Calcifying fibrous tumour torsion: a rare cause of abdominal pain

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DESCRIPTION

A previously healthy 20-year-old man presented with a 2-day history of right lower quadrant pain radiating into the right groin and testis. He was afebrile with no other associated symptoms. He had no previous operations and no known allergies. On examination, he had right iliac fossa tenderness and a positive Rovsing's sign. Urinalysis was unremarkable. He had a leucocytosis ($14.5 \times 10^9/L$) with neutrophilia ($12.0 \times 10^9/L$), and elevated serum C reactive protein (104 mg/L). Pelvic and scrotal ultrasonography had no significant findings.

He was thus taken for laparoscopic appendectomy for clinical acute appendicitis. On laparoscopy, a mass surrounded by omental adhesions was found in the right paraumbilical region. Gentle blunt dissection revealed a pedunculated, engorged, haemorrhagic soft tissue mass arising from the anti-mesenteric border of the proximal jejunum ([figure 1](#)). The lesion was excised without the need for bowel resection and a normal-appearing appendix was removed. His postoperative recovery was unremarkable.

Figure 1 Intraoperative image of the soft tissue mass attached to the anti-mesenteric border of the jejunum with torsion around a narrow stalk.

Figure 2 (A) H&E-stained representative slide of resected calcifying fibrous tumour (acquired with 4× objective). The image is dominated by stroma containing bland spindle-shaped cells, which, in areas, are arranged in whorled formation. Psammoma bodies are also seen. (B) H&E-stained representative slide demonstrating intratumoral haemorrhage (acquired with 2× objective).

Learning points

- ▶ Calcifying fibrous tumour is a rare benign tumour which may be identified intra-abdominally.
- ▶ Uncommon pathologies can mimic more common causes of acute abdominal pain, then identified on laparoscopy, necessitating clinician judgement intraoperatively regarding possible diagnoses and the optimal operative approach.

Histopathology demonstrated a well-circumscribed 60×30×15 mm piece of haemorrhagic tissue. Classical features of calcifying fibrous tumour (CFT) were present with paucicellular, hyalinised stroma containing bland spindle cells and multiple psammomatous and dystrophic calcifications. Evidence of torsion with haemorrhagic infarction was present ([figure 2](#)).

CFT is a rare benign mesenchymal tumour mainly observed in children and young adults.^{1,2} In a review of the 157 cases reported internationally, there is an estimated one case of abdominal CFT per year worldwide.³ There is a slight predominance in females (1:1.27) and a trimodal distribution at 0–4 years, mid-20s and mid-30s. The tumour is solitary in 94% of reported cases, with the most common location being the stomach, followed by the small intestine.^{1,3} Abdominal CFTs are proposed to be the late, 'burned out' manifestation of inflammatory myofibroblastic tumours.^{1,4,5} Consensus on the treatment for abdominal CFT is surgical excision, usually via an open approach.³

Abdominal CFT can become symptomatic and present as an acute condition. In this setting, it is characterised by pain with other possible gastrointestinal symptoms, depending on its location along the gastrointestinal tract and the extent of intraluminal and peritoneal involvement.³ This is the first report of torsion and haemorrhagic infarction of a CFT on the small intestine. Previously, clinicians have encountered torsion of a related benign tumour of the omentum, also following a clinical presentation consistent with acute appendicitis.⁶ We present the rare diagnosis of CFT, which may present as a mimic of other pathologies resulting in acute abdominal pain. Identification and recognition can assist clinicians to plan for its resection and the operative approach required to achieve this.

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