

# Metastatic Squamous Cell Carcinoma of the Urachus

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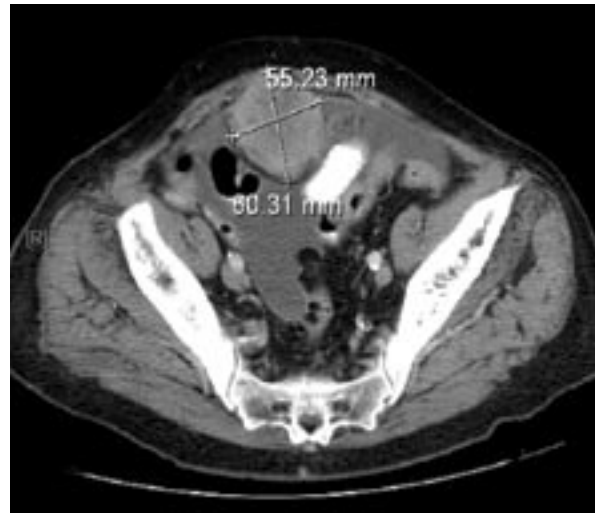
## Case Report

An 87-year-old man with a history of coronary artery disease presented to the emergency room with progressive lower abdominal pain and hematuria of 3 weeks' duration. Physical examination revealed a palpable hard nontender mass measuring 10 × 12 cm in the lower abdomen. No hepatomegaly or splenomegaly was found. Hemoglobin was 9.1 g/dL and urinalysis revealed counts of 468 red blood, 20 squamous, and 1,321 white blood cells per high power field. Cystoscopy was normal except for dimpling of the dome of the urinary bladder. A recent screening colonoscopy was normal. A computed tomography (CT) scan of the abdomen and pelvis revealed a solid mass measuring 5.5 × 6 cm (Figure 1) adjacent to the dome of the urinary bladder with areas of necrosis; multiple low-density lesions in the liver, the largest one measuring 1.5 cm; thickening of the proximal small bowel; significant ascites; nodular densities; thickening of the peritoneum (Figure 2); and multiple lytic and sclerotic changes in the left iliac bone consistent with metastatic disease.

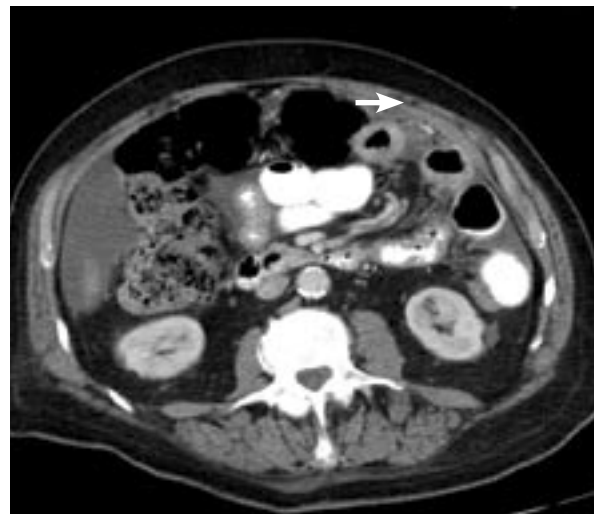
CT-guided biopsy of the pelvic mass was performed and pathology revealed a carcinoma with squamous cell differentiation (Figure 3). The patient was treated with systemic chemotherapy using carboplatin and paclitaxel. Despite best efforts, the patient survived for only a month from the date of diagnosis.

## Discussion

Squamous cell carcinoma of the urachus is very rare; a search of the literature found only eight cases reported from 1870 to 2006. This is the first reported case of metastatic disease at the time of diagnosis. The urachus is a remnant of the allantois, an excretory organ in the fetus. In adulthood it persists as a fibromuscular band plugged with desquamated epithelial cells. It is located in the space of retzius (spatium retropubicum) between the

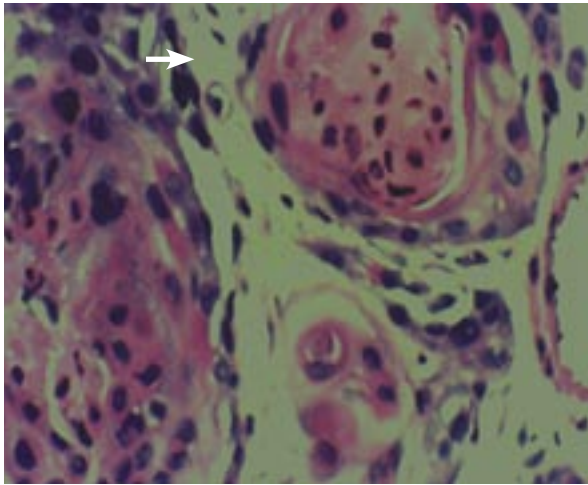


**Figure 1.** Computed tomography scan of the abdomen and pelvis. A solid mass (urachal tumor) adjacent to the dome of the urinary bladder.



**Figure 2.** Computed tomography scan of the abdomen and pelvis showing thickening of the peritoneum.

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**Figure 3.** Urachal squamous cell carcinoma with keratin pearl formation.

fascia transversalis ventrally and the peritoneum dorsally. The urachus measures about 3–10 cm in length and is 8–10 mm in diameter at the dome of the urinary bladder and 2 mm at the apex at the umbilicus.<sup>1</sup>

Carcinoma of the urachus accounts for 0.01% of all malignancies and less than 1% of all bladder tumors. The most common types of urachal cancer are adenocarcinomas (85–90%); transitional and squamous cell carcinomas are rare. The urachal epithelium undergoes squamous metaplasia to give rise to squamous cell carcinoma, as explained by the totipotential theory of Mostofi and associates.<sup>2</sup> The urachal tumors were divided into 7 classes by Begg; of these, intramucosal, intramural, and supravescical are the most important.<sup>3</sup>

Squamous cell carcinoma of the urachus is three times more common in men than in women and patient age ranges from 27 to 87 years. Urachal tumors usually remain undiscovered for a long period and are often found incidentally or at an advanced stage. The common presenting symptoms of squamous cell urachal carcinoma are abdominal pain (75%), bladder symptoms (75%), and hematuria (50%).<sup>4</sup> Clinical symptoms vary with the pattern of local growth of the tumor. Intramural tumors present with hematuria, dysuria, pollakisuria, and symptoms similar to those of bladder cancer, whereas supravescical tumors present as a suprapubic mass and hematuria.<sup>5</sup> Urachal calculi were documented in two cases.<sup>1,6</sup> The most common sites of metastasis of urachal cancers are bone (50%), lung (46%), liver (27%), peritoneal carcinomatosis (23%), and spine (23%), followed by brain and lymph-node manifestations.<sup>7</sup>

Cystoscopy with biopsy and radiographic evaluation with CT or magnetic resonance imaging of the abdomen

and pelvis are needed for the diagnosis. Chest CT scan or radiography may also be needed to exclude metastasis. The traditional staging of urothelial tumors, which arise from the surface epithelium and grow outward, do not apply, as urachal tumors arise in the muscle wall or externally and grow inward.<sup>8</sup> Sheldon and colleagues classified urachal carcinomas into 4 stages: stage I, tumor in urachal mucosa; stage II, invasion confined to the urachus; stage III, local extension to the bladder, abdominal wall, peritoneum, or local viscera; and stage IV, regional or distant metastasis.<sup>4</sup>

Of the eight reported cases of squamous cell carcinoma, 6 patients were treated with excision of the tumor.<sup>5,9</sup> One patient received chemoradiation.<sup>1</sup> Most of the patients died within 1 year of diagnosis and only 1 patient was tumor-free 18 months after partial cystectomy and excision of the tumor.<sup>9</sup> Local control of the tumor is crucial and is usually treated with partial or total cystectomy and en bloc excision of the urachal mass with pelvic lymphadenectomy, peritoneal resection, and umbilectomy. The first operation must control the disease locally and is crucial for the success of the treatment. If there is a local recurrence, the patient is usually not salvageable.<sup>3</sup> Partial cystectomy with en bloc resection of the urachal ligament and umbilicus can lead to long-term control in surgically resectable urachal carcinomas.<sup>8,10</sup> Squamous cell carcinoma of the urachus is very rare and has a high potential for metastasis; prognosis is poor. At present there is no standard chemotherapy regimen for metastatic carcinoma of the urachus. Multi-institutional clinical trials are needed to study the effect of various combination chemotherapy regimens in these patients.

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# Review

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Kodali and colleagues report a very rare tumor: an 87-year-old man presenting with a metastatic squamous cell carcinoma of the urachus.<sup>1</sup> Although urachal tumors are nearly always adenocarcinomas,<sup>2-4</sup> sarcomatoid, squamous, or transitional cell elements have been infrequently reported.<sup>2</sup> Given the rare reports of squamous cell tumors of the urachus, it is important to distinguish this diagnosis from other squamous cell tumors of the bladder that may invade the urachus secondarily or otherwise involve the midline of the bladder, such as a midline squamous tumor arising from a chronic suprapubic catheter.

Wheeler and Hill<sup>5</sup> and Mostofi and colleagues<sup>6</sup> previously published stringent criteria for diagnosing urachal tumors. However, these criteria may be overly restrictive and, if implemented, would exclude many of the known cases of urachal cancer. Investigators at The University of Texas M. D. Anderson Cancer Center (MDACC) have developed a more practical approach for the diagnosis of urachal tumors (Table 1).<sup>7</sup>

It is also important to note that typical bladder staging does not apply in the presence of a urachal tumor, as the bulk of the tumor is often outside the lumen of the bladder. Frequently, the disease is clinically silent until sufficient time has passed to allow growth to penetrate the bladder, thereby causing the urinary symptoms typically associated with this malignancy. In cases in which the tumor is still localized to the bladder and urachal ligament without evidence of metastases, it is appropriate to pursue surgical resection via partial cystectomy with en bloc resection of the urachal ligament and umbilicus. However, even with surgical resection, survival has historically been measured at 44–49% at 5 years.<sup>3,5,8</sup> Recently several factors increasing the risk of relapse have been reported: positive margins at the time of surgery,<sup>3,4</sup> lack of en bloc resection of the urachal ligament and umbilicus,<sup>3,4</sup> peritoneal involvement/carcinomatosis,<sup>3,9</sup> and lymph node or other metastases at the time of surgery.<sup>3</sup>

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There is no standard chemotherapy combination for the treatment of this tumor. Responses have been reported with combinations of paclitaxel with cisplatin.<sup>3,10</sup> A recent MDACC report suggested three responses out of 9 patients treated using combinations of 5-fluorouracil (5-FU) and cisplatin.<sup>3</sup> Currently, a clinical trial of a combination of 5-FU, leucovorin, gemcitabine (Gemzar, Eli Lilly), and cisplatin (Gem-FLP) is ongoing at MDACC for patients with metastatic or surgically unresectable urachal cancer.

The clinical case reported by Kodali and colleagues brings to mind an important question: should we base our treatment upon anatomy or histology? From a surgical point of view, it would be important to err on the side of caution and do en bloc resection of the bladder with the urachal ligament and umbilicus whenever urachal carcinoma is suspected. From the chemotherapy point of view, however, the treatment is not as clear. Most of the larger series have reported chemotherapy responses in the setting of adenocarcinoma histology.<sup>3,10</sup> Our current strategy has been to choose chemotherapy regimens based upon histology (eg, 5-FU–based chemotherapy for adenocarcinomas, ifosfamide/adriamycin combinations for sarcomatoid).<sup>11</sup> Given the responsiveness of other squamous tumors, such as those of the head and neck, to 5-FU and platinum-based combinations, Gem-FLP may still provide a reasonable alternative. Likewise, taxane and platinum combinations may also be active,<sup>9</sup> based upon data with squamous bladder tumors. However, it is unlikely that an 87-year-old patient would be able to tolerate these chemotherapy combinations, especially any that contain high-dose ifosfamide.

Unfortunately, once metastases develop, the prognosis remains very poor, with retrospective series reporting a median survival anywhere from 12 to 24 months.<sup>3,4</sup> Clearly, prospective studies are indicated to determine any potential benefits from systemic chemotherapy. However, given the rarity of urachal cancer, multi-institutional col-

**Table 1.** The University of Texas M. D. Anderson Cancer Center Criteria for Diagnosis of Urachal Tumors

Primary Criteria	Supportive Criteria
Location in the bladder dome or elsewhere in the midline of the bladder	Enteric-type histology
	Absence of urothelial dysplasia
Sharp demarcation between tumor and normal surface epithelium	Absence of cystitis cystica or cystitis glandularis transitioning to the tumor
	Absence of primary adenocarcinoma of another organ

(Continued on page 919)

laboration is necessary in order to fully explore the impact of systemic chemotherapy.

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