J Hong Kong Col Radiol. 2010;13:218-21

CASE REPORT

Mesenteric Fibromatosis: an Uncommon Cause of Abdominal Pain

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ABSTRACT

Mesenteric fibromatosis is a fibroblastic proliferation of the mesentery and is often called an intra-abdominal desmoid tumour. It is a rare condition with the potential to cause significant morbidity and death due to its locally invasive pattern of growth. The non-specific clinical presentation of this condition makes preoperative diagnosis difficult. We present a case of mesenteric fibromatosis with cavity formation and review its clinical and radiological features.

Key Words: *Abdominal pain; Fibromatosis, abdominal; Fibromatosis, aggressive; Gardner syndrome; Mesentery*

中文摘要

腸系膜纖維瘤病：腹痛的罕見病因

陸嬈、馮啟雄、馮成海

腸系膜纖維瘤病是指腸系膜中成纖維細胞增殖的現象，一般被稱為腹內硬纖維瘤。這種罕見疾病可能會有局部的侵入式生長，而導致較高的發病率和死亡。這病臨床上並無特異性，術前診斷相當困難。本文報告腸系膜纖維瘤形成膜腔的一個病例，並討論此病的臨床及放射學特徵。

INTRODUCTION

Mesenteric fibromatosis is a deep infiltrative fibroproliferative process, primarily involving the mesentery and constitutes a rare type of fibromatosis.[1,2] It is characterised by a locally infiltrative pattern of growth, with a tendency for local recurrence.[3] Most cases are sporadic but some are associated with trauma or Gardner's syndrome (colonic polyposis, osteomas, and soft tissue tumours).[2] Due to its infiltrative nature, it can cause significant complications and even death. We report a case with mesenteric fibromatosis and cavity formation, presenting with acute abdominal pain and fever.

CASE REPORT

A 31-year-old woman presented with a two-day history of acute abdominal pain and fever in May 2010. There was no history of abdominal trauma or surgery. She was febrile and had abdominal tenderness. Urgent computed tomography (CT) of the abdomen and pelvis showed a relatively well-marginated large lobulated mass (20.3 cm x 18.3 cm x 8.2 cm) in the abdomen (Figure 1a). A 6.3 cm x 6.9 cm x 5.4 cm rim-enhanced cavity was noted at the inferior aspect of the large mass lesion (Figure 1b). CT features were suggestive of soft tissue tumours such as gastrointestinal stromal tumour (GIST) or lymphoma.

Emergency laparotomy revealed a large mass appearing to arise from the distal stomach and involving the transverse colon. Distal gastrectomy and right hemicolectomy were performed and the large mass was

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Submitted: 20 Sep 2010; Accepted: 26 Oct 2010.

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resected. Evidence of abscess formation with soiling of the peritoneal cavity by pus was noted. Subsequently, *Bacteroides fragilis*, *Escherichia coli*, and *Streptococcus anginosus* were cultured from the peritoneal swab. The provisional diagnosis was GIST. The postoperative course was uneventful.

Pathological examination revealed that the right colon and the stomach were partially encased by a huge tumour mass, with a smooth brownish outer surface with areas of fibrinous deposition (Figure 2a). Serial slicing yielded a soft-to-firm brownish to greyish tissue. Areas of necrosis were noted in the tumour mass. There was no definite fistulous communication with the surrounding bowel loops. Microscopic examination yielded a proliferation of spindle cells in a loose array or fascicles, with infiltration into the mesenteric fat, gastric and colonic muscle wall (Figure 2b). There was variable myxoid change and collagenous stroma. Characteristic prominence of thick-walled medium-sized blood vessels was noted, but there was no significant cytological atypia. Immunohistochemically, the tumour cells were negative for CD34, CD117 and DOG1 (GIST markers), SMA (smooth muscle marker), and S100 (Schwann cell marker). Areas of necrosis and acute inflammatory infiltrate were identified. These features were compatible with mesenteric fibromatosis. In view of the association of mesenteric fibromatosis with Gardner's syndrome, the patient was scheduled for oesophagogastroendoscopy and colonoscopy. No other manifestations of Gardner's syndrome were identified in this patient.

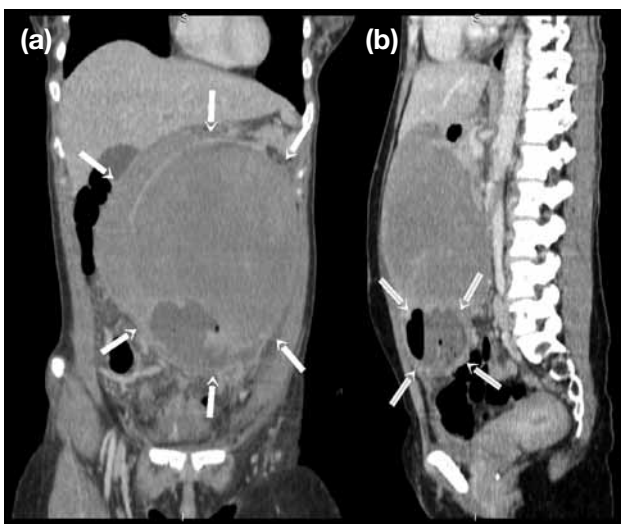

Figure 1. (a) A coronal computed tomographic (CT) image of the abdomen and pelvis shows a relatively well-defined large lobulated mass in the abdomen (arrows) displacing loops of bowel. (b) A sagittal CT image of the abdomen and pelvis shows a rim-enhancing cavity (arrows) containing an air-fluid level.

DISCUSSION

Mesenteric fibromatosis is a rare condition with an estimated annual incidence of two to four cases per million people and accounts for about 0.03% of all neoplasms.[4-6] It is the most common primary tumour of the mesentery and accounts for 8% of all fibromatoses.[4,7] Mesenteric fibromatosis is often locally aggressive but does not metastasise.[5] It may infiltrate surrounding tissue and cause significant morbidity and

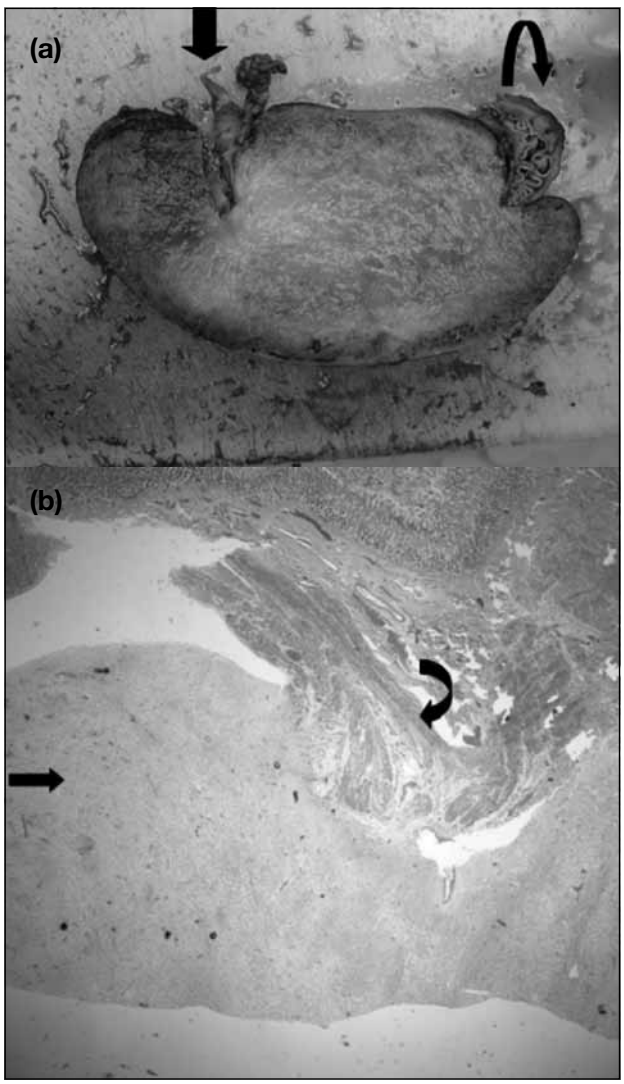

Figure 2. (a) Cut surface of the large tumour mass of mesenteric fibromatosis showing the tumour mass composed of brownish to greyish tissue, partially encasing the colon (straight arrow) and the stomach (curved arrow). (b) Microscopic examination showing spindle cells in a loose array / fascicles (straight arrow) with an irregular junction at the muscularis propria of the stomach (curved arrow).

death.[5] It is most often located in the mesentery of the ileum, and sometimes in the mesocolon or gastrohepatic ligament.[2,8]

A slightly higher incidence in women has been reported and the peak age of onset is 25 to 35 years.[5,6,9] It may be secondary to trauma, or hormonal stimulation, and has been associated with Gardner's syndrome and multiple pregnancies.[5,8,10,11]

Most patients with mesenteric fibromatosis are asymptomatic until late in its course.[7] Patients may present with abdominal pain, vomiting, abdominal mass, weight loss, and fever.[4,5,10] It may be complicated by mesenteric ischaemia, intestinal obstruction, perforation and haemorrhage, ureteric obstruction, and tumour necrosis.[4,5,12]

The radiological features of these tumours are variable; there are no specific imaging features to distinguish between mesenteric fibromatosis and other mass lesions in the abdomen.[9,11] On sonography, mesenteric fibromatosis have variable echogenicity with well-defined or irregular borders. On CT, mesenteric fibromatosis is either ill-defined or well-circumscribed, with variable attenuation and contrast enhancement.[7,11] Magnetic resonance imaging (MRI) shows low signal intensity relative to muscle on T1-weighted images, with variable signal intensity relative to muscle on T2 images.[11] CT and MRI are useful for evaluating the size of the tumour and involvement of surrounding structures before surgical resection. CT and MRI are also useful for detecting recurrence of disease in surgically managed patients. An interesting CT feature in this case was a rim-enhancing cavity with air-fluid levels in the large tumour mass. Cavity formation in mesenteric fibromatosis has not been well-documented in the literature. Possible causes of cavity formation include tumour necrosis or fistulous communications with surrounding bowel loops. No definite fistula was identified in our case.

Histopathological examination is essential for diagnosis of mesenteric fibromatosis.[4,9] On gross pathological examination, the tumour is usually well circumscribed and is white and coarsely trabeculated on cross-section. Histologically, it is composed of spindled or stellate fibroblastic cells embedded in a collagenous stroma.[7]

The differential diagnoses of mesenteric fibromatosis include GISTs, lymphomas, and fibrosarcomas. As in this case, the most common misdiagnosis is GIST.[3] When mesenteric fibromatosis involves the wall of the stomach or bowel, it may appear to arise from that site, mimicking a GIST. Histopathological examination helps differentiate mesenteric fibromatosis and GIST.[3] In differentiating GIST from mesenteric fibromatosis, the latter is suggested when there is prominent vasculature, infiltrative growth pattern, and collagenous stroma. Negative immunostaining for CD34, CD117 and DOG1 also makes GIST less likely. Correct distinction of mesenteric fibromatosis from GIST has significant clinical implications, as these two conditions have different prognostic and therapeutic connotations. Mesenteric fibromatosis is relatively benign and without the capacity to metastasise, whereas GIST frequently has malignant potential.

The clinical course of mesenteric fibromatosis is variable, ranging from rapid growth to spontaneous regression.[10,12] Treatment of choice is complete surgical excision with a margin of uninvolved tissue. Adjuvant therapy includes radiotherapy, non-steroidal anti-inflammatory drugs, antioestrogens and cytotoxic chemotherapy.[3,5,7,9,13] Recurrence rates after excision have been reported to be 10 to 90%.[2,5] A mortality rate of 6% has been reported.[13]

CONCLUSION

Mesenteric fibromatosis is a rare benign tumour that is locally aggressive and is a possible cause of acute abdominal pain. Clinical presentation and radiological features are often non-specific. This case illustrates that cavitation is a feature. Mesenteric fibromatosis should be considered a differential diagnosis in patients presenting with abdominal pain and fever.

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