IMJ Official Journal of the Irish Medical Organisation

Ir Med J; March 2023; Vol 116; No. 3; P748

March 2023

Crohn's disease presenting as acute bowel perforation and superior mesenteric vein thrombosis

M. Lefter[1], E. Heffernan[2], N. Nolan[3], A. Hanly[4]

1. School of Medicine. University College Dublin, Belfield, Dublin 4.
2. Department of Radiology, St. Vincent's University Hospital, Elm Park, Dublin 4.
3. Department of Pathology, St. Vincent's University Hospital, Elm Park, Dublin 4.
4. Department of Colorectal surgery, St. Vincent's University Hospital, Elm Park, Dublin 4.

Abstract

Presentation:
A 51-year-old male presented with acute severe abdominal pain and rapidly developing septic shock.

Diagnosis:
Pneumoperitoneum was revealed on the CXR. Subsequent CT abdomen demonstrated hollow viscus perforation, features of chronic bowel inflammation and chronic superior mesenteric vein thrombosis with collaterals.

Treatment:
He underwent a small bowel resection of necrotic perforated jejunum and end-jejunostomy formation. Histology revealed features in keeping with Crohn's disease (CD). Two years post-resection, he underwent a jejunostomy reversal and is due to commence Vedolizumab for his CD.

Discussion:
This case highlights the need for a high index of clinical suspicion for rare but life-threatening presentations of CD, even in the absence of a prior history of CD.

Introduction

Crohn's disease (CD) can affect any part of the gastrointestinal tract from mouth to anus, and is characterized by transmural inflammation. It can present with a myriad of clinical manifestations at initial presentation and over the ensuing years. Free intestinal perforation of the small bowel is uncommon in CD with an incidence of 1-3%.[1] The consequences of such perforations can be life-threatening; hence, it is critical that such are diagnosed early. We present a case of a rare first presentation of CD, manifesting with a free small bowel perforation with consequent septic shock.

Case Report

A 51-year-old male presented to the Emergency Department (ED) with a 12-hour history of sudden onset, progressive severe abdominal pain, radiating to the right shoulder tip, associated with

vomiting, dry retching, and movement intolerance. He had no diarrhoea, blood per rectum, or fever. Apart from irritable bowel syndrome symptoms for 10 years, he had no medical/surgical history and was on no medications. On arrival to ED, he appeared acutely unwell and pale. Vital signs: BP 112/78, regular pulse 98 beats/minute, temperature 35.4^{0}C, respiratory rate 18/min, oxygen saturation 96%. He had upper abdominal guarding and tenderness. CXR revealed a pneumoperitoneum (Figure 1). Laboratory studies: low WCC (1.6) and neutrophils (1.1), high CRP (26.1) and lactate (4.8), indicating sepsis.

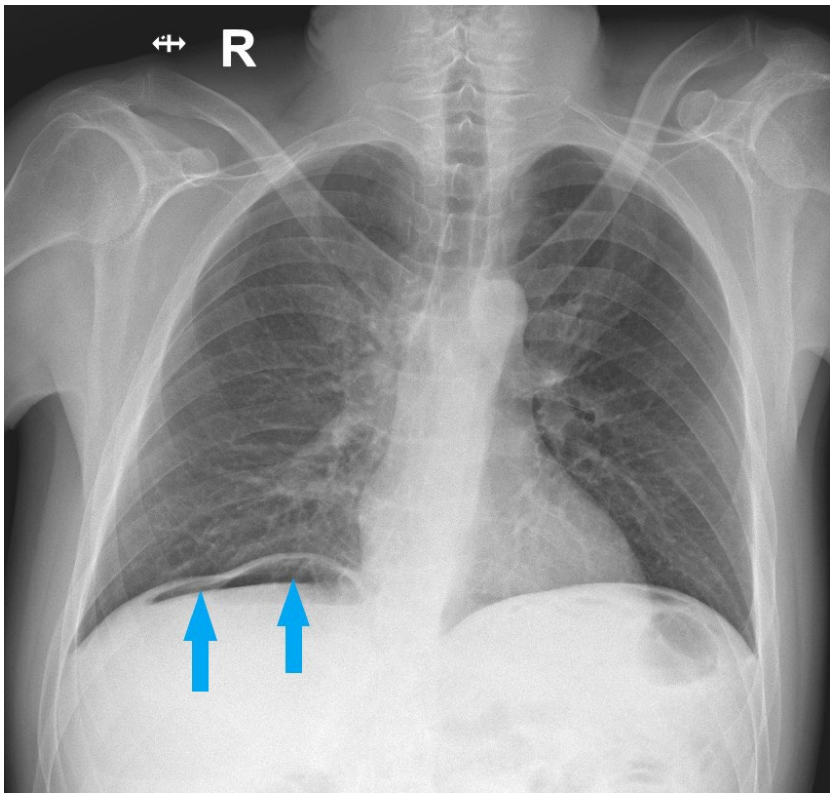

Figure 1: Frontal erect chest radiograph showed a moderate volume pneumoperitoneum with free gas below the right hemidiaphragm (arrows).

Computed Tomography (CT) imaging showed features of hollow viscus perforation, mural thickening of the proximal jejunum, consistent with an inflammatory process (Figure 2) and chronic superior mesenteric vein (SMV) thrombosis with collaterals, possibly due to chronic CD.

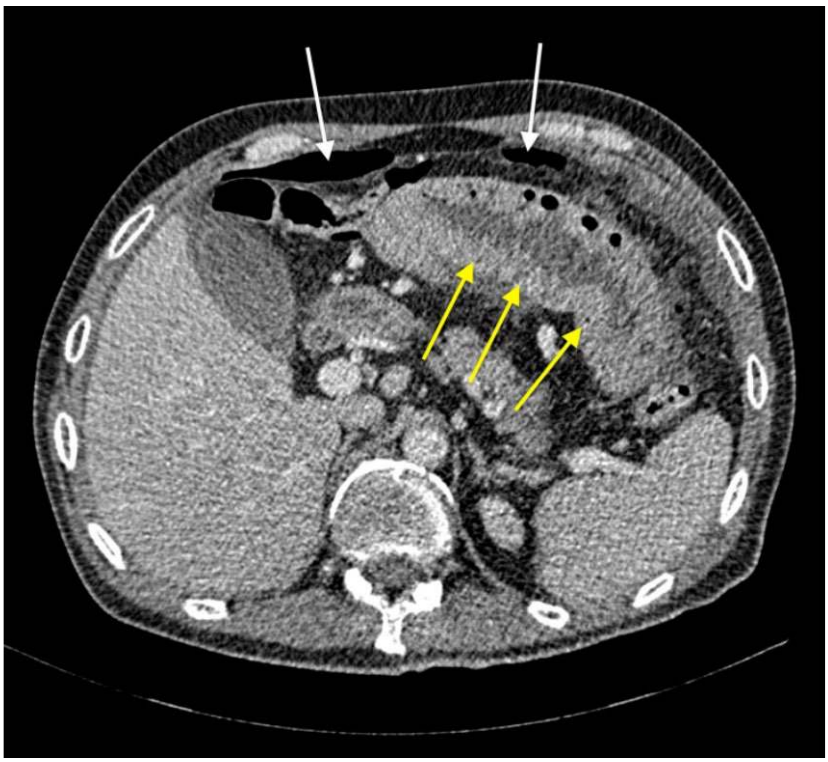

Figure 2: Transverse CT image shows free intraperitoneal gas in the upper abdomen consistent with hollow viscus perforation (white arrows). There is a segment of proximal jejunum which demonstrates marked mural thickening (yellow arrows), consistent with an inflammatory process.

Within the first ninety minutes, he deteriorated rapidly with hypotension (95/67 mmHg) and increasing lactic acidosis (lactate 7 and pH 7.26). Despite sepsis resuscitation, he remained hypotensive and became hypothermic (temperature 35.2^{0}C).

An emergency laparotomy with 7cm small bowel resection of necrotic perforated jejunum and end-jejunostomy formation was performed. The surgery was difficult due to four quadrant faeculent peritonitis, SMV thrombosis and a short mesentery which resulted in venous engorgement. Intraoperative findings included diseased distal small bowel, fat encroachment and multiple patent strictures, indicative of CD. Resected small bowel histology is illustrated in Figure 3.

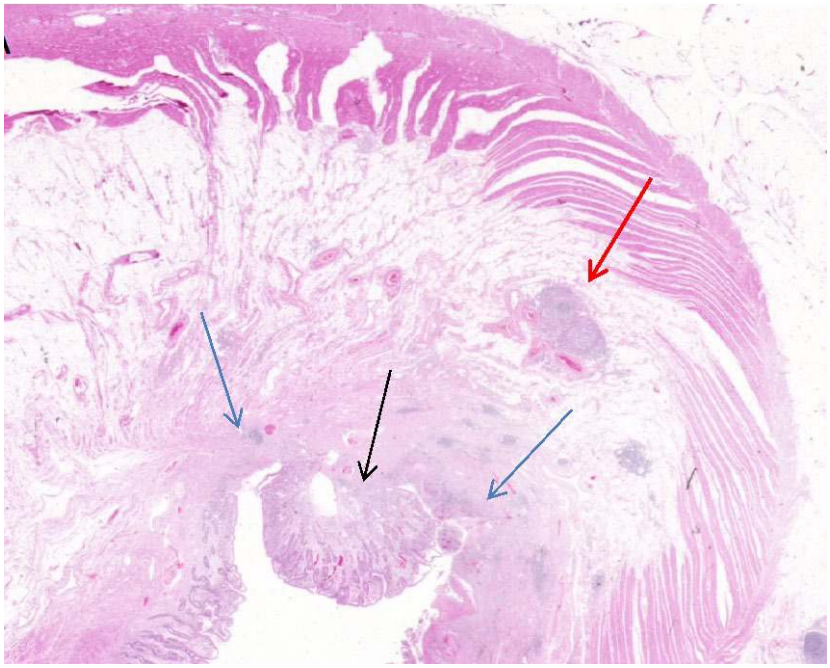

Figure 3: Low power histology section from the patient's resected small bowel specimen demonstrating ulcers and fissuring (blue arrows), inflammatory pseudopolyp (black arrow) and granuloma (red arrow).

The patient had a prolonged postoperative ICU stay for several weeks due to difficult to control sepsis, associated with delirium. Two years post-resection, he successfully underwent a jejunostomy reversal, is asymptomatic from a luminal perspective and is due to commence Vedolizumab for his CD.

Discussion

CD is an immune-mediated relapsing-remitting inflammatory bowel disease (IBD), affecting any part of the gastrointestinal tract with transmural inflammation. Granulomas, the hallmark of CD, are found in 40-60% of resected bowel specimens and suggest a more aggressive form of IBD[2].

IBD, while a chronic condition, can present as acute surgical emergencies e.g., toxic colitis, haemorrhage, intra-abdominal masses/abscesses, intestinal obstruction, perforation[3]. Microperforations and frank free perforation both occur in CD, with the latter being more uncommon (1-3%) and requiring emergency operation to reduce mortality[1, 3-5], as in our case.

Our patient had pneumoperitoneum on CXR, seen in only 20% of patients with intestinal perforation from CD[1]. Therefore, a keen awareness of bowel perforation needs to be maintained even in the absence of pneumoperitoneum.

The risk of thromboembolism is three times higher in patients with IBD[6].Our patient had SMV thrombosis with collaterals indicating chronicity, a rare thromboembolic complication of CD (1.7% cases), often associated with a severe and complex clinical course[7-9].

In summary, this case highlights a number of important points. Even at first presentation, CD can cause free bowel perforations and overwhelming sepsis, representing a life-threatening emergency.

Furthermore, CD is associated with an increased thromboembolic risk, as seen in our case. A high index of clinical suspicion for acute presentations of CD, even with no prior history of CD, is the key take-home point.

Conflicts of Interest:
None declared.

Corresponding author:
Madalina Lefter,
University College Dublin,
School of Medicine,
Dublin 4,
Ireland.

E-Mail: Madalina.lefter@ucdconnect.ie

References:

1. Leal RF, Ward M, Ayrizono Mde L, et al. Free peritoneal perforation in a patient with Crohn's disease - Report of a case. Int J Surg Case Rep 2013;4:322-324.
2. Molnár T, Tiszlavicz L, Gyulai C, Nagy F, Lonovics J. Clinical significance of granuloma in Crohn's disease. World J Gastroenterol 2005;11:3118-3121.
3. Berg DF, Bahadursingh AM, Kaminski DL, Longo WE. Acute surgical emergencies in inflammatory bowel disease. Am J Surg 2002;184:45-51.
4. Ishihara S, Watanabe T, Nagawa H. Free Colonic Perforation in a Patient With Crohn's Disease and Loop Ileostomy: Report of a Case. International Surgery 2011;96:159-161.
5. Chaikof EL. Nontraumatic perforation of the small bowel. The American Journal of Surgery 1987;153:355-358.
6. Papa A, Gerardi V, Marzo M, Felice C, Rapaccini GL, Gasbarrini A. Venous thromboembolism in patients with inflammatory bowel disease: focus on prevention and treatment. World J Gastroenterol 2014;20:3173-3179.
7. Jackson CS, Fryer J, Danese S, Vanagunas A, Polensky S, Buchman AL. Mesenteric vascular thromboembolism in inflammatory bowel disease: a single center experience. J Gastrointest Surg 2011;15:97-100.
8. Naïk Vietti N, Vietti Violi N, Schoepfer AM, et al. Prevalence and clinical importance of mesenteric venous thrombosis in the Swiss Inflammatory Bowel Disease Cohort. AJR Am J Roentgenol 2014;203:62-69.
9. Kopylov U, Amitai MM, Lubetsky A, Eliakim R, Chowers Y, Ben-Horin S. Clinical and radiographic presentation of superior mesenteric vein thrombosis in Crohn's disease: a single center experience. J Crohns Colitis 2012;6:543-549.