

Appendiceal mucinous neoplasm– a radiological conundrum

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LS

Ultrasound scan

This is a case presentation of a 54-year-old female who was referred from her GP on a 2WW pathway with a suspected pelvis mass. The Ca125 was normal (17). On reviewing symptoms with the patient, she reported pain and numbness in her upper right thigh.

A 18x9x8cm, hypoechoic, solid, intra-abdominal mass was identified which caused medial displacement of the right iliac vessels and protruded into the femoral canal. Transvaginal scan indicated an avascular mass of smooth outline and heterogenous echopattern. The suspicion of a psoas muscle sarcoma was raised, and a CT scan for staging and MR for further evaluation was recommended with the request to include the upper leg within the imaging.

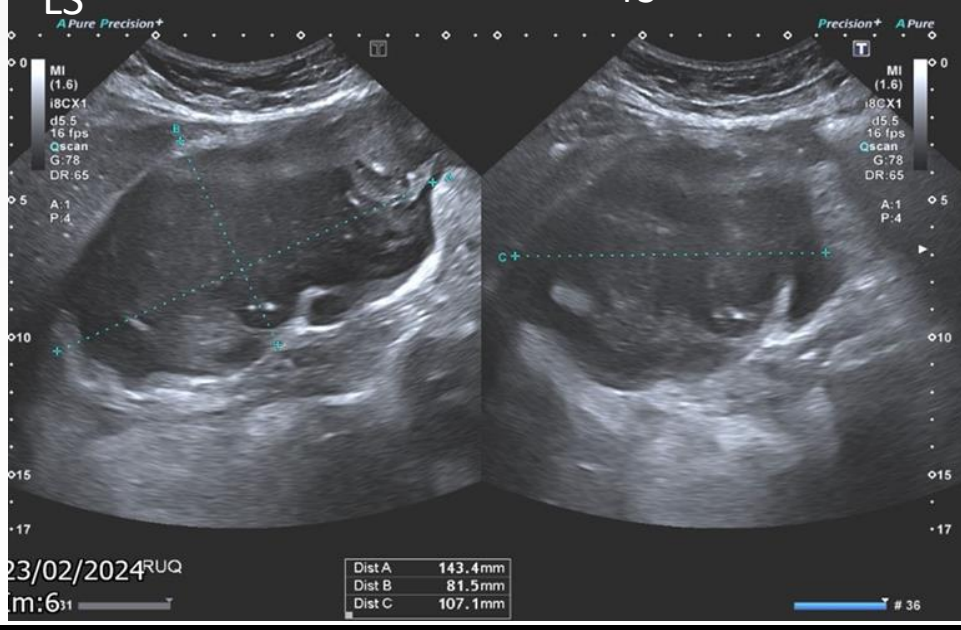


Image 1: Solid pelvic mass extending to liver

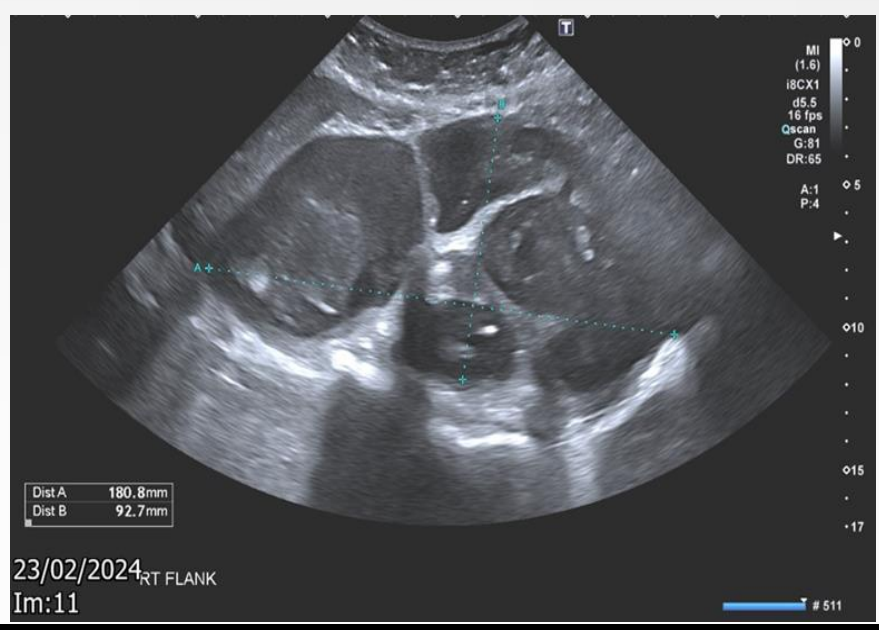


Image 2: Lobulated outline and septations seen

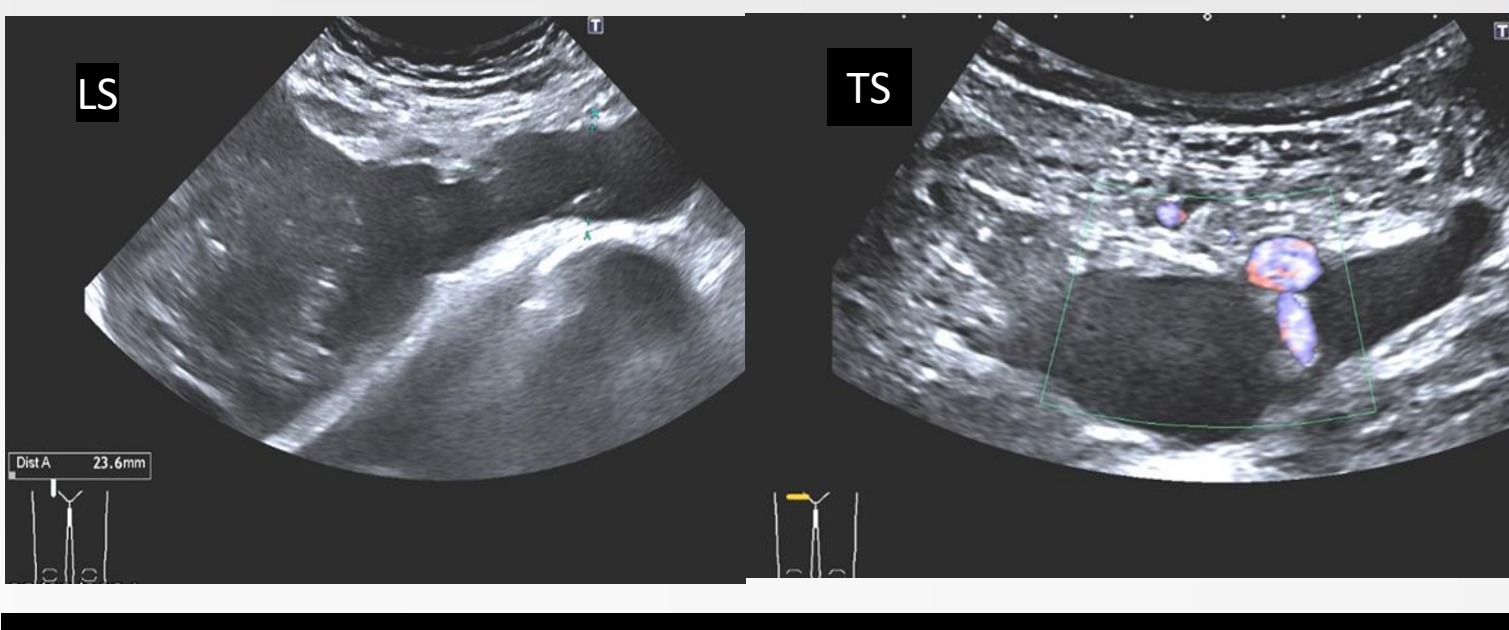


Image 3: Extension of mass into upper thigh via femoral canal

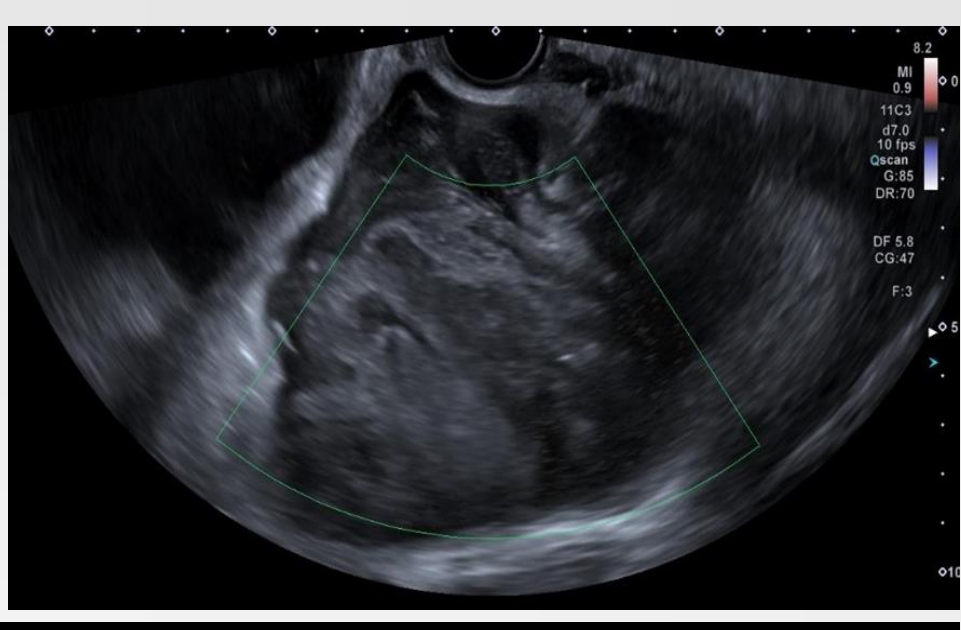


Image 4: Heterogenous appearance of mass

CT scan with contrast

CT confirmed a 25x12cm lobulated tumour within the right pelvis sidewall containing specks of calcium. The tumour involved the right iliopsoas muscle displacing the right iliac vessels medially and a tongue of tumour extending into the right femoral canal. A gynaecological tumour was excluded, and retroperitoneal sarcoma was suspected.



Image 5: Fleck of calcification within the mass

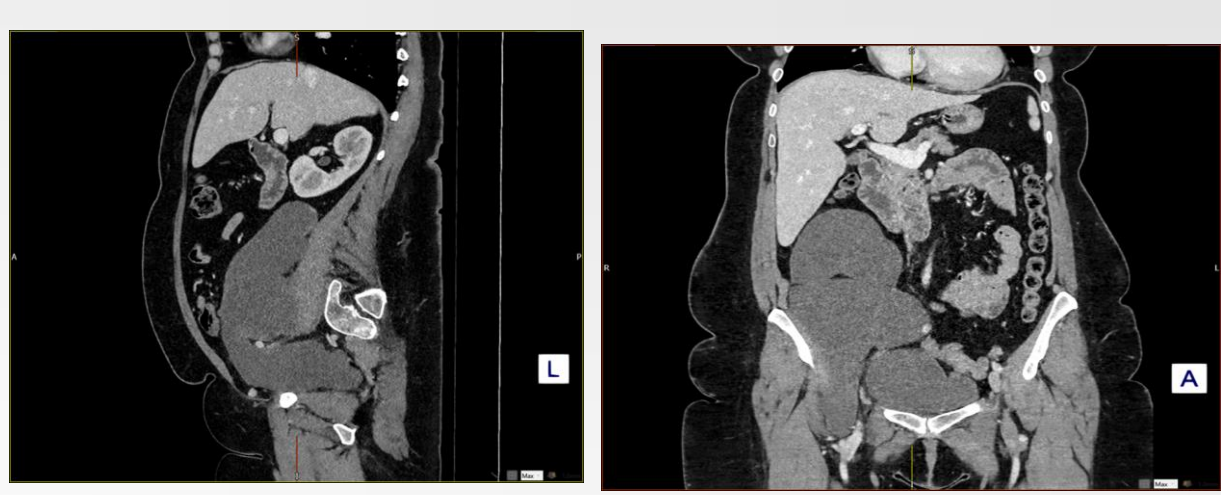


Image 6+7: Mass effect of lesion and extension down through right femoral canal

DVT Ultrasound

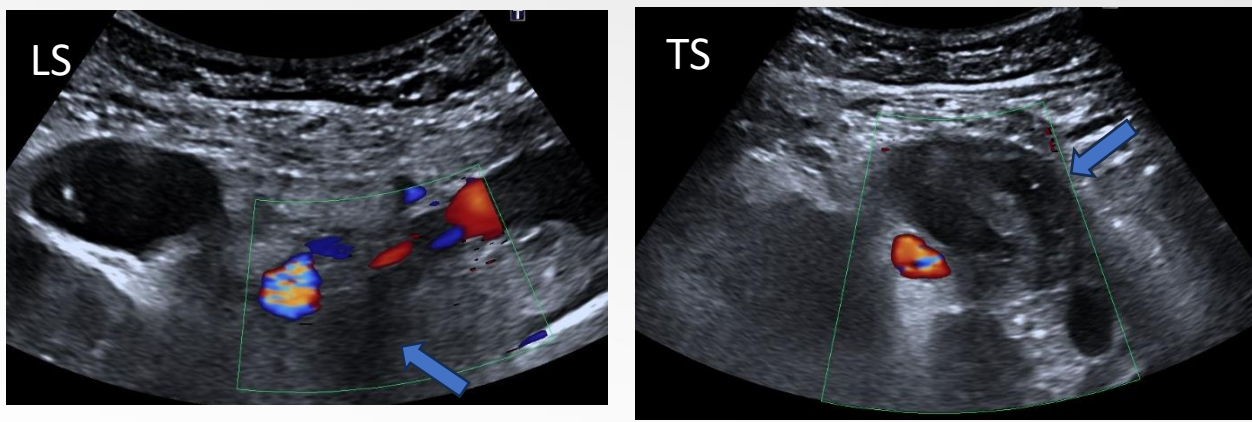


Image 8+9: Ultrasound Doppler of the right leg, showing no Doppler flow in the distal EIV (blue arrow on the LS image) and an extrinsic mass compressing the vein (blue arrow on the TS image)

The patient developed a swollen right leg and had a Wells score of 3. A DVT scan confirmed non-occlusive thrombus within the distal external iliac vein

Sarcoma MDT

In view of ongoing uncertainty, this case was further discussed at a sarcoma MDT and it was agreed following discussion between a retroperitoneal consultant surgeon and a consultant interventional radiologist to perform a CT-guided biopsy.

CT-guided biopsy results, Sarcoma and HPB MDTs

The CT-guided biopsy performed on the 3rd of April by a retroperitoneal consultant came back as a gelatinous material and was confirmed by histopathology as benign. The Sarcoma and HPB MDTs considered the biopsy and imaging results and identified the condition as a low-grade appendiceal mucinous neoplasm (LAMN), graded as pT4N0M1c (see table 1). This diagnosis is consistent with the raised CEA (flagged by the April Gynae MDT) which is noted in literature to be associated with mucinous neoplasm of appendiceal origin (Khan et al., 2017).

p	T4 (tumour stage)	N0	M1	c
primary tumour	tumour penetrates visceral peritoneum, including mucinous peritoneal tumour within the right lower quadrant	no regional lymph node metastasis	distant metastasis	further characterisation

Table 1. TNM stage classification (Shaib et al., 2017)

Post-operative CT to assess for recurrence

CT follow-up to exclude recurrence was performed and areas of soft tissue thickening were noted around paracolic gutters and the central laparotomy scar. It is difficult to say if this represents postsurgical change or early local recurrence. Therefore, a further CT in 3 months is recommended.



Images 15-17: 15: Soft tissue thickening at the superior left paracolic gutter. 16: Soft tissue thickening anteriorly on the far-right anterior abdominal wall. 17: Soft tissue thickening on right lateroconal (renal) fascia

Discussion

This case illustrates that a low-grade appendiceal mucinous neoplasm (LAMN) can pose a diagnostic challenge, and is commonly mistaken for an ovarian or bowel tumour or a sarcoma (Santos et al., 2022). LAMN are a diverse group of epithelial neoplasms which can cause, as in our patient, cystic dilation of the appendix due to accumulation of gelatinous material, morphologically referred to as a mucocele.

LAMN of the appendix is rare and although can be asymptomatic, it may cause symptoms that can also be present in a sarcoma, including localised mass, pain, venous compression and weakness of hip movements (Hammouche et al., 2010). If rupture occurs it can seed mucin and neoplastic epithelium into the peritoneum causing pseudomyxoma peritonei (PMP), which can cause life threatening complications and change the prognosis dramatically (Santos et al., 2022).

Of note, an 'onion-skin' appearance on ultrasound due to internal concentric echogenic layers of mucin (images 18+19) is pathognomonic for LAMN (Tirumani et al., 2013), although it was not observed in our patient. The internal echotexture of the mucocele of the appendix varies and can include dystrophic mural calcifications that may produce acoustic shadowing (Santos et al., 2022). These calcifications were not seen on ultrasound in this case but were noted on CT.

Conclusion

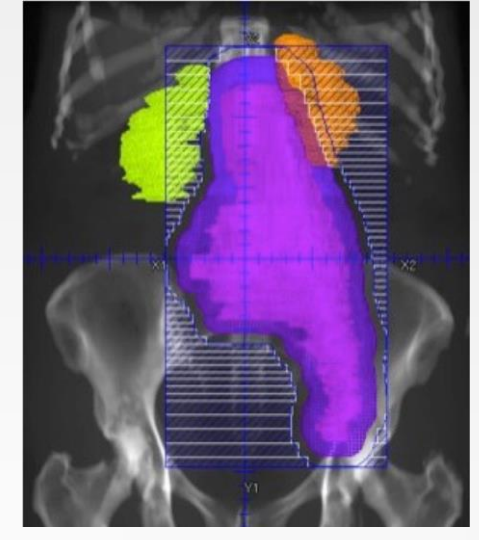
When presented with a well defined, avascular abdominal-pelvic mass, consider LAMN, which although rare can have significant morbidity/implications. Although not always present, the onion-skin appearance is pathognomonic for making a diagnosis. Presence of calcification and raised CEA may also give further clues.

Reporting the ultrasound scan

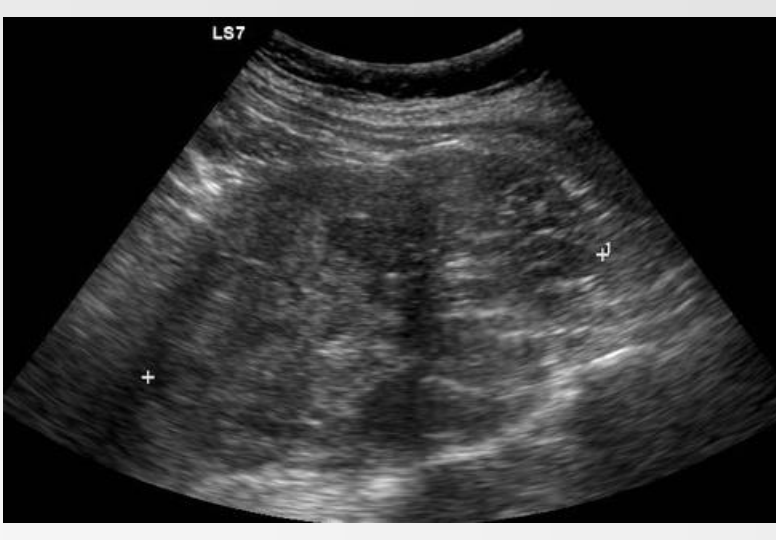
This unusual mass was difficult to characterise, and we therefore performed a literature search to help form a differential diagnosis. The search identified cases presenting masses of similar appearance which resulted from iliopsoas sarcoma (see images below). This discovery informed us when we made a differential diagnosis of iliopsoas sarcoma.



Coronal CT + C portal venous phase for a retroperitoneal sarcoma (Haque, 2024)



Psoas sarcoma digitally reconstructed radiograph, kidneys are shaded in green and orange, whilst the radiotherapy target volume is in purple (McKay et al., 2017)



Iliopsoas sarcoma ultrasound (Knipe, 2024)

Gynaecology MDT

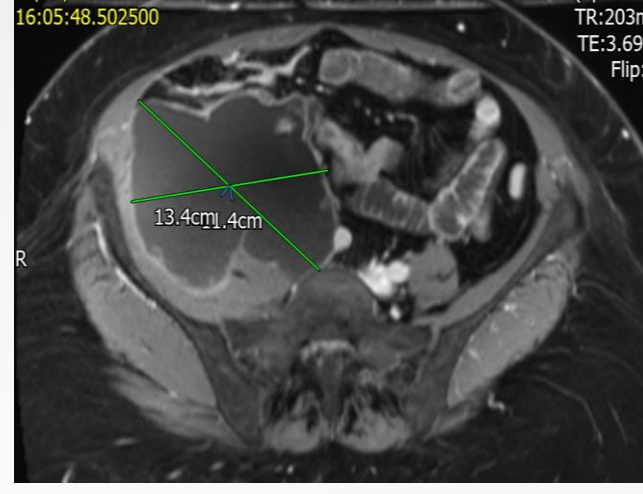
The images were reviewed at gynaecology MDT which agreed that the large mass centred on the right iliopsoas was in keeping with a retroperitoneal sarcoma, and not arising from the uterus. It was agreed that the mass extended into the thigh and retropubic space. The case was referred to the Sarcoma MDT.

MRI scan with contrast

MRI showed a 25x13x11cm non-infiltrative lesion with well-defined margins. Some debris was noted within the lesion, otherwise no definite solid elements were identified. MRI was not able to accurately characterise this mass however raised suspicion of a large iliopsoas bursa or a myxoid tumour



Images 10+11: MRI with contrast showing post contrast enhancement of the wall of the lesion with possibly some debris within the lesion and no definite solid elements



Images 12+13: MRI without contrast showing the shape of this large mass

Gynaecology MDT

CT images were re-visited at the gynae MDT and a fleck of calcification was noticed along with a subtly thickened appendix, which was abutting, and likely inseparable, from the mass. A further gynae MDT in April also flagged up markedly elevated carcinoembryonic antigen (CEA).

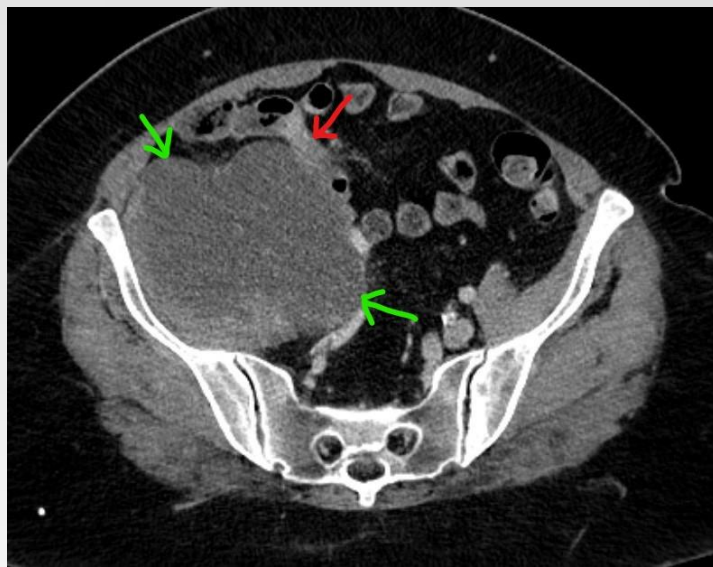


Image 14: Thickening of the appendix: the red arrow points at the thickened appendix. The green arrows point to the massive cystic mass abutting the appendix, in keeping with a LAMN

Treatment and management

According to NICE a patient with this type of diagnosis requires specialist treatment at a dedicated treatment centre of which there are only two in the UK: our patient was treated at the Peritoneal Malignancy Institute at Basingstoke and North Hampshire NHS Foundation Trust, (the other centre is the Colorectal and Peritoneal Oncology Centre in Manchester). Our patient underwent cytoreductive surgery (CRS), which involves careful removal of the appendiceal mucinous neoplasm with complete stripping of the peritoneum, followed by hyperthermic intraperitoneal chemotherapy (HIPEC), in which the surgeon uses heated chemotherapy drugs to bathe the peritoneal space of the entire abdomen.



Images 18-19: 'Onion-skin' appearance of an appendiceal mucocele (Caspi et al., 2004)