

CASE REPORT

Urachal Mucinous Adenocarcinoma of the Bladder

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Mucinous adenocarcinoma of the bladder is rare, occurring in 0.5-2.0% of bladder cancer. Most patients present with hematuria, suprapubic pain and dysuria. Presented is a case of a 44 year old male with 4 month history of hematuria and terminal dysuria who underwent radical cystoprostatectomy with histopathology findings of mucinous adenocarcinoma. Upon diagnosis, 25% of patients have distant metastases and 50% have stage IV disease with 50% five years survival rate for stage I-III tumor with no stage IV patients surviving beyond two years. Hence, for this type of cancer, early diagnosis is crucial. Surgery is the mainstay of treatment and is resistant with chemotherapy and radiation. Thus, for patient with confirmed or highly suspicious mucinous adenocarcinoma of the bladder, timely radical resection is warranted.

Keywords: mucinous adenocarcinoma, urachal adenocarcinoma, bladder cancer

Introduction

Bladder cancer is the second most common tumor of genitourinary tract. Majority of bladder cancer cases are urothelial carcinomas (90-95%)¹, with bladder adenocarcinoma comprising only 0.5-2.0%, with mucinous subtype being extremely rare.^{2,3} Urachal remnant is one of major risk factors. It is formed early in embryonic life. In two-thirds of the population, it coalesces with umbilical arteries to form umbilical ligament and obliterated at birth. It persisted in one-third, forms as vestigial remnant, which connects the dome or anterior wall of urinary bladder to the umbilicus and may become primary site of tumor. According to studies, there is progressive transformation from mucinous metaplasia to mucinous adenoma to mucinous adenocarcinoma. Almost all patients presents with hematuria. Other symptoms include voiding difficulties, suprapubic

pain and mucous discharge upon micturation. It has aggressive behavior and noted to be resistant to chemotherapy and radiation, with surgery considered as the most effective treatment option. Hence, early diagnosis is crucial.

Due to the rarity of this tumor, the authors present here a case of primary urachal mucinous adenocarcinoma of the bladder emphasizing its diagnosis and management.

The Case

This is a case of a 44-year old male, smoker who presented with intermittent gross hematuria and terminal dysuria for four months. Abdominal exam was unremarkable without palpable abdominal mass. Digital rectal examination revealed normal prostate without nodules. CT scan revealed a 3 cm irregular mass at the anterior

bladder wall. (Figure 1). Transurethral resection of bladder tumor was done with biopsy result of mucinous adenocarcinoma. Colonoscopy was done which showed normal result. He underwent radical cystoprostatectomy with pelvic lymph node dissection, appendectomy and ileal conduit. Intraoperatively, mass was noted at the bladder dome extending to the urachus measuring 3cm. (Figure 2) Histopathology showed mucinous adenocarcinoma with transmural invasion into

perivesical fatty tissue and negative nodal invasion. (T3N0Mx). The authors plan to follow up patient every 3-6 months for 2 years then annually with repeat imaging.

Discussion

Primary mucinous adenocarcinoma of urinary bladder arises in middle aged men associated with

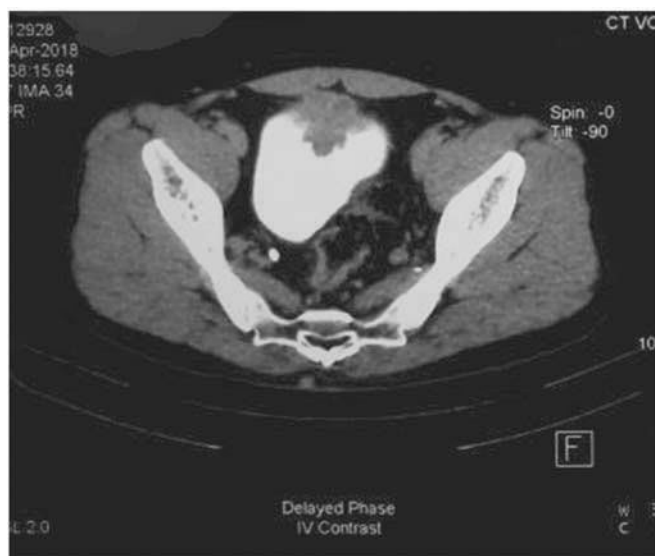


Figure 1. A mass located in the anterior bladder wall identified on computed tomography, sized 3.0cm. Surrounding bladder wall was normal. No abnormal findings in the pelvic cavity.



Figure 2. A 3cm mass noted at the dome of bladder wall extending to the urachus. Bladder wall was smooth.

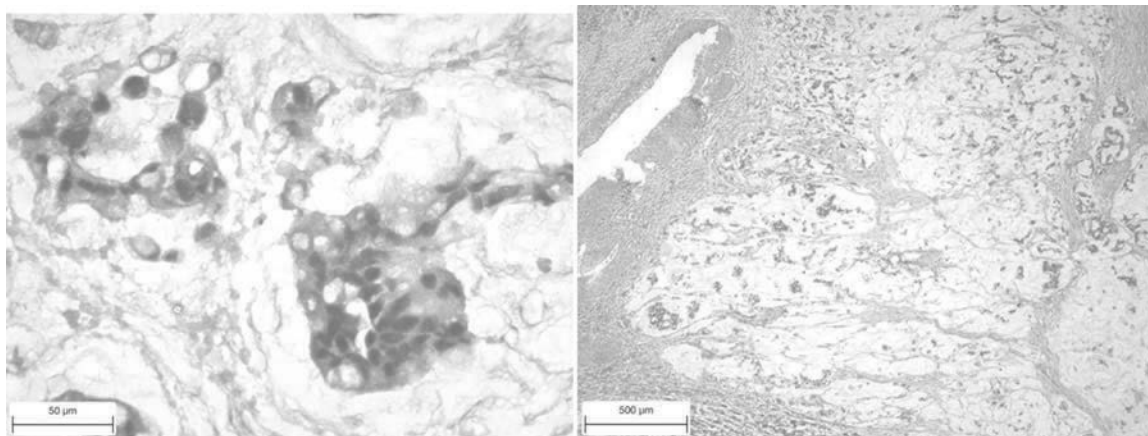


Figure 3. On histological examination, bladder tumor discloses a malignant neoplasm composed of nests of neoplastic epithelial cells with large, round to ovoid, eccentric hyperchromatic nuclei, some with prominent nucleoli and eosinophilic cytoplasm. These cells are floating in lakes of mucin and invades into the muscularis propria of the bladder and extends into the perivesical fatty tissues.

urachal remnant. It arises due to intestinal metaplasia caused by telomerase shortening with cytogenetic abnormalities. This tumor has overlapping histologic characteristic with primary adenocarcinoma arising from colon, prostate and gynecologic tract. Risk factors include persistent urachal remnant, glandular cystitis and bladder exstrophy.² The accumulation of secretions and associated infection may promote its development. The growth pattern mainly involves infiltration into deep muscular layer. Hence, majority of bladder mucinous adenocarcinoma patients are stage T2 or T3 at diagnosis.¹ Ninety percent of patients present with hematuria. Others present with voiding difficulties and suprapubic pain similar to other bladder tumors or mucous discharge from bladder during micturition.² Urinary bladder adenocarcinoma includes different histologic variants: a) Non-specific adenocarcinoma, b) Adenocarcinoma of enteric origin, c) Adenocarcinoma with signet-ring cells and d) Mucinous adenocarcinoma² and is categorized as urachal and non-urachal adenocarcinoma because of different history, pathologic diagnosis, and prognosis.⁴ Urachal adenocarcinoma presents at earlier median age than non-urachal adenocarcinoma (56 vs 69 years), frequently affects women than men (45 vs 36%), less likely to be high grade (35 vs 66%), and shows a better 5 year overall survival rate (48 vs 35%). Criteria proposed to distinguish the 2 entities include presence of urachal remnant, an intact or ulcerated urothelium, invasion of muscularis wall of the bladder, anterior abdominal wall, or umbilicus.⁴

Radical cystectomy with enbloc surgical resection of the urachal ligament for muscle invasive disease is the treatment of choice, the only curative option and showed superiority over partial cystectomy for urachal tumor due to possibility of undetectable local invasion on imaging.^{2,4} Survival strongly correlates with the stage, grade and subtypes of disease. It approaches

to 75-100% among tumors confined to the urinary bladder. Upon diagnosis, 25% of patients have distant metastases and 50% have stage IV disease. Recent study showed the 50% five-year survival rate for stage I-III tumor with no stage IV patients surviving beyond two years. Bladder adenocarcinoma is resistant to chemotherapy and radiation; thus, for patients with confirmed or highly suspicious mucinous adenocarcinoma of the bladder, timely radical resection is warranted.¹

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