

Background

- Urachal carcinoma (UC) is a rare, aggressive malignant neoplasm that arises from the urachal remnant of the urachus¹.
 - During embryological urinary development, the allantois connects the superior bladder to the umbilicus and eventually develops into the urachus. Typically, the urachus closes forming the median umbilical ligament, however this structure can fail to close in approximately 30% of adults, leaving a persistent space or tract².
 - Tissue within the remnant can persist and has the potential to become neoplastic along the tract through transitional epithelial metaplasia, commonly arising in the bladder dome or anterior wall¹.
 - Adenocarcinoma is the most common type (>90%) and 70% are mucin producing³.
- Patient Aspect:**
- UC accounts for approximately 0.01% of all cancers and <1% of all bladder⁴.
 - Patients are typically asymptomatic, often presenting non-specific symptoms later when metastasis has spread. Symptoms include; hematuria, mucinuria, and suprapubic masses⁴.
 - Due to late presentation and typically diagnosed in advanced stages, UC has a poor prognosis with a 5-year survival rate of 50%¹.
 - Treatment typically involves surgical intervention which includes an en bloc resection of urachus with umbilical cord and partial cystectomy with individualized neoadjuvant/adjuvant therapy in advanced cases⁵.

Clinical History

- Initial Presentation:**
- Female patient in fifth decade.
 - Progressing pelvic-abdominal pressure, with irregular vaginal bleeding and mucinous discharge.
 - A palpable mass was identified below the umbilicus.
- Imaging:**
- MRI of the umbilical mass revealed a 2.2 x 2.3 cm cystic mass superior to the bladder dome.
 - Bilateral mucinous ovarian metastases were also identified, (Right; 6.7 x 4.7 cm and Left; 3.0 x 3.5 cm) with scattered mucinous peritoneal metastases in the pelvis including the posterior cul-de-sac.
 - There was no significant change between subsequent imaging.
- Interventions:**
- A biopsy from the pelvic mass, showed abundant mucin and atypical mucinous epithelium, consistent with mucinous adenocarcinoma.
 - The immuno-profile showed a positive CK7 and focal SATB2 expression, favoring mucinous urachal adenocarcinoma.
 - After six cycles of neoadjuvant chemotherapy a laparoscopic cytoreductive surgery was performed, including the removal of the urachal cyst, total hysterectomy and bilateral salpingo-oophorectomy, and rectosigmoid resection.
 - Followed by heated intraperitoneal chemotherapy.

Gross Examination

Dome of Bladder:

- Unoriented piece of tissue with a dark brown lesion on the peritoneal surface with mucin.
- Sectioning reveled an ill-defined, cystic, and mucinous mass, continuous with the peritoneal lesion.

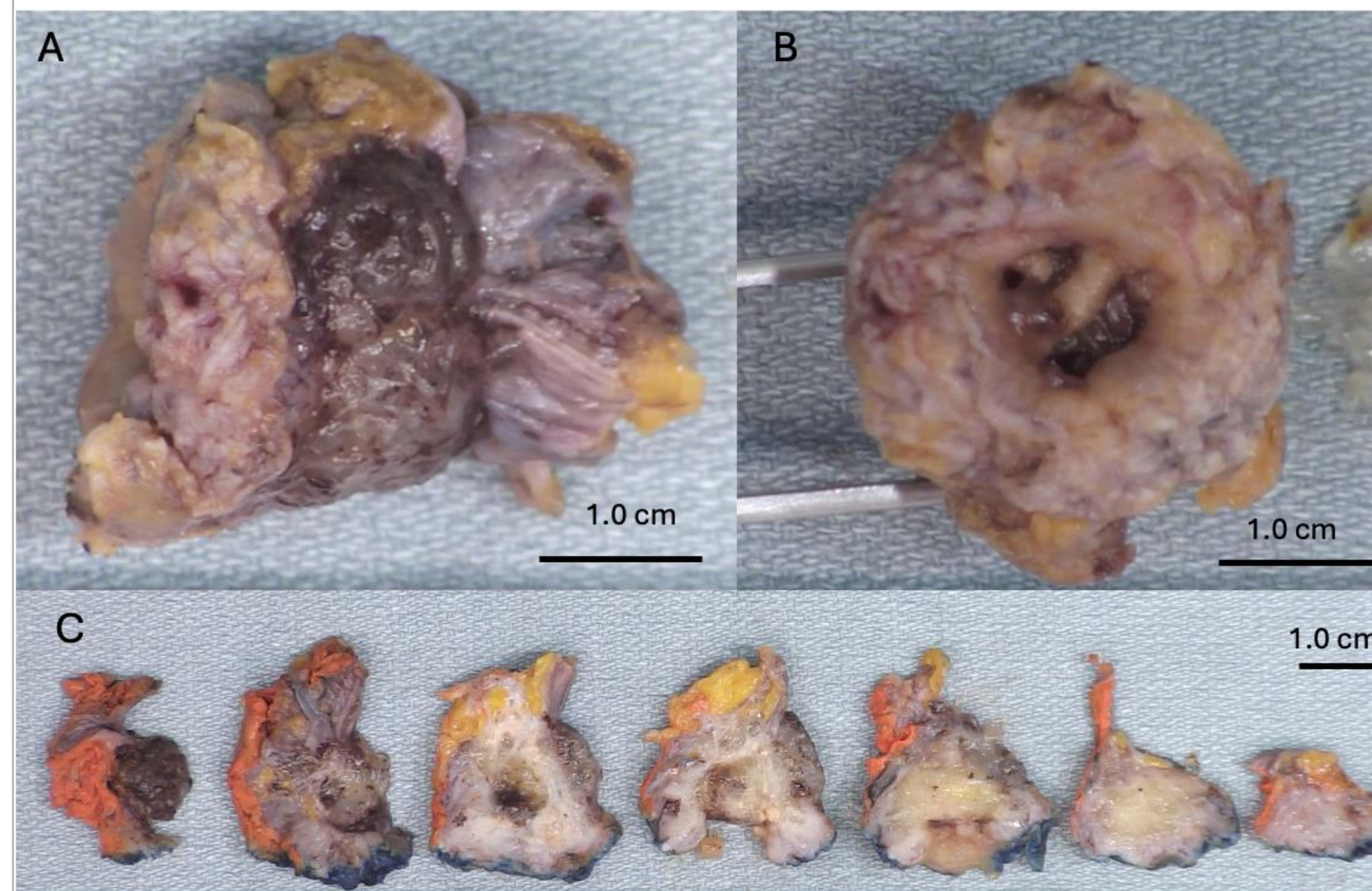


Figure 1. Urachal cyst specimen from bladder dome. **A:** Side view of the urachal cyst. **B:** Superior view of the urachal cyst. **C:** The specimen was serially sectioned to demonstrate the relationship of the cyst/mucinous mass with the surrounding soft tissue and peritoneal surface. **Inking Code:** Orange: External surface(excluding peritoneal surface), and Blue: Mucosal resection margin.

Uterus, Vagina, Rectosigmoid, and Rectum:

- Upon sectioning of the uterus, a mucoid mass was identified on the posterior vaginal cuff (1.8 x 1.8 x 1.2 cm) with extension through the soft tissue without gross invasion into the rectal wall. (**Figure 2**)
- A mucoid mass was attached to the rectal serosa (1.4 x 0.8 x 0.3 cm).

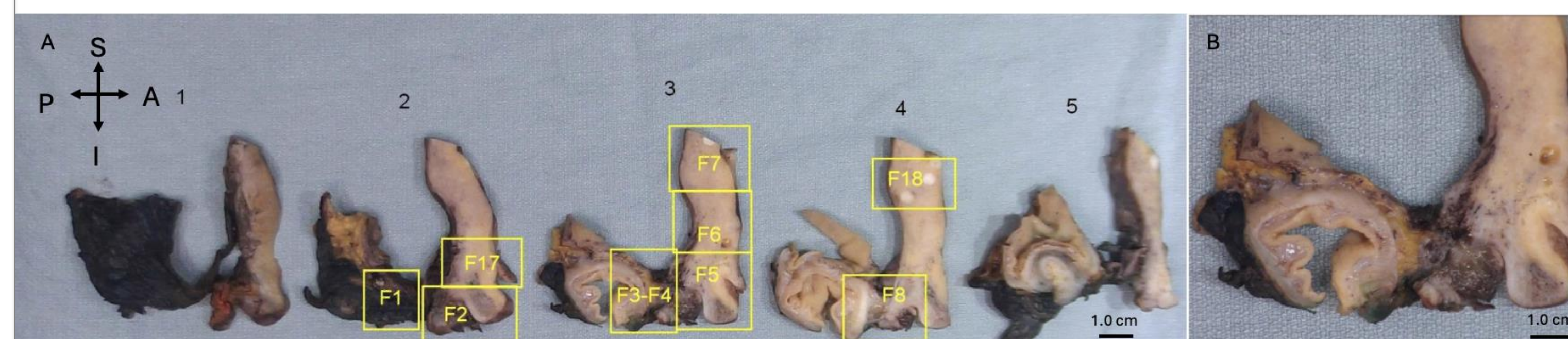


Figure 2. Uterus and rectum with mucoid mass. **A:** The specimen was serially sagittal sections demonstrating the relationship of the uterus/vagina to the rectum demonstrating the involvement of the mucoid mass. A blocking map of where sections were taken is included. **B:** Demonstrates one of the slices with extension of the mucoid mass to surrounding tissue.

Bilateral Fallopian Tubes and Ovaries:

- Multiple mucoid deposits were present on the external surface of both ovaries.
- The cut surface of both ovaries revealed a cystic structure containing yellow-white mucus material.

Results

- **Microscopic evaluation:** shows acellular mucin pools with focal malignant cells in the primary tumor bed and metastatic sites.
- **Histological Type:** Urachal Adenocarcinoma.
- **Margins:** All margins were negative for invasive tumor.
- **Tumor Extent:** Lesion invades adjacent structures including perirectal tissue, posterior vaginal cuff, and rectum. Lymphatic and vascular invasion was not identified.
- **Staging:** The tumor corresponds with Sheldon Stage IIID, Mayo Stage IV, and Ontario Stage T4.
- **Treatment Response:** Strong response to neoadjuvant chemotherapy (TRG2), with predominant fibrosis of the tumor bed and <50% residual cancer cells present.

Discussion

Staging:

- No AJCC TNM staging system for UC.
- Application of the bladder cancer TNM system is limited as UC has distinct clinical and pathological characteristics¹.
- There are three proposed staging systems for UC, Sheldon, Mayo and Ontario staging systems (**Table 1**), however these systems have limitations, including limited sample sizes, lack of tumor sizing, and lack of validation¹.

Table 1. Comparison of the different staging systems for urachal carcinoma.

Sheldon ⁶	Mayo ⁷	Ontario ⁸
I. Confined to urachal mucosa	I. Confined to urachus/bladder	T1. Confined to urachus submucosa
II. Invasion confined to urachus	II. Extends beyond urachus/bladder	T2. Confined to muscular bladder wall
III. Extension to: A. Bladder B. Abdominal wall C. Peritoneum D. Surrounding viscera	III. Extends to regional lymph nodes	T3. Extends to peri-urachal or vesical soft tissue
IV. Metastases A. Lymph node B. Distant sites	IV. Extends to distant lymph nodes or sites	T4. Invasion to adjacent organs

Pathologists' Assistant (PA) Role:

- There is no standard grossing protocol for UC, the PA must adapt and conduct a thorough dissection, including photo documentation and block mapping.
- Conducting a thorough examination and appropriate tissue selection is needed to accurately determine staging and prognosis.

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