Short reports

Crohn's disease presenting with acute colonic perforation

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Summary

Crohn's disease is well known for its multiple intestinal complications. The chronic nature and fibrous inflammation which invariably accompanies this disease explains the relative rarity of free intestinal perforation. The true incidence of free bowel perforation is difficult to assess but 1–2% is the anticipated occurrence during the course of illness. We present a case of Crohn's disease which presented for the first time with acute colonic perforation, an extremely rare entity, and review the current world literature.

Keywords: Crohn's disease, colonic perforation

A 30-year-old man suffering from Becker's muscular dystrophy and mental retardation, presented with sudden-onset generalised abdominal pain and vomiting. He was afebrile but tachycardic. His abdomen was not distended but there was generalised tenderness with mild guarding in the upper abdomen. Bowel sounds on presentation were normal. Routine blood investigations revealed a white cell count of $14.7 \times 10^9/l$ and normal serum amylase; the initial abdominal X-ray was reported as having normal gas pattern (figure 1).

The patient subsequently became pyrexial with worsening abdominal pain and passage of loose blood-stained stools. After a few hours he suddenly became hypotensive and the subsequent chest X-ray demonstrated free gas under the diaphragm (figure 2). Prompt laparotomy revealed free perforation at the splenic flexure, infarction of the adjacent colon and gross faecal peritonitis. The rest of the large and small bowel was entirely normal. A left hemicolectomy was performed removing the perforated segment and the adjacent oedematous bowel. The viable end of the transverse colon was brought out in the right upper quadrant as an end colostomy. The patient underwent a stormy postoperative period which eventually terminated in multiple organ failure and death.

The excised colon revealed longitudinal mucosal ulceration and the histology confirmed extensive ulceration of aphthoid and fissuring type on a background of diffuse inflammatory infiltrate affecting the length of the bowel excised and which was transmural in nature. Numerous discrete non-caseating granulomas were present throughout the bowel

wall and regional lymph nodes. The overall feature was of Crohn's disease.

Discussion

Crohn's disease is characterised by a chronic course and a transmural inflammation. The accompanying fibrous reaction and adhesion to adjacent viscera appears to limit the complication of free perforation. Although the true incidence of free perforation is difficult to assess³ due to inclusion of reports of perforated intra-abdominal abscesses, incomplete reports, and unreported cases, it is generally accepted that 1–2% of patients¹ with Crohn's disease will present as free perforation.

The majority of the cases cited in the literature refer to small bowel perforation; free colonic perforation is considered to be an extremely rare entity.³ A review of the English literature reveals 39 fully described cases of free colonic perforation.^{2–4} Three additional cases of free colonic perforation have been cited in a study dealing with the results of surgery for acute Crohn's colitis⁵ while another study concerned with the extent of colectomy

Figure 1 Abdominal X-ray on admission

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Figure 2 Subsequent chest X-ray showing pneumoperitoneum

Clinical manifestations

- *intestinal*: abdominal pain, diarrhoea, intestinal obstruction, abscess, fistula formation (entero-enteric, enterocutaneous), anal diseases (fissure, fistula, skin tags)
- *liver*: cholangitis, cirrhosis, cholangiocarcinoma
- *joints*: arthritis, sacro-ileitis/ankylosing spondylitis
- *skin*: erythema nodosum
- *cardiovascular*: pericarditis, Takayasu's disease
- *pulmonary*: bronchiectasis, fibrosing alveolitis
- *kidneys*: renal calculi, pyelonephritis, obstructive uropathy
- *eyes*: uveitis, episcleritis

Box 1

in large bowel Crohn's disease mentioned six other cases.⁶

Bowel perforation generally occurs during the course of the illness in a patient with established Crohn's disease⁷ and is frequently observed in association with toxic dilatation. In the absence of toxic dilatation many reasons have been advanced to explain the occurrence of free perforation. The ulcer/fissure does not, on occasions, penetrate close enough to the serosal surface to invite the usual adhesive

Learning points

- free intestinal perforation can occur at any stage of Crohn's disease
- free perforation can occur in the absence of toxic dilatation, concomitant steroid therapy or distal obstruction
- in the presence of an unexplained intestinal perforation, specific inspection of the bowel should be made to look for macroscopic signs of Crohn's disease
- in the initial stages, perforation of the splenic flexure may not produce the typical signs of peritonitis

Box 2

peritoneal inflammatory reaction. In acute Crohn's disease the lack of fibrosis and adhesions facilitate free perforation. The inflammatory process of the acute disease may bring about an obliterative inflammation of the arteries, causing a thrombosis of intramural vessels and resulting in infarction and perforation. Distal obstruction by increasing the intraluminal pressure and steroid therapy by diminishing the immune response have also been proposed as possible mechanisms for precipitating bowel perforation.

Our patient was not known to have Crohn's disease. To our knowledge, Crohn's disease presenting for the first time with free colonic perforation is a rare occurrence and only five such well-described cases have been reported previously.^{2,3,8,9} Interestingly, there was no evidence of toxic dilatation or distal stenosis in our patient, who had not received any steroids after his admission to the hospital. In the previously mentioned five cases, only two patients had toxic dilatation⁸ and none of them had distal obstruction.

Our case serves to illustrate the protean manifestations of Crohn's disease. These are, however, best exemplified by the experience of Crohn himself. In 1957 he stated that 'free perforation of ileitis into the peritoneal cavity never occurs or at least I have not seen it' while in 1965 he reported seven cases of free perforation.¹⁰ Be that as it may, this case lends further credence to the belief that colonic perforation can occur in any age group, at any stage of the disease, and at any site of the intestinal tract.²

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