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Case 10714
Pelvic Lymphangioleiomyomatosis

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Section: Abdominal Imaging
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Patient: 33 year(s), female

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Clinical History

A 32 year-old woman presented to Accident and Emergency, in the evening, due to an acute exacerbation of chronic right iliac fossa pain. The pain had started two years previously. There was no relevant medical or drug history. On examination there were no significant findings.

Imaging Findings

Ultrasound (US) (Fig. 1) revealed a large septated cystic mass that occupied the right iliac fossa and the right side of the pelvic cavity.

Computed tomography (CT) (Fig. 2) demonstrated a low-density, well-defined mass that pushed the pelvic organs towards the contra-lateral side.

Magnetic resonance (MR) imaging (Fig. 3) showed a high signal mass on T2WI, with septa lacking invasive characteristics.

The right ovary was not identified on any of the imaging techniques.

Further imaging studies did not reveal any other lesions, namely lung or kidney involvement.

The patient underwent surgery in order to remove the cystic mass. Histopathologic analysis was performed, showing smooth muscle cells proliferation encasing lymphatic tissue.

Discussion

Lymphangioleiomyomatosis (LAM) is a rare systemic disorder characterised by the proliferation of bundles of smooth muscle cells around vessels and lymphatics. It affects almost exclusively women during childbearing years. There is no clear hereditary pattern, but it seems likely that the pathogenesis is associated with a mutation in the somatic TSC2 gene. Increased oestrogen states, such as pregnancy or therapy with exogenous oestrogens, seem to be associated with disease progression. However, oestrogen blockers have, thus far, shown little effect on disease control [1, 2].

LAM's most common manifestations, such as exertional dyspnoea and pneumothorax, are due to pulmonary involvement. Approximately half of the patients diagnosed with LAM have abdominal involvement [1]. Abdominal symptoms can, rarely, predate lung disease [2].

Abdominal manifestations of the disease include [1-3]:

- Renal angiomyolipomas, which are usually multiple, bilateral and symmetrical, occur in 20-54% of patients [1]. US, unenhanced CT and MR can accurately diagnose angiomyolipomas, due to the fat content. However, the absence of fat does not exclude the diagnosis. Multiple tumours and those larger than 4cm are more likely to increase and be symptomatic. Symptoms are due to haemorrhage resulting in pain, haematuria and hypovolaemic shock.

- Lymphangioleiomyomas are cystic masses resulting from the dilatation of lymphatic vessels due to obstruction by the proliferation of smooth muscle cells. They are more common in the abdomen, retroperitoneum and pelvis, but can occur in the mesdiastinum and neck. These occur in 20% of patients diagnosed with LAM[1]. Their size and wall thickness vary. They differ from lymphangiomas in that the latter lack smooth cells on their walls. Patients usually present with nausea, bloating, abdominal distension, peripheral oedema and urinary symptoms. These symptoms may worsen during the day, due to the increase in size of the lymphangioleiomyomas. About 10% of patients will develop chylous ascites, which results from overdistension and consequent rupture of lymphatic cysts. The patient presented with a pelvic lymphagioleiomyoma, prior to pulmonary involvement.

-Abdominal lymphadenopathies, normally retroperitoneal or retrocural and, occasionally, pelvic are visible on CT in approximately 33% of patients [3]. These can present, on unenhanced CT, low attenuation areas indicative of chylous collections or hamartomas. Hamartomas will enhance after contrast injection.

LAM is a slowly progressive illness that usually results in death due to respiratory failure. Currently there is no effective treatment.

Cystic septated masses located in the abdomen or pelvis can be seen in LAM, however the most common abdominal findings are renal angiomyolipomas.

Final Diagnosis

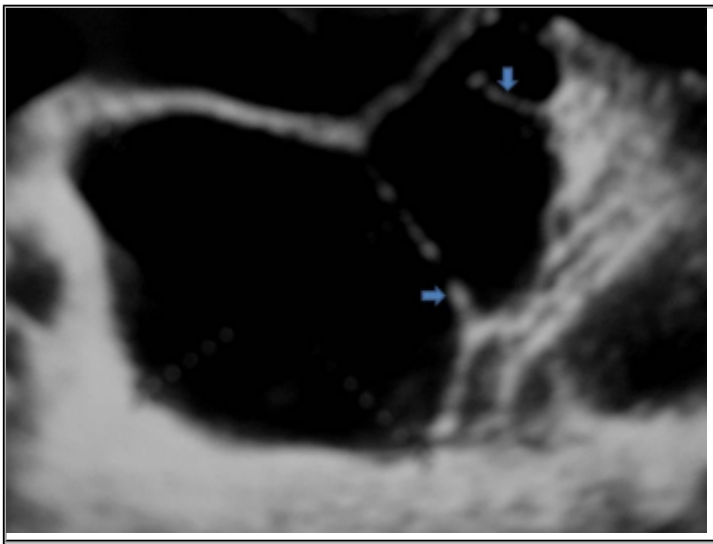
Pelvic Lymphangioleiomyomatosis

Differential Diagnosis List

Ovarian cyst, Lymphocele

Figures

Figure 1 Pelvic US



Pelvic US showing anechoic mass, with fine septa (arrows).

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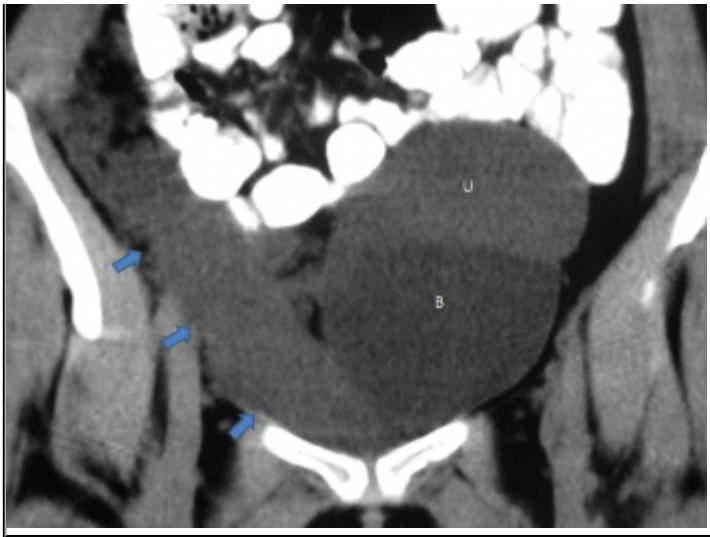
Area of Interest: Abdomen;
Imaging Technique: Ultrasound;
Procedure: Diagnostic procedure;
Special Focus: Cysts;

Figure 2 Pelvic CT (Coronal)



CT coronal reconstruction showing hypodense mass (blue arrows) on the right illiac fossa and right side of the pelvis, which pushes the bladder (B) and uterus (U) to the contralateral side.

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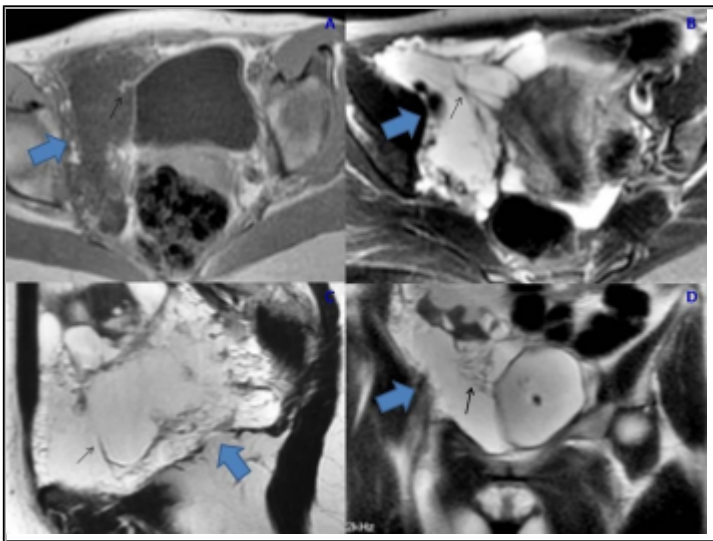
Area of Interest: Abdomen;
Imaging Technique: CT;
Procedure: Diagnostic procedure;
Special Focus: Cysts;

Figure 3 Pelvic MR



Cystic septated mass (blue arrow) with enhancing septa (black arrow). (A) Axial T1WI post paramagnetic contrast injection, (B) axial T2WI with fat saturation, (C) sagittal T2WI and (D) coronal T2WI.

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Area of Interest: Abdomen;
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MeSH

Lymphangiomyomatosis [C04.557.375.460.465]
A progressive disorder of women of child-bearing age, marked by nodular and diffuse interstitial proliferation of smooth muscle in the lungs, lymph nodes, and thoracic duct. (Dorland, 27th ed)

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[3] Meria R, Wikenheiser-Brokamp KA, Young LR, McCormack FX (2012) Lymphangioleiomyomatosis: new concepts in pathogenesis, diagnosis, and treatment Semin Respir Crit Care Med. 33(5):486-97

Citation

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