Emergency Surgery for a Ruptured Intra-abdominal Desmoid Tumour

Dear Editor,

Desmoid tumours are fibrous benign tumours that are often indolent until local symptoms evolve.[1] We highlight a rare case of a ruptured intra-abdominal desmoid tumour presenting as acute abdomen.

A 58-year-old female presented to our institution with a 1-day history of abdominal pain and vomiting. This was associated with fever, chills and rigors. On examination, the abdomen was tender in the peri-umbilical area with some guarding. Laboratory investigations were largely unremarkable except for a mildly raised white blood cell count of 10.9 x 10^9.

Computed tomographic (CT) scan of the abdomen was performed and it revealed a mass in the small bowel mesentery with significant ascites (Fig. 1). Our initial impression was either gastrointestinal stromal tumour or mesenteric teratoma.

In view of the persistent abdominal signs, exploratory laparotomy was performed. During the surgery, there was a ruptured tumour in the jejunal mesentery with 1 L of purulent ascites (Fig. 2). The tumour was located 20 to 25 cm from the duodeno-jejunal flexure with surrounding induration. Even though the bowel was not involved, resection of the tumour would result in vascular compromise of the small bowel. Hence, small bowel resection with hand-sewn end-to-end anastomosis was performed.

She was discharged well on the 6th postoperative day. Histological evaluation of the jejunal specimen confirmed the diagnosis of a desmoid tumour. The lesion extended to the resection margin and abutted the serosal surface.

The fundamental principles in managing any patient with life-threatening intra-abdominal septicaemia must be the preservation of life and the containment and eradication of the source of sepsis. In our patient, after commencement of intravenous antibiotics and ample fluid resuscitation, exploratory laparotomy was warranted as the source of the peritonitis was localised to the abdomen as delineated by the CT scan. Copious amount of irrigation was necessary in view of the extensive contamination. It was only after histological evaluation that the perforated tumour was diagnosed as an intra-abdominal desmoid tumour.

Desmoid tumour is a rare lesion representing <3% of all soft tissue tumours with an estimated incidence of 2 to 4 new cases per million per year.[2] Its association with familial adenomatous polyposis and Gardner's syndrome has been well described.[2]

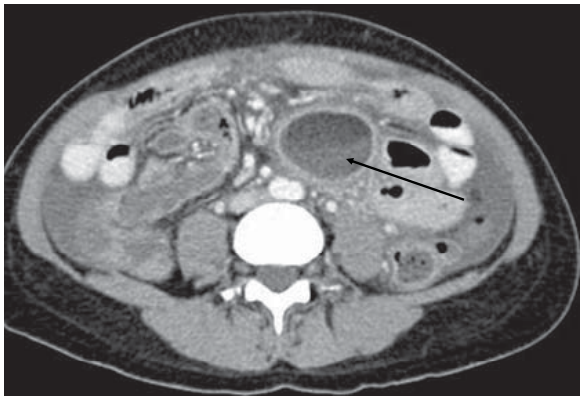

Fig. 1. Computed tomographic (CT) scan showing the cystic lesion in the mesentery of the small bowel (arrow).

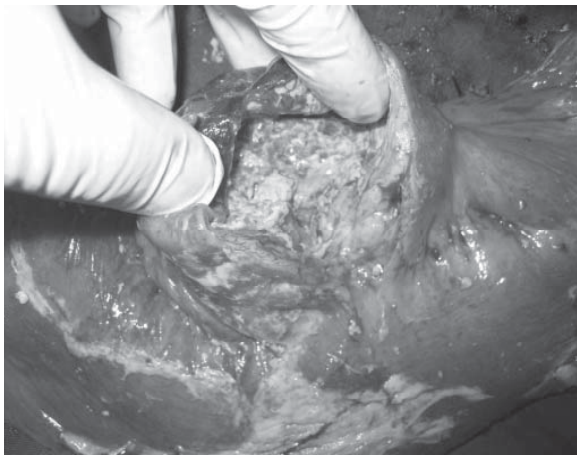

Fig. 2. Image of the resected specimen highlighting the perforated desmoid tumour.

However, the clinical behaviour and natural history of desmoid tumours remain unpredictable. Even though most are indolent or slowly enlarging, the disease could progress rapidly and aggressively in some patients.[2] Some of these complications include intestinal obstruction, ischaemic bowel secondary to vascular compression and hydronephrosis from ureteric compression.

Surgery is often indicated to manage any life-threatening complications such as bowel perforation, significant gastrointestinal haemorrhage and peritonitis,[3] as seen in our patient.

Wide surgical resection is recommended to achieve disease free margins as this has been shown to be associated with a significant reduction in the local recurrence rate.[4] However, some of the intraoperative factors that would determine the

type of operative intervention include the extent of small bowel involvement, adhesions and the involvement of the base of the mesentery.[5] In our patient, the treatment of choice was small bowel resection as resection of the tumour alone would have resulted in significant bowel ischaemia due to its proximity to the mesenteric vessels.

However, the complications of any extensive bowel surgery such as short gut syndrome and long-term parenteral nutrition are significant considerations.[5] If the disease is too extensive and not suitable for complete resection, palliative bypass or resection should be performed. Unfortunately, despite complete resection, recurrences are not uncommon and remain a sizeable problem.[5] Adjuvant chemotherapy has been shown to reduce the risk of tumour recurrence,[5] and is strongly recommended in patients who have widespread intra-abdominal contamination and/or positive resection margin, such as our patient.

Though intra-abdominal desmoid tumour is usually asymptomatic, it can present as a surgical emergency requiring immediate operative intervention.

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