Urachal Carcinoma: Contemporary Surgical Outcomes

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Purpose: We determined surgical outcomes in a contemporary series of patients who underwent surgery for urachal carcinoma.

Materials and Methods: A cohort of 50 patients with clinically staged localized urachal carcinoma underwent extended partial cystectomy, including the urachal tumor mass and entire urachus. The patients were followed a median of 5+ years for cancer-free survival.

Results: Of the 50 patients 93% (26 of 28) with tumor confined to the urachus and bladder survived compared to 69% (9 of 13) with extravesical or peri-urachal tumor invasion and none (9 patients) with tumor invasion into the peritoneal cavity. Local recurrence was noted in 9 patients (18%) and 2 had salvage therapy. The most significant predictors of survival were pathological tumor stage and negative surgical margins.

Conclusions: Wide resection of the tumor mass and entire urachus resulting in negative soft tissue and bladder margins cures the majority of nonmetastatic urachal cancers.

Key Words: urachus, carcinoma, salvage therapy

Urachal carcinoma is rare, accounting for less than 1% of bladder neoplasms. Urachal remnants persist in and above the bladder, and urachal carcinoma may arise from any of these segments. Most are adenocarcinomas but other histological subtypes are described. The recommended treatment is primarily surgical, with extended partial cystectomy, en bloc excision of the urachal mass, urachal tract and umbilicus, and pelvic lymph node dissection. While some have advocated radical cystectomy as definitive therapy, this procedure can usually be reserved for larger tumors that involve more than the upper hemisphere of the bladder. Radiation and chemotherapy are ineffective against urachal carcinoma.

Although most urachal cancer invades the bladder, it is associated with a poor prognosis due to advanced stage at diagnosis, and extravesical tumor growth favoring local recurrence and distant metastases. Unfavorable tumor features coupled with a lack of effective chemotherapy for recurrent disease emphasize the importance of surgery as crucial for the survival of patients with nonmetastatic urachal cancer. In this report we focus on surgical outcomes in a contemporary cohort of patients with urachal carcinoma who were treated in a consistent fashion with extended partial cystectomy and pelvic lymphadenectomy.

PATIENTS AND METHODS

We operated on 50 patients who presented with a presumed urachal carcinoma between 1984 and 2004. All patients were diagnosed with transurethral biopsy of a solitary tumor located at or near the dome of the bladder. Cases were staged as nonmetastatic cancer on radiographic scans of the chest, abdomen and pelvis. Because adenocarcinoma of the dome cannot be distinguished clinically from urachal carcinoma, the cases were treated as urachal cancer. Each patient underwent extended partial cystectomy with en bloc resection of the entire urachus including the umbilicus, the posterior rectus fascia, and a generous portion of overlying peritoneum and perivesical soft tissue extending out to the lateral pelvic sidewalls. The superior vesical arteries were sacrificed and the upper half of the bladder above this level was removed. Wide margins of bladder were incorporated in the surgical specimen to ensure removal of tumor in the bladder wall and microscopic tumor involving perivesical fat. Resection of involved adjacent organs was done when needed to remove all disease. A bilateral pelvic lymph node dissection was performed, removing at a minimum all distal common iliac, external iliac, obturator and hypogastric nodes. Six surgeons performed the operations. Final pathological stage was based on the Sheldon staging system for the primary urachal tumor. However, node status and distant metastases were considered separate categories.[1]

The end points of our study were cancer-free survival and local recurrence. Survival was defined as the time from surgery until death from urachal cancer. Patients alive at last followup or who died of other causes were censored at that point. Local recurrence was defined as tumor recurrence within the pelvis or bladder. Associations between patient, tumor and surgical variables were tested for their

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Nothing to disclose.

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impact on survival by Pearson's chi-square test. Kaplan-Meier survival curves were constructed and compared using the log rank test. Cox proportional hazards regression was used in a multivariate analysis for predictors of post-cystectomy survival. Statistical tests were 2-sided and p values less than 0.05 were considered significant. The Institutional Review Board approved the study.

RESULTS

Clinical characteristics of the 50 patients presenting with urachal carcinoma are shown in table 1.[1–13] The male-to-female ratio was 1.8:1. Median patient age was 50 years (range 16 to 87). The majority of urachal tumors (88%) was high grade adenocarcinoma. All but 4 urachal cancers showed local extension (stage III). Half of these were confined to the urachus and bladder (IIIA) and half invaded adjacent fat (IIIB) or extended into the peritoneal cavity (IIIC, D). In addition to extended partial cystectomy, tumor resection involving the colon (2 cases), small bowel (1 case), portion of omentum (2 cases), anterior abdominal wall (1 case) and peritoneal implants (1 case) was also performed. Six patients (12%) had positive soft tissue surgical margins (local stage IIIB, C or D tumors). None of the patients had a positive bladder margin. Bilateral pelvic lymph node dissection was performed in all patients. Median number of nodes retrieved was 14 (range 4 to 34). Nodal metastases (median 3 positive nodes) were found in 8 (16%) patients. In 2 patients distant metastases were found at surgery in the liver and ovary despite clinically negative studies. In both cases the metastases were resected along with the primary urachal tumor. None of the patients received adjuvant chemotherapy.

TABLE 1. *Characteristics of patients with urachal carcinoma*

	No. (%)
Sex:	
Male	32 (64)
Female	18 (36)
Age:	
Younger than 55	28 (56)
Older than 55	22 (44)
Tumor size (cm):	
Less than 4	23 (46)
4 or Greater	27 (54)
Adenocarcinoma:	44 (88)
Enteric type	38
Signet-ring cell	4
Lymphoepithelioma-like Ca	2
Undifferentiated Ca:	6 (12)
Squamous cell features	2
Transitional cell/adenomatous features	3
Clear cell features	1
Tumor grade:	
Low	3 (6)
High	47 (94)
Pathological tumor stage (Sheldon):	
I (confined to urachal mucosa)	2 (4)
II (invasion confined to urachus)	2 (4)
III (local extension)	46 (92)
IIIA (to bladder)	24 (48)
IIIB (peri-urachal + vesical fat)	13 (26)
IIIC (to peritoneum)	6 (12)
IIID (to viscera other than bladder)	3 (6)
IVB (metastasis to other organs)*	2 (4)
Lymph node metastasis:	
Yes	8 (16)
No	42 (84)
Pos surgical margins	6 (12)
Local recurrence:	9 (18)
Pelvis/bladder	6
Bladder only	3
Distant metastases:	16 (32)
Liver	8
Lung	6
Bone	5
Peritoneal	2

* Discovered at surgery (1 each in liver and ovary).

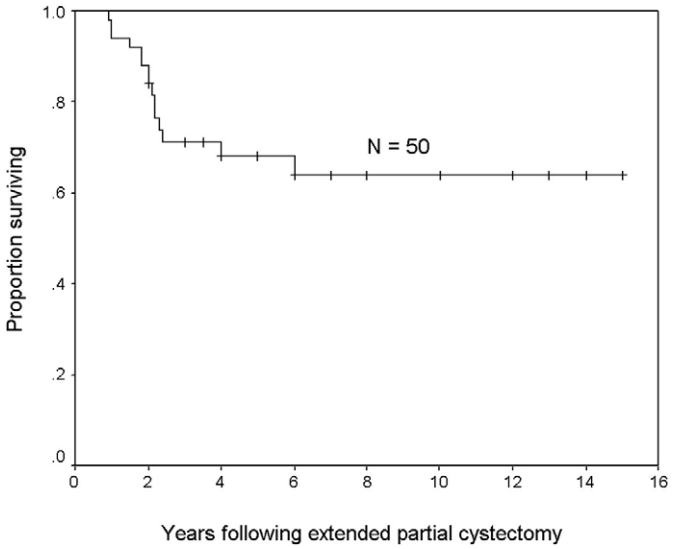

FIG. 1. Cancer-free survival of patients with urachal carcinoma

Figure 1 shows cancer-free survival in all 50 patients presenting with urachal cancer. A total of 35 patients (70%) survived. Median survival was not reached with a median followup of 5.1 years (6.8 years for survivors and 2.2 years for patients who died of disease). Table 2 shows patient outcomes by clinical and tumor features. Gender, age, tumor size, histological pattern and tumor grade did not correlate with 5-year survival. Worse survival was associated with advanced pathological stage, presence of nodal metastases and positive surgical margins. Figure 2 shows cancer-free survival stratified by pathological groups. Of 28 patients with tumor confined to the urachus and bladder (stage IIIA or less), 26 (93%) survived 5 years compared with 9 of 22 (41%) with local invasion of surrounding fat (stage IIIB) or peritoneal cavity (median survival 2.3 years, 95% CI 1.8–2.7). Figure 3 shows cancer-free survival by surgical margins. Surgical margins were positive in soft tissue at the lateral pelvic side walls or overlying peritoneum. Only 1 of 6 patients (17%) with a positive surgical margin survived (median survival 1.5 years, 95% CI 0.86–2.1) compared with 34 of 44 (77%) patients who had negative surgical margins. Of 8 patients with node positive disease 2 (25%) survived compared with 33 of 42 (78%) with node negative disease. Of the 29 patients having 14 or more nodes removed 21 (72%) survived vs 14 of 21 patients (67%) with fewer than 14 nodes ($p = 0.66$). Although the number of nodes examined did not correlate with survival, 6 of the 8 patients found to have nodal metastases had more than 14 nodes resected, including the 2 who survived. Both patients with node positive disease (2 and 3 positive nodes, respectively) did not receive adjuvant chemotherapy and were disease-free after 4 and 8 years of followup.

In 9 cases (18%) local recurrence was noted in the pelvis (6 cases, 3 invading the bladder) and bladder only (3 cases) within the first 2 years of followup. Of the 9 cases 4 had