Organ system: Genitourinary Tract

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HISTORY

80-year-old woman presented with gross hematuria and was found to have a large palpable and non-mobile abdominal mass. Cystoscopy revealed a 5.5 cm hemorrhagic tumor in bladder dome. Patient subsequently underwent partial cystectomy, based on the histological findings on biopsy.

Gross and microscopic Images
Macroscopic image: 5.5 cm mass with a tan-brown, cystic and solid cut surface
Tumor cells with tubular and papillary architecture lined by cuboidal cells with pleomorphic nuclei and prominent nucleoli (H&E, 100X)
Occasionally tumor cells show prominent hobnail morphology (H&E, 400X)

Tumor cells show clear cytoplasm (H&E, 400X)

**Immunohistochemistry**

Positive stains: PAX8, CK7, Napsin-A, HNF1beta, wild-type p53 and high proliferation index by Ki67

Negative stains: p63, CK20, GATA3, ER, and PR

**Diagnosis**

Clear cell adenocarcinoma of bladder, Mullerian type, in a background of endometriosis, pT2bN0.

**Differential diagnoses**

- Nephrogenic adenoma
- Clear cell variant of urothelial carcinoma
- Metastatic clear cell renal cell carcinoma
- Cervical or vaginal clear cell adenocarcinoma
Discussion

Primary clear-cell bladder adenocarcinoma is rare, mainly affecting middle-aged women. Association with bladder endometriosis suggests Mullerian origin. The tumor displays papillary, cystic or solid growth pattern. The papillary fibrovascular cores may show hyalinization. Myxoid stromal reaction is common. The tumor cells are flattened, cuboidal, and hobnail, with clear or eosinophilic cytoplasm. Cytologic atypia is moderate to severe, with variable mitotic activity and necrosis. Tumor cells are usually positive for CK 7, CA125, PAX8, AMACR, HNF-B, p53 and Napsin.

Clear cell adenocarcinoma is considered an aggressive tumor, with a 5-year survival rate of 40% for urethral or bladder CCA. Advanced stage and poor prognosis are reported for clear cell adenocarcinoma in prostatic location, whereas low stage exophytic tumors are associated with better outcome.

Awareness of the entity and diagnostic criteria is important for the pathologists and clinicians to provide accurate diagnosis and optimal treatment to the patient.

References


