

Case 9868

Leiomyomatosis Peritonealis Disseminata: a case report

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Section: Abdominal Imaging

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Patient: 32 year(s), female

Authors' Institution

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Clinical History

A 32-year-old woman was referred to our Institution with a persistent abdominal distension lasting for two weeks.

On the admission she showed a normal serum haemoglobin level and thrombocytopenia.

She had a history of abdominal discomfort and uncommon pelvic pains.

These symptoms had been diagnosed and treated as a gastro-intestinal infection.

Imaging Findings

Digital radiography revealed a reduced expansion of the right costo-phrenic angle.

The ultrasound examination showed the presence of three heterogeneous masses characterised by hypervascularity and anechoic areas, determining dislocation of the bowel loops.

Further Imaging investigations, such as CT, confirmed the presence of many other solid masses in the peritoneal cavity, with heterogeneous attenuation and a c.e. similar to uterine leiomyomas.

The tumour in the retro-umbilical region measured 70 mm, the one in the sub-hepatic region reached an extension of 170 x 110 x 180 mm.

On the MR examination these tumours showed low signal intensities on T1 weighted (T1w) images, heterogeneous signals, including numerous hyperintense foci, on T2 weighted (T2w) images, depending on various degrees of oedema and cystic degeneration, and intense and diffuse enhancement after the administration of a Gadolinium contrast material.

Biopsy was performed and the histological diagnosis was smooth muscle tissue without necrosis.

Discussion

Leiomyomatosis Peritonealis Disseminata (LPD) was described first by Wilson and Peale in 1952; Taubert identified LPD as a pathological entity in 1965 and only about 100 cases were described in the English literature since then [1]. It affects mainly women in reproductive age, even if few cases affecting males have been described. Recently a familial case of LPD has been reported [2].

LPD resemble typical uterine leiomyomas at both gross and microscopic level and represent a rare benign proliferative process, characterised by multiple smooth muscle nodules developing through the peritoneum probably from Mueller's epithelium, which is distributed throughout the subperitoneal mesenchyme. The main hypothesis is that the smooth muscle nodules are derived from metaplasia of submesothelial cells.

A hormonal influence has been hypothesised (contraceptives, hormone replacement therapy, tamoxifen) [3]. Pregnancy has been reported as an independent factor. Many patients have a history of previous fibroid excision, with possible intra-abdominal deposition during surgery and a parasitic mechanism involved. Nevertheless the exact pathophysiology of LPD remains uncertain.

Most cases are asymptomatic and discovered incidentally during surgery. Symptoms include abdominal and pelvic pain, abdominal distension, urinary frequency and less commonly palpable mass, gastrointestinal bleeding and peritonitis.

Ultrasound and CT imaging features are useful to identify and stage the disease. The typical signal intensity patterns on MR images can be used to confirm the diagnosis: the masses of LPD are isointense relative to muscle on T1w sequences, have low signal intensity on T2w sequences and enhance heterogeneously following intravenous administration of Gadolinium-based contrast media. Like uterine fibroid tumors, large masses can show hemorrhagic, necrotic, myxoid and cystic degeneration.

Biopsy is requested and the histopathologic examination shows oxyphilic spindle-shaped cells without signs of malignancy, namely cytological atypia, mitotic features, coagulative type necrosis or invasiveness. Tumour cells are positive for vimentin, desmin and -SMA (Smooth muscle actin), and negative for keratin, S-100, CD34, C-kit.

Once the diagnosis is histologically confirmed, withdrawal of hormonal influence is often sufficient; alternatively patients can undergo surgical excision and/or therapy with Gonadotropin releasing hormone (GnRH) agonists.

Recurrence is possible and each recurrence is more likely to produce sarcomatous degeneration, which is uncommon (10%) [4]. In most reports of malignant LPD, no history of oestrogenous exposure or relationship with uterine leiomyomas was found, suggesting a different pathogenesis with a higher risk of malignant transformation. In such cases, surgical treatment is mandatory; adjuvant chemotherapy, bilateral salpingo-oophorectomy and hormonal therapy can be chosen [5].

Final Diagnosis

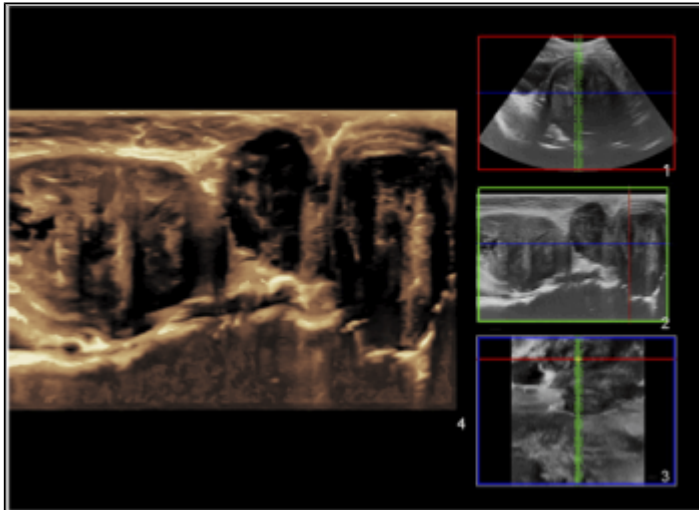
The final diagnosis was Leiomyomatosis Peritonealis Disseminata.

Differential Diagnosis List

Primary peritoneal serous carcinoma, Peritoneal carcinomatosis, Malignant mesothelioma

Figures

Figure 1 Ultrasonography



Panoramic imaging of the three main lesions showing the spatial relationship of nodules in the midsagittal plane.

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Area of Interest: Abdominal wall;
Imaging Technique: Ultrasound;
Procedure: Imaging sequences;
Special Focus: Neoplasia;

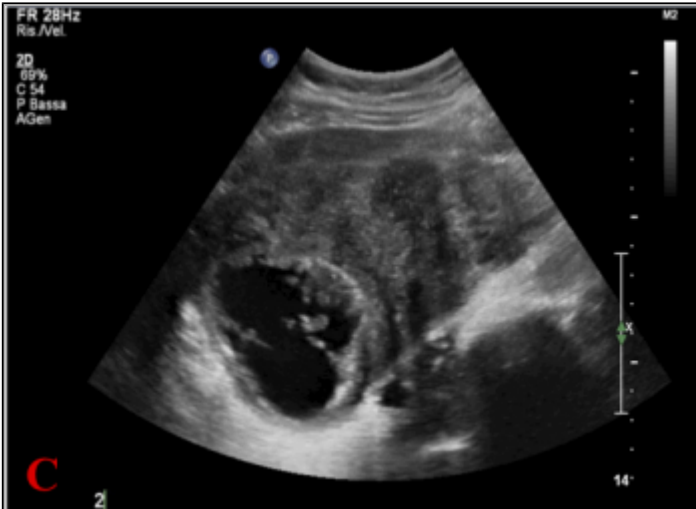


Transverse sonogram of the biggest nodule (13.6x9.7 cm) in the subhepatic region.

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Area of Interest: Abdominal wall;
Imaging Technique: Ultrasound;

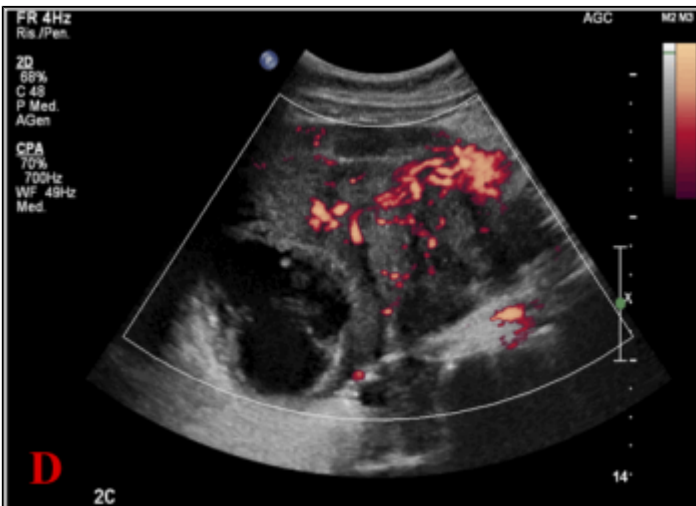
Procedure: Imaging sequences;
Special Focus: Neoplasia;



Grey-scale transverse sonogram revealing a heterogeneous structure and anechoic areas.

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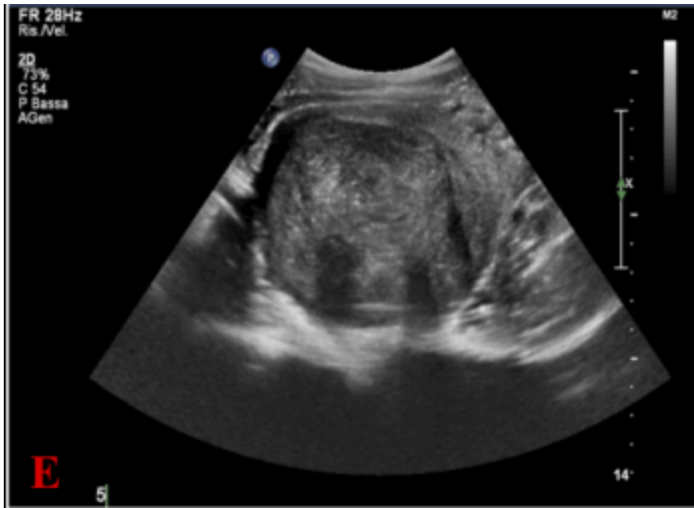
Area of Interest: Abdominal wall;
Imaging Technique: Ultrasound;
Procedure: Imaging sequences;
Special Focus: Neoplasia;



Power-Doppler sonogram of the same lesion showing high vascular supply.

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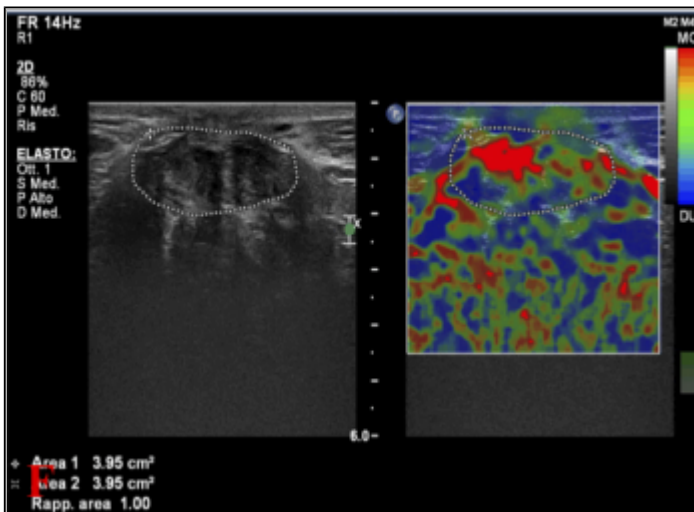
Area of Interest: Abdominal wall;
Imaging Technique: Ultrasound-Power Doppler;
Procedure: Imaging sequences;
Special Focus: Neoplasia;



Another mass arising from the uterus with the same echo-pattern.

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Area of Interest: Abdominal wall;
 Imaging Technique: Ultrasound;
 Procedure: Imaging sequences;
 Special Focus: Neoplasia;



The elasticity image of one of the lesions shows a low tissue stiffness, typical of soft tissues.

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Area of Interest: Abdominal wall;
 Imaging Technique: Elastography;
 Procedure: Imaging sequences;
 Special Focus: Neoplasia;

Figure 2 Venous phase CT

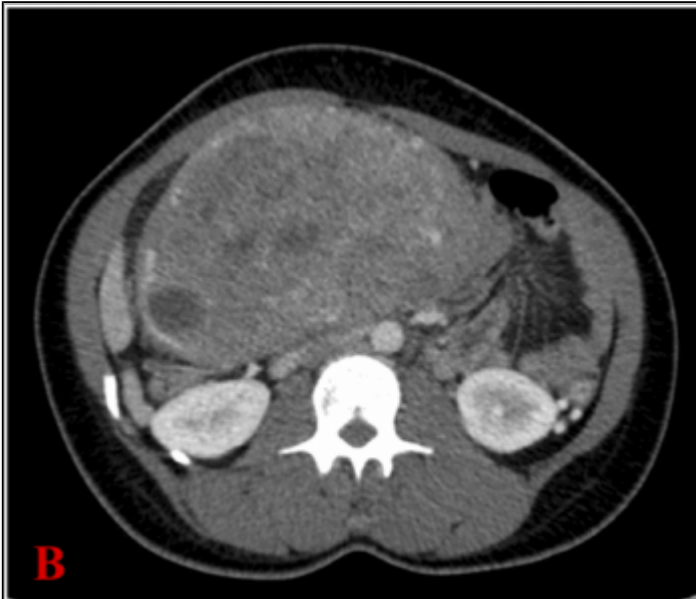




MPR reconstruction in the sagittal plane showing the continuity of the peritoneal lesions with the uterine ones.

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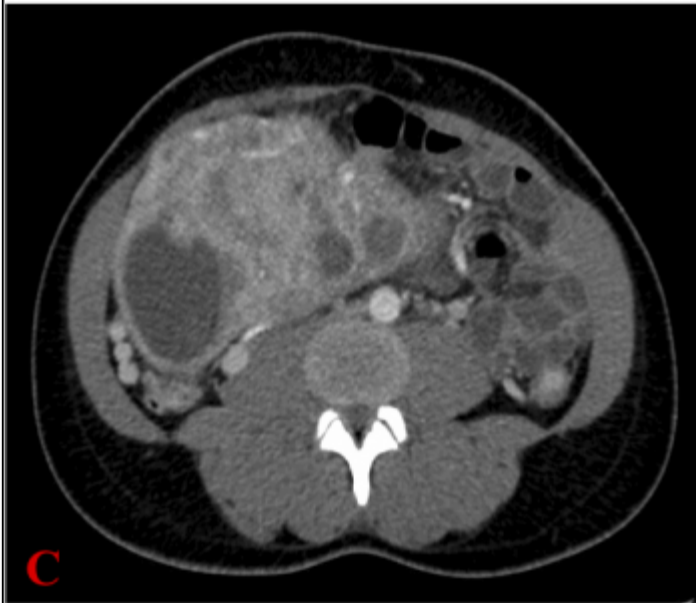
Area of Interest: Abdominal wall;
Imaging Technique: MR;
Procedure: Diagnostic procedure;
Special Focus: Neoplasia;



Here the lesion in the subhepatic region shows a heterogeneous attenuation and contrast enhancement with dislocation of the bowel loops.

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Area of Interest: Abdominal wall;
Imaging Technique: CT;
Procedure: Imaging sequences;
Special Focus: Neoplasia;



(cfr 1c-d) The lesion shows an extremely heterogeneous pattern with fluid collections.

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Area of Interest: Abdominal wall;
Imaging Technique: CT;
Procedure: Imaging sequences;
Special Focus: Neoplasia;



A lesion in the uterus mimicking a leiomyomatous uterus.

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Area of Interest: Abdominal wall;
Imaging Technique: CT;
Procedure: Imaging sequences;
Special Focus: Neoplasia;

Figure 3 MRI of the abdomen and the pelvis after Gadolinium injection

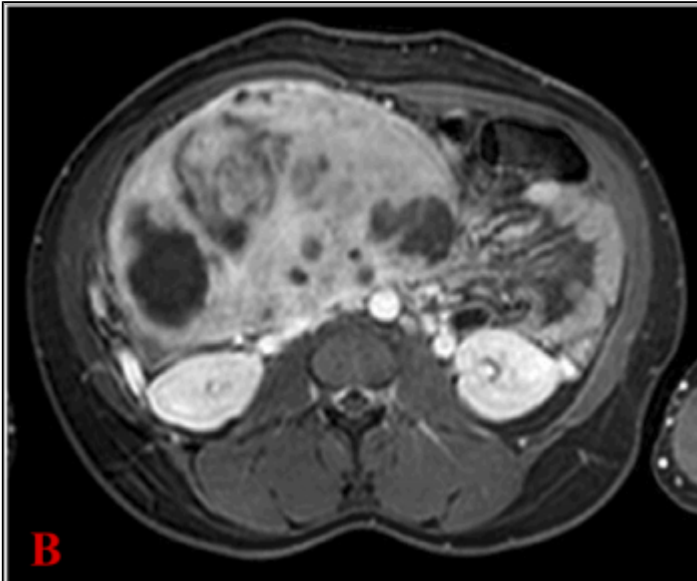




(cfr 2b) T2-weighted sequence showing heterogeneous hypointensity of the lesion and hyperintense fluid areas, representing cystic degeneration.

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Area of Interest: Abdominal wall;
Imaging Technique: MR;
Procedure: Diagnostic procedure;
Special Focus: Neoplasia;



(cfr 2b) Fast GE "THRIVE" sequence in the portal-venous phase showing heterogeneous c.e. of the lesion with dislocation of the bowel loops.

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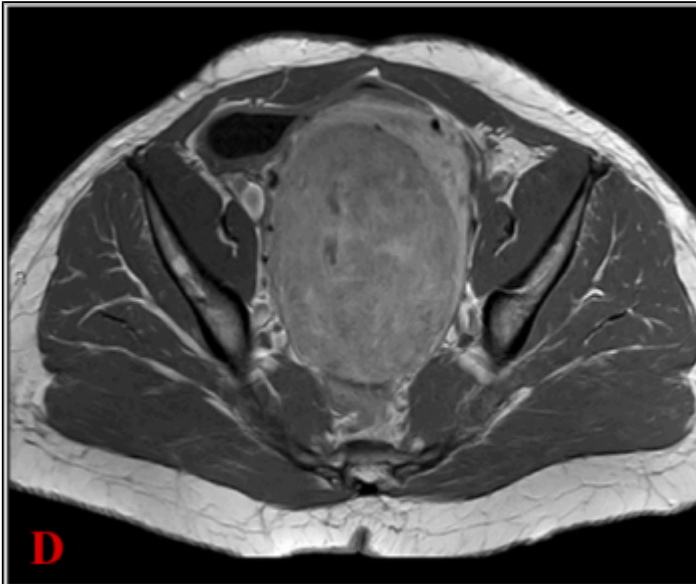
Area of Interest: Management;
Imaging Technique: MR;
Procedure: Imaging sequences;
Special Focus: Neoplasia;



T1-weighted sequence after Gadolinium injection in the sagittal plane showing the lesion within the uterus and other lesions in the peritoneal cavity

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Area of Interest: Management;
Imaging Technique: MR;
Procedure: Diagnostic procedure;
Special Focus: Neoplasia;

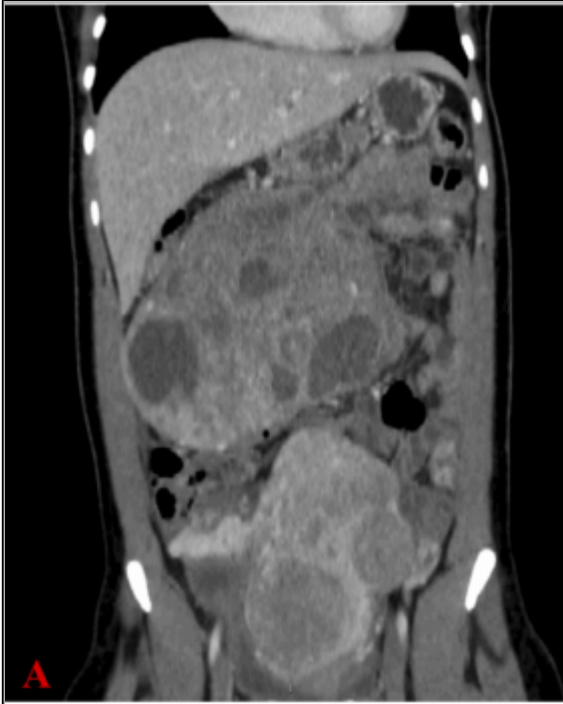


(cfr 2d) T1-weighted sequence after Gadolinium injection showing the lesion in the uterus.

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Area of Interest: Abdominal wall;
Imaging Technique: MR;
Procedure: Imaging sequences;
Special Focus: Neoplasia;

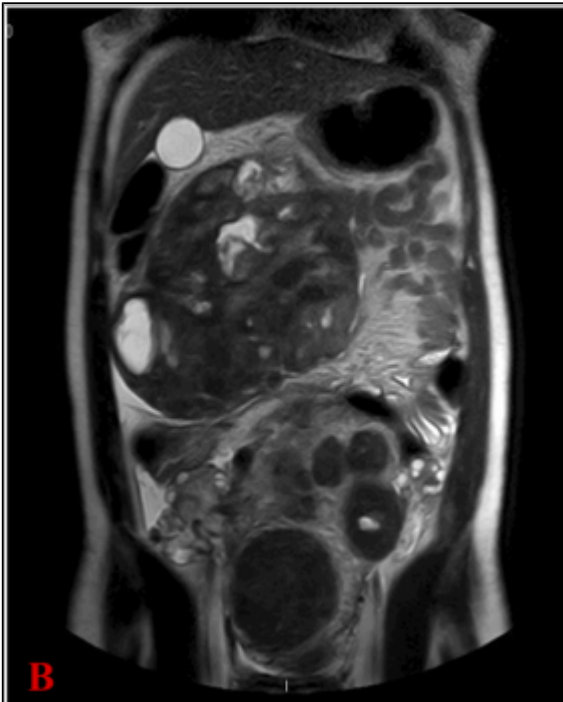
Figure 4 Coronal images



Venous phase CT showing the lesion occupying most of the abdomen, with dislocation of the bowel loops.

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Area of Interest: Abdominal wall;
Imaging Technique: CT;
Procedure: Imaging sequences;
Special Focus: Neoplasia;



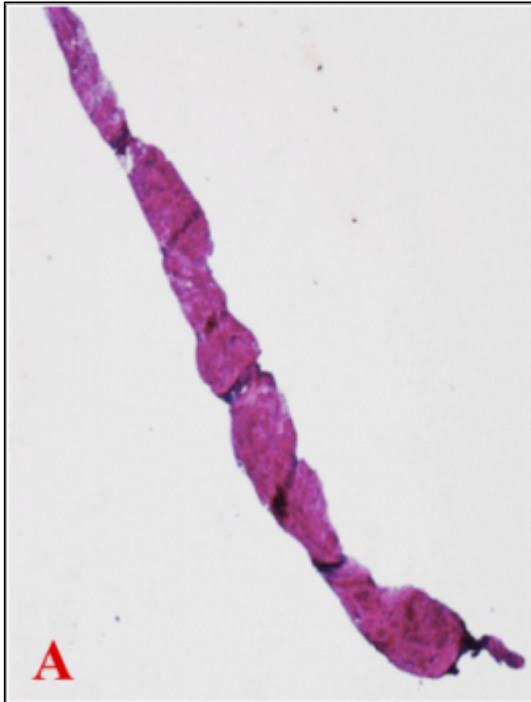
Coronal T2-weighted MR image demonstrating well defined multiple hypointense nodules with a heterogeneous pattern and fluid collections. No signs of infiltration are seen.

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Area of Interest: Abdominal wall;

Imaging Technique: MR;
Procedure: Imaging sequences;
Special Focus: Neoplasia;

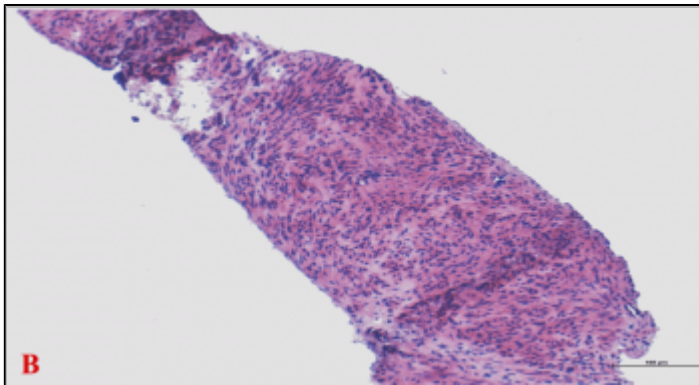
Figure 5 Histological examination, haematoxylin and eosin



The tissue sample, 2x

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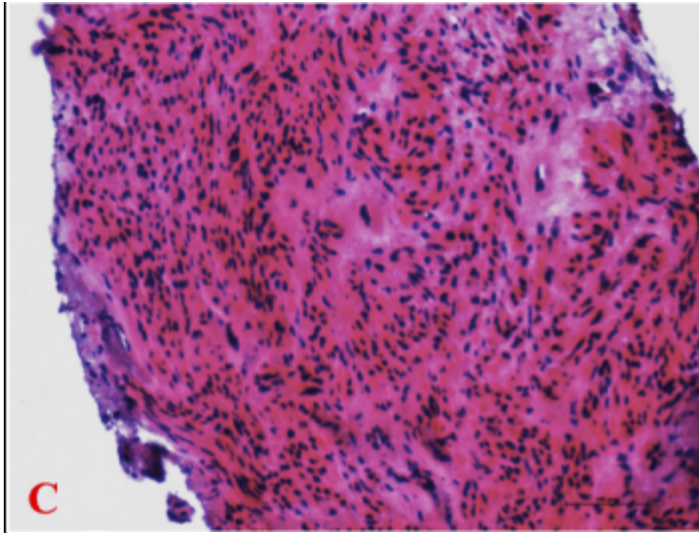
Area of Interest: Abdominal wall;
Imaging Technique: Experimental;
Procedure: Biopsy;
Special Focus: Neoplasia;



Microscopic appearance showing interdigitating fascicles of smooth muscle cells, 10x

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Area of Interest: Abdominal wall;
Imaging Technique: Experimental;
Procedure: Biopsy;
Special Focus: Neoplasia;



No cytological atypia and few mitoses are seen, 20x

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Area of Interest: Abdominal wall;
Imaging Technique: Experimental;
Procedure: Normal variants;
Special Focus: Neoplasia;

MeSH

Leiomyomatosis [C04.557.450.590.450.465]

The state of having multiple leiomyomas throughout the body. (Stedman, 25th ed)

Peritoneal Neoplasms [C04.588.033.513]

Tumors or cancer of the PERITONEUM.

References

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Citation

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