

Adenocarcinoma of the Urachus Masquerading as a Right Lower Quadrant Mass:

A Case Report

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Abstract

We report a patient who presented initially with a right lower quadrant mass which was attached to the anterior abdominal wall. The final pathological diagnosis was adenocarcinoma of the urachus, an exceedingly rare bladder tumor. **Key Words:** Urachal anomalies, urachal carcinoma, lower quadrant mass, adenocarcinoma.

URACHAL CARCINOMA IS AN UNUSUAL CANCER with an incidence of approximately 1% of all bladder cancers. Occurring in approximately 1 in 5 million adults, it represents approximately 0.01% of all cancers (1). It may occur at any age from the late teens to the eighth decade, with a predilection for males (2). While the typical presentation for this condition involves hematuria and irritative voiding symptoms, we present such a case which presented initially as a right lower quadrant mass with attachments to the anterior abdominal wall. It is this atypical presentation of an exceedingly rare tumor that challenges clinicians to consider adenocarcinoma of the urachus in the differential diagnosis of a right lower quadrant mass.

Case Report

The patient is a 57-year-old man who complained of a 3-month history of white discharge in the urine without sediment. He also claimed decreased force of stream and occasional double voiding, as well as intermittent perineal pain. He

denied prior medical history. He had had a cholecystectomy in 1980.

On physical examination, the patient was afebrile, with a blood pressure of 130/70 mm Hg and a pulse of 86/minute. Cardiac and pulmonary examinations were unremarkable. Abdominal examination revealed normally active bowel sounds without evidence of guarding or rebound tenderness. Palpation in the right lower quadrant revealed a palpable soft mass between the anterior superior iliac spine and the umbilicus. Auscultation over the mass revealed no evidence of bowel sounds. The testicles were descended bilaterally and the phallus was circumcised. On digital rectal examination, the prostate was without masses and was estimated to be approximately 25 grams. The stool was hemoccult negative.

Prior symptoms of urinary frequency and straining to void were evaluated by uroflowmetry. His uroflow was 8 cc/sec (normal range: greater than 15 cc/sec). Urinalysis revealed 5 rbc/hpf. Urine cytology was negative. Flexible cystoscopy revealed normal bladder mucosa and the suggestion of an inflammatory lesion in the bladder apex possibly consistent with an extrinsic mass or a colonic fistula. This inflammatory lesion could not be biopsied cystoscopically. The differential diagnoses considered at this point are listed in Table 1. CT scan of the abdomen and pelvis revealed a right lower quadrant mass

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TABLE 1
Differential Diagnosis of Right Lower Quadrant Mass

Adenocarcinoma of cecum
Adenocarcinoma of right colon
Cholecystitis
Colovesical fistula
Inflammatory bowel disease
Intussusception of the cecum
Mesenteric adenitis
Pancreatitis
Urachal remnant

thought to be of gastrointestinal origin abutting the bladder. The location of the mass on CT scan allowed for simple needle biopsy to be undertaken. CT-guided needle biopsy of the mass revealed fibrosis and microcalcification with no tumor cells seen.

The patient's complaint of persistent right lower quadrant discomfort over the ensuing weeks prompted laparoscopic exploration of the pelvic lesion. Operative findings noted a 1.0 cm by 1.0 cm mobile lesion at the right anterior bladder wall-peritoneal surface without attachments to the bowel or abdominal wall. Frozen section was consistent with colonic adenocarcinoma. The procedure was converted from laparoscopy to an open retroperitoneal surgical approach. Partial cystectomy was performed through a Pfannenstiel incision approximately 2 cm above the pubis. Pathology revealed moderately differentiated colloid-type urachal adenocarcinoma which extended from the perivesical fat to the mucosa within the urachal remnant. The lesion was intramural, with the bladder mucosa being ulcerated and showing no primary changes (Figure). The final pathological diagnosis was adenocarcinoma of the urachal remnant and not a bladder wall adenocarcinoma. A postoperative CT scan at 3 months failed to reveal any mass lesions. Cystoscopic follow-up revealed no evidence of tumor.

Discussion

Urachal carcinoma accounts for less than 1% of bladder cancers and approximately 40% of vesical adenocarcinomas. More common in men, typical signs and symptoms include abdominal pain and mass, umbilical discharge, dysuria and/or hematuria (1–3). This patient had a diminished uroflow as part of his preoperative workup. Since urachal lesions typically involve the dome of the bladder and spare the bladder neck, it is unlikely that this patient's diminished uroflow is related to the urachal carcinoma. Cystourethroscopy revealed mild prostatic

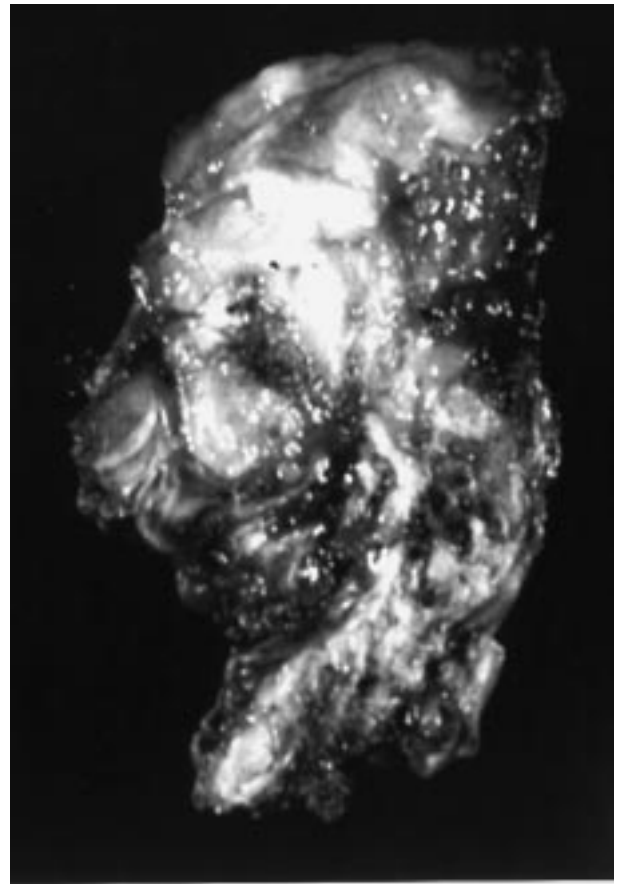


Figure. Gross specimen demonstrating a glistening mucoid tumor mass involving the bladder wall and adjacent soft tissues.

enlargement, which is the likely reason for the observed decreased uroflow.

To evaluate the significance of the findings on CT scan, colonoscopy was performed. Colorectal polyps were not observed, but some small internal hemorrhoids were noted. The patient's continuing abdominal pain and right lower quadrant mass led to the diagnostic laparoscopy, which revealed an extracolonic mass at the anterior bladder wall. The frozen section biopsy was consistent with colonic adenocarcinoma. Taken together, the most likely diagnosis was urachal adenocarcinoma, particularly since the underlying bladder mucosa was normal. The surgical procedure was converted from laparoscopy to an open retroperitoneal surgical approach, in order to perform a partial cystectomy.

The evaluation for a suspected urachal tumor should be similar to evaluation for any bladder tumor. Urinalysis, urine culture and cytology should be performed (3, 4), and a careful history and physical examination should be completed. The most common symptom is hematuria due to erosion of the tumor into the dome of the bladder.

Mucus, although common in the pathologic specimen, is only found in the urine in 15–33% of cases (4). Mucus or bloody discharge from the umbilicus has also been reported (3, 4). In general, lesions are often silent until they have reached an advanced stage.

Diagnostic cystoscopy is an important examination. Chest x-ray, computed tomograph (CT) of the abdomen and transurethral biopsy (if possible) are necessary prior to surgical resection. CT scan provides the most accurate diagnosis and staging for urachal carcinoma. This modality can demonstrate intra- and extravesical involvement, as well as surrounding pelvic lymph nodes (5). Approximately 60% of urachal tumors have regions of low attenuation, which suggests the high mucin content of these lesions. Nearly 50% of scans demonstrate curvilinear, punctate or scattered peripheral calcifications (4, 5). The CT scan can differentiate other lesions that may mimic urachal carcinoma (Table 2).

Cystoscopic appearance can be as follows: a polypoid or papillary mass on the bladder dome, or a bulge with normal overlying urothelium. The urachal opening may reveal bloody or gelatinous discharge (1, 2, 6). Histologic patterns of these tumors include glandular, colloid, papillary or signet ring configuration. Approximately 69% of urachal tumors are mucin-positive adenocarcinomas, 15% mucin-negative adenocarcinoma, 8% sarcoma, 3% squamous cell carcinoma and 2% others. Tumors can spread by direct extension to the space of Retzius, abdominal wall, peritoneum and bladder. Staging of this lesion has been proposed by Sheldon et al. and by others (Table 3) (1, 3). Muscle invasion is nearly always present. Local recurrence is noted in up to 80% of cases, while distant metastasis occurs late (3, 4).

Potential sites of urachal metastases include lungs, liver, bone and regional lymph nodes. CT scan may evaluate the liver and nodes. Chest x-ray, alkaline phosphatase, and/or bone scan should also be performed (2, 3).

The preferred treatment is wide local excision with partial or radical cystectomy. This approach

TABLE 2
Lesions That May Mimic Urachal Carcinoma

Adenocarcinoma of the cecum
Adenocarcinoma of the colon
Dermoid myoma
Ovarian carcinoma
Uterine myoma

TABLE 3
Staging of Urachal Carcinoma

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

From Sheldon C, Clayman RV, Gonzalez R, et al. (1).

should also include en bloc resection of the umbilicus and bilateral pelvic lymphadenectomy (3). To prevent local recurrence, it is very important to maintain wide margins during the resection. Sheldon et al. (1) noted that 21% of recurrences are in the pelvis while 16% occur in the bladder. Local recurrence typically occurs within the two years following initial surgery. Others have suggested radical cystectomy with pelvic node dissection as a preferred treatment. However, there is no significant survival advantage over partial cystectomy alone. Herr (7) suggests that survival is related to the stage of disease at presentation rather than the extent of surgical procedure. The overall five-year survival rate for urachal carcinoma is approximately 50%. Preoperative radiotherapy has also been examined with unproven usefulness. These tumors are relatively resistant to radiation therapy and chemotherapy (8, 9).

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Added in page proof: One year later, a local recurrence was removed. Radiotherapy and chemotherapy were administered.