A case of perforated chronic idiopathic megacolon
Benjamin Cribb, Rukshan Ranjan, Nigel Henderson

A 61-year-old male, with long-standing abdominal distension, presented with generalised abdominal pain, shortness of breath, and marked abdominal distension. The patient reported increasing abdominal distension over a 6-week period with associated infrequent bowel motions. No other significant medical or surgical history of note.

X-ray and CT imaging showed extensive colonic distension and a large volume pneumoperitoneum (Figures 1 and 2). There was also marked elevation of the right hemidiaphragm with associated volume loss of the right lung.

He proceeded acutely to theatre. An attempt at pre-oxygenation prior to induction of anaesthesia resulted in respiratory distress with oxygen desaturation. Needle decompression of the abdomen was required to relieve the tense pneumoperitoneum. At laparotomy, the patient was found to have a massively distended colon (see Figure 3), and a large amount of free intra-peritoneal gas without gross faecal contamination. Colonic micro-perforation was suspected and subtotal colectomy with end ileostomy performed. Histopathological analysis showed stercoral ulceration in the descending colon (the presumed area of micro-perforation). There was no evidence of aganglionosis or other intrinsic neuromuscular abnormality.

The patient had a protracted post-operative course due to respiratory failure, which necessitated tracheostomy insertion and ventilatory assistance. This gradually improved and the patient made a full recovery.

The causes of chronic megacolon can be divided into congenital, acquired and idiopathic causes. In general, chronic megacolon develops from refractory constipation from any cause. Idiopathic megacolon is a rare and poorly understood condition. This condition has many similarities with other conditions associated with chronic megacolon, such as Hirschsprung’s disease, chronic idiopathic intestinal pseudo-obstruction and idiopathic megarectum. However, idiopathic megacolon is unique in that the onset of symptoms can occur in early or late childhood or in adult life. The aetiology of this condition may be different between those patients with an early onset of symptoms and those with a late onset.

The symptoms of idiopathic megacolon also differ between these two groups. Patients with an adult onset do not always complain of constipation, though it is the most common symptom. Abdominal pain, abdominal distension, palpable abdominal mass, irregular bowel habit and sometimes overflow diarrhoea are the other most common symptoms in patients with an adult onset of idiopathic megacolon. Faecal impaction is uncommon.

Pathological features of idiopathic megacolon are thickening of the enteric smooth muscle, but with an intact enteric nervous system. Conservative management of patients with idiopathic megacolon is frequently ineffective. For the majority of patients with an onset of symptoms later in life, medical treatment is unsuccessful.
Surgery is reserved for those who fail conservative management or who develop a complication necessitating urgent surgical intervention, such as perforation or ischemia. The surgical options for idiopathic megacolon in the elective setting have previously been evaluated. Subtotal colectomy with ileorectal anastomosis appears to be the optimum procedure in patients with a non-dilated rectum. In patients with a dilated rectum, restorative proctocolectomy with ileal pouch reconstruction is recommended. However, in the acute setting of perforation there are few surgical options available, other than subtotal colectomy with end ileostomy formation, as performed in this case.

Although faecal impaction with sterile perforation is not uncommon, this case report of perforated idiopathic megacolon is a rare complication of this interesting condition.

**Figure 1:** Axial computed tomography image of the abdomen demonstrating a massive pneumoperitoneum with the abdominal viscera pushed posteriorly. Note the marked wasting of the anterior abdominal wall.

**Figure 2:** Coronal computed tomography image demonstrating the marked colonic distension with associated elevation of the right hemidiaphragm and reduced right lung volume.

**Figure 3:** The resected specimen demonstrating the generalised distended colon.
CLINICAL CORRESPONDENCE

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Author information:
Benjamin Cribb, Department of general surgery, Taranaki Base Hospital, New Plymouth, New Zealand; Rukshan Ranjan, Department of general surgery, Taranaki Base Hospital, New Plymouth, New Zealand; Nigel Henderson, Department of general surgery, Taranaki Base Hospital, New Plymouth, New Zealand.

Corresponding author:
Nigel Henderson, Department of general surgery, Taranaki Base Hospital, New Plymouth, 123 Vivian St, New Plymouth 4310, New Zealand.
Nigel.Henderson@tdhb.org.nz

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