AMSER Case of the Month
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“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

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Appropriateness Category</th>
<th>Relative Radiation Level</th>
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<tbody>
<tr>
<td>US abdomen</td>
<td>Usually Appropriate</td>
<td>○</td>
</tr>
<tr>
<td>CT abdomen with IV contrast</td>
<td>Usually Appropriate</td>
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<tr>
<td>MRI abdomen without and with IV contrast</td>
<td>Usually Appropriate</td>
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<tr>
<td>MRI abdomen without IV contrast</td>
<td>May Be Appropriate</td>
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<tr>
<td>CT abdomen without and with IV contrast</td>
<td>Usually Not Appropriate</td>
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<tr>
<td>FDG-PET/CT skull base to mid-thigh</td>
<td>Usually Not Appropriate</td>
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<tr>
<td>Radiography abdomen</td>
<td>Usually Not Appropriate</td>
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<td>Fluoroscopy contrast enema</td>
<td>Usually Not Appropriate</td>
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<tr>
<td>Fluoroscopy upper GI series</td>
<td>Usually Not Appropriate</td>
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<tr>
<td>Fluoroscopy upper GI series with small bowel follow-through</td>
<td>Usually Not Appropriate</td>
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This imaging modality was ordered by the surgeon.
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)
References:


