OPEN ACCESS

Bowel perforation in chronic idiopathic megarectum and megacolon

Chukwuebuka Anyaegbuna, Alexandros Apostolopoulos, Hitesh Patel

General Surgery, Newham University Hospital, London, UK

Correspondence to
Dr Alexandros Apostolopoulos, Alexandros.Apostolopoulos@bartshealth.nhs.uk

Accepted 30 May 2018

DESCRIPTION

A 24-year-old man with autism spectrum disorder presented to accident and emergency complaining of a 1-week history of abdominal pain and diarrhoea. He had a history of chronic constipation with multiple previous admissions stretching 4 years back. On examination, he had a tender, distended abdomen with quiet bowel sounds. His CT scan on admission (figure 1) showed a dilated (up to 18 cm) rectum and sigmoid colon filled with faeces. The patient was managed conservatively with oral laxatives and regular enemas. However, the patient refused the enemas and was maintained on oral laxatives. Two days later, the patient complained of worsening abdominal pain. On examination, he had a peritonitic abdomen and reduced consciousness level. Biochemical investigations revealed worsening renal function and that the patient was now acidotic. A chest X-ray revealed free air beneath the diaphragm. A repeat CT scan (figures 2 and 3) showed a substantial perforation of the dilated sigmoid colon. The patient was sent to intensive care unit before undergoing an emergency laparotomy where a Hartmann procedure was performed.

Megacolon and/or megarectum are rare conditions defined as dilatation of the colon not caused by mechanical obstruction.[1] It is classified by different values depending on the location of the dilatation in the colon. In the rectosigmoid region, a diameter greater than 6.5 cm is used.[2] It can be divided up into acute, chronic or toxic megacolon. Chronic megacolon occurs when this dilatation is permanent secondary to a long-standing disorder of colonic motility. This condition can be congenital, acquired or idiopathic. Congenital megacolon may be caused by genetic abnormalities such as those associated with Hirschsprung's disease or MEN2B.[3] Acquired megacolon is associated with neuropathies, myopathies and connective tissue disease. Idiopathic megacolon are cases of megacolon where no organic cause is found. This patient had chronic, idiopathic megacolon and megarectum. Patients with this condition report recurrent episodes of constipation, abdominal pain, distension and bloating starting in childhood or adolescence. Due to increased intraluminal pressures, these patients are at an increased risk of ulceration of the bowel wall. Hence, they are at an increased risk of perforation. The literature on managing these patients is not consistent and, thus, these patients present a serious management challenge to surgeons.

For most patients with chronic megacolon and constipation, a non-surgical approach can be effective. This includes high colonic water enemas, water-soluble contrast enemas and oral polyethylene glycol-based

Figure 1 Sagittal view of the sigmoid colon filled with faeces.

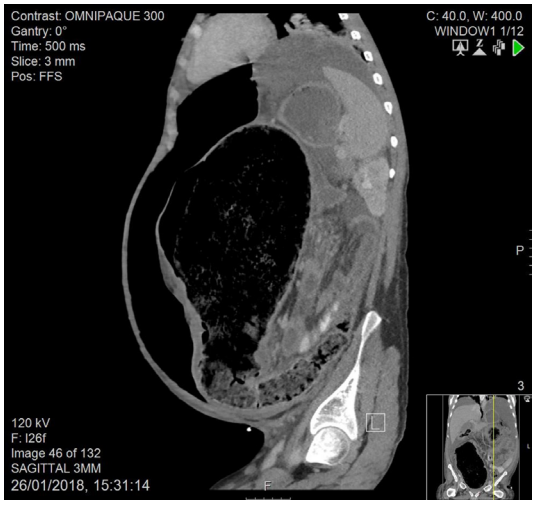

Figure 2 Sagittal view of sigmoid colon post-perforation, with free air in the abdomen.

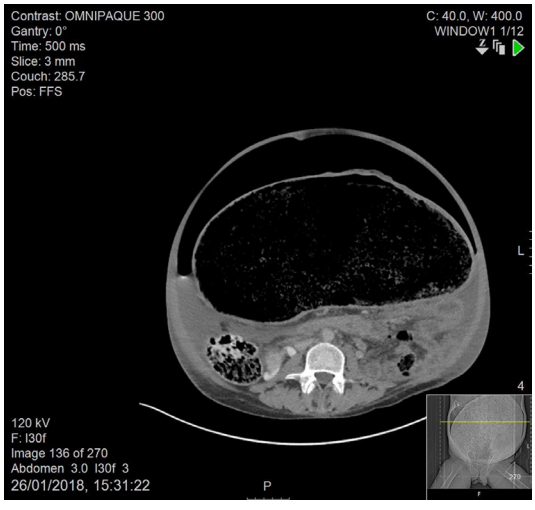

Figure 3 Transverse view of sigmoid colon post-perforation, with free air and faeces in the abdomen.

To cite: Anyaegbuna C, Apostolopoulos A, Patel H. *BMJ Case Rep* Published Online First: [*please include* Day Month Year]. doi:10.1136/bcr-2018-225406

solutions. If these methods succeed, patients can be discharged with dietary advice and a regular colonic regime. However, if these methods fail, it is important to consider early surgical therapy. This includes colectomy with ileoanal anastomosis in those with normal anorectal function and a diverting loop ileostomy in patients with abnormal anorectal function. This is associated with good outcomes in the majority of patients.[2]

In this case, an earlier appreciation during a previous admission that this was a rare presentation of chronic idiopathic megacolon and megarectum may have prevented the perforation and elective surgery could have been planned.

Learning points

- Early recognition of chronic idiopathic megacolon is important.
- Close monitoring of patients with megacolon is essential as these patients are at increased risk of perforation.
- Conservative measures include high colonic water enemas, water-based contrast enemas, oral polyethylene glycol-based solutions and dietary modifications. When conservative measures fail, surgical intervention is associated with good outcomes.

Contributors AA and CA were responsible for planning, conducting, reporting and conception. Design of the manuscript was completed by AA and HP.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

1 Camilleri M. Acute and chronic pseudo-obstruction. In: Feldman M, Friedman LS, Sleisenger MH, eds. *Sleisenger & Fordtran's gastrointestinal and liver disease*. 8th edn. Philadelphia, USA: Saunders, 2007:2679–702.
2 O'Dwyer RH, Acosta A, Camilleri M, *et al*. Clinical features and colonic motor disturbances in chronic megacolon in adults. *Dig Dis Sci* 2015; 60:2398–407.
3 Heuckeroth RO. Hirschsprung disease—integrating basic science and clinical medicine to improve outcomes. *Nat Rev Gastroenterol Hepatol* 2018; 15:152–67.