Case Report

Extended partial cystectomy with augmentation cystoplasty in urachal adenocarcinoma: An oncologically favorable but underutilized alternative to radical cystectomy

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Abstract

Partial/extended partial cystectomy (EPC) is the most common surgery done for localized urachal malignancies. However, sometimes, after EPC, patients may be left with small bladder remnant, reconstruction of which will result in very small capacity bladder with resultant severe storage voiding symptoms. We report a case of urachal adenocarcinoma, in which when a small bladder remnant was left post-EPC, instead of proceeding with radical cystectomy (RC) and neobladder (the standard alternative), bladder augmentation was done with good oncological and voiding outcome. Augmentation cystoplasty has many advantages over neobladder and we suggest it as an oncologically comparable alternative to RC with neobladder, which has been underutilized in urachal malignancies as we found on literature review.

Key Words: Augmentation cystoplasty, extended partial cystectomy, radical cystectomy, urachal adenocarcinoma

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INTRODUCTION

Primary urachal adenocarcinoma (UA) is a rare bladder cancer with 350 cases reported until 2007.[1] Extended partial cystectomy (EPC) along with excision of urachal remnants is the most common surgery performed for localized urachal tumors. Post EPC, a very small bladder remnant may be left occasionally. As an alternative to radical cystectomy (RC), we opted instead to do augmentation cystoplasty (AC) with good oncological and voiding result. There are few cases where this option has been exercised but with good result, and we feel bladder augmentation may be a better alternative to RC.[2-4]

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CASE REPORT

A 62-year-old diabetic male presented with two episodes of painless hematuria in 6 months. Physical examination was essentially normal. Investigations showed creatinine

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of 1.1 mg/dL, hemoglobin of 14.2 gm/dL, and urine examination showed microhematuria. Urine cytology was negative for malignant cells. Computed tomography (CT) scan showed single heterogeneously enhancing bladder mass [Figure 1], no pelvic lymphadenopathy, and normal kidneys. After negative metastatic workup, the patient underwent cystoscopy that showed a single solid bladder tumor on dome and anterior wall with a bladder capacity of 250 mL. Rest of the bladder mucosa was normal. Transurethral resection of bladder tumor was performed. Histopathology confirmed UA [Figure 2]. Cystoscopically guided laparoscopic EPC along with excision of urachal remnants and umbilicus along with pelvic lymphadenectomy was done. After confirming negative surgical margins by frozen section, the patient was left with very small bladder (almost whole of the supratrigonal bladder was excised) [Figure 3]. Approximation of bladder remnant would have been difficult, if possible, and resulting bladder would have been like a thimble bladder with very small capacity. Hence, the patient underwent open AC using 15 cm of ileum in U configuration in a standard technique (after extending the incision by which umbilicus was excised and specimen extracted). Final histopathology confirmed UA with negative margins and pelvic nodes being free of tumor. Postoperative recovery was uneventful, with a cystogram postoperatively that showed a bladder capacity of 250 mL. The patient is voiding well every 2 hourly and well-satisfied with voiding, though no formal questionnaire-based scale was used. Uroflowmetry showed voided volume of 190 mL with Qmax of 10 mL/min with postvoid urine of 50 mL. The patient never had any nocturnal incontinence, but had nocturnal frequency of 2–3 times. After 1 year of follow-up, there was no evidence of local or systemic recurrence of tumor on cystoscopy, CT abdomen, liver function, and X-ray chest.

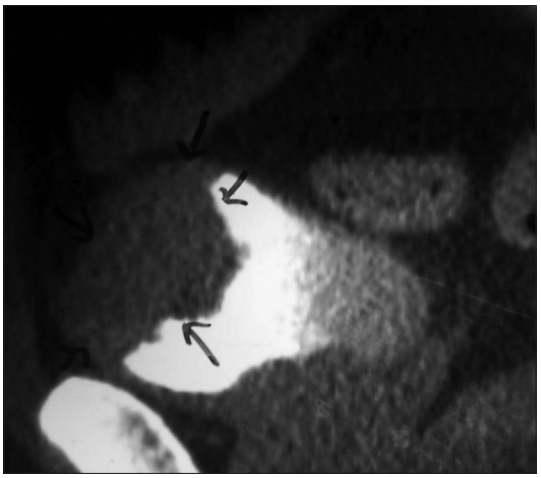

Figure 1: Computed tomography scan showing dome and anterior bladder wall malignancy

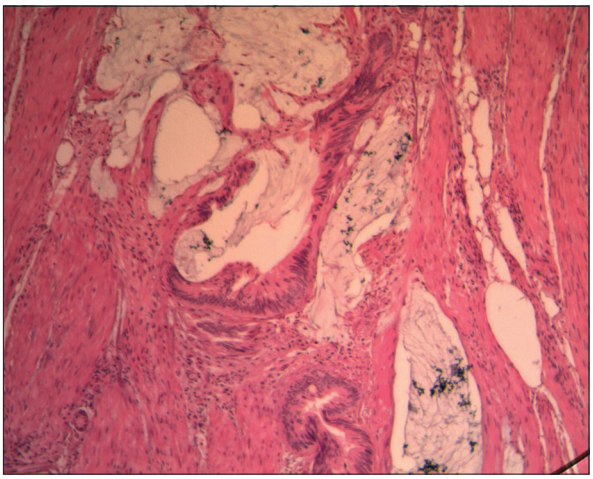

Figure 2: Histopathology: Urachal adenocarcinoma with mucin lakes

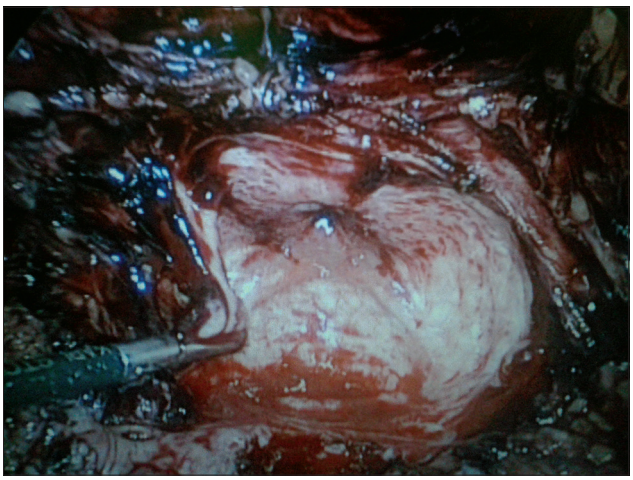

Figure 3: Small bladder remnant after laparoscopic extended partial cystectomy

DISCUSSION

Remnants of urachus (fetal excretory organ) in adults may persist as tubular or cystic structure in one-third cases, consisting of mucosa, smooth muscle, and connective tissue from which urachal cancers can develop.[5]

Adenocarcinoma (UA) is the most common urachal malignancy. It is a highly malignant cancer with poor prognosis as compared to transitional cell carcinoma (TCC).[5] Chemotherapy is the mainstay of metastatic and inoperable (5–17%) cases, but chemotherapy is less standardized and less effective as compared to TCC.[5] Surgery is the most effective option for localized disease. Due to focal nature of disease, with no field changes in rest of the bladder mucosa, these tumors are ideally suited for EPC.[6] It is important not to compromise on margin, as positive margin is a worse prognostic factor.[7] Extension of microscopic disease beyond palpable margin is common and hence margin of 2–3 cm with multiple frozen sections is advocated by Donat and Harry.[8] Hence, there should be no hesitation in resecting adjacent organs/abdominal wall to achieve R0 status. Once

negative margin is achieved, survival is correlated to stage and it is not affected by surgical procedure (EPC/RC).[7] Two staging systems are used; Sheldon [Table 1][5] and Mayo Clinic.[5] Higher stage is associated with worse survival.[4] In the largest series of 60 cases, out of which 80% had hematuria, (indicating IIIA disease) 5 years survival of 49% with recurrence rate of 15% was observed.[7]

EPC with excision of urachal remnant and umbilicus is the most common surgical procedure (92% cases) in localized disease.[1] Historically, earlier RC was recommended due to poor results with partial cystectomy.[6] Failure to take wide margins as realized now may be the reason for these results. The term EPC is emphasized to differentiate it from less radical procedure of partial cystectomy which involves excision of only dome of bladder.[8] RC is recommended if extent of resection results in inadequate functional urinary reservoir.[9] In our case, RC would have the standard procedure along with neobladder.

Neobladder is associated with many disadvantages, notably high rates of persistent night time incontinence, particularly in the first 2 years.[10] Further, with a chance of recurrence of 50%, for which no uniformly effective chemotherapy is available, neobladder seems a less suitable option for UA than TCC, which has better prognosis and better chemotherapy options.

Oncological outcome after EPC/RC, once negative margin is achieved is found to be equivalent.[8] We thus chose to perform augmentation after EPC rather than RC. It has many advantages over neobladder: Technical ease, use of smaller length of intestine, no need of ureter reimplantation, good continence, and less chance of requiring intermittent catheterization (6% when augmentation for nonneurogenic bladders are considered).[11]

AC has been used less commonly after EPC. Certainly, it has been advocated as an option after EPC,[8] unlike a recent recommendation to proceed with RC in the event of a small bladder remnant post-EPC in a review.[9] In an extensive PubMed search using words, "urachal adenocarcinoma," "augmentation," "augmentation cystoplasty," and "ileocystoplasty," we could find three cases from 1991 to date including one case of postrenal transplant detected UA with good oncological result on follow-up of 2 years.[2-4]

Table 1: Sheldon classification of urachal adenocarcinoma[5]

	Description of urachal malignancy
I	Confined to urachal mucosa
II	Urachal invasion present
IIIA	Extension to bladder
IIIB	Abdominal wall extension
IIIC	Peritoneal extension
IIID	Extension to viscera other than bladder
IVA	Lymph node metastasis
IVB	Distant metastasis

Evidence for the use of bladder augmentation as our case is rather limited due to few cases reported. Further, it will be difficult to have a large series of such cases for comparison of bladder augmentation to RC with neobladder in view of rarity of urachal malignancies that are candidates for RC. Lack of long-term follow-up is a limitation in our case and the cases reported earlier. We could not find any standardized protocol for follow-up of UA on literature search. We plan to keep our patient on follow-up of 6 monthly X-ray chest, liver functions, and ultrasound abdomen, along with annual cystoscopy and CT abdomen.

To summarize, AC after EPC may be a good alternative to RC with neobladder when a very small bladder remnant is left, but this option has been underutilized in UA. It gives a good capacity bladder with excellent continence and good voiding with good short-term oncological result and provides oncological principles of resection as discussed that are followed.

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Conflicts of interest
There are no conflicts of interest.

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