

AMSER Case of the Month

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57-year-old female with increased abdominal girth and a palpable abdominal mass

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Patient Presentation

- HPI: 57 y/o female G2P2 presents to her PCP with the chief complaint of increasing abdominal girth for 6 months and a palpable abdominal mass. She denied any abdominal or pelvic pain, nausea, vomiting, urinary urgency or change in her bowel movements.
- PSHx: Two Caesarean sections, endometrial ablation, and R salpingo-oophorectomy for an ovarian torsion.
- OB/GYN: Pap smear (2/21) was negative for intraepithelial lesion or malignancy. LMP was 2017 following endometrial ablation.
- Fam Hx: Uterine Cancer (Mother), Ovarian cysts (Sister)
- Allergy: iodinated contrast material

Pertinent Labs

- CA-125: 38.4 U/mL (reference range: 0-35 U/mL)

What Imaging Should We Order?

Select the applicable ACR Appropriateness Criteria

Variant 1:

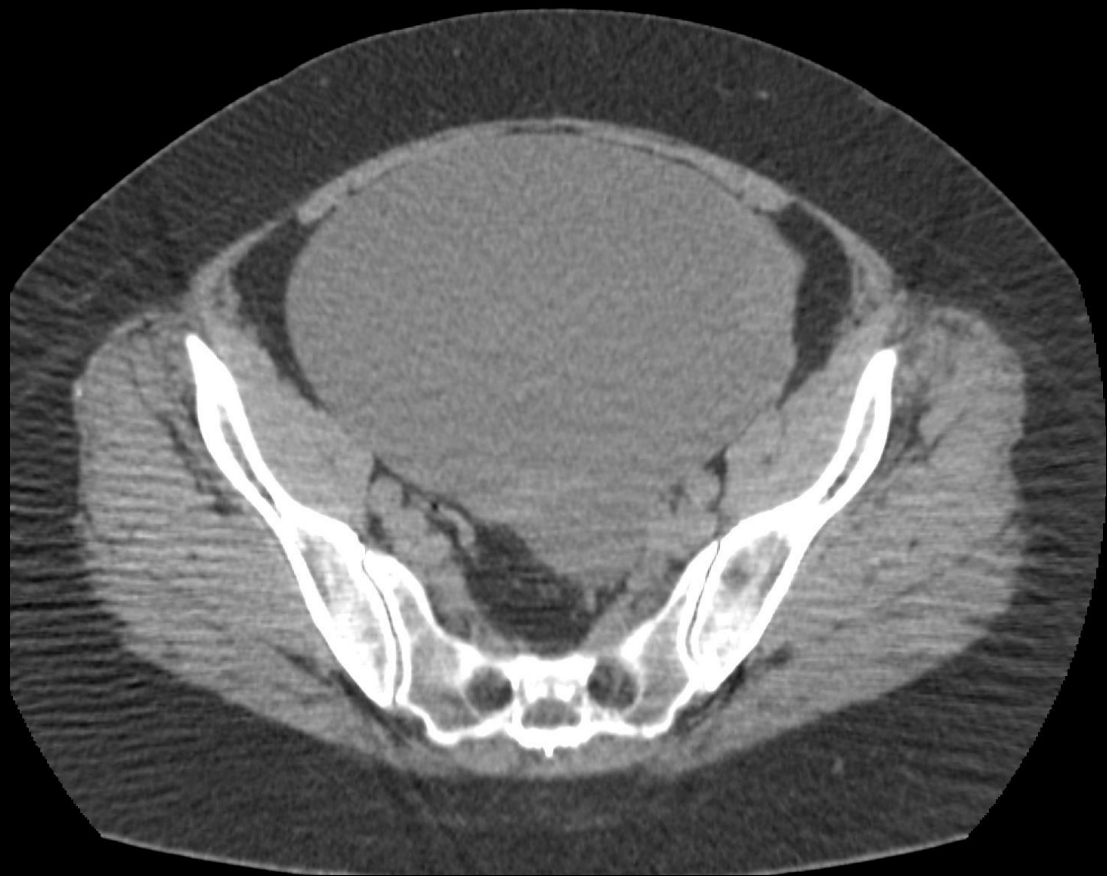
Palpable abdominal mass. Suspected intra-abdominal neoplasm. Initial imaging.

Procedure	Appropriateness Category	Relative Radiation Level
CT abdomen with IV contrast	Usually Appropriate	⊕⊕⊕
US abdomen	Usually Appropriate	○
MRI abdomen without and with IV contrast	May Be Appropriate	○
CT abdomen without IV contrast	May Be Appropriate	⊕⊕⊕
MRI abdomen without IV contrast	May Be Appropriate	○
CT abdomen without and with IV contrast	Usually Not Appropriate	⊕⊕⊕⊕
FDG-PET/CT skull base to mid-thigh	Usually Not Appropriate	⊕⊕⊕⊕
Radiography abdomen	Usually Not Appropriate	⊕⊕
Fluoroscopy contrast enema	Usually Not Appropriate	⊕⊕⊕
Fluoroscopy upper GI series	Usually Not Appropriate	⊕⊕⊕
Fluoroscopy upper GI series with small bowel follow-through	Usually Not Appropriate	⊕⊕⊕

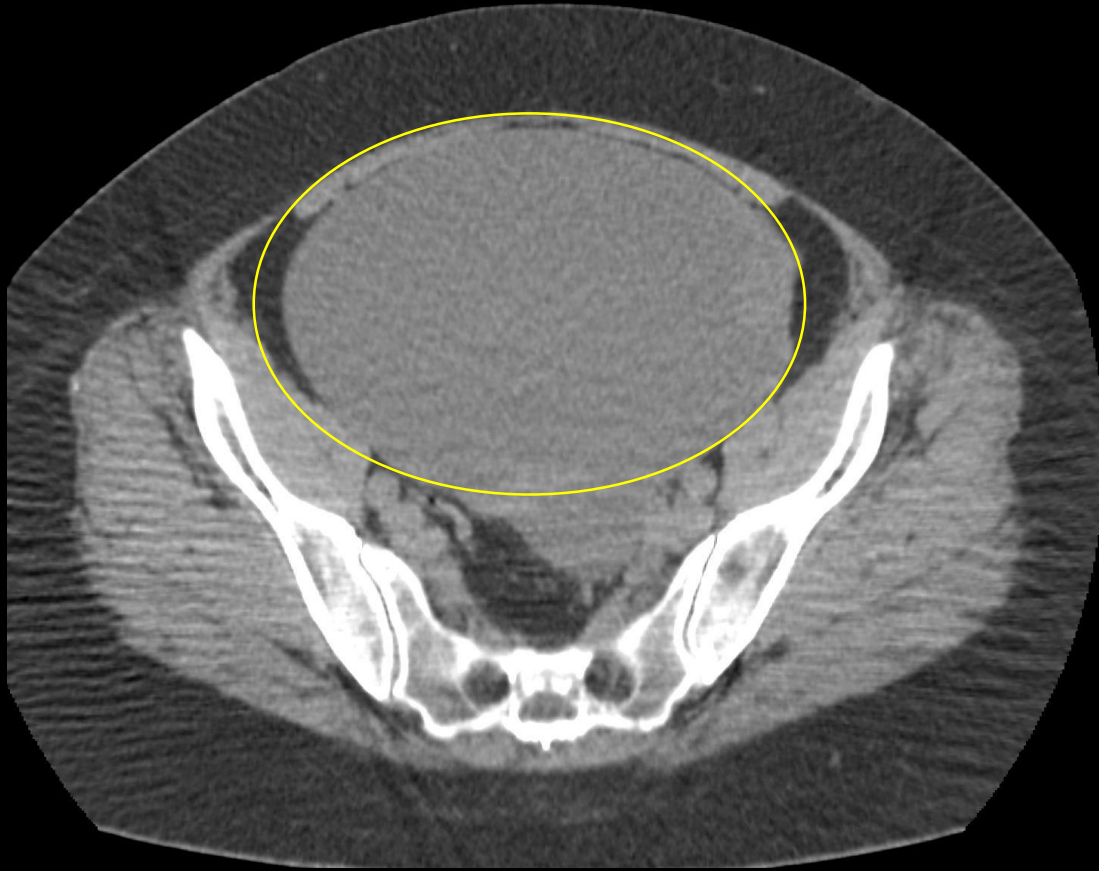
This imaging modality was ordered by the ER physician



Findings (unlabeled)



Findings: (labeled)



CT scan of the abdomen and pelvis revealed a low-density 18.9 x 13.1 x 16.7 cm ovarian mass with significant mass effect on the small bowel w/o obstruction located cephalad to the anterior aspect of the uterus. No adenopathy was noted. Ideally the scan would have been performed with IV contrast, but due to patient allergy, oral contrast was utilized. In the sagittal image, the * represents the bladder, the # represents the uterus, and the ovarian mass is circled.

Surgical Specimen



Patient underwent an exploratory laparotomy with a TAH and L salpingo-oophorectomy with radical dissection for neoplasm debulking, omentectomy, pelvic and periaortic lymphadenectomy, appendectomy and resection of peritoneal implants.

Left ovarian mass containing both cystic and solid components measuring 25cm.

Final Diagnosis

- Pathology revealed epithelial **clear cell carcinoma** confined to the L ovary.
- Pathology also revealed benign left and right pelvic lymph nodes as well as benign left and right para-aortic lymph nodes.
- Appendectomy and resection of peritoneal implants along the small bowel and sigmoid colon demonstrated no metastases.
- Cytology of peritoneal washings was negative for malignancy.

Case Discussion: Ovarian Clear Cell Carcinoma (OCCC) Presentation and Pathology

- OCCC may present with
 - acute symptoms: ascites, pleural effusion, bowel obstruction
 - subacute symptoms: bloating, abdominal distension, urinary urgency, early satiety, or pelvic/abdominal pain
- OCCC is responsible for ~10% of all epithelial ovarian carcinomas.
- It has a higher frequency in the East Asian population.
- It often presents at an early stage without distant metastases
 - poor prognosis if advanced due to its resistance to platinum based-chemotherapy
- OCCC is often associated with endometriosis and, less often with clear cell adenofibromas. It has also been associated with Lynch syndrome (DNA mismatch repair deficiency).
- The most common gene alterations in OCCC are mutations of AT-rich interaction domain1A (ARID1A) and phosphatidylinositol-4,5-bisphosphate 3-kinase catalytic subunit alpha (PIK3CA).

Case Discussion: Ovarian Clear Cell Carcinoma (OCCC) Presentation and Pathology (cont.)

- Typically presents as a large mass with an average size of 15cm. The tumor consists of thick-walled uni- or multilocular cysts with protruding solid nodules. Morphologic features include a combination of papillary, tubulocystic and solid patterns, combined with clear and eosinophilic cells and stromal hyalinization.
- It is associated with thromboembolic events and paraneoplastic hypercalcemia.
- OCCC expresses napsin-A, hypoxia-inducible factor 1 alpha, glypican-3 and hepatocyte nuclear factor 1-beta (HNF-1 beta) which is a sensitive and specific marker.

Radiology Diagnosis and Staging

Findings are non-specific but often present as:

- A unilocular large cyst with one or more solid nodular protrusions into the cavity of the mainly cystic mass.
- High-attenuated cystic portion on CT and MRI.

Staging is the same as general ovarian cancer staging with an emphasis on metastases to the lymph nodes and distant spread.

- Standard staging procedure consists of peritoneal cytology, visual assessment of the upper abdomen, peritoneal surfaces, and large and small bowel mesentery as well as other abdominal organs (appendix), hysterectomy and bilateral salpingo-oophorectomy, pelvic and paraaortic lymph node dissection and infracolic or infragastric omentectomy.

Treatment

- For women with high-risk ovarian carcinoma (stage IC or II, clear cell histology, or high tumor grade) adjuvant chemotherapy is recommended.
- Adjuvant chemotherapy is typically given to women with early-stage ovarian carcinoma approximately 21-35 days following debulking surgery.
- The National Comprehensive Cancer Network (NCCN) recommends IV platinum-based therapy with paclitaxel/carboplatin. Because of OCCC resistance to platinum-based chemotherapy, clinical trials are exploring the effectiveness of other agents like temsirolimus in combination with paclitaxel and carboplatin.

References:

- Iida, Yasushi, et al. “Clear Cell Carcinoma of the Ovary: a Clinical and Molecular Perspective.” *International Journal of Gynecologic Cancer*, 2020, doi:10.1136/ijgc-2020-001656.
- Taylor, Erin C., et al. “Multimodality Imaging Approach to Ovarian Neoplasms with Pathologic Correlation.” *RadioGraphics*, vol. 41, no. 1, 2021, pp. 289–315., doi:10.1148/rg.2021200086.
- Weerakkody, Yuranga. “Clear Cell Ovarian Carcinoma: Radiology Reference Article.” *Radiopaedia Blog RSS*, radiopaedia.org/articles/clear-cell-ovarian-carcinoma#nav_epidemiology.