A mystery wrapped in an enigma: the abdominal cocoon syndrome

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Abstract

Abdominal cocoon syndrome is a rare cause of intestinal obstruction that often presents as an incidental finding at surgery, creating a management dilemma for those unfamiliar with its appearance. Surgical excision of the ‘cocoon’ is the mainstay of treatment. This report describes a 42-year-old patient who successfully underwent surgery in which the fibrous peritoneal membrane was dissected free from the serosal surface of the bowel.

Case report

A 42-year-old man, who was a recent immigrant to New Zealand from Tuvalu, was admitted acutely with a 3-day history of central abdominal pain, constipation and 30 kg weight loss. He had a reduced appetite and abdominal distension, but no associated nausea or vomiting. He was passing flatus. There was no history of fever, dysuria, recent diarrhoea, rectal bleeding or previous abdominal surgery. There was no family history of inflammatory bowel disease or bowel cancer. His past medical history included type 2 diabetes mellitus only.

On clinical examination a non-tender mass was palpable in the right iliac fossa, with an empty rectum. Laboratory results revealed a haemoglobin of 170 g/L, white cell count of 13.9 × E9/L and gamma GT of 89 U/L. Abdominal radiograph did not show any dilated bowel loops and chest radiograph was normal.

With a provisional diagnosis of an inflammatory or neoplastic right iliac fossa mass, a computed tomography (CT) scan was performed (Figure 1). This demonstrated no mass lesion, but multiple dilated small bowel loops with interloop free fluid and no clear transition point. On the basis of a presumed diagnosis of small bowel obstruction, the patient proceeded to surgery.

At surgery the small bowel appeared markedly shortened, measuring 80 cm, with a normal-appearing duodenojejunal (D-J) flexure and terminal ileum (Figure 2). The small bowel mesentery demonstrated marked fibrosis and scarring with extension over the serosal surface of the bowel, creating a fibrous ‘cocoon’ around the entire small bowel. The colon and superior liver surface were also covered by a similar thickened fibrous layer. No obstructing point was identified. Peritoneal washings were taken for histology and microbiology.

A decision was made to close the laparotomy and complete autoimmune, microbiological, cytological and endoscopic investigations, all of which were normal. Parenteral nutrition was commenced and a decision made to proceed to re-laparotomy at day 9.
Figure 1. Computed tomography scan demonstrating what appear to be multiple dilated loops of small bowel with no transition point

Figure 2. Entire length of small bowel contained within a fibrous inflammatory ‘cocoon’
At second laparotomy, the membranous cocoon (Figure 3) was incised and dissected free from the serosal surface of the small bowel and mesentery, using sharp dissection, from the D-J flexure to the terminal ileum.

Prophylactic appendicectomy was performed, as described in other similar cases. Histopathology of the excised peritoneal membrane demonstrated fibrosis with increased vascularity, haemosiderin deposition and mild inflammatory cell infiltrate, consistent with idiopathic sclerosing encapsulating peritonitis.

**Figure 3. Fibrous peritoneal membrane dissected free from the serosal surface of the bowel**

Postoperatively the patient required return to theatre on day 2 for intra-abdominal haemorrhage from a small mesenteric arterial vessel. He subsequently made an uneventful recovery and at 3 months postoperatively is now asymptomatic.

**Discussion**

Abdominal cocoon syndrome is a rare condition, in which the bowel and mesentery become encased in a fibrous membrane.\(^1,2\) The aetiology remains uncertain, although peritoneal dialysis, β-blockers, fungal infection, tuberculosis, cirrhosis, retrograde menstruation and gynaecological infection have all been implicated.\(^2,7\) It is more
commonly reported in females and patients are often from sub-tropical or tropical regions.

Two main types of abdominal cocoon syndrome have been described: sclerosing encapsulating peritonitis (SEP)—idiopathic or secondary; and peritoneal encapsulation: the membrane may originate from the yolk sac during return of the organs intra-abdominally during embryological development.\textsuperscript{5,8}

Clinical presentation is often with acute or subacute small bowel obstruction, associated with weight loss, abdominal pain or an abdominal mass. Diagnosis is often made at surgery, although CT imaging may be helpful.\textsuperscript{9}

Division ± excision of the fibrous cocoon appears to lead to successful resolution of symptoms.\textsuperscript{10} However morbidity and mortality related to this procedure is reported as 38–90% and 60–71% respectively.

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