AMSER Case of the Month August 2020

"Umbilical Hernia"

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Patient Presentation

- HPI: 33 yo male presenting with progressively enlarging umbilical region mass felt to represent an umbilical hernia, first noticed 8 months prior to visit
- ROS: Denies abdominal pain, N/V, fevers, chills, SOB, jaundice, change in bowel habits or appetite, urinary symptoms, weight loss
- PMH/Meds: None, no medications
- PSH: None
- SH: Never smoker, social alcohol use
- PE: Reducible umbilical hernia, otherwise within normal limits
- LABS: No pertinent labs

What Imaging Should We Order?
ACR Appropriateness Criteria

Variant 2: **Palpable abdominal mass. Suspected abdominal wall mass. Initial imaging.**

Procedure	Appropriateness Category	Relative Radiation Level
US abdomen	Usually Appropriate	O
CT abdomen with IV contrast	Usually Appropriate	☢☢☢
MRI abdomen without and with IV contrast	Usually Appropriate	O
MRI abdomen without IV contrast	May Be Appropriate	O
CT 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 surgeon

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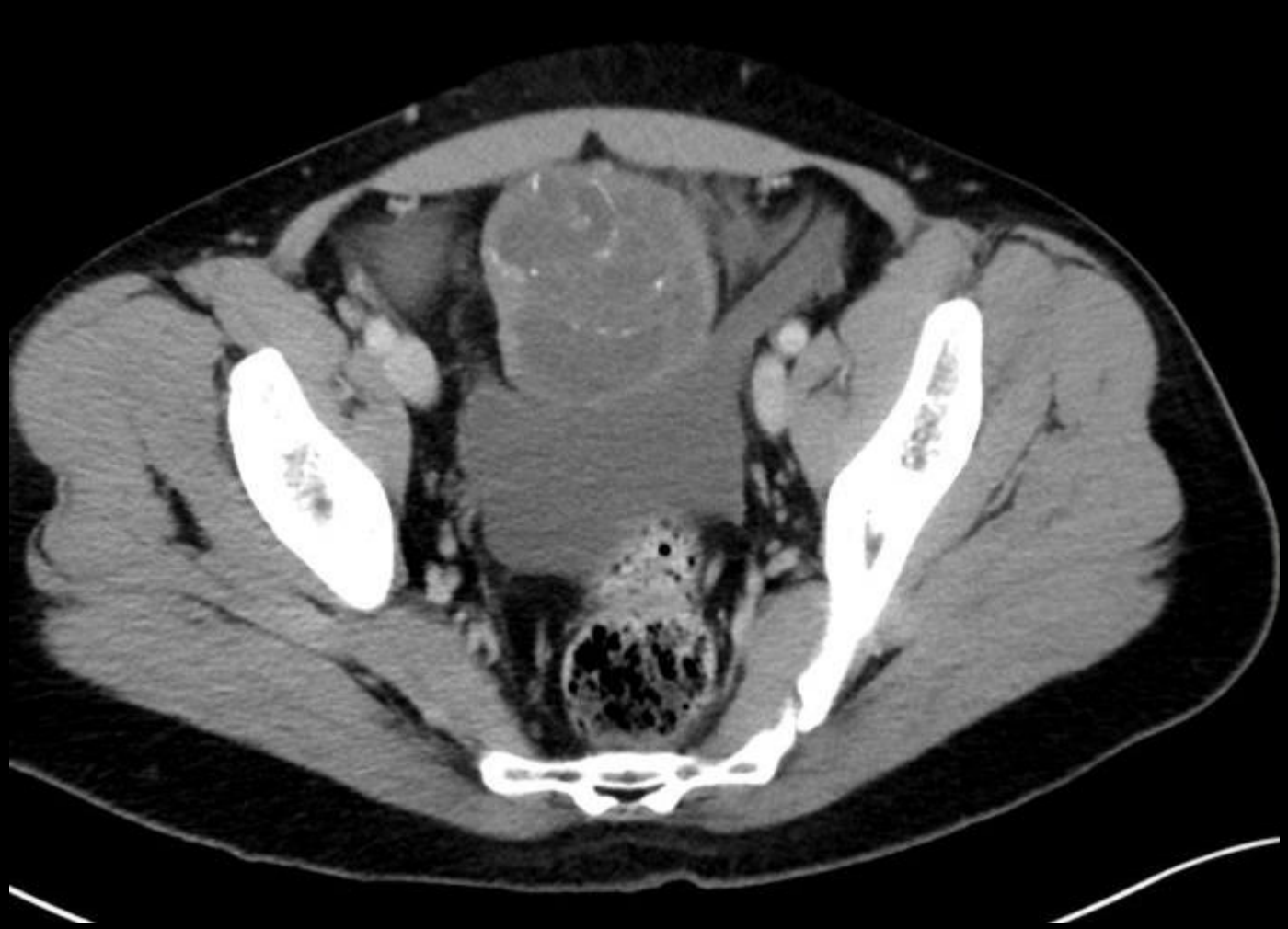
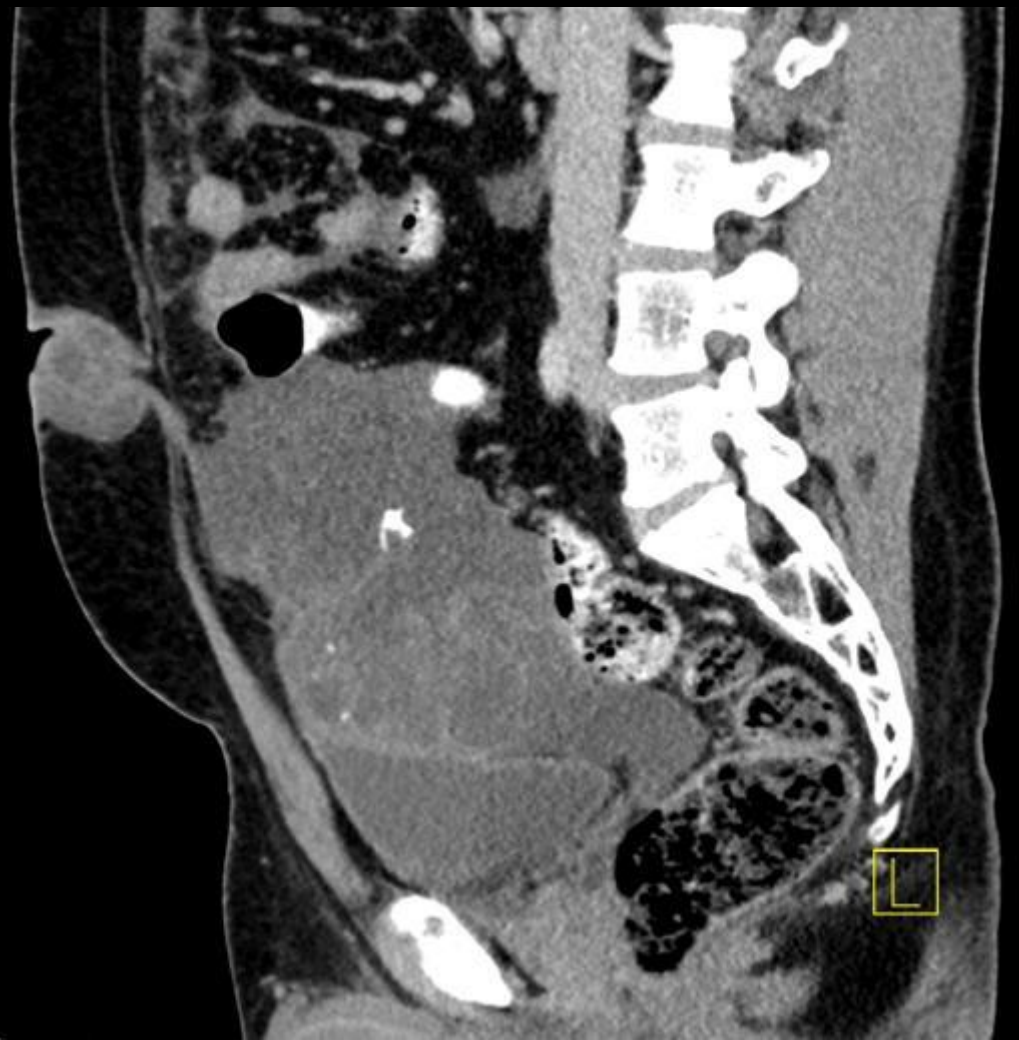
Findings (unlabeled)

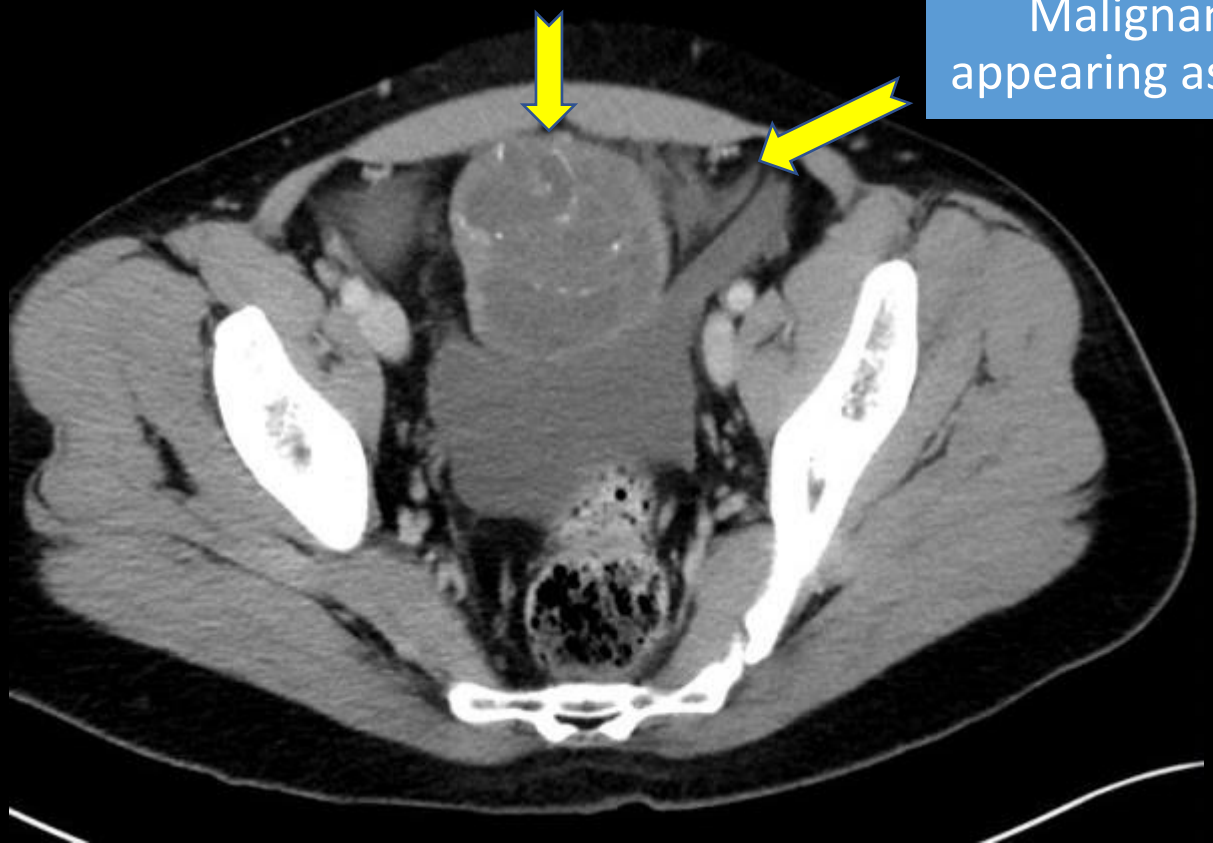




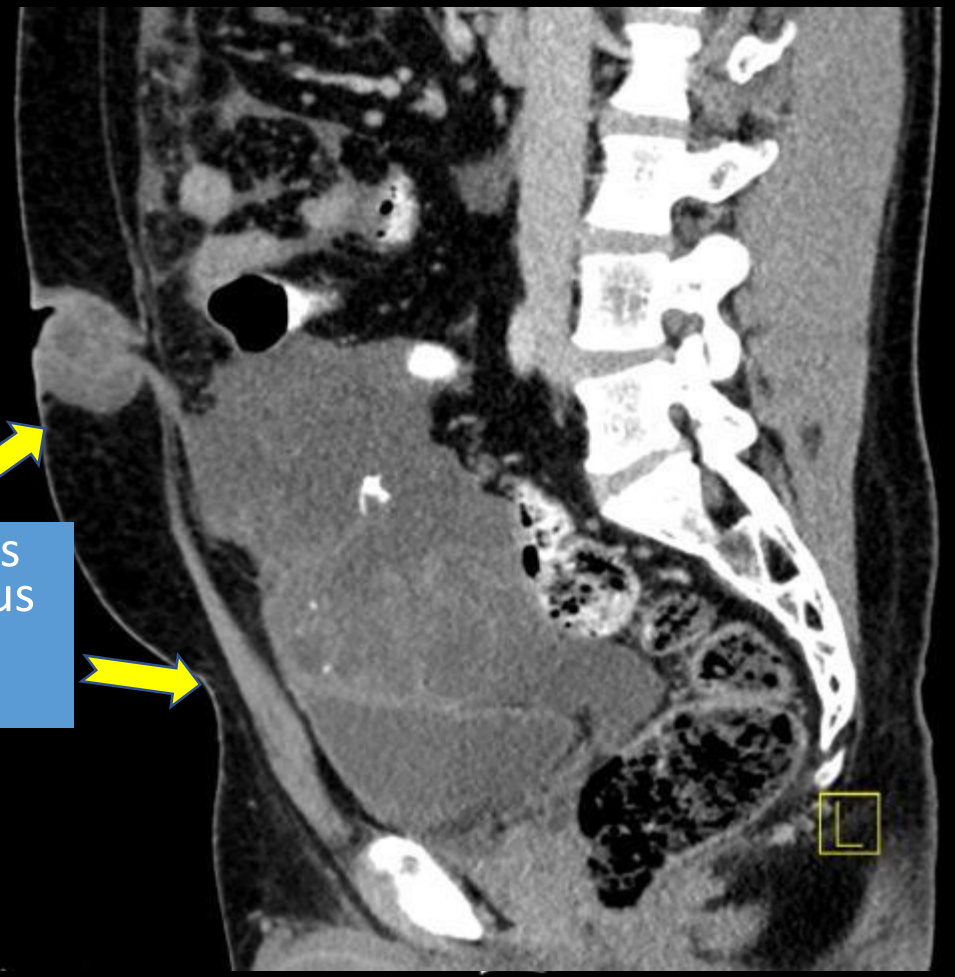
Findings (labeled)

Mixed solid and cystic mass with pathognomonic (punctate, stippled, curvilinear and peripheral calcifications

Malignant appearing ascites



Mass extends from umbilicus to dome of bladder



Final Dx:

Metastatic Urachal Adenocarcinoma

Case Discussion

- Follow-up laboratory workup revealed CEA (carcinoembryonic antigen) elevated at 32.6 ng/mL
- Diagnostic laparoscopy confirmed well-differentiated urachal adenocarcinoma, mucinous type
- Surgical resection 10 days later revealed extension beyond bladder into peritoneum and focally involving liver capsule (stage pT4)

Urachal Neoplasms

- Very rare, <1% of all bladder cancer → limited literature
- Initial presenting symptom usually hematuria, others include mucinuria, recurrent UTIs, local pain, or umbilical discharge
- Urachus: tissue of origin
  - Residua of the embryological allantois and extends from the bladder dome to the umbilicus within the extraperitoneal, retropubic space (cave of Retzius)
  - Usually seals off into a fibrous cord (median umbilical ligament)
  - Can persist with 4 possible remnant types: patent urachus (unsealed along entire extent), umbilical-urachal sinus (unsealed at umbilicus), vesicourachal diverticulum (unsealed at bladder), urachal cyst (unsealed along its midportion)
- Remnants at risk for recurrent infections and neoplasm formation – most common malignancy is adenocarcinoma, mainly mucinous type (type associated with calcifications)

Staging

- No AJCC TNM staging system accepted
- Sheldon Staging System best known/most used
  - pT1 – no invasion beyond the urachal mucosa
  - pT2 – invasion confined to the urachus
  - pT3 – local extension to the (a) bladder, (b) abdominal wall, (c) viscera other than the bladder
  - pT4 – metastasis to (a) regional lymph nodes, (b) distant sites

Management

- Silent early lesions with propensity for local growth and tendency to metastasize – usually present at stage pT3 or higher
- High likelihood of recurrence documented in the literature
- Primary therapeutic approach = surgical resection with partial or radical cystectomy and *en bloc* resection of the urachal ligament with the umbilicus (+/- chemotherapy)

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