the HP through laparotomy. A previous report discussed use of a minimal-access HP to deal SV. A single skin incision of ≤ 4 cm at the proposed colostomy site was used to remove the redundant mesosigmoid and sigmoid colon. The same incision was used to mature the end colostomy [26]. In this way, the operation was incisionless, and there was no laparoscopy performed. This might be a preferred approach for surgery to decompress SV in elderly patients.

Conclusions

SV is a potentially life-threatening situation predominantly affecting the elderly. Clinical features typically present with the triad of constipation, severe abdominal pain, and progressive abdominal distention. The “coffee bean sign” can be observed in imaging examinations. Although HD is most often diagnosed in the perinatal period and infancy, it can present during childhood, adulthood, and even in old age. SV may lead to difficult-to-control BP, which can increase short-term risk. Then, the management of SV is to relieve the obstruction and prevent recurrence no matter which therapy was used, especially in the elderly.

Conflict of interest

None.

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