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SQUAMOUS CELL CARCINOMA OF THE URACHUS

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Urachal tumors account for only 0.01% of all malignancies and 0.3% of all bladder tumors. Of the urachal tumors described previously most were adenocarcinoma. To our knowledge only 7 cases of squamous cell carcinoma of the urachus have been reported in the literature with no tumor-free survival after the first year. We report a case of urachal squamous cell carcinoma discovered on cystoscopic examination with the longest postoperative tumor-free survival to date of longer than 18 months at last followup.

CASE REPORT

A 70-year-old man presented with dysuria, frequency and painless hematuria 3 months in duration. Urinalysis demonstrated 3 to 7 red blood cells per high power field, while urinary cytology and excretory urography were negative. Cystoscopy detected a calcified 1 × 1 cm. mass at the bladder dome and simultaneous bimanual examination revealed another 3 × 3 cm. palpable suprapubic mass, which were suspicious of urachal tumor. Transurethral biopsy and resection of the bladder dome tumor extracted 2 tan tissue fragments, eventually diagnosed as transitional cell papilloma. The larger, tubular soft tissue mass was 3 × 3 × 12 cm. on computerized tomography (CT) and extended from the superior part of the bladder to the umbilicus (fig. 1, *A*). Partial cystectomy was performed with urachal tumor, umbilicus, adjacent peritoneum and omentum, and lateral umbilical ligaments removed en bloc. (fig. 2, *A*).

Grossly, the suprapubic mass was 8 × 6 × 6 cm. and tan yellowish with focal necrosis. Multiple sections were sampled with a section taken for each cm. of greatest dimension of tumor. Microscopically, a pure well differentiated squamous cell carcinoma of the urachus was seen with partial invasion to the muscle layer of the bladder but no involvement of the bladder mucosa and submucosa (fig. 2, *B*). After discharge from the hospital on postoperative day 9 the patient remained symptom-free for 18 months. Followup abdominal CT at 7 months showed no tumor recurrences (fig. 1, *B*).

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DISCUSSION

Located in the space of Retzius the urachus is a vestigial remnant of the allantois that develops into a fibromuscular band, plugged with desquamated epithelial cells by adulthood. Urachal carcinomas are rare malignant epithelial tumors involving the suprapubic and intravesical regions of the bladder dome and anterior wall, are mostly intramural with deep ramifications in the bladder wall and are not secondary.[1] This rare entity accounts for only 0.01% of all malignancies and 0.3% of all bladder tumors.

Most cases described previously were adenocarcinoma. According to the totipotential cell theory of Mostofi et al, the transitional cell urachal lining epithelium can undergo squamous metaplasia, giving rise to squamous cell carcinoma.[1] Begg categorized urachal tumors into 7 classes with intramucosal (class 1), intramural (class 2) and supravesical (class 3) being the most important.[2] On cystoscopy most cases of urachal carcinoma are stage B2 due to the invariable deep muscle involvement, and stages C and D1 represent perivesical fat involvement and local metastasis, respectively.[3]

To our knowledge only 7 cases of squamous cell carcinoma of the urachus have been reported in the literature from 1870 to 1986.[4] Patient age ranged from 27 to 77 years (median 60) and the male-to-female ratio was 6:2. Of the cases juxtavesical location was reported in 5 and intracystic location in 2. Urachal cysts were observed in 6 cases. Ulcers were also detected in 3 cases by cystoscopy. Local growth patterns were divided into intramural type (4 cases) with bladder cancer related symptoms, and supravesical type (3 cases) with hematuria and palpable suprapubic mass. Treatment included excision in 7 cases, and radiation and chemotherapy in 1.

Metastasis was not observed but disease recurred in 2 cases. Urachal tumors are well known for a poor prognosis

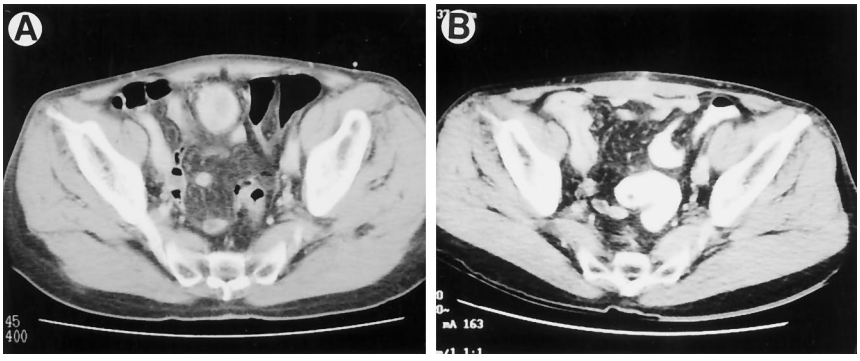

FIG. 1. Abdominal CT. *A*, urachal tumor. *B*, at 7-month followup no tumor recurrence is evident

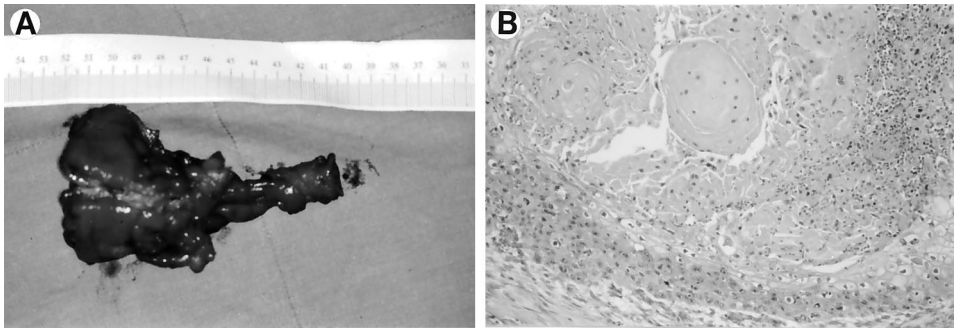

FIG. 2. *A*, urachal tumor and adjacent tissue after resection. *B*, urachal squamous cell carcinoma exhibits keratin pearl formation. H & E, reduced from ×100.

with the tumor-free survival rate uniformly reported to be less than 1 year. Most patients died within 8 months secondary to postoperative complications and disease recurrence.[4] Most cases were incorrectly diagnosed clinically before surgery, resulting in a poor prognosis due to delayed diagnosis and surgical intervention.

CONCLUSIONS

Like most bladder tumors correct diagnosis can be accomplished with a thorough history and physical examination, diagnostic imaging studies, such as CT, magnetic resonance imaging, or excretory urography, urinary cytology and direct cystoscopic visualization with special emphasis on the bladder dome. With early detection and well planned surgical intervention our patient remained complication-free for longer than 18 months.

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