

Leiomyomatosis peritonealis disseminata presenting as omental torsion

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ABSTRACT Leiomyomatosis peritonealis disseminata is usually asymptomatic or mimics widespread malignancy; acute presentation is rare. We describe a patient with right iliac fossa and lower abdominal pain. Two masses were detected via computed tomography, but at surgery, one of these implanted leiomyomas had undergone acute omental torsion. This case illustrates a rare complication of omental leiomyoma torsion clinically mimicking acute appendicitis.

Keywords: abdomen, appendicitis, leiomyomatosis, peritoneal neoplasms, torsion abnormality
Singapore Med J 2012; 53(4): e71–e73

INTRODUCTION

Leiomyomatosis peritonealis disseminata (LPD) is a rare disorder in which multiple leiomyoma-like nodules are implanted on the peritoneal surface.⁽¹⁾ This is generally a benign entity; however, rare cases of malignant change have been reported.⁽²⁾ LPD occurs mainly in premenopausal women. In exceptional cases, it has occurred in men with malignant change⁽³⁾ and in postmenopausal women.⁽⁴⁾ This condition is often asymptomatic and more commonly an incidental finding during imaging or surgery. We present a case of LPD with clinical and imaging features mimicking acute appendicitis, which was later found to have omental torsion instead.

CASE REPORT

A 44-year-old Chinese woman presented with sudden onset of colicky right iliac fossa and lower abdominal pain. The patient had no other significant history. She had undergone a Caesarean section several years ago, followed by laparoscopic myomectomy of uterine fibroids two years prior to the current admission. During physical examination, suprapubic and right iliac fossa rebound tenderness was elicited. Digital examinations of the rectum and vagina were normal. The patient was afebrile, with a blood pressure of 96/65 mmHg and a normal heart rate. Laboratory tests showed mild neutrophilic leucocytosis, and urinalysis was normal. Urgent chest and abdominal radiographs did not reveal any evidence of pneumoperitoneum or bowel obstruction.

Emergency computed tomography (CT) revealed two well-circumscribed soft tissue masses in the lower abdomen and pelvis. The first was a soft tissue mass located in the midline of the pelvis (Fig. 1a), separate from the uterus, ovaries and bladder, which indented the dome of the bladder. The second mass was observed along the deep surface of the left rectus abdominis muscle (Fig. 1b). These were initially considered to represent incidental

findings due to a different underlying process given the relatively benign appearance of the masses. The vermiform appendix was mildly dilated (diameter 10 mm, Fig. 1a), with wall enhancement. There was a small amount of free fluid within the pelvis, in keeping with inflammatory response. The radiological impression at that time was early acute appendicitis.

In view of the CT findings of the two soft tissue masses, laparotomy rather than laparoscopic surgery was performed. Intra-operatively, the appendix appeared inflamed and was thus resected. The first, midline mass seen on CT was found to lie at the tip of the greater omentum and connected to a vascular pedicle that had twisted upon itself (Figs. 1c & 2). This, together with the second mass deep to the rectus abdominis muscle mass, was completely excised. The patient recovered well and was discharged two days post surgery. Follow-up CT at three months showed no evidence of recurrence. The patient remains on imaging surveillance.

Microscopic examination of the omental mass revealed a well-circumscribed proliferation of fascicles of bland spindle cells with cigar-shaped nuclei and ample eosinophilic cytoplasm, with minimal mitotic activity and lacking atypia or necrosis (Fig. 3). These spindle cells were immunopositive for smooth muscle actin and desmin, in keeping with smooth muscle differentiation. Large parts of the omental mass showed oedema, haemorrhage and infarction. In addition, these smooth muscle cells were focally positive for progesterone receptor. Immunohistochemistry for CD34, c-kit, S100 and HMB45 was negative. These findings were consistent with a benign leiomyoma. Similar histological features were identified in the rectus abdominis muscle mass. Histological examination of the appendix showed no evidence of mural inflammation. Instead, a 7-mm serosa-based nodule, also in keeping with a benign leiomyoma, was detected on the resected appendix. This nodule was, however, not visible in the CT images.

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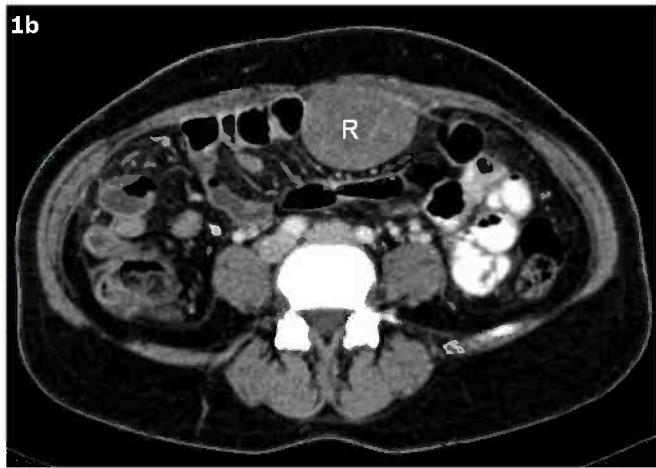


Fig. 1 Axial contrast-enhanced CT images show (a) a dilated and thickened appendix (arrows), with a small amount of adjacent free fluid and the upper extent of a soft tissue mass (O) that is visible in the midline; the mass was separate from but indenting the dome of the bladder (see Fig. 1c); and (b) a second soft tissue mass (R) deep to the left rectus abdominis muscle, with density that is similar to muscle. (c) Coronal contrast-enhanced CT image shows the two masses (O & R). The first tissue mass (O) is attached to a vascular pedicle (arrows), and at laparotomy, both this mass and its pedicle were found to arise from the tip of the greater omentum (see Fig. 2).

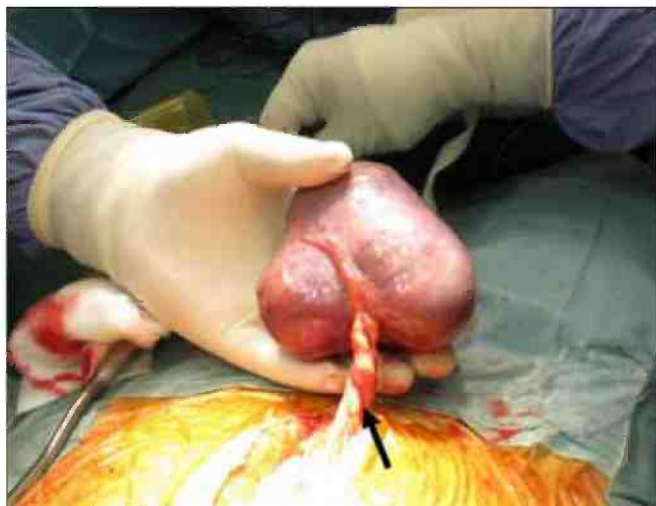


Fig. 2 Intra-operative photograph shows the omental mass (labelled O in Figs. 1a & c), connected to a twisted vascular pedicle (arrow). Fig. 1c shows the corresponding CT image.

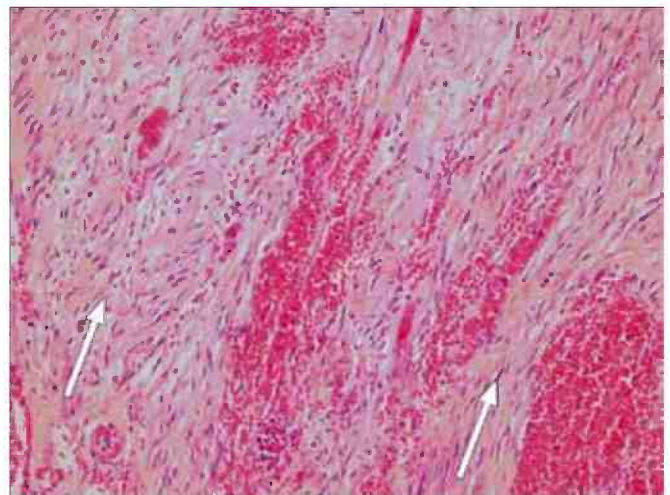


Fig. 3 Photomicrograph of non-infarcted areas of the omental mass shows bland spindle cell proliferation arranged in fascicles (arrows). These spindle cells were immunopositive for smooth muscle actin (not shown) (Haematoxylin & eosin, $\times 200$).

DISCUSSION

LPD is a rare entity that is characterised by multiple smooth muscle implants in the peritoneal cavity. Due to the rarity of the condition, the exact pathophysiology is unknown. In a histopathological review of 20 cases, Tavassoli and Norris postulated that LPD may result from the predisposition of pluripotent mesenchymal cells to form smooth muscle.⁽⁵⁾ LPD has largely been associated with conditions such as pregnancy,⁽⁵⁾ oral contraception,⁽⁵⁾ assisted reproduction,⁽⁶⁾ endometriosis⁽⁷⁾ and oestrogen-secreting ovarian

fibrothecomas.⁽⁸⁾ This suggests that oestrogen exposure is a possible aetiology, similar to that for uterine leiomyomas. LPD is also reported to be associated with laparoscopic resection of uterine leiomyomas,⁽⁹⁾ suggesting transcoelomic dissemination rather than de novo peritoneal metaplasia. In our patient, it is highly possible that prior laparoscopic uterine myomectomy was a predisposing factor.

LPD is usually asymptomatic and found incidentally during imaging or surgery, and a presentation with acute abdomen is

rare. LPD implants on the surface of the ovaries have, on rare occasions, resulted in ovarian torsion.⁽²⁾ However, the available literature on complications of omental leiomyoma implantation and LPD presenting with omental torsion is limited. In our patient, the slightly thickened vermiform appendix and free peritoneal fluid made the pre-operative diagnosis more difficult, and a false positive diagnosis of acute appendicitis was made. Since no histological evidence for acute appendicitis was found and the subcentimetre implant on the appendix could not be seen at the retrospective review of the CT examination, we postulate that the imaging appearance of the appendix was secondary to reactive inflammation surrounding the torsed omental implant.

In several reports, LPD has manifested as widespread peritoneal masses, mimicking carcinomatosis and leading to unnecessary radical treatment.^(1,10,11) In our patient, only two lesions were detected radiologically; this would be considered an unusual manifestation of LPD. Aggressive fibromatosis (desmoid) may be a differential diagnosis for multiple solid intra-abdominal masses, although the relative lack of mesenteric retraction suggests otherwise. Tuberculosis infection and peritoneal carcinoma are other conditions resembling LPD.⁽¹⁾

Pre-operative diagnosis of LPD is challenging, as it typically appears as well-circumscribed solid nodules on cross-sectional imaging, which is a nonspecific finding.⁽¹⁾ However, co-existing uterine leiomyomas and the absence of omental caking and ascites suggest LPD.⁽¹⁾ Similar to uterine leiomyomas, the lesions are often heterogeneous on CT attenuation, with enhancement that is similar to uterine leiomyomas.⁽¹⁾ On magnetic resonance imaging, the nodules are isointense to muscle, enhance and show low signal on T2-weighted images.^(1,6)

When nodules of LPD are of a sufficient size (approximately > 6 mm), fluorine 18 fluorodeoxyglucose (¹⁸F-FDG) positron emission tomography (PET) may be used to distinguish isometabolic activity of LPD from hypermetabolic uptake of leiomyosarcoma.⁽¹²⁾ However, a recent review has found that FDG PET cannot be used to reliably differentiate uterine leiomyosarcoma from uterine leiomyoma, as the former rarely shows mild FDG uptake and the latter rarely shows high FDG uptake.⁽¹³⁾

When a distinction cannot be made pre-operatively, intra-operative frozen section analysis may be necessary.⁽¹¹⁾ Histologically, these nodules are composed of fascicles of eosinophilic spindle-shaped cells lacking nuclear atypia or necrosis, with few to no mitotic figures and an absence of vascular invasion.⁽¹⁾ Treatment methods include surgical excision,⁽⁹⁾

oophorectomy,⁽¹⁴⁾ gonadotropin-releasing hormone analogues⁽¹⁵⁾ and aromatase inhibitors.⁽¹⁶⁾ Follow-up imaging is recommended due to the risk of recurrence and malignant transformation.⁽⁹⁾

In conclusion, most reported cases of LPD resemble peritoneal carcinomatosis on imaging but are clinically silent. Acute presentation is rare. We have presented a highly unusual case of LPD resulting in omental torsion, with clinical and radiological signs that mimicked acute appendicitis.

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