CASE REPORT

Mesenteric desmoid tumour masquerading as a fat-containing cystic mass

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ABSTRACT. Mesenteric desmoid tumour (MDT) is an uncommon neoplasm that typically presents as a solid soft-tissue mass on cross-sectional imaging. MDT manifesting as a fat-containing cystic mass on CT has not been described in the literature. We report such an unusual case with clinicopathological correlation.

Received 5 January 2009
Revised 17 February 2009
Accepted 12 March 2009

DOI: 10.1259/bjr/68468861

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Desmoid tumour is an uncommon benign neoplasm. It can occur in association with either familial adenomatous polyposis (FAP) or Gardner's syndrome or can occur sporadically [1]. Mesenteric desmoid tumour (MDT) is a form of intra-abdominal desmoid tumour with the classic CT appearance of a homogeneous solid soft-tissue mass, displacing surrounding structures and vasculature. Atypical CT appearance such as cystic change (due to abscess formation) is rare [2, 3]. We report a uniquely unusual case of a ruptured, infected MDT presenting as a fat-containing cystic mass. To the best of our knowledge, this CT appearance has not been previously reported.

Case report

A 58-year-old nulliparous and post-menopausal woman presented with an acute onset of lower abdominal pain, vomiting and pyrexia over 3 days. She had no significant medical history, previous surgery or family history of FAP. Clinical examination revealed lower abdominal tenderness and mild guarding. Blood tests revealed neutrophilic leukocytosis with an elevated total white cell count of $11.0 \times 10^9 \text{ l}^{-1}$ (normal $3.6\text{--}9.3 \times 10^9 \text{ l}^{-1}$) and a differential count of 94.1% neutrophils, consistent with an infection. The urea and electrolyte levels and liver function were normal.

Contrast-enhanced multidetector computed tomography (SOMATOM Sensation Cardiac 64-slice scanner, Siemen Medical Solutions, Erlangen Germany; 120 kV, 180 mA, 0.33 s per rotation, reconstructed slice thickness 5 mm) of the abdomen and pelvis was performed. CT revealed a solitary, 45×34 mm well-defined cystic mass arising from the small bowel mesentery causing

displacement of the surrounding vessels. A smaller cystic component extruding from the superior portion of the mass was also present. Surrounding inflammatory stranding was seen around the entire lesion with high-density free fluid in the abdomen. Interestingly, the mass was almost entirely fat-containing (average CT attenuation value of -45 Hounsfield units (HU)) with a small internal fluid level (Figure 1). There was no associated soft-tissue mass, calcification or regional lymphadenopathy. In view of the predominance of intralesional fat, an inflamed mesenteric teratoma and cystic lymphangioma were the working diagnoses at the time of the CT scan.

Intra-operatively, the mass was confirmed to arise from the jejunal mesentery, closely related to a segment of the jejunum with no gross bowel wall involvement. A focal area of perforation was seen, corresponding with the small extruded component seen on CT (Figure 1b). The mass was firm on palpation, measuring $45 \times 45 \times 40$ mm. Following *en bloc* resection of the mass and an adjacent segment of jejunum, *ex situ* incision revealed a large cystic component with the cavity measuring 40 mm in maximal dimension (Figure 2). The content of the cyst was soft in consistency, mixed with purulent contents. Pus was found in the peritoneal cavity. The findings confirmed the CT finding of a predominantly cystic mass and the purulent fluid alluded to an underlying infective process. The patient recovered uneventfully and was discharged on the seventh post-operative day. She was noted to be well at 6 months follow-up.

Histopathological examination of the tumour wall (Figure 3) reported moderate cellular proliferation, composed of sweeping fascicles of plump spindle-shaped cells with bland uniform nuclei and pale eosinophilic cytoplasm. In the periphery, the fascicles of spindle cells enclosed lobular aggregates of normal-looking mesenteric adipose tissue. The wall of the lesion showed a lining of inflamed granulation tissue with adjacent

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(a)

(b)

Figure 1. (a) Axial and (b) coronal reconstructed CT image of the abdomen showing a predominantly fat-containing cystic mass centred in the small bowel mesentery, adjacent to a loop of small bowel. A small internal fluid level is present (long white arrow in (a), and surrounding inflammatory stranding is noted. High-density free fluid is present in the paracolic gutters on both sides (short arrows in (a)). A small extruded component is seen in the superior aspect (arrow in (b)) and was found to represent a point of rupture on surgery.

organising abscess and evidence of acute chronic inflammation. Immunohistochemistry stained positive for β -catenin, desmin and smooth muscle actin. Histological features and immunohistochemistry were consistent with an MDT.

Discussion

Desmoid tumours represent part of a clinicopathological spectrum of deep fibromatoses and are classified by anatomical location. In the abdominal region, desmoid

Figure 2. Photograph of the mass after *ex situ* incision and drainage of the fluid, confirming the cystic nature of the mass. The cystic cavity measures 4 cm in size and was encapsulated by the small bowel mesentery. Focal area of perforation (not shown) was noted in the superior aspect corresponding to the area of extrusion seen in Figure 1b.

Figure 3. Micrograph of the tumour: haematoxylin and eosin (H&E) stain at 40 $\times$ magnification. Abundant lobules of entrapped mesenteric adipose tissue (arrows) are present within tumoral tissue. The tumour demonstrates sweeping fascicles of plump spindle cells containing bland nuclei. Positive staining for β -catenin, smooth muscle actin and desmin (not shown) was also observed. The findings confirm the diagnosis of a mesenteric desmoid tumour.

tumours can arise from the deep abdominal wall soft tissue or intra-abdominally (*e.g.* small bowel mesentery and pelvis). Small bowel mesentery is the most common site for intra-abdominal desmoid tumours [4]. Desmoid tumours, although non-metastasising, are known to be locally aggressive, with notoriety for recurrence after surgical resection. Although desmoid tumours are frequently associated with polyposis syndromes, such as FAP and Gardner's syndrome, most cases of MDT occur sporadically, with only 13% of patients having FAP [4, 5]. Additionally, unlike FAP-related cases, the incidence of which is higher in middle-aged women [6], sporadic MDTs do not show any age or sex predilection [5, 7]. Furthermore, patients with FAP and MDT tend to present with multiple but smaller lesions, whereas sporadic cases tend to present with a solitary large lesion, as in our case [1, 4]. Symptoms from MDT can arise owing to local pressure and obstruction of the bowel or its blood supply [6]. Retroperitoneal extension of MDT can cause hydronephrosis [1], and abscess formation with rupture is rare [2].

The pathogenesis of desmoid tumour is unknown, but links have been made to hormonal, genetic and traumatic factors (*e.g.* previous surgery) [8]. Notably, previous surgery has been shown to be a less important risk factor in sporadic cases, with only 10% of patients having previous surgery, compared with 83% in FAP [9]. Furthermore, post-resection recurrence tends to be less common in sporadic cases than in patients with FAP [4]. Although the treatment algorithm remains debatable, the above forms the rationale for surgical resection being preferred in sporadic cases, whereas medical therapy (chemotherapy or hormonal therapy) is advocated in patients with FAP [4, 8, 10].

Cross-sectional modalities such as CT and MRI are the preferred imaging modalities for mesenteric masses. The CT and MRI appearance of MDT is directly related to the histological characteristics, with homogeneity related to the amount of collagenous *vs* myxoid stroma, as well as to tumour vascularity [11–13]. The typical appearance of MDT is a soft-tissue mass displacing the bowel loops and surrounding vasculature, with or without mesenteric involvement and bowel serosal changes. The soft-tissue component is such a dominant feature that it often simulates solid tumours, such as gastrointestinal stromal tumour (GIST), lymphoma or soft-tissue sarcoma [4].

In our case, the absence of a significant soft-tissue component typical of MDT is striking. We postulate this paucity of the soft-tissue component to be the result of spontaneous tumour regression. Spontaneous regression of desmoid tumour is believed to be related to the withdrawal of oestrogenic stimulation and has been documented in post-oophorectomy and post-menopausal women [14, 15]. This causal relationship is particularly relevant in our patient, who is 2 years post menopause. Tumour regression with cystic degeneration could also result from focal infarction secondary either to a compromised vascular supply within the tumour or to superimposed infection, or to a combination of both factors. This would not be surprising given the invasive nature of MDTs and their propensity for causing vascular compromise; however, it is difficult to ascertain in our case if infection and tumour regression occurred as sequential or concurrent events.

Cavitation resulting from abscess formation can occur in MDT, giving rise to the appearance of a cystic mass on CT [2, 16]. However, this case differs in that MDT is not known to contain a conspicuous amount of fat density on CT (Figure 1). In the literature, a fat-containing cystic mesenteric mass with internal fluid level has been described only in teratomas [17] and mesenteric cavitory lymph node syndrome [18]. In our case, this previously undescribed CT manifestation made MDT an extremely remote consideration. The fat densities were found to represent fluid mixed with purulent contents on incision of the tumour. Histological evidence of entrapped mesenteric fat lobules along the tumoral margin (Figure 3) leads us to think that fat necrosis and liquefaction of previously enclosed mesenteric adipose tissue is one possible source of the fat densities. Fat necrosis could have been incited by ischaemic insults from vascular compromise and/or from the superimposed infection. The internal fluid level seen on CT is probably due to layering of less dense fat on top of denser purulent fluid. Abscess formation leading to raised intralesional pressure is probably the cause for the tumour rupture. As our patient was previously well, acute events such as infection and tumour rupture are the most likely cause for her acute presentation.

To conclude, we report the first case of an MDT appearing as a fat-containing cystic mass on CT, and illustrate this rare and perhaps misleading mimicry.

Acknowledgments

The authors would like to thank NUH-NUS Medical Publications Support Unit, Singapore, for assistance in the preparation of this manuscript.

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