Free perforation in regional enteritis

K. NASR, D. A. MOROWITZ, J. G. D. ANDERSON, AND J. B. KIRSNER

From the University of Chicago, and Billings Hospital, Chicago, Illinois, USA

The inflammatory reaction in regional enteritis is characteristically transmural, with affected loops of intestine frequently becoming attached to adjacent structures. As a result, deep ulcerations and fissures commonly lead to fistula formation rather than to free perforation into the peritoneal cavity. Over the past three years, we have encountered five cases of regional enteritis with free perforation. These cases and a review of the literature are presented in an attempt to clarify the pertinent factors leading to this complication.

CASE REPORTS

CASE 1 (89-26-98) This 11-year-old white girl was admitted to the University of Chicago Hospital with a two-year history of lower abdominal cramps and bloody diarrhoea. Barium x-ray studies revealed regional enteritis involving the distal ileum, sigmoid colon, and rectum. While on intravenous fluids, antibiotics, and corticosteroids, she experienced recurrent episodes of partial small intestinal obstruction. With sudden worsening of her symptoms, a laparotomy was performed, revealing diffuse peritonitis (mixed Gram-negative organisms). She underwent a resection of 29 cm of terminal ileum and adjacent 5 cm of ascending colon, creating a diverting ileocolostomy. Pathological examination revealed severe regional enteritis with superficial and deep ulcerations of the entire terminal ileum. A small perforation was found in the diseased ileum, proximal to a markedly thickened distal segment and 8 cm from the ileocaecal valve. Two months after the resection, the ileocolostomy was closed. Six months later, there was radiological evidence of disease in the distal 15 cm of ileum. Over the next two years, she has continued to present evidence of disease in both the distal ileum, the sigmoid, and the rectum. Though she has remained asymptomatic, she has failed to grow.

CASE 2 (79-82-58) This 23-year-old white man was seen at the University of Chicago Hospital with a two-year history of abdominal pain and diarrhoea. Barium x-ray studies revealed regional enteritis involving the terminal ileum. Treatment included a bland diet, phenobarbital, belladonna, and azulphidine. Symptoms continued and the patient subsequently was admitted to hospital on two occasions with partial small intestinal obstruction. On the second admission to hospital, attempts at intestinal decompression were unsuccessful; previously active bowel sounds disappeared and a plain film of the abdo-

men revealed free air under both diaphragms. Laparotomy revealed diffuse peritonitis (*E. coli*). Resection of 48 cm of terminal ileum and 9 cm of ascending colon was accompanied by an ileocolostomy. Though the pathological examination confirmed the diagnosis of regional enteritis with moderate constriction of the lumen, no overt perforation could be found. During the five months following resection, the patient has continued well.

CASE 3 (96-70-94) This 50-year-old white woman was admitted to the University of Chicago Hospital with a one-year history of abdominal pain, diarrhoea, and weight loss increasing in severity and accompanied by fever for the six weeks preceding her admission. Barium studies revealed regional enteritis involving the terminal ileum and the proximal ascending colon. She was treated with antibiotics and corticosteroids. Four days after admission, she developed increasing abdominal pain and distention with active bowel sounds. Abdominal radiographs demonstrated dilated loops of small intestine suggestive of partial obstruction. Four days later, following an unsuccessful attempt at decompression, the pain worsened suddenly and was accompanied by hypoactive bowel sounds. Abdominal radiographs showed free air under both diaphragms. Peritonitis (*Proteus morgani*) and a perforation of the diseased ileum (25 cm from the ileocaecal valve) were found at laparotomy. She underwent resection of 131 cm of ileum and 11 cm of adjacent ascending colon with an end-to-end ileocolic anastomosis. Pathological examination confirmed the diagnosis of regional enteritis with very extensive ulcerations and an area of constriction distal to the perforation. She has been well over the three months following resection.

CASE 4 (76-43-11) In September 1960 this 65-year-old white man was diagnosed as having regional enteritis involving the distal ileum. Subsequently, in January 1961, he underwent a resection of 76 cm of terminal ileum with an end-to-side ileotransverse colostomy, leaving an ileal stump, caecum, and the colon proximal to the point of the anastomosis as a blind loop. Pathological examination confirmed the diagnosis of regional enteritis. Five months postoperatively, with recurrent symptoms, he was found to have recurrent disease in the 30 cm of the ileum proximal to the ileotransverse colostomy. Despite corticosteroids, the abdominal symptoms continued and increased a few days before his final admission in May 1962. On arrival at the hospital, he was febrile and hypotensive. Increased steroids and antibiotics failed to alter his condition and he died several hours after admission. Necropsy confirmed the presence of peritonitis with

several small perforations of the ileum proximal to a thickened and narrowed distal ileal segment of the ileo-transverse colostomy. The ileum was severely ulcerated for 35 cm proximal to the anastomosis. The proximal colon was normal while the colon distal to the anastomosis was diseased.

CASE 5 (80-21-43) At age 26, this 31-year-old white man was diagnosed as having regional enteritis involving the terminal ileum and ascending colon. With the development of pneumaturia, he had resection of an ileovesical fistula along with 100 cm of diseased ileum

and 15 cm of ascending colon. He did well for four years, but with a recurrence of disease, not responsive to corticosteroids, and the development of partial intestinal obstruction, he underwent a bypassing ileotransverse colostomy. With continuing symptoms, repeat barium studies showed involvement of the ileum proximal to the ileotransverse colostomy. Because of abdominal pain and distension accompanied by active intestinal bleeding and loss of bowel sounds, the patient was transferred to the University of Chicago Hospital. An abdominal radiograph showed air under both diaphragms. Laparotomy revealed peritonitis and perforations in the

TABLE I
REVIEWED CASES OF FREE PERFORATION IN REGIONAL ENTERITIS

Author	Duration of Symptoms	Area of Involvement	Area of Perforation	Steroids	Therapy, Follow up, Comments
Armitage and Wilson (1950)	10 yr	Ileum	Ileum	No information	Died, information incomplete
Crohn (1965)	Acute	Ileum	Ileum	No	Resection, no follow up
(6 cases)	Acute	Ileum	Ileum	Unknown	Incomplete information
Cummings (1962)	2½ yr	Ileum	Ileum	No	First operation closure of perforation, 2nd operation resection six weeks; well
Garner <i>et al</i> (1951)	1½ yr	Jejunum	Jejunum	No	Closure of perforation and decompression, within six months recurrent symptoms
Gow and Walsh (1962)	Acute	Ileum	Ileum	No	Closure of perforation, died six days postoperatively with second perforation
	2 yr	Ileum	Ileum	No	Closure of perforation, five days postoperatively, second perforation, second operation bypass; died 12 hr postoperatively
Greene <i>et al</i> (1962)	Acute	Ileum	Ileum	No	Resection eight years; well
Halligan and Halligan (1937)	Acute	Ileum	Ileum	No	Closure of perforation, six months; well
Harjola <i>et al</i> (1965)	2 yr	Jejunum, ileum	Jejunum	Yes 3 weeks	Resection; died one hour postoperatively
	1 month	Ileum	Ileum	No	Resection one month; well
Homb (1946)	Unknown	Jejunum	Jejunum	No	Closure of perforation; died three days postoperatively
Horn and Rhoads (1944)	3 yr	Ileum, caecum, Meckel's diverticulum	Meckel's diverticulum	No	Resection; well postoperatively
Hurwitz and Zovickian (1952)	3½ yr	Jejunum, Ileum	Not found	No	First operation resection, second operation resection and ileostomy, four years; well
	2 yr	Ileum, caecum	Ileum	No	First operation closure of perforation and proximal ileostomy, second operation bypass three months; well
Lavelle (1964)	4 yr	Jejunum, ileum, ascending colon	Perforation found, exact site unknown	No	Closure of perforation; died 42 hours postoperatively
Linden (1958)	Acute	Ileum	Ileum	No	Closure of perforation three months; well
	Acute	Ileum	Ileum	No	Closure of perforation; no follow up
Menin and Behrend (1959)	1½ yr	Ileum	Ileum	No	Resection several months; well
Milwidsky and Barzilay (1954)	8 yr	Ileum	Ileum	No	First operation decompression, second operation bypass one year
Neely (1960)	1 yr	Ileum	Ileum	No	First operation drainage, second operation resection; no follow up
	Acute	Ileum	Ileum	No	Closure of perforation and bypass one month; well
	Acute	Ileum	Ileum	No	First operation resection with double enterostomy; second operation ileostomy; no follow up
Rappaport <i>et al</i> (1951)	Several months	Ileum	Ileum	No	Resection; no follow up
	Acute	Ileum	Ileum	No	Exteriorization of ileum; died
	4 yr	Jejunum, ileum, caecum	Ileojejunosomy	No	Resection and bypass; died
Robinson and Craft (1967)	Several yr	Ileum	Ileum	No	Resection 14 months; well
	7 yr	Ileum	Ileum	No	Resection five months; well
Rosenberg <i>et al</i> (1963)	Acute	Jejunum	Jejunum	No	First operation drainage, second operation resection; died eight days postoperatively
Sewell (1967)	3 mth	Ileum	Ileum	No	Resection six months; well
Wayne and Lithgow (1967)	2 yr	Ileum	Bypassed ileum	Yes	Resection, died, leakage at suture line

diseased ileum. He underwent resection of 103 cm of ileum and the colon to a point 5 cm proximal to the splenic flexure with the creation of a double enterostomy. Pathological examination confirmed the diagnosis of regional enteritis with many ulcerations and six perforations in the bypassed ileal loop. Eight months post-operatively, with considerable clinical improvement, the patient subsequently underwent re-anastomosis of the ileum to the colon.

DISCUSSION

Though the incidence of free perforation in regional enteritis remains unknown, the association is not infrequent. To 36 cases from the literature, we have added five patients seen by us in the past three years, no attempt having been made to review our entire patient population with regional enteritis.

The review of the literature (Table I) indicates the high frequency of acute regional enteritis in cases presenting with free perforation. Though none of the reviewed cases was definitively diagnosed as having obstruction, 10 cases were incompletely reported and 21 patients were seen after having already perforated. Our five patients emphasize the importance of obstruction as well as fulminant disease as contributory factors. All perforations occurred in the diseased segment, usually the ileum. In the presence of obstruction, the perforations always were found proximal to the diseased and narrowed bowel. Though four of five patients were receiving corticosteroids at the time of perforation, the finding that in 34 of the 36 reviewed cases corticosteroids were not being used suggests that steroids are not a major predisposing factor.

From these observations, it appears that free perforation would occur in two sets of circumstances. First, acute or fulminant disease, in which a channel rapidly forms between mucosa and serosa but is not protected by adherent omentum or bowel. Secondly, there could exist pressure changes, such as accompany intestinal obstruction, which would lead to free perforation in the areas of disease, there not being sufficient time for the development of the protective mechanisms which lead to the formation of abscesses and fistula. It could be predicted that the emphasis on the medical therapy of regional enteritis will lead to more frequent obstructing

lesions and perhaps more frequent free perforations. With a 20 to 25% mortality, free perforation should be considered a serious potential hazard of partial intestinal obstruction in regional enteritis.

SUMMARY

Five cases of free perforation with regional enteritis have been added to 36 already noted in the literature. Factors predisposing to free perforation include acute or fulminant disease as well as partial small intestinal obstruction.

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