A rare benign condition that mimics malignancy: disseminated peritoneal leiomyomatosis

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Learning Objective

Disseminated Peritoneal Leiomyomatosis (DPL) is a rare benign condition that can mimic disseminated peritoneal malignancy.

Its presentation often creates significant diagnostic challenges and therefore highlights the importance of a multidisciplinary approach to ensure optimal patient care.

Background

- Disseminated Peritoneal Leiomyomatosis (DPL) is a rare benign condition, with fewer than 200 cases reported worldwide [1].
- It is characterised by multiple leiomyomas (smooth muscle nodules) in the pelvic cavity and lower abdomen. The condition predominantly affects women of reproductive age, with a reported average age of 38.5 years [1].
- Macroscopically, DPL presents as multiple nodules of varying sizes located on or beneath the peritoneal surface, often mimicking peritoneal malignancy. However, histological evaluation typically demonstrates benign features [1,2].
- This highlights the importance of a multidisciplinary team approach for accurate diagnosis and appropriate management for the patient.
- Definitive diagnosis is confirmed through histological examination.

Pathogenesis

- Pathogenesis remains incompletely understood. Two principal aetiological theories have been proposed: the hormone theory and the iatrogenic theory [1,2].
- The hormone theory postulates that high levels of exogenous or endogenous female gonadal steroids stimulate smooth cell differentiation. This hypothesis is supported by the increased prevalence of DPL in women of reproductive age and its association with pregnancy or use of oral contraceptives [1,2].
- The iatrogenic theory suggests that DPL may result from the dissemination of uterine leiomyoma tissue during surgical procedures, particularly following morcellation at the time during myomectomy, leading to implantation and subsequent growth of leiomyomatous tissue within the peritoneal cavity [1,2].

Case Presentation

- 37-year-old female (Gravida 2, Para 1).
- No relevant past medical or surgical history.
- No relevant family history.
- 11/40 weeks pregnant was referred for suspicious appearance of her uterus on a routine antenatal dating scan.

Imaging Findings

- **Figure 1 and 2:** Initial antenatal ultrasound imaging:
  11/40 weeks' gestation.
  Heterogenous and suspicious appearance of the uterus and enlarged bilateral adnexal lesions.

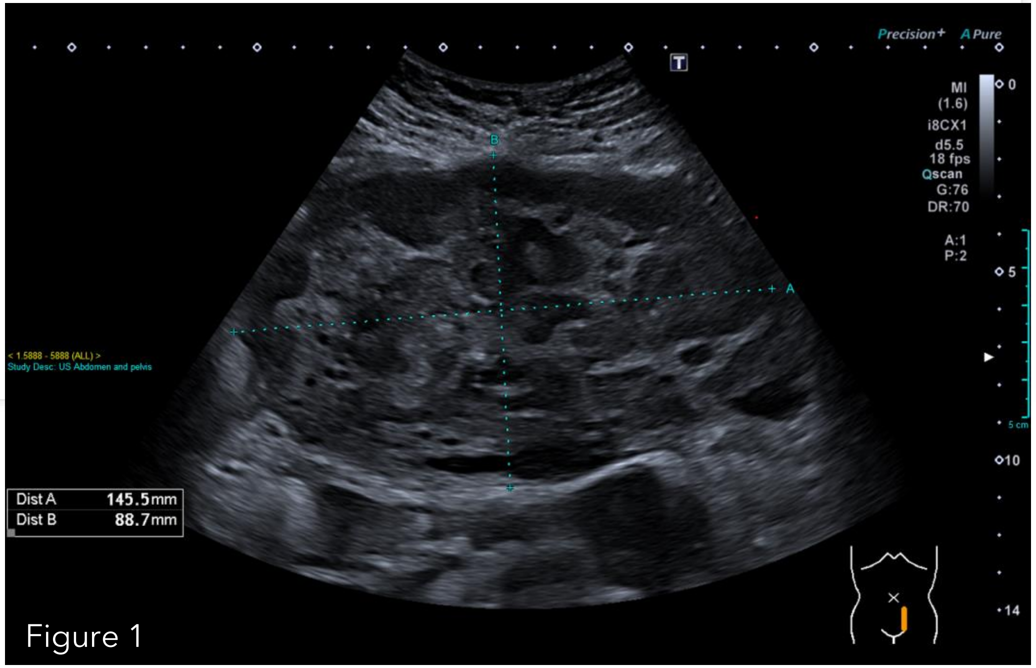

Figure 1

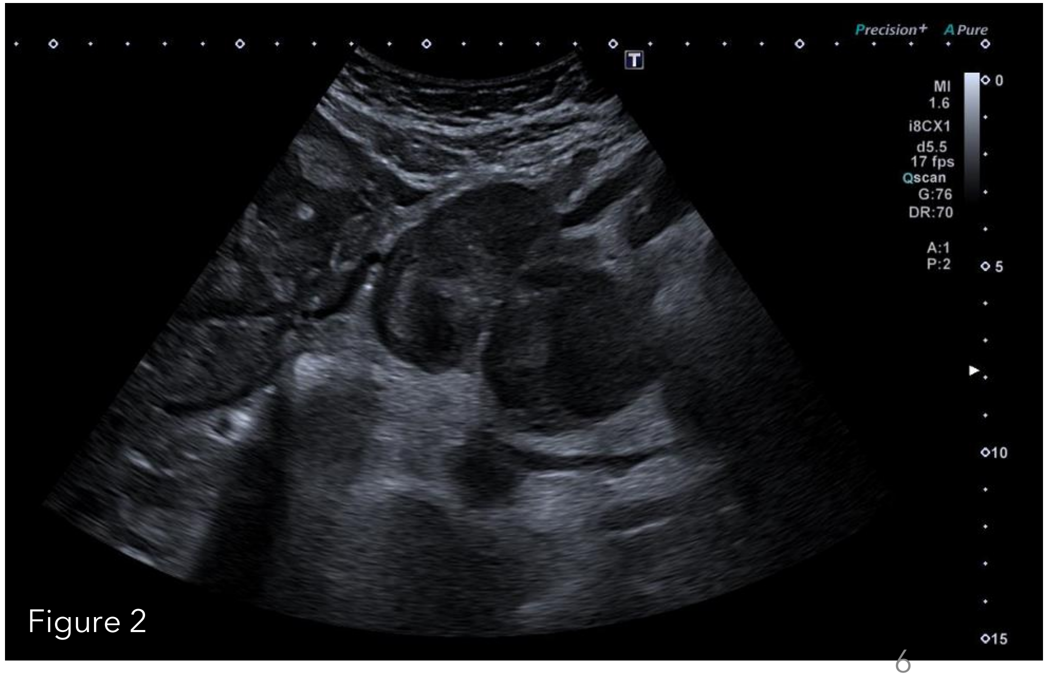

Figure 2

Imaging Findings

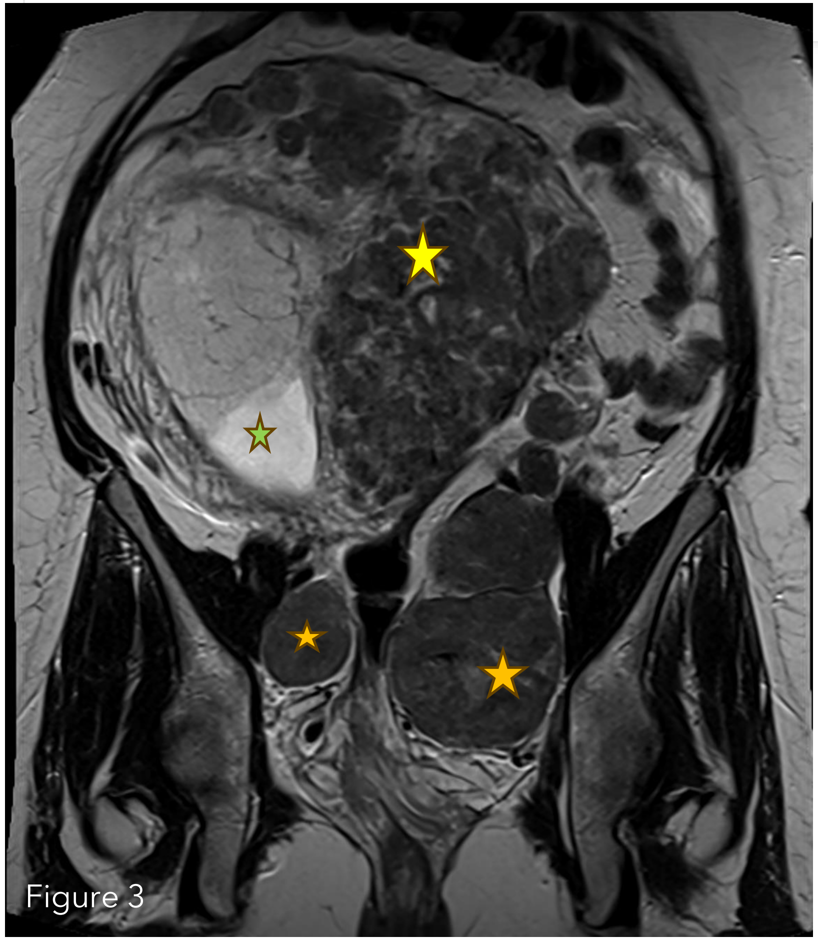
Figure 3

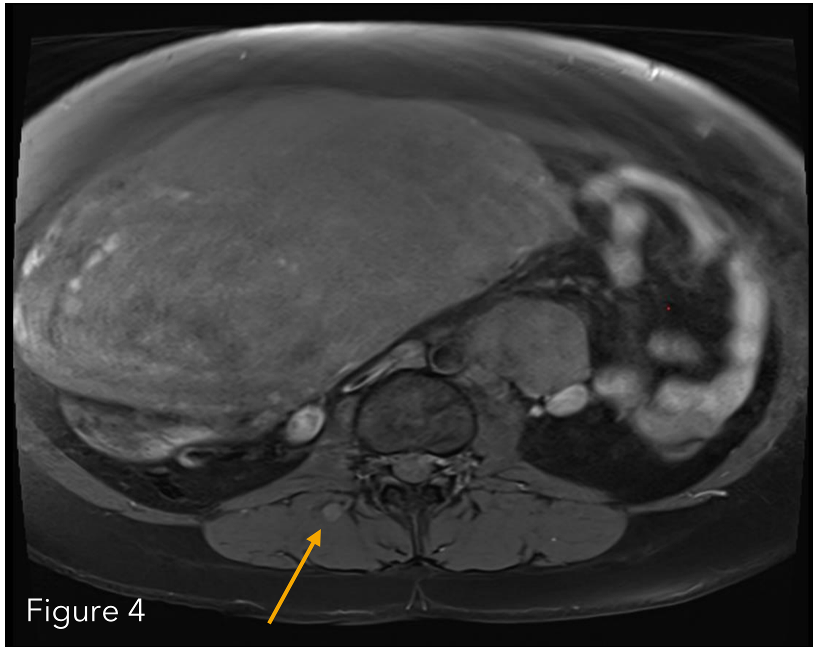
Figure 4

- **Figure 3:** T2w non-contrast MRI abdomen and pelvis: gravid uterus with a single foetus in the endometrial cavity (green star). Large heterogenous uterine mass (yellow star) and extensive bilateral pelvic lymphadenopathy (orange stars).

- **Figure 4:** T1w fat saturated non-contrast MRI abdomen and pelvis: intramuscular soft tissue deposit (yellow arrow).

- **Other findings:**
  - Non-contrast MRI abdomen and pelvis: 5cm right iliac fossa fat containing focus could represent right ovarian dermoid cyst. Extensive left iliofemoral DVT.
  - Non-contrast CT chest: several bilateral tiny nonspecific pulmonary nodule

Sarcoma MDT Discussions

- Multiple MDT discussions with diagnosis of uncertainty and a wide range of differentials from imaging include metastatic germ cell tumour, sarcoma and carcinoma.

- Other consideration:
    - To continue with pregnancy with 4 weekly interval MRI scan during pregnancy for monitoring.
    - To liaise with Obstetrics, Haematology and Vascular Surgery regarding plan for delivery.

- **Initial MDT outcome:** tumour markers, US guided biopsy of abnormal uterine fibroid.

Imaging Findings

- **Figure 5 and 6:** US guided biopsy of abnormal uterine wall (yellow star) adjacent to engorged uterine vessels and left pelvic sidewall lymph node.

- **This yielded the antenatal diagnosis.**

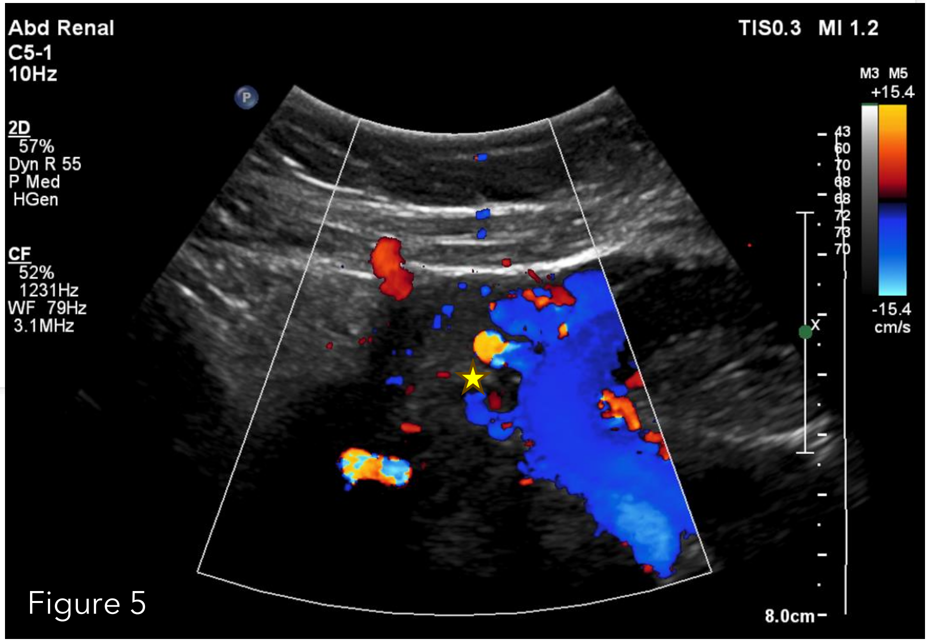

Figure 5

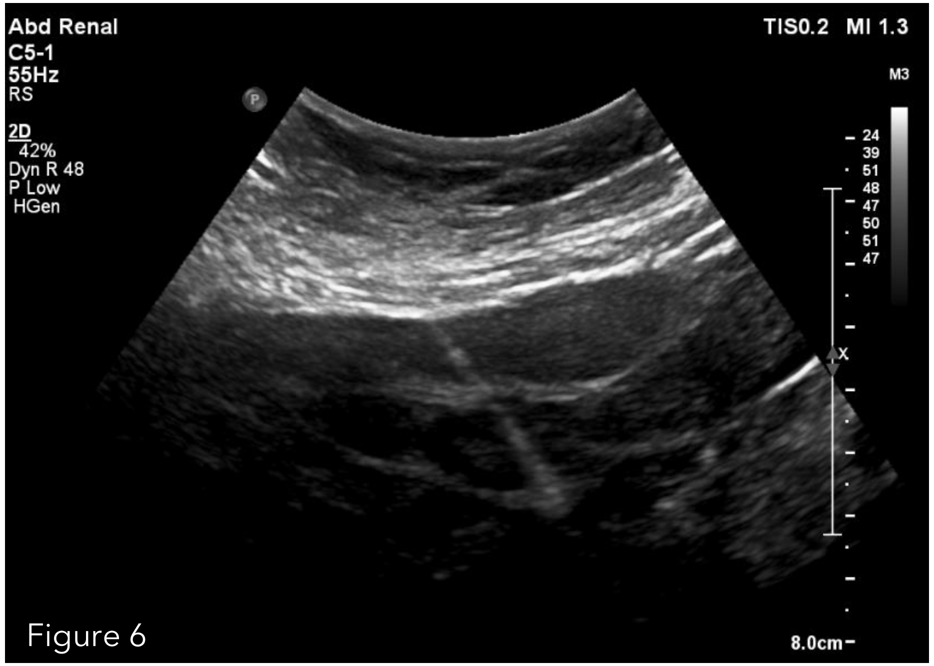

Figure 6

Histopathology

- Core biopsy of left pelvic side wall lymph node and abnormal uterus
- The features in both biopsies are suggestive of smooth muscle neoplasm with no histological evidence of malignancy.
- The possibility of metastasising leiomyoma should be considered.

Management

- **Biochemical tumour markers:** CA-125, CA15-3, CA19-9 are negative. Slightly raised LDH 347 U/L

- **Surgical management:** interval monitoring MRI during pregnancy and for elective surgery at 34+2 gestation
  - Midline laparotomy for Caesarean section, total abdominal hysterectomy, bilateral salpingo-oophrectomy, PA node dissection, bilateral pelvic node dissection, ureterolysis, adhesiolysis and right iliac thrombectomy (due to use of aortic occlusion balloon during surgery)

Further Sarcoma MDT Discussion

- Final histology of surgical specimens shows multifocal bland smooth muscle proliferation, which is present in all the specimens. The overall features fits most with disseminated peritoneal leimyomatosis.
- No histological evidence of malignancy.

- **MDT outcome:** GnRH analogue injection as it is hormone sensitive tumour and post-partum 3 months CT surveillance.

Summary

- This case highlights the diagnostic challenges of DPL in pregnancy, its potential to mimic malignancy and consideration of the foetus.
- It underscores the importance of histological confirmation and the MDT approach to optimise maternal and foetal outcomes.

References

1. Bucuri CE, Ciortea R, Malutan AM, Oprea V, Toma M, Roman MP, Ormindean CM, Nati I, Suciu V, Simon-Dudea M, Mihu D. Disseminated Peritoneal Leiomyomatosis-A Challenging Diagnosis-Mimicking Malignancy Scoping Review of the Last 14 Years. Biomedicines. 2024 Aug 3;12(8):1749. doi: 10.3390/biomedicines12081749. PMID: 39200214; PMCID: PMC11351519.

2. Al-Talib A, Tulandi T. Pathophysiology and possible iatrogenic cause of leiomyomatosis peritonealis disseminata. Gynecol Obstet Invest 2010;69:239–44. doi:10.1159/000274487.