Mesenteric fibromatosis: a rare cause of peritonitis

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Case presentation

A 65-year-old Chinese man presented with a 2-day history of left-sided abdominal pain with fever and watery diarrhoea in February 2016. Systemic enquiry was unremarkable and he had no recent travel or contact history. On admission, his blood pressure was 127/67 mm Hg, pulse rate was 110 beats/min, and body temperature was 37.8°C. Abdominal examination revealed peritoneal signs over the left side of the abdomen and evidence of a tender irregular firm mass. There was no organomegaly or ascites. Blood tests demonstrated leukocytosis with white cell count of 11.8 × 10^9/L but findings were otherwise normal. Both chest and abdominal X-rays were unremarkable.

Urgent contrast-enhanced computed tomography (CT) of the abdomen and pelvis revealed a circumscribed mass (10.2 cm x 11.1 cm x 10.3 cm) located in the left abdominal cavity that could not be delineated from adjacent small bowel loops. A 1.6-cm thick layer of rim-enhancing collection with gas density was closely related to the left posterolateral aspect of the mass and there was a small amount of peritoneal fluid at the pelvic and left side of the abdominal cavity (Fig 1a and 1b). Radiological features were consistent with a gastrointestinal stromal tumour (GIST) complicated by abscess formation.

Broad-spectrum empirical antibiotic was started and emergency laparotomy was arranged. At laparotomy, there was generalised peritonitis with purulent peritoneal fluid. An 11 cm x 13 cm tumour was found at the mesenteric side of the proximal jejunum which had ruptured with abscess formation. The tumour involved the jejunal wall but there was no mucosal lesion (Fig 1c). Laparotomy was otherwise unremarkable. En-bloc resection of the tumour with the adjacent jejunum was performed followed by primary anastomosis.

Gross examination showed a multinodular tumour with an area of purulent material on the surface at the serosa, measuring up to 11 cm in diameter. The tumour showed a fibrotic and whitish cut surface (Fig 1d). On light microscopy, a circumscribed and non-encapsulated spindle cell neoplasm was seen centred at the subserosa and muscularis propria (Fig 2a). The spindle cells were arranged in vague fascicles and possessed elongated nuclei with a small amount of amphophilic cytoplasm set in a collagenous background. There was no significant nuclear atypia and mitotic figures were present at up to 1 per 50 high-power field. Scattered linear blood vessels were noted among the spindle cells. Ulceration with fibrinous exudation, granulation tissue reaction, and mixed inflammatory cell infiltration were noted at the serosal surface. On immunohistochemical staining, tumour cells exhibited a beta-catenin nuclear translocation pattern and were weakly positive for c-Kit (Fig 2b and 2c). They were negative for DOG-1, CD34 (GIST markers) [Fig 2d], MNF116 (cytokeratin marker), S100 (Schwann cell marker), and actin and desmin (smooth muscle markers). The overlying small intestine mucosa was unremarkable. The features were compatible with a diagnosis of mesenteric fibromatosis (MF).

The patient had an intra-abdominal collection postoperatively that was successfully treated by ultrasound-guided drainage and antibiotics. He was discharged 2 weeks later.

Discussion

Mesenteric fibromatosis is a rare sporadic mesenchymal neoplasm of the small bowel mesentery that arises from myofibroblasts. It is a histologically benign disease and lacks the capacity to metastasise.[1-3] Nonetheless, MF is locally aggressive with a high recurrence rate after surgical resection. Symptomatic MF mostly presents with abdominal pain or a palpable mass on physical examination. Gastrointestinal perforation is a rare manifestation. The first case of peritonitis secondary to MF reported in the literature was by Gorlin and Chaudhry in 1960.[4] The case presented here was initially misinterpreted as a ruptured GIST. Although MF and GIST are completely different entities, their clinical, radiological, and histological features frequently overlap and may confuse clinicians.

Computed tomography is the mainstay of diagnosis and typically demonstrates an infiltrative homogeneous soft tissue mass that abuts or extends into the gastrointestinal wall.[3] The case presented here demonstrated a mass that could not

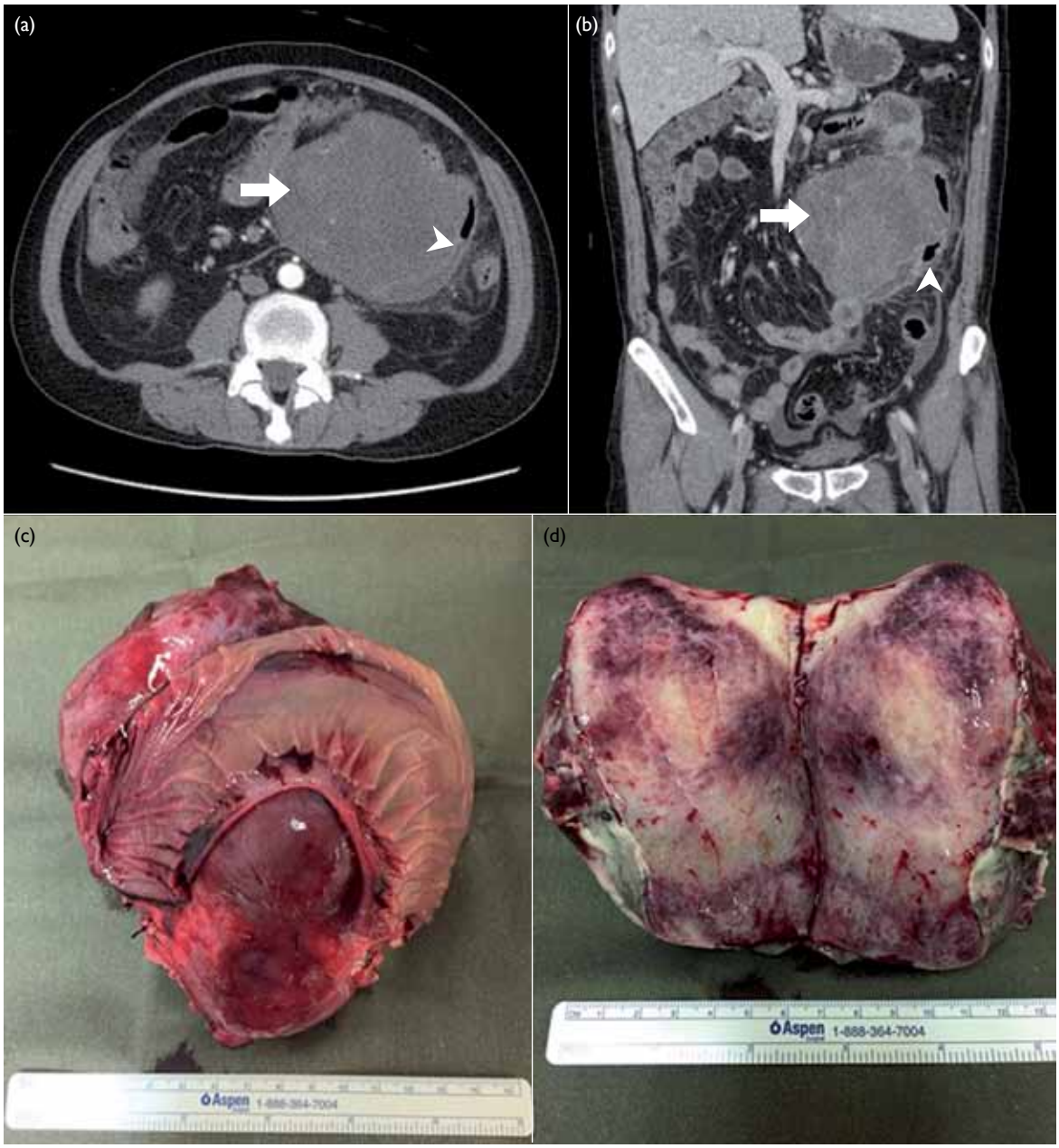

FIG 1. (a) Axial and (b) coronal sections of contrast-enhanced computed tomography showing a 10.2 cm x 11.1 cm x 10.3 cm heterogeneous hypodense mass located at the left abdomen (arrows) with an adjacent rim-enhancing cavity with gas (arrowheads). (c) Tumour located at the mesenteric side of jejunum. (d) White fibrotic cut surface

be delineated from adjacent small bowel wall thus mimicking a small-bowel GIST. To distinguish MF from GIST on CT, Zhu et al[1] suggested a number of differentiating features in favour of MF including extra-gastrointestinal location, ovoid or irregular contour, homogeneous enhancement, absence of intralesional necrosis, lower degree of enhancement and lesion-to-aorta CT attenuation ratio. Magnetic resonance imaging of MF typically demonstrates low-signal intensity relative to muscle on the T1-weighted image and variable signal intensity on the T2-weighted image. On the contrary, GIST typically has high-signal intensity on T2-weighted images.[3]

Gross pathological examination of MF usually shows a well-circumscribed hard-to-firm mass with white glistening on the cut section. Microscopically, MF has a number of characteristics similar to GIST, with frequently overlapping immunophenotypes. Distinction of MF from GIST is clinically important, as they are different entities with a different clinical course, treatment options, and prognosis. On light microscopy, MF samples typically demonstrate homogeneous spindle cells without atypia, infrequent mitotic figures, and abundant

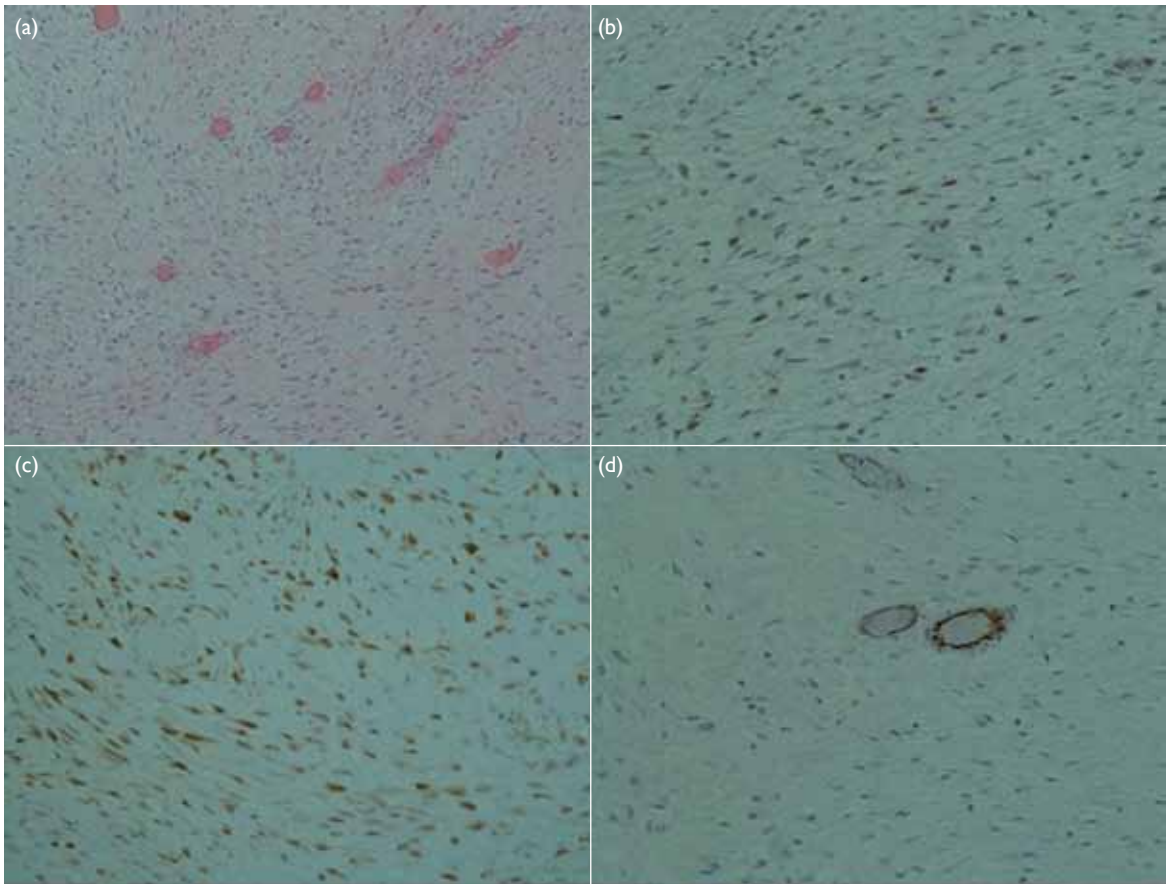

FIG 2. (a) Spindle cells in a collagenous background (H&E, x 100). (b) Weakly positive immunostaining for c-Kit. (c) Positive immunostaining for nuclear beta-catenin. (d) Negative immunostaining for CD34

collagen among dilated vessels.[2,3] In contrast, GIST samples demonstrate spindle cells forming fascicles commonly with atypia and higher cellularity with necrosis often present. Both MF and GIST may manifest overexpression of c-Kit.[2,3] Nonetheless, nuclear beta-catenin is expressed in MF but not in GIST, and MF is negative for CD34.

Treatment of MF should be tailored to the individual patient. Although watchful waiting may be offered for asymptomatic MF, surgical resection is usually indicated in large symptomatic cases of MF or in MF with complications.[5] Such MF is known to be locally aggressive and tends to recur when incompletely resected.[2,3,5] The decision for radiotherapy or systemic treatment with chemotherapy or hormonal therapy should be made after discussion with oncologists. Recently the use of imatinib, a tyrosine kinase inhibitor, has shown success in the treatment of locally advanced MF.[5]

Declaration

The authors have disclosed no conflicts of interest.

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