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Dr. Sergei Mashkov
General Surgery Registrar, Palmerston North Hospital, New Zealand

Dr. Emma McKay
Anatomical Pathology Registrar, Palmerston North Hospital, New Zealand

Richard Coutts
General Surgeon, Palmerston North Hospital, New Zealand

Corresponding Author:
Dr. Sergei Mashkov
General Surgery Registrar, Palmerston North Hospital, New Zealand

Calcifying abdominal tumour, a diagnostic dilemma: Case report

Dr. Sergei Mashkov, Dr. Emma McKay and Richard Coutts

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Abstract
Calcifying fibrous tumor is a rare type of benign mesenchymal tumor that presents in various locations including gastro-intestinal (GI) tract, that can become a diagnostic dilemma for clinicians due to its rarity and histopathologic features that overlap with other, more common stromal lesions of the GI tract. This case report presents a manifestation of multifocal peritoneal calcifying fibrous tumor in a female patient with an acute abdomen.

Keywords: Calcifying fibrous tumor, CFT, mesenchymal tumor, stromal tumor, benign gastro-intestinal tumor, abundant hyalinized collagen

Introduction
Calcifying fibrous tumor (CFT) is a rare, benign mesenchymal mass lesion that was first described in the literature in 1988. Subsequent studies documented the variety of anatomic locations including gastrointestinal tract, mediastinum, pleura, heart, lungs, neck, and others [1]. Between 1988 until 2015, just under 160 cases of calcifying fibrous tumor were reported in the literature. CFT arising from the gastrointestinal tract was documented in the small and large bowel, stomach, peritoneum, oesophagus and appendix. Most GI calcifying fibrous tumors are the subject of single case report. The tumor has a slight prevalence for females and commonly seen in adults with median age of 49 years. One study reported gastrointestinal CFTs occurrence in siblings. The presentation of CFT in the gastrointestinal tract may present a diagnostic dilemma, due to its rarity and histopathologic features that overlap with other, more common stromal lesions of the gastrointestinal tract.
Our case report describes a manifestation of multifocal peritoneal calcifying fibrous tumor in a middle-aged female patient who presented with acute onset of severe abdominal pain and fever.

Clinical features
According to a systematic review of all up to date reported CFT cases in the literature, it can occur virtually anywhere in body. The most common locations in in the gastrointestinal tract are stomach, small bowel, mesentery and peritoneum. The majority of calcifying fibrous tumors were found as a solitary lesion, with around 5-25% cases revealed to have multiple lesions [1].
Gastrointestinal CFTs are most commonly asymptomatic and discovered incidentally, although can present in various and sometimes unique ways. Abdominal pain and discomfort are the most common symptoms, followed by lack of appetite, fever, fatigue, dyspepsia and vomiting. Gastric CFTs may manifest as gastric ulcers [4]. Small intestine and mesenteric lesions can present with symptoms of bowel obstruction and intussusception. The majority of symptomatic patients have had chronic symptoms related to CFTs (82%) according to the systematic review. Only 18% patients presented with acute symptoms [2].

Diagnostic tests
Imaging findings are not specific and typically show a mass with a clear border, well circumscribed with mild enhancement and calcification on ultrasound and CT. MRI examination was also described in literature, however findings are not characteristic and usually show isosignal intensity on T1-weighted imaging and hyposignal intensity on T2-weighted imaging [2]. On direct laparoscopic or endoscopic visualisation, CFTs typically appear as a subserosal or submucosal nodules.

Differential diagnosis

The major diagnostic dilemma in the preoperative stage is to determine the character of the tumor and distinguishing malignant from benign for optimal operative management. The differential diagnosis for CFT of the gastrointestinal tract includes gastrointestinal stromal tumor (GIST), schwannoma, leiomyoma, solitary fibrous tumor, fibromatosis, sclerosing mesenteritis, and reactive nodular fibrous pseudotumor (RNFP) [3].

Gross and microscopic pathology

Gastrointestinal CFTs are mostly located in submucosal layer and macroscopically reveal a well-circumcised, unencapsulated spherical to lobulated mass with variable calcification. CFTs have a firm texture and average size is 2.6 cm with a range from 0.5 to 10 cm [2].

Histologically, there are three features characteristic of CFTs: abundant hyalinized collagen, lymphoplasmacytic inflammatory infiltrates and scattered calcifications with necrosis invariably absent [4].

Etiology

Currently the aetiology and pathogenesis of CFT is uncertain. Possible causes include trauma, surgical intervention, and infection. It was also hypothesised by some authors that CFT might be a manifestation of IgG4-related disease, due to cases of CFT with increased IgG4-positive plasma cells and elevated serum IgG4 levels. Associations with Castleman disease, inflammatory myofibroblastic tumor (IMT) and sclerosing angiomatoid nodular transformation of the spleen (SANT) were also described. It is possible that CFT may simply represent an end-stage manifestation of a variety of inflammatory and sclerotic processes, without regard to a specific aetiology [1, 2].

Management and prognosis: CFTs are benign lesions and the overall prognosis is excellent. Patients with diagnosed CFTs should be offered local surgical excision by either endoscopic, laparoscopic or open technique. The local recurrence rate for CFTs has been estimated to be approximately 10%. To date there have been no malignant transformation or death due to CFT reported [1].

Case report

A 30-year-old female presented with a 2-day history of central abdominal pain and nausea. Her medical and surgical history were unremarkable apart from previous stomach ulcers and caesarean section. There was no reported family history of gastrointestinal disease. Clinical examination revealed mild fever of 38 ^{0}C and abdominal tenderness in peri-umbilical and right iliac fossa areas. Initial laboratory workup including haematology and biochemistry panels and tumour markers (CEA, Ca 125, Ca 19-9) were unremarkable apart from a mildly elevated C-reactive protein of 63 mg/L (<5 mg/L). Abdominal ultrasound was not diagnostic and showed free abdominal fluid adjacent to loops of bowel in right lower abdomen. Diagnostic laparoscopy revealed multiple omental and lesser sac nodules and peritoneal disease suspicious for an advanced malignancy without obvious primary source. Intraoperative omental biopsies as well as abdominal cavity washings were taken. In anticipation of the histology report, an urgent CT of the chest, abdomen and pelvis was arranged and showed a poorly defined soft tissue mass within the transverse mesocolon with omental and peritoneal deposits though with no evidence of metastatic disease in the liver or chest.

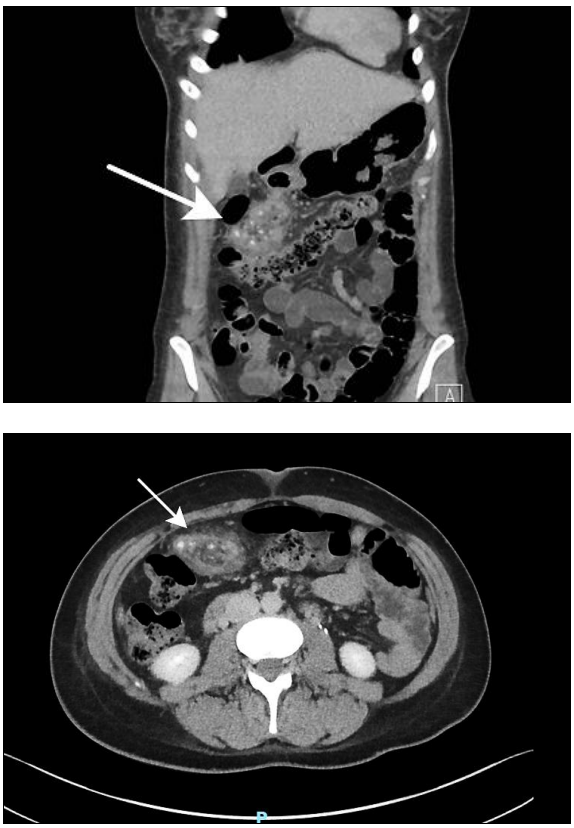

Fig 1: CT scan showing poorly defined soft tissue mass with small dense foci centrally within the right-sided transverse mesocolon. The lesion has contact on the inferior aspect of the pylorus and lies just medial to the hepatic flexure (white arrows). Coronal and axial views

There were no signs of upper gastrointestinal tract malignancy identified on subsequent gastroscopy.

A formal resection of multiple intraperitoneal nodules was arranged after the diagnosis of multifocal peritoneal calcifying fibrous tumor was confirmed on pathology report. Laparotomy with resection of multiple CFT from the lesser and greater omentum, diaphragm, spleen capsule and anterior stomach wall went remarkably well.

The intraoperative specimen was submitted for histopathological examination.

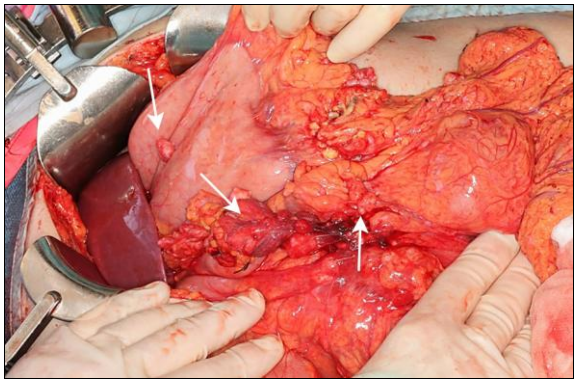

Fig 2: Intraoperative view. The greater omentum and anterior wall of the stomach with multiple calcified fibrous tumours (white arrows).

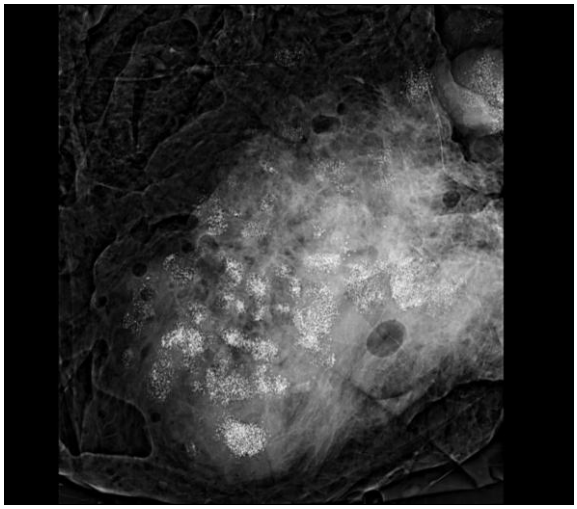

Fig 3: Intraoperative specimen, section of the greater omentum containing multifocal calcifying fibrous tumor using Hologic Trident specimen X-ray machine. Multiple foci of calcification noted.

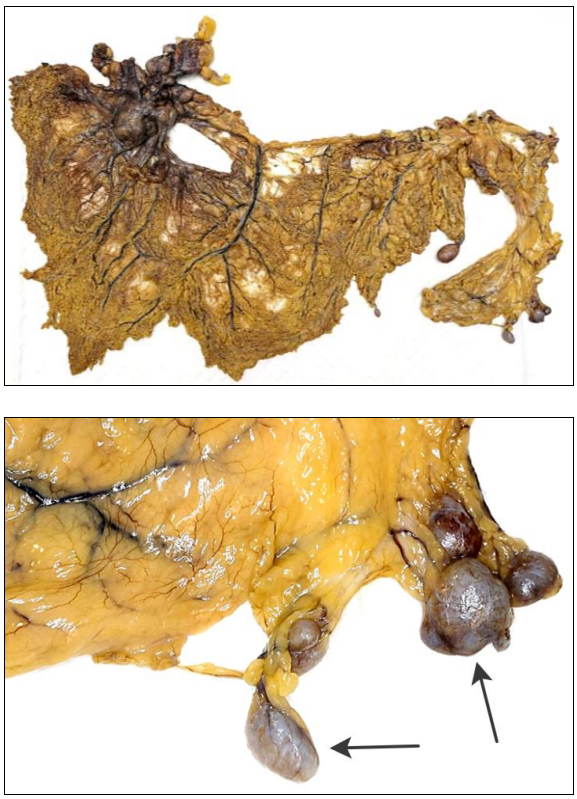

Fig 4: Macroscopic photos of the greater omentum with multiple calcified fibrous tumor nodules (Black arrows)

Microscopically, the sections showed hypocellular nodules, comprising scattered stellate to spindled cells with bland oval nuclei and pale wispy cytoplasm set within densely hyalinised collagenous stroma. Scattered throughout the nodules were numerous lamellated psammomatous and dystrophic calcifications. The nodules contained thin-walled blood vessels with open lumina that were associated with a mild lymphoplasmacytic inflammatory cell infiltrate. There was no cytological atypia, mitotic activity or necrosis identified.

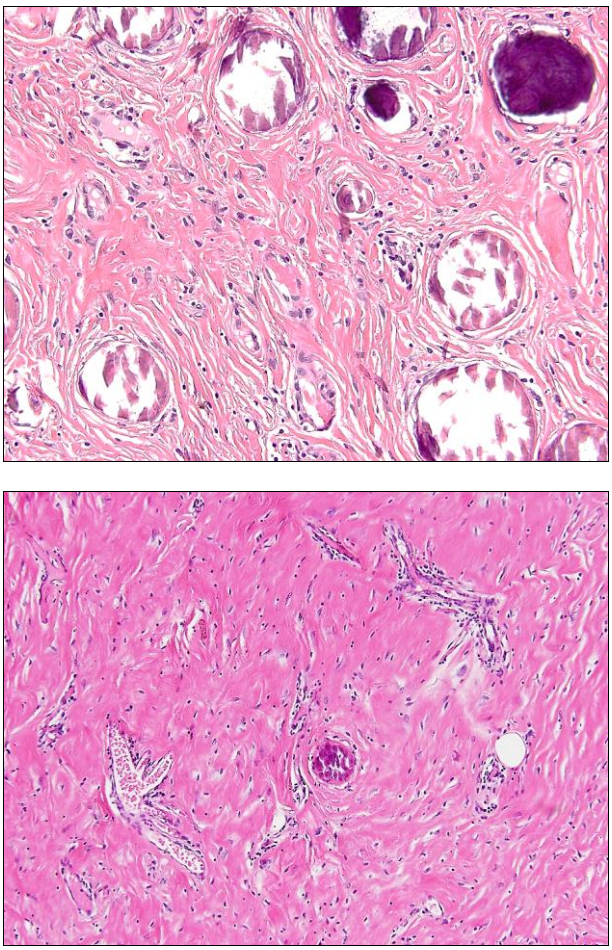

Fig 5: Microscopic features of calcifying fibrous tumor (10x and 20x magnification).

Immunohistochemical studies revealed the fibroblasts of the tumour expressed factor XIIIa and were negative for actin, caldesmon, CD34, CD117, SMA, ALK1 and DOG1. IgG4 was found to be focally expressed in the plasma cells only.

The above morphological appearance and immunophenotype were consistent with multifocal calcifying fibrous tumour.

The patient underwent unremarkable recovery and remained symptoms free at the first clinic follow up the subsequent surveillance scan were also unremarkable. Patient sister was recommended to obtain screening MRI scan of abdomen and pelvis, considering a reported case of CFT occurrence in siblings. The scan was negative.

Conclusion

Calcifying fibrous tumour is rare type of soft tissue tumour. These are usually solitary, although incidences of multifocal disease have been reported. The gastrointestinal tract is a common site of involvement by CFT. These are benign

tumours with no metastatic potential described in the literature. Local resection is typically curative, however occasional recurrence following resection has been described.

Conflict of Interest
Not available

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Not available

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