A Rare Presentation of Crohn's Disease

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Abstract:
Free peritoneal perforation is a rare complication of Crohn’s disease with a report of only 100 cases in the literature. It needs an emergency exploration and an unaware general surgeon is confounded in intraoperative decision-making. We present our experience when this rarity struck us in a district hospital and briefly review the guidelines of optimal management of this complication of Crohn’s disease.

Key Words: Free perforation, Crohn’s disease, Ileal perforation

Case Report:
A 19-year old girl was referred to our hospital for management of acute abdomen. She had a history of colicky pain abdomen with distension and vomiting for four months. All these complaints became more severe for the past 3 days. Vomiting was bilious and associated with decreased urine output. Her past and personal histories were not contributory. Family history was positive for tuberculosis. On examination, she was malnourished, dehydrated and pale. BP was 100/70 and pulse rate was 110 per minute. Abdomen was distended and warm with loss of bowel sounds.

An erect x-ray abdomen was taken which showed gas under diaphragm with dilated bowel loops. She underwent exploratory laparotomy under general anesthesia. Peritoneal fluid was feculent. Entire length of the bowel was inspected and findings accurately recorded. Multiple strictures and 3 perforations (Figure 1 and Figure 2) were present in the distal ileum all within 60 cm from ileocecal junction. Ileal mesentery was thick and friable with no palpable adenopathy. Large bowel and rest of the small bowel was grossly normal. Around 70 cm of ileum was resected and a hand-sewn ileo-ileal anastomosis was performed in two layers. Patient had an uneventful postoperative recovery.

Histopathological examination of the resected ileum showed Crohn’s disease. After postoperative recovery, the medical gastroenterology department was involved in her care. Immunosuppression (6-Mercaptopurine, 1.5mg/kg) was started. The patient is currently on follow-up with no clinical evidence of disease recurrence.

Discussion:
Crohn’s disease (CD) is a chronic relapsing and remitting inflammation of the bowel involving all its layers and characterized by noncaseating granulomas. It has an unpredictable course and surgical management is challenging. Majority of patients with CD require surgery as time progresses, such that 78% and 90% undergo surgery after 20 years and 30 years of symptoms, respectively.(1) The most common indication for surgery in case of small bowel CD is obstruction due to a terminal ileal stricture while perforation, fistulae and abscess formation account for the remaining. The indications for surgery in Crohn’s colitis include abscesses (25%), perianal disease (23%), failure to thrive (21%), toxic megacolon (19%) and intestinal obstruction (12%).(2) Failure of medical management in the form of persisting symptoms despite aggressive therapy, recurring symptoms on tapering and worsening symptoms during therapy is another indication for surgical intervention.

Though contained ileal perforation with septic features and inflammatory abdominal mass is common, free perforation into the peritoneal cavity with generalized peritonitis is a rare presenting feature of Crohn’s disease with an incidence of 1-3% in Western countries.(3) Only 100 cases have been reported in literature.(4) In patients with intra-abdominal sep-
sis, a delay in surgery for presurgical nutritional restoration may be detrimental, especially because the benefit of presurgical nutrition is unproven in prospective studies.(5) Therefore an emergency surgery is warranted.

Resection is the procedure of choice for Crohn's ileitis. Most commonly, the disease involves the terminal ileum and caecum requiring ileocecal resection. When caecum is spared and adequate length of healthy ileum, approximately 7-10cm proximal to ileocecal valve remains, an ileal resection with end-to-end ileal anastomosis can be performed. This preserves the ileocecal valve and helps to minimize diarrhoea post-surgically.(6)

A study by Fazio et al (7) resolved the controversy regarding optimal margin width at resection of Crohn's disease. It showed that large margins are not beneficial and there was no statistically significant difference in recurrence rates between patients with histologically involved and uninvolved margins. However, the surgeon has to choose the line of resection based on intraoperative assessment of extent of disease. Proximal level of involvement is determined by palpation of mesenteric border of the bowel. In involved areas, the phenomenon of fat-wrapping (or fat hypertrophy), a feature peculiar to Crohn's disease (8) will obscure the bowel wall, which becomes palpable when normal intestine is reached. A few centimeters away from this point will be an appropriate line of resection and small aphthous ulcers seen inside the bowel lumen shall not require further extension of resection.(6)

The technique of intestinal anastomosis in Crohn's disease has been a matter of debate for quite sometime. A longer side-to-side anastomosis may be beneficial over an end-to-end anastomosis as it is believed that a large caliber anastomosis will take long time to produce stenosing obstruction. However, published data to date does not show superiority of one particular type of intestinal anastomosis.(9) Anastomosis can be hand-sewn or performed with a stapler. Under appropriate conditions like minimal peritoneal contamination in a patient with hemodynamic stability and good nutrition, resection and primary anastomosis is a safe procedure with a leak rate of less than 1 percent. (10) Nevertheless, in high surgical risk candidates and patients with free colonic perforation or fulminant peritonitis, resection with proximal ileostomy is the treatment of choice.(11) Primary closure of the perforation is absolutely contraindicated under any circumstances.

As no single surgery for Crohn's disease is curative, these patients always have a very high risk of recurrence, the prevention of which should be in the mind of every surgeon operating on Crohn's disease. Young age, short disease duration and perforating disease are risk factors for early postoperative recurrence.(12) Unfortunately, our patient had all these risk factors. There is evidence to suggest the use of some types of medical therapy to prevent both endoscopic and symptomatic recurrence of Crohn's disease after surgery. Besides being inappropriate for maintenance therapy in nonsurgical Crohn's patients, steroids have proved inefficient for prophylaxis against disease recurrence. Although there is evidence and support for the efficacy of other 5-acylsalicylate preparations in the maintenance of postsurgical remission, the overall beneficial effect of mesalamine is small. Only a modest benefit has been shown with azathioprine and 6-Mercaptopurine. But since there is much stronger evidence supporting their use in maintenance therapy after medically-induced remission, their use is probably justified in high-risk post-operative patients.(13) Finally, smoking cessation is strongly advocated in all post-operative patients as it has been found that smokers have double the rate of recurrence.(13)

We wish that the above-mentioned facts will be helpful for surgeons in intraoperative decision making when such a rare cause of ileal perforation is encountered. We conclude by stating that a team with a conservative surgeon and an aggressive physician will be ideal for optimal management of Crohn's disease.

References:


