

A Case of Urachal Carcinoma

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Abstract

Urachal carcinoma is a rare, non-urothelial carcinoma that represents less than one percent of all bladder cancers. We present the case of a 58-year-old man who presented with the acute onset of painless hematuria. Cystoscopy revealed a bladder mass, and subsequent biopsy of a liver mass confirmed the diagnosis of metastatic urachal adenocarcinoma. The patient initially underwent a chemotherapeutic regimen of 5-fluorouracil, leucovorin, gemcitabine, and cisplatin (GEM-FLP) given every four weeks. Our case illustrates the importance of recognizing and diagnosing urachal carcinoma correctly since the therapeutic options differ compared to traditional urothelial cancers.

Keywords: Urachal cancer; Urachus; Carcinoma; Rare bladder tumors

Introduction

The urachus is the fibrosis remnant of the allantois, a canal that drains the fetal bladder from the dome of the bladder to the umbilical cord. During fetal development, as the genitourinary system develops, the urachus regresses, becoming a fibrous tissue between the bladder dome and umbilicus along the midline. The urachal remnant may persist in approximately one-third of patients [1]. Urachal carcinoma is a rare non-urothelial carcinoma involving this vestigial remnant and accounts for about 0.2% of all bladder cancers [2]. Disseminated metastatic urachal cancer is even rarer.

Histologically, urachal tumors are almost universally adenocarcinomas, whereas the histology of traditional bladder cancer is typically transitional cell carcinoma or more rarely squamous carcinoma.

Diagnostic criteria from M.D. Anderson Cancer Center include the presence of an adenocarcinoma at the location of the bladder dome or elsewhere in the midline of the bladder and a sharp demarcation between tumor and normal surface epithelium. Supportive criteria include enteric-type histology, absence of urothelial dysplasia, absence of cystitis cystica or cystitis glandularis transitioning to the tumor, and absence of primary adenocarcinoma of another organ.

Although rare, the diagnosis of urachal cancer is important not miss, as there are critical differences in treatment and management compared to standard urothelial cancers. We present a case of metastatic urachal cancer and briefly review common diagnostic and treatment considerations.

Case

A 58-year-old male with a history of borderline type 2 diabetes mellitus presented to the urology department with a one-week history of painless, gross hematuria. He denied a history of bleeding, urinary tract infection, nephrolithiasis, or trauma. Relieving factors included drinking fluids. Review of systems was positive for abdominal pain, constipation, urinary urgency and frequency, and back pain. Family history was unremarkable.

His physical examination revealed fullness at the dome of the bladder. The remainder of the exam, including the genitourinary exam, was unremarkable. Laboratory examination revealed a white blood cell count of 6740 cells/uL, a hemoglobin of 14.5 g/dL, a platelet count of 362,000 platelets/uL, a normal basic metabolic panel, and a prostate specific antigen of 0.85 ng/mL.

A flexible cystoscopy revealed an extensive mass involving

the dome of the bladder. A computed tomography (CT) scan of the abdomen and pelvis showed an irregular, partially cystic mass extending off the superior bladder wall, nodular peritoneal thickening consistent with peritoneal seeding, enlarged lymph nodes in the iliac and paraesophageal regions, and a large, 6-cm rim-enhancing mass within the liver (Figures 1 and 2). A subsequent biopsy of the liver mass revealed adenocarcinoma with immunohistochemistry positive for antibodies to CDX2 and cytokeratin 7. These findings were consistent with a diagnosis of metastatic urachal carcinoma.

The patient was started on a therapeutic regimen, developed at M.D. Anderson, consisting of 5-fluorouracil, leucovorin, gemcitabine, and cisplatin (Gem-FLP), essentially combining a cisplatin-based regimen such as those used to treat traditional bladder cancer with 5-fluorouracil for adenocarcinomas. He completed four cycles of this regimen. Complications of treatment included nausea and non-cardiac chest pain. Restaging scans after two cycles showed evidence of interval improvement with a more cystic appearance to the liver metastatic lesion and peritoneal implants, consistent with a response to therapy. However, his scans after four cycles revealed progression (Figure 3). He was switched to second-line therapy, which included cetuximab and



Figure 1: Irregular, partially cystic mass with calcifications extending off the superior bladder wall on computed tomography, transverse view.

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Figure 2: Irregular, partially cystic mass with calcifications extending off the superior bladder wall on computed tomography, sagittal view.

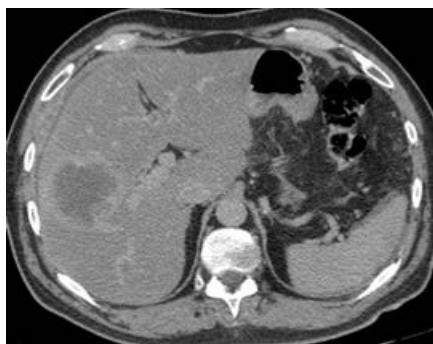


Figure 3: Rim-enhancing mass in the right lobe of the liver on computed tomography.

Stage I	No invasion beyond the urachal mucosa
Stage II	Invasion confined to the urachus
Stage III	Local extension into the bladder (IIIA), abdominal wall (IIIB), peritoneum (IIIC), or viscera other than the bladder (IIID)
Stage IV	Metastases to the regional lymph nodes (IVA) or distant sites (IVB)

Table 1: The sheldon staging system for urachal carcinoma.

irinotecan. While his tumor markers carcinoembryonic antigen (CEA) and serum carbohydrate antigen 19-9 (CA 19-9) have decreased, imaging revealed further progression. He is currently being evaluated for a phase-one trial as third-line/palliative treatment.

Discussion

Urachal carcinoma is a rare and aggressive form of bladder cancer that typically originates in the adult remnants of the fetal allantois, the urachus. Histologically, urachal tumors are almost always adenocarcinomas, in contrast with traditional bladder tumors which tend to be transitional cell carcinomas (urothelial) or rarely squamous-cell carcinomas.

Initially, diagnostic criteria had been proposed that were so strict and numerous that they would have excluded most of the cases of urachal cancer that had been published. In 2006, M.D. Anderson Cancer Center published more practical diagnostic criteria that included:

1. Location in the bladder dome or elsewhere in the midline of the bladder
2. Sharp demarcation between tumor and normal surface epithelium

Supportive criteria: enteric-type histology, absence of urothelial dysplasia, absence of cystitis cystica or cystitis glandularis transitioning

to the tumor, and absence of primary adenocarcinoma of another organ [3].

Presenting symptoms are nonspecific and may include hematuria and/or an abdominal mass/fullness, although many cases are asymptomatic until later stages. Computed tomographic (CT) imaging may show a midline abdominal-wall mass; stippled calcification in the mass is a pathognomonic finding. Diagnosis should be strongly suspected when cystoscopy or surgical resection of tumor reveals an adenocarcinoma in the midline or dome of the bladder.

The Sheldon staging system for urachal carcinoma is comprehensive; however, many patients present with advanced disease due to lack of symptoms at earlier stages. The Sheldon staging system [4] is detailed in Table 1.

It is important to diagnose urachal carcinoma preoperatively in order to determine the proper surgical procedure. In non-metastatic, operable disease, surgical management involves a complete or partial cystectomy with en bloc resection of the urachal ligament and umbilicus. For patients who underwent definitive surgical treatment, five-year overall survival was between 43 and 50% [5]. There is currently no proven role for neo adjuvant or adjuvant chemotherapy for urachal cancer preceding or following a complete surgical resection with negative margins.

For inoperable patients, including those with metastatic disease, systemic chemotherapy traditionally has shown poor efficacy in the treatment of urachal tumors. Earliest reports showed a partial response in with the regimen of 5-fluorouracil, doxorubicin, and mitomycin; however, the responses were not durable [6]. Recently, M.D. Anderson completed a phase II trial of 5-fluorouracil, leucovorin, gemcitabine, and cisplatin (Gem-FLP); results of this prospective trial have not been published at this time. In a review of 26 patients with metastatic disease treated at M.D. Anderson, the median survival for patients was 24 months, compared to 12 months at other institutions [7,8].

Negative prognostic factors include tumor growth in the abdominal wall, peritoneum, and/or adjacent organs; lymph node metastasis; distant metastasis; and macroscopic residual tumor [9].

In conclusion, urachal carcinoma is a rare, aggressive adenocarcinoma of the bladder. Our case illustrates the importance of recognizing and diagnosing urachal carcinoma correctly since both surgical and systemic therapeutic options differ compared to traditional urothelial cancers.

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