

ORIGINAL ARTICLE

Is partial cystectomy an oncologically viable option for primary urachal adenocarcinoma?

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Abstract

Aim: Urachal adenocarcinomas (UA) of the urinary bladder constitute 2% of all urinary bladder malignancies and arise at the dome of the bladder. Most UA are treated with a partial cystectomy (PC) with en bloc resection of the median umbilical ligament and umbilicus. We aim to evaluate the oncological outcome of patients treated for UA using partial cystectomy in our department.

Patients and methods: Twelve were diagnosed with UA on initial transurethral resection (TUR) biopsies from January 2001 until October 2010. The mean age at the time of diagnosis was 57.67 years (Range 37-82) amongst whom 67% (8) were male. Two patients had previous malignancies which included TCC of the ureter and Prostate Adenocarcinoma. The mean duration from diagnosis to PC was 5.6 weeks (Range 3-11). Fifty percent (6) of patients underwent a cystogram and the all catheters were removed at 2 weeks following PC. Histology in all 12 patients confirmed UA and the incidence of serosal positive margin rate was 33% (4). All patients were followed up with sequential cystoscopies and radiological imaging.

Results: At a mean follow up of 36.9 months (4.4-97.2), the incidence of local recurrence was 8.3% (1) and this patient had a subsequent radical cystectomy. Distant metastasis was seen in 33% (4) of patients amongst whom 2 had positive margins with no evidence of prior local recurrence. Four patients (33%) died of UA and all cases with metastatic disease belonged to stage pT3 or higher.

Conclusion: Partial cystectomy is an oncologically viable option in patients with Urachal adenocarcinomas. The final stage and serosal positive margin status on the final histopathological analysis of the specimen at partial cystectomy are significant predictive factors on the risk of metastasis and local recurrence. Patients with both the above factors should be followed up very closely to identify local recurrence and metastasis.

Key words

Urachal adenocarcinoma, Partial cystectomy, Oncological outcome

1 Introduction

Urachal adenocarcinoma (UA) is a rare bladder malignancy and is estimated to account for 0.5-2% of all newly diagnosed urinary bladder tumours^[1-4]. In addition to being rare, UA is aggressive with current 5 year survival rates ranging from

6.5% to 61%^[5-9]. Surgery is the only option for curative treatment, in the form of either partial cystectomy (PC) or radical cystectomy (RC)^[9]. Given the rarity of this malignancy, no prospective clinical trials exist to compare the two surgical procedures^[4]. With a lack of definitive guidelines, the choice of management is based largely on single institution reports.

Traditionally patients undergoing a RC for UA had an 80% 5 year cancer survival rate. A RC involves the patient undergoing a concomitant urinary diversion (Ileal conduit or neo-bladder) which has clinical, metabolic and quality of life repercussions for patients. A partial cystectomy has the benefit of bladder preservation. In previous literature the oncological outcome of patients with UA undergoing PC were poor with data from the 1970s indicating very high recurrence upto 50-78 %^[10, 11]. However, data over the past twenty years is more reassuring and indicates both surgical procedures have comparable 5 year survival rates^[12-14]. Therefore, the current trend is for bladder preservation with PC with en bloc resection of the median umbilical ligament and umbilicus^[9]. Partial cystectomy has certain advantages over radical cystectomy, such as preserving a functionally continent native urinary reservoir and sparing of potency in males^[9, 12].

We aim to evaluate the oncological viability of open partial cystectomy (PC) with en bloc resection of the median umbilical ligament and umbilicus in the management of UA in our institution making this on the largest single center series from the United Kingdom (UK).

2 Materials and methods

From January 2001 until October 2010, Twelve patients (n=12) were diagnosed with primary UA of the urinary bladder on initial cystoscopy and transurethral resection biopsies. All patients underwent CT and MRI staging followed by case discussion at our centers uro-oncology multi-disciplinary meeting (MDT) meeting.



Figure 1. Creation of initial of cystostomy

Patients were carefully counseled pre-operatively highlighting the potential incidence of tumour recurrence and need for future radical cystectomy and urinary diversion. Our surgical technique illustrated in Figure 1-3 involved an initial midline laparotomy incision followed by the mobilization of the medial umbilical ligament and creation of the extraperitoneal space. On identification of the bladder an open cystostomy is created and the tumour is identified. Using the LigaSure Impact device[®] a circumferential incision is made with the bladder ensuring a minimal clearance margin of 2cm. The specimen is sent for frozen section and once the margin status is confirmed to be clear the bladder is closed in 2 layers. A

catheter is left insitu. We perform a cystogram in 2 weeks followed by a removal of catheter. All patients from 2007 underwent a partial cystectomy using the LigaSure Impact device[®] (n=7).

As this report spans over ten years, the surgical technique has evolved during the period to account for modernization in technology. All patients (n=12) underwent an open partial cystectomy with en bloc resection of the median umbilical ligament and umbilicus plus pelvic lymph node dissection upto the level of the external iliac lymph nodes. We used the LigaSure Impact device[®] to reduce the risk of intra operative bleeding.



Figure 2. Application of LigaSure Impact device[®] circumferentially around bladder with 2 cm margin from visible Urachal Adenocarcinoma at dome



Figure 3. Excised Urachal adenocarcinomas

3 Results

Of the study population, two patients had previous malignancies prior to being diagnosed with UA. These included transitional cell carcinoma (TCC) of the Ureter and Prostate Adenocarcinoma. The male to female ratio was 8:4 with mean age at diagnosis being 57.67 years (range 37-82). Table 1 summarizes the characteristics of the study population.

Table 1. Study Population Characteristics

Race	Caucasian N=12
Male: Female Ratio	8:4
Mean age at diagnosis	57.67 Years
History of previous malignancy	N=2

The most common presenting symptom was visible haematuria (n= 9, 75%), other presenting symptoms are summarized in Table 2.

Table 2. Presenting symptoms

Presenting Symptom	Number of patients
Haematuria	9
Haematuria/Dysuria	1
Haematuria/Suprapubic Pain	1
Incidental finding on follow up for previous malignancy	1

The mean duration from diagnosis to PC was 5.6 weeks (Range 3-11). Thirty percent (n=6) had a cystogram post-operatively (based on individual surgeon preference), and all catheters were removed at 2 weeks following PC. Histology confirmed UA in all 12 patients and staging wise, 66.7% (n=8) of the tumours were T3 (see Table 3).

Table 3. Tumour Staging

Tumour Stage	Percentage of Cases
T1	16.67 % (n=2)
T2	0%
T3	66.67% (n=8)
T4	16.67% (n=2)

One patient (n=1) developed a small leak from the bladder during surgery but this was immediately oversewn and fixed intraoperatively, other than this incident, there were no intraoperative complications. Postoperatively one patient (n=1) suffered a complication; an anastomotic leak with a wound infection but made subsequent good recoveries.

All patients were followed up with cystoscopies at 3 months and subsequent regular cystoscopies along with radiological imaging.

The positive serosal margin rate was 33% (n=4) on final histology. Distant metastasis were seen in 33% (n=4) of whom two had positive margins with no evidence of prior or local recurrence. All cases with metastatic disease were stage T3 or higher on final histology.

At a mean follow up of 36.9 months (4.4-97.2), the incidence of local recurrence was 8.3 % (1), and this patient had a subsequent radical cystectomy. A total of four patients died from UA.

4 Discussion

Urachal adenocarcinomas (UA) is a rare malignancy worldwide and there are approximately details of 300 cases published^[3]. In view of the above there continues to be the need to establish definitive guidelines for the ideal surgical technique to manage these patients. The aim of any type of surgical intervention either with a partial or radical cystectomy is to achieve cancer cure. Current literature highlights that surgery is the only oncological viable option to cure patients with UA^[9]. The response to chemotherapy in patients with UA has been traditionally thought to be poor, making this a chemoresistant tumour^[4, 15-17]. In recent literature however a partial response has been seen in patients with UA with

cisplatin combination (S-1/CDDP) chemotherapy and combination 5 fluorouracil and irinotecan^[1, 18]. No long term data is currently available and hence the role of chemotherapy needs to be fully evaluated.

Urinary Bladder UA tumours often tend to be insidious in onset and gradually progressive. Patient develops symptoms when the UA is large making haematuria the commonest presentation^[4]. This time delay is a risk factor for local invasion and metastasis^[3, 9, 19]. The debate on whether to perform a RC or PC for UA goes back to the 1930s^[15]. From a patient quality of life perspective a RC has the distinct disadvantage of requiring the formation of an ileal conduit but this procedure has traditionally been associated with better 5 year survival rates^[10]. However, data from the past twenty years has demonstrated on numerous occasions that RC and PC have no difference in terms of survival rates^[3, 13, 15].

The largest patient series of UA was published by Ashley et al. in 2006^[3]. In this paper the authors reviewed 60 cases of UA which were surgically treated with either RC or PC. It was again confirmed that no difference in survival was noted between patients who underwent either procedure. They cited 5 year survival rates of 49% and recurrence rates of 15%. In addition to these findings, Ashley et al. identified two new key prognostic factors which were positive surgical margins and tumour stage. Previously the influence of tumour stage had not been reported^[7, 20].

Our data demonstrates an 80% 5 year survival rate and a recurrence rate of 8.3%. These results are very reassuring in supporting the choice of PC in managing UA. This 80% survival rate is comparable with the outcomes of RC as published by Anderstrom et al. in 1983. On this basis, it is reasonable to conclude PC has an oncologically comparable outcome to RC. PC also has the advantage of bladder preservation and since most UA occur at the dome of the bladder, surgical removal of the tumour often leaves enough tissue to reconstruct the bladder with adequate bladder capacities of approximately 250ml.

Our results also support the importance of positive serosal surgical margins and tumour grade. In our series, 33% (n=4) of patients developed distant metastasis of whom two had positive margins with no evidence of prior or local recurrence. All cases with metastatic disease were stage T3 or higher. There is currently no established management for patients with metastatic UA due to its highly aggressive nature^[3].

Unfortunately, the recurrence rates of UA in patients having undergone RC are not readily available therefore could not be compared. However, when compared to previously published series involving PC, our rates are lower (5% vs. 15% as reported by Ashley et al.). Given the importance of margin status, this lower recurrence rate may perhaps be linked to our usage of frozen sections intraoperatively for 5 patients.

In terms of clinical presentation, 75% (n=9) of patients had haematuria. This compares well with Ashley et al. which cited rates of roughly 80% and confirm previous studies that haematuria is the most common clinical presentation^[2, 3]. Not only is it a common presentation, but Ashley et al. also identified it as an important predictor of malignancy as their study demonstrated a 17-fold risk of cancer in patients presenting with haematuria.

Despite the reassuring data this study presents in terms of supporting PC as an oncologically viable option for UA, several limitations exist. As UA is rare, our sample size is limited (n=12). Any study with small sample sizes faces the sample problem larger scale applicability. In addition, over the 10 year period this study was undertaken, surgical technique has improved with the use of better technologies such as frozen sections and the Ligasure device. We will publish further results when we have longer follow up and larger patient numbers available to verify our results further.

5 Conclusion

Given the rarity of Urachal Adenocarcinoma the value of single institution reports cannot be underestimated. We have demonstrated Partial Cystectomy is an oncologically comparable to Radical Cystectomy in patients Urachal Adenocarcinoma with of 5 year cancer specific and overall survival rates. We have also demonstrated the lowest published rates of recurrence which further supports the use of PC. In our series the two most important factors to predict recurrence

and progression are positive serosal margin rate and T stage (>T3) on final histopathology. Further research is needed to firmly establish the role of chemotherapy and other multidisciplinary forms of management for UA given its aggressiveness.

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