Glandular lesions and tumors in Uropathology

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Clinical presentation

• 45-year-old woman with no previous tumor history

• Symptoms: Mucous discharge from an umbilical orifice and abdominal pain

• Physical examination: midline suprapubic mass located in the abdominal wall with cutaneous communication

• RM: multilocular cystic mass 5 cm in diameter with no connection with other organs

• Surgery: excision of the mass together with umbilicus, the surrounding abdominal skin, and a tubular structure located below and connected with the mass.
Macroscopic appearance
Macroscopic appearance
Macroscopic appearance
Microscopic appearance
Low grade atypia
Papillary formations
Skin connection
Tubular structure
Morphologic Features

- Cystic formation
- Mucinous content
- Intestinal type epithelium
- Growth pattern: flat to cuboidal to typical mucinous columnar to pseudostratified and papillary structures
- Low grade atypia
Differential Diagnosis

- Cystic neoplasm of the urachus
- Cystic neoplasm of intestinal origin
- Cystic neoplasm of ovarian origin
Urachus

Anatomical dissection of bladder and prostate during an autopsy of a stillborn (37 weeks) performed in our institution.
Fetal Urachus

Transversal section

Urachus

Bladder lumen

Umbilical arteries
Classification of urachal anomalies

- Juxtaumbilical
- Intermediate
- Juxtavesical
- Giant Cyst
- Multiple cysts
Urachus

The remnant epithelium is mostly transitional (urothelial) cells

The majority of urachal neoplasms are of the glandular type

Hypothesis:
• 1) urothelial cells go through intestinal metaplasia that evolve in adenocarcinoma
• 2) the intestinal-type epithelium originates from cloacal inclusions or from enteric rests

Presence of foci of intestinal-type epithelium or scattered goblet cells within a transitional epithelium in 10.9%
Glandular Neoplasms of the Urachus

A Report of 55 Cases Emphasizing Mucinous Cystic Tumors With Proposed Classification

Mahul B. Amin, MD,* Steven C. Smith, MD. PhD,* John N. Eble, MD,† Priya Rao, MD,*
William W. L. Choi, MD,‡§ Pheroze Tamboli, MD,∥ and Robert H. Young, MD¶

<table>
<thead>
<tr>
<th>Benign Mucinous Cystic Tumors</th>
<th>Borderline and Malignant Mucinous Cystic Tumors</th>
<th>Malignant (Noncystic) Neoplasms</th>
</tr>
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<tbody>
<tr>
<td>Mucinous cystadenoma</td>
<td>Mucinous cystic tumor of low malignant potential (MCTLMP)*</td>
<td>Mucinous adenocarcinoma</td>
</tr>
<tr>
<td></td>
<td>Mucinous cystadenocarcinoma 1. with microinvasion</td>
<td>Enteric-type adenocarcinoma</td>
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<tr>
<td></td>
<td>2. frankly invasive</td>
<td>Adenocarcinoma, with mixed features†</td>
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<tr>
<td></td>
<td></td>
<td>Adenocarcinoma, not otherwise specified</td>
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*May or may not show intraepithelial carcinoma, which should be noted.
†Report should itemize the various features and estimate the amount of each.

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Updates in the Pathologic Diagnosis and Classification of Epithelial Neoplasms of Urachal Origin

Gladell P. Paner, MD,* † Antonio Lopez-Beltran, MD, PhD,‡ Deepika Sirohi, MD,§ and Mahul B. Amin, MD§

- Glandular neoplasms
  - Adenomas
    - Villous adenoma
    - Mucinous cystadenoma
    - Mucinous cystic tumor of low malignant potential
  - Adenocarcinomas
    - Noncystic adenocarcinomas
      - Enteric (intestinal)
      - Mucinous (colloid)
      - Signet ring cell
      - Not otherwise specified
      - Mixed
    - Cystic adenocarcinomas
      - Mucinous cystadenocarcinoma
        - With microinvasion
        - Frankly invasive
  - Mixed carcinomas

- Nonglandular neoplasms
  - Urothelial neoplasms
  - Squamous cell neoplasms
  - Neuroendocrine neoplasms
  - Mixed type neoplasms

Benign

Borderline

Malignant
Mucinous adenocarcinoma with superficial stromal invasion and villous adenoma of urachal remnants: a case report

R Mazzucchelli, M Scarpelli, R Montironi


Cyst

Canal

Villous adenoma
# Mucinous Cystic Tumors

<table>
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<tr>
<th>Tumor</th>
<th>Definition</th>
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<tr>
<td>Mucinous cystadenoma</td>
<td>Cystic tumor lined by a single layer of mucinous columnar epithelium devoid of atypia</td>
</tr>
<tr>
<td>Mucinous cystic tumor of low malignant potential</td>
<td>Cystic tumor with areas of epithelial proliferation, including papillary formation and low-grade atypia</td>
</tr>
<tr>
<td>Mucinous cystic tumor of low malignant potential with intraepithelial carcinoma</td>
<td>Cystic tumors with significant epithelial stratification and unequivocal malignant cytological features and often with stroma-poor papillae and cribriform pattern</td>
</tr>
<tr>
<td>Mucinous cystadenocarcinoma with microinvasion</td>
<td>Stromal invasion $&lt; 2$ mm and comprising $&lt; 5%$ of the tumor</td>
</tr>
<tr>
<td>Frankly invasive mucinous cystadenocarcinoma</td>
<td>More extensive invasion</td>
</tr>
</tbody>
</table>

*Adopted from Amin et al.\(^5\)*
In the series presented by Amin et al. in 2014 of 55 cases, mucinous cystic tumors ranged from 0.8 to 8 cm in maximum dimension and were mostly unilocular.

- 87% of tumors showing an extravasation of acellular mucin into the stroma. **Potential pitfall**

- Dystrophic calcification were present in 39% of the tumors.

- None of patients with mucinous cystic tumors experienced recurrence or metastasis, in contrast with the 45% 5-year survival rate of invasive urachal adenocarcinomas.
Mucinous cystic tumor of low malignant potential

- Mucinous cystic tumor of low malignant potential may show areas resembling mucinous cystadenoma but also has **stratified cyst lining** of usually not more than 3 cell layers

- The lining in mucinous cystic tumor of low malignant potential is more **proliferative** than cystadenoma

- Architecture varies from flat to tufted, **pseudo papillary** and villous to tubule-villous

- Mild to moderate nuclear **atypia**
Immunophenotype of urachal glandular lesion

- CK 20 : 100%
- CDX 2 : 80%
- CK 7 : 30%
- β catenin negative
- ER negative
- PR negative
- p63 negative
Ovarian neoplasms:
- Ck7 positive in 88%
- Ck20 positive in 65%
- CDX2 positive in 38%
- PAX8 positive in 75%
- ER positive in 13%

Colonic adenocarcinoma:
- Ck 7 usually negative (8%)
- Ck 20 positive in 90%
- CDX2 positive in 92%
- β catenin positive (nuclear) in 50%

Urothelial carcinoma:
p63 positive in 80% of cases
IHC results

- CDX2+
- CK20+
- CK7-
- PAX8
- P63-
- βcatenin
- UPK-
Final diagnosis

• Midline location
• Histologic appearance
• Immunohistochemical results

Mucinous cystic tumor of low malignant potential

No recurrence or metastatic spread was recorded after 3 years of follow up.