Radical cystoprostatectomy to treat urachal carcinoma

Ahmed Ebrahim, BS^a , Nitin Kondapalli, MD^b , and W. Scott Webster, MD^c

^aCollege of Medicine, Texas A&M Health Science Center, Dallas, Texas; ^bDepartment of Pathology, Baylor University Medical Center, Dallas, Texas; ^cUrology Clinics of North Texas, Baylor University Medical Center, Dallas, Texas

ABSTRACT

Urachal carcinoma is a highly uncommon malignancy with an estimated prevalence of 0.01% to 0.02% of all adult cancers. Due to its rarity, no standardized management protocol for urachal cancer has been developed. Surgery is often the main therapeutic measure. A 48-year-old man presented with hematuria for 8 months. Imaging revealed a mass at the bladder dome. Biopsy indicated mixed adenocarcinoma with a small cell component. Radical cystoprostatectomy with ileal urostomy was performed. After surgical resection, he was diagnosed with urachal adenocarcinoma (mixed type). The patient tolerated surgery and was discharged home uneventfully. Follow-up computed tomography at 6 months was negative.

KEYWORDS Cystoprostatectomy urachal; urachal carcinoma; urachus cancer

The early fetal bladder drains via the allantois into the yolk sac. During fetal development, the allantois becomes the urachus. The urachus normally obliterates; its remnant—a fibromuscular band running from the bladder dome to the umbilicus—is known as the median umbilical ligament. Incomplete obliteration of the urachus can occur. These remnants can result in complications such as urachal cyst and sinus.¹ The urachal remnants can also, in rare cases, become cancerous. Malignancy of the urachus was first described by Hue and Jacquin in 1863.² Further study has revealed that 90% of urachal cancers are adenocarcinomas,³ although sarcomas and carcinomas of squamous, urothelial, and neuroendocrine origin have also been described.^{4,5} Urachal carcinomas are extremely rare; their incidence rate is estimated to be less than 1% of all bladder cancers.⁶

CASE DESCRIPTION

A 48-year-old man presented with an 8-month history of intermittent gross hematuria. Prior workup included urinalysis, which showed increased red and white blood cells but negative cultures. A trial of ciprofloxacin had been ineffective. Imaging via computed tomography (CT) revealed a mass at the bladder dome. Biopsy via transurethral resection of bladder tumor showed adenocarcinoma with small cell

features. This raised suspicion for a urachal primary tumor, which was clinically stage 3 based on CT.

The patient was taken to the operating room for a radical cystoprostatectomy with umbilectomy and pelvic lymphadenectomy. A midline incision was made from the pubic symphysis up to the umbilicus. The umbilicus was circumscribed and the lateral aspects of the urachal remnant were carefully excised. While mobilizing the bladder down to the external iliac artery, the mass was found at the bladder dome. After completely mobilizing the bladder, the vas deferens were transected. The ureters were mobilized, clipped, transected, and packed away. The prostate was carefully freed from the endopelvic fascia and the dorsal venous complex was ligated and transected. The prostate was removed off the anterior aspect of the rectum and the seminal vesicles were removed from their lateral attachments. A LigaSure device was used to control the vascular pedicles at the level of the bladder. The prostate and bladder were removed en bloc (*Figure 1a*). A pelvic lymphadenectomy of the nodes was performed from the aortic bifurcation inferiorly past the internal and external iliac arteries into the obturator fossa. The vascular supply to the ureters appeared to arise from the region of the presacral nodes, and the decision not to excise them was made. To create an ileal urostomy, a 12-cm segment of bowel was identified 15 cm from the ileocecal junction. The segment was stapled off, and side-to-side anastomosis of the bowel was

Corresponding author: Ahmed Ebrahim, BS, Baylor University Medical Center, 3500 Gaston Ave., Roberts Suite 1013, Dallas, TX 75246 (e-mail: aebrahim@medicine.tamhsc.edu)

Color versions of one or more of the figures in the article can be found online at www.tandfonline.com/ubmc.

Received May 9, 2019; Revised July 9, 2019; Accepted July 18, 2019.

Figure 1. (a) Gross specimen with umbilicus (dark arrow) and prostate (light arrow). (b) Urachal adenocarcinoma with mucinous component (4×). (c) Small cell component in urachal carcinoma (4×). (d) Incidental finding of prostatic adenocarcinoma (Gleason score 6 [3 + 3])—involving the prostate.

performed in standard fashion. The anastomosis was reinforced with 3-0 silk suture. The ureters were anastomosed to the ileal conduit using 7.5-French stents. The ileal conduit was brought out through a marked stoma site and secured. A drain was placed in a separate stab incision and the wounds were closed in multiple layers. The patient was transferred to recovery in stable condition.

The umbilicus, urachus, bladder, prostate, distal segments of the distal ureters, and 34 lymph nodes were collected by pathology. Gross examination of the bladder demonstrated a palpable mass involving the fibrofatty tissue of the urachus, which extended inferiorly to involve the bladder dome. Opening the bladder revealed a 2.5-cm area of mucosal ulceration overlying the mass. Upon microscopic examination, the tumor was found to be an adenocarcinoma with enteric and mucinous features (*Figures 1b, 1c*). A detached segment of urothelium at the ulcer showed in situ urothelial carcinoma. No invasive high-grade urothelial carcinoma or diffuse cystitis cystica/glandularis was noted in the adjacent urothelium. All lymph nodes and ureter segments were found to be negative for metastatic carcinoma. In accordance with the Sheldon classification for urachal adenocarcinomas (*Table 1*),⁷ the current tumor was staged IIIa. Incidentally, an acinar adenocarcinoma was detected on examination of the prostate (*Figure 1d*). Perineural invasion was present, although surgical margins were found to be negative and no extraprostatic extension was identified. The Gleason score was 6 (3 + 3) and the grade group was 1.

The patient's postoperative course was uneventful. He reported having bowel movements on postoperative day 3. His urostomy output averaged over 2 L per day and was clear without hematuria or pyuria. He was discharged home on postoperative day 6. Follow-up CT at 6 months showed no evidence of recurrent or residual neoplasm or metastatic disease.

Table 1. Sheldon staging system for urachal adenocarcinomas^a

pT1	No invasion beyond the urachal mucosa
pT2	Invasion confined to the urachus
pT3	a. Local extension to the bladder b. Local extension to the abdominal wall c. Local extension to the viscera other than the bladder
pT4	a. Metastasis to regional lymph nodes b. Metastasis to distant sites

pT indicates pathologic tumor stage.

^aAdapted from Sheldon et al.⁷

DISCUSSION

The most common symptom of urachal carcinoma is hematuria. Urachal carcinomas typically present late because they have already invaded the bladder and ulcerated its mucosa by the time hematuria is grossly apparent. Due to the rarity and relatively late presentation of urachal carcinomas, survival data are sparse, and evidence-based medical therapies are lacking. International guidelines suggest that physicians use their discretion in prescribing radiotherapy and chemotherapy regimens on a case-by-case basis.⁸ Chemotherapy in the treatment of urachal carcinoma is commonly used in patients whose disease burden precludes the possibility of surgical intervention.⁹ Data supporting adjuvant chemotherapy in the postoperative setting are also emerging. Two patients with metastatic urachal carcinomas after resection were given multiple courses of titanium silicate/cisplatin; 9 months after chemotherapy discontinuation, the metastases had diminished in size and there were no signs of disease progression.¹⁰ More recently, a patient with a urachal carcinoma with metastatic disease and peritoneal dissemination was reported to be alive and without evidence of disease recurrence 5 years after treatment. Treatment included surgical resection as well as gemcitabine, cisplatin, and FOLFIRI chemotherapy.¹¹

Urachal carcinomas have significant morbidity and mortality, due in large part to their late presentations. Although the rarity of this malignancy has precluded the development of an oncologic protocol, there is a growing amount of evidence in favor of chemotherapy—both as an adjuvant and as a first-line treatment.

ORCID

Ahmed Ebrahim <http://orcid.org/0000-0002-1076-4835>

Nitin Kondapalli <http://orcid.org/0000-0003-2941-9123>

- Choi YJ, Kim JM, Ahn SY, et al. Urachal anomalies in children: a single center experience. *Yonsei Med J*. 2006;47:782–786. doi:10.3349/ymj.2006.47.6.782.
- Hue L, Jacquin M. Cancer colloïde de la lombille et de paroi abdominale antérieure ayant envahi la vessie [Colloidal cancer of the lumbar

- and anterior abdominal wall having invaded the bladder]. *Union Med de la Siene-Inf Rouen*. 1863;6:418–420.
3. Paner GP, Lopez-Beltran A, Sirohi D, et al. Updates in the pathologic diagnosis and classification of epithelial neoplasms of urachal origin. *Adv Anat Pathol*. 2016;23:71–83. doi:10.1097/PAP.0000000000000110.
 4. Szarvas T, Módos O, Niedworok C, et al. Clinical, prognostic, and therapeutic aspects of urachal carcinoma—a comprehensive review with meta-analysis of 1,010 cases. *Urol Oncol*. 2016;34:388–398. doi:10.1016/j.urolonc.2016.04.012.
 5. Behrendt M, Jong J, Rhijn BW. Urachal cancer: contemporary review of the pathological, surgical, and prognostic aspects of this rare disease. *Minerva Urol Nefrol*. 2016;68:172–184.
 6. Bruins HM, Visser O, Ploeg M, et al. The clinical epidemiology of urachal carcinoma: results of a large, population based study. *J Urol*. 2012;188:1102–1107. doi:10.1016/j.juro.2012.06.020.
 7. Sheldon CA, Clayman RV, Gonzalez R, et al. Malignant urachal lesions. *J Urol*. 1984;131:1–8. doi:10.1016/s0022-5347(17)50167-6.
 8. National Comprehensive Cancer Network. Guidelines in oncology: bladder cancer, https://www.nccn.org/professionals/physician_gls/f_guidelines.asp. Published 2018.
 9. Collins DC, Velázquez-Kennedy K, Deady S, et al. National incidence, management and survival of urachal carcinoma. *Rare Tumors*. 2016;8:97–101. doi:10.4081/rt.2016.6257.
 10. Ozawa M, Kuromoto A, Morozumi K, et al. Two cases of urachal carcinoma treated by TS-1/CDDP as adjuvant chemotherapy. *Hinyokika Kyo*. 2017;63:413–419.
 11. Yasui M, Jikuya R, Tatenuma T, et al. Urachal carcinoma with peritoneal dissemination treated with chemotherapy and surgical resection leading to prolonged survival with no recurrence. *Case Rep Urol*. 2018;2018:9836154. doi:10.1155/2018/9836154.

Avocations

Port Vell (Old Harbor) is a waterfront harbor in the port of Barcelona, Catalonia, in Spain. The port is at the end of the famous boulevard La Rambla. Photo taken by Alejandro C. Arroliga, MD, with a Nikon D7100 F 13, ISO 200, 1/50. Dr. Arroliga (Alejandro.Arroliga@BSWHealth.org) is chief medical officer of Baylor Scott & White Health.