

CASE REPORT

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A case of sigmoid volvulus secondary to Hirschsprung's disease

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Abstract We report a case of sigmoid volvulus accompanied by undiagnosed Hirschsprung's disease (HD). A 3-year-old boy who had had chronic constipation since birth was admitted because of progressive abdominal distention and obstipation. A plain abdominal film was compatible with megacolon and distal obstruction. Sigmoid volvulus secondary to HD was found at emergency laparotomy, and a colostomy was performed after detorsion. A Soave procedure performed 3 months later yielded complete loss of symptoms. Whenever an operation for sigmoid volvulus is performed in children, a frozen biopsy section of distal colon to exclude HD is recommended.

Key words Hirschsprung's disease · Volvulus · Sigmoid colon

Introduction

The signs and symptoms of Hirschsprung's disease (HD) are highly variable. While it may present in the newborn period with acute signs of abdominal distention such as vomiting and failure to pass meconium, on rare occasions the disease may persist undiagnosed into childhood. A child

with undiagnosed HD may appear to have a mechanical obstruction, megacolon, perforation, and peritonitis. In extreme cases, megacolon from unrecognized HD may lead to colonic volvulus, particularly in the sigmoid colon. We report a case of sigmoid volvulus (SV) accompanying HD in a child.

Case report

A 3-year-old boy was admitted to our department with abdominal distention and obstipation for the last 3 days. His past history revealed that the initial passage of meconium had been delayed until the 3rd day of life and was achieved by rectal irrigations and enemas. Unfortunately, the treating physician had dismissed the possibility of HD at that time. Since then, the boy had had chronic constipation that could be managed by enemas. On admission, he had marked abdominal distention without peritoneal tenderness or rebound. The rectum was empty on digital examination. All laboratory tests were normal. There were hugely dilated loops of colon indicating distal obstruction on a plain upright film of the abdomen (Fig. 1).

Despite rectal irrigations for 36 h the abdominal distention persisted and peritoneal tenderness developed. An emergency laparotomy was planned for a colostomy. At exploration, a megacolon with distention of the sigmoid colon was observed with a 360° clockwise volvulus above the transitional zone. After detorsion the colon appeared viable, not requiring resection. Biopsy specimens were obtained below the transitional zone for frozen section histopathologic examination, and confirmed the diagnosis of HD. A loop colostomy was carried out at the ganglionic proximal sigmoid colon. In the 3rd postoperative month the patient underwent a Soave procedure, followed by closure of the



Fig. 1 Abdominal radiograph showing huge, dilated loops of colon

colostomy 2 months later. He was doing well at 1-year follow-up.

Discussion

Dean and Murry reported the first case of HD presenting as a SV in 1952 [2]. Since then, six cases of SV with HD have been reported in children [1, 3–5]. All patients were male, ranging in age from 2 days to 12 years (Table 1). We have presented the eighth case of SV with HD. The age at presentation of the patients with volvulus is usually older than that reported with other presentations of HD.

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Table 1 Review of cases with sigmoid volvulus secondary to Hirschsprung's disease

Case no.	Reference	Age	Sex	Treatment
1	Dean et al. [2]	4 years	M	Sigmoid detorsion
2	Shepherd [4]	12 years	M	Sigmoid detorsion
3	Ciardimi et al. [1]	3 years	M	Not stated
4	Ciardimi et al. [1]	5 years	M	Not stated
5	Valla et al. [5]	5 days	M	Sigmoid resection
6	Valla et al. [5]	5 years	M	Sigmoid resection
7	McCalla et al. [3]	2 days	M	Sigmoid detorsion
8	Erdener et al.	3 years	M	Sigmoid detorsion and colostomy

As the child grows older the delay in diagnosis allows development of a megacolon, which may lead to volvulus. Although nonoperative reduction by barium enema or sigmoidoscopy is recommended for adult patients with SV, it is less often successful in children [3]. In addition, because SV in children often occurs secondary to an underlying primary disease, surgical intervention should be considered without delay. After the volvulus is reduced and if the bowel is viable, detorsion alone is adequate.

In children, whenever performing an operation for SV it is advisable to take a frozen section biopsy of the distal colon for ganglion cells to exclude HD.

References

1. Ciardimi A, Vizzoni L, Dimelli P (1977) Il volvolo del sigma in eta infantile. *Osp Ital Ped* 12: 663-675
2. Dean GO, Murry JW (1952) Volvulus of the sigmoid colon. *Ann Surg* 135: 830-840
3. McCalla TH, Arensman RM, Falterman KW (1985) Sigmoid volvulus in children. *Am Surg* 9: 514-519
4. Shepherd JJ (1969) The epidemiology and clinical presentation of sigmoid volvulus. *Br J Surg* 56: 353-359
5. Valla JS, Louis D, Berard J, et al (1982) Volvulus du sigmoide chez l'enfant. *Chir Pediatr* 23: 93-96