



Oncology

Umbilical sparing robotic partial cystectomy for localized urachal adenocarcinoma: A case report

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ABSTRACT

Urachal adenocarcinoma, the third most common histopathological type of non-urothelial bladder cancer, is often aggressive, presenting in advanced stages. Increased understanding of the embryologic origin of the tumor with concurrent advances in surgical technique have allowed partial cystectomy to become the gold standard of surgical treatment. However, the benefit of en bloc umbilectomy remains questionable. Here we present the diagnosis and management of 67-year old patient diagnosed with mucinous cystadenocarcinoma of the urachus treated with umbilical-sparing robotic partial cystectomy. We also provide a review of the existing literature on this rare tumor and its management.

Introduction

The urachus is a remnant of a fetal structure that normally obliterates early in gestation to form the median umbilical ligament. A urachal remnant is detected in about one-third of adults and while its presence is usually a benign finding, it may lead to the development of both benign and malignant lesions. Of all diagnosed bladder tumors, urachal adenocarcinomas compromise less than 1%.¹

Non-metastatic urachal adenocarcinoma is generally managed surgically with partial or radical cystectomy. Negative surgical margin is the most important prognostic marker.² Historically, en bloc umbilectomy with urachal remnant excision has been advocated. However, we present a case of umbilicus sparing robotic partial cystectomy and urachal excision for a small incidentally discovered urachal adenocarcinoma located at the bladder dome.

Case presentation

A 67-year-old with past medical history of renal cell carcinoma (PT1b, G2) status post open partial nephrectomy underwent surveillance computed tomography of his abdomen and pelvis (CTAP). The CTAP illustrated bladder wall thickening and a well-defined mass near the dome of the bladder with tethering to the abdominal wall (Fig. 1 and Fig. 2). Review of prior imaging showed subtle changes in the urachal

area on a CTAP from 6 months prior. Medical history was otherwise significant for type 2 diabetes, hypertension, chronic kidney disease, colon cancer status post sigmoid colectomy, and polysubstance abuse.

The patient then underwent flexible cystoscopy during which areas of hypervascularity were visualized at the dome of the bladder. No other masses or lesions were appreciated. Urine cytology at this time was negative for high grade urothelial carcinoma. The patient was experiencing mild lower urinary tract symptoms including frequency and urgency. No hematuria gross or macroscopic was noted. Preoperative MRI was obtained to better define the lesion and did not show evidence of urachal cyst, diverticulum, or any other urachal anomalies besides bladder wall thickening at the dome (Fig. 3). Preoperative levels of CEA and CA 19-9 were not obtained.

The patient was recommended to undergo robotic partial cystectomy. Given the incidental finding of the urachal mass on imaging, the patient elected to proceed with robotic partial cystectomy without en bloc umbilectomy because of the small size and asymptomatic nature of the lesion.

Pathological specimen revealed a cystic mucinous tumor, 1.8 cm in largest dimension, originating within the bladder dome, associated with a urachal remnant. The cyst was found to contain abundant luminal mucin with a lining of mucinous epithelium lacking nuclear atypia. Pseudostratification and tufting architecture were present. No invasive components were appreciated. All surgical margins and lymph nodes

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Fig. 1. CTAP (sagittal and transverse views) illustrating well defined mass at the bladder dome and tethering to the abdominal wall.



Fig. 2. Pre-operative MRI showing bladder wall thickening at the dome.

were negative. Surveillance CT revealed no evidence of disease at 6 months post operatively and the patient was doing well at time of his annual follow up.

Discussion

Historically, radical cystectomy with pelvic lymph node dissection was considered the optimal treatment for urachal adenocarcinoma.² Today, increased understanding of the embryologic origin of the tumor with concurrent advances in surgical technique have allowed partial cystectomy to become the gold standard of surgical treatment for patients with urachal adenocarcinoma with the largest, most recent series showing no significant difference in survival between partial vs. radical cystectomy.^{1,2}

Because synchronous tumor may occur along the urachal tract, the recommendation is to treat these tumors with en bloc resection of the urachal ligament with the umbilicus and anterior abdominal wall in

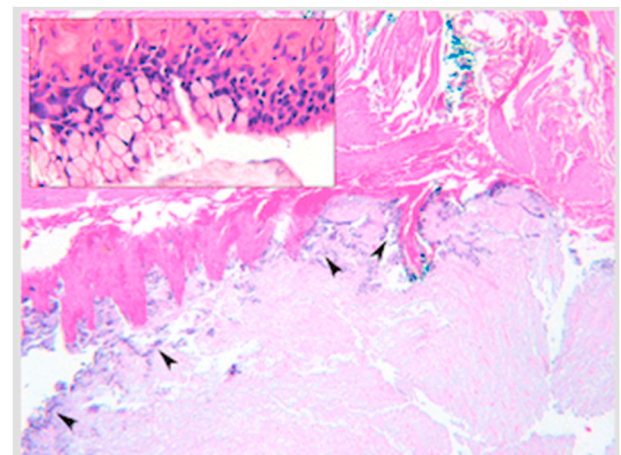


Fig. 3. These images of the bladder lesion show a cystic cavity embedded in smooth muscle and filled with light blue mucinous debris. The arrowheads show the location of the goblet cells in the epithelial lining. (Hematoxylin & eosin, original magnification $\times 40$). The inset is a higher power magnification of a segment of the mucosal lining showing non-mucinous urothelium on the right and the goblet cell metaplasia on the left. (Hematoxylin and eosin, original magnification X250). (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

addition to partial cystectomy and pelvic lymph node dissection when indicated [2]. This recommendation is based partially upon a retrospective study of 42 patients with urachal carcinoma spanning 17 years published in 2003 that showed that long-term survivors were more likely to be treated with en bloc resection [2]. 13 of 16 long-term survivors were those treated with both en bloc resection and umbilectomy, this finding, was however not statistically significant [2]. Furthermore, Sheldon et al. reported navel invasion on autopsy in only 7% of patients who died from urachal carcinoma.³

Ashley et al.'s analysis of 130 patients with urachal carcinoma found that positive surgical margins, high tumor grade, positive nodes, metastasis at time of diagnosis, and failure to perform umbilectomy were independent predictors of death.⁴ However, in their multivariate analysis, only tumor grade and positive margins remained significant predictors of death.⁴

While the above studies provide rationale for umbilical resection, its utility has been questioned by other groups. Umbilectomy is not without morbidity, in addition to its significant impact on post-operative perception of body image. The Ashley study has a large proportion of patients who had very advanced stage tumors at time of resection which

may partially explain the finding that lack of umbilectomy was an independent predictor of death.⁴ In 2019, Pavelescu and colleagues described their findings on 16 patients who underwent surgical resection of urachal carcinoma between 2005 and 2015, 11 of 16 did not undergo umbilectomy.⁵ They report no cases of umbilical recurrence after a mean follow-up of 2.5 years in their series of localized urachal adenocarcinoma.⁵

En bloc umbilectomy increases surgical complexity and potential morbidity, in addition to cosmetic issues with removing the umbilicus. Given the unclear benefit of an umbilectomy for localized urachal adenocarcinoma, our patient elected to forego umbilectomy. Foregoing umbilectomy allowed the partial cystectomy to be completed robotically.

Conclusion

En bloc umbilectomy for urachal adenocarcinoma increases the scope of the surgical procedure for localized urachal carcinomas. We have presented a case where a clinically localized urachal

adenocarcinoma was successfully managed while omitting umbilectomy. Review of the literature indicates that for smaller, more localized masses without obvious umbilical involvement, umbilectomy may be safely omitted. Foregoing umbilectomy may also make minimally invasive partial cystectomy more feasible without compromising oncologic outcomes.

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