Human Pathology (2015) **46**, 1808–1814

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Human PATHOLOGY

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Original contribution

Urachal carcinoma: a pathologic and clinical study of 46 cases☆,☆☆

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Received 30 March 2015; revised 28 July 2015; accepted 29 July 2015

Keywords:
Urachus;
Urachal carcinoma;
Adenocarcinoma;
Tumor staging system;
Differential diagnosis;
Patient outcome

Summary Urachal carcinoma is a rare tumor that has not been well studied. To determine the pathologic and clinical features of this disease, we retrospectively evaluated 46 cases from our surgical pathology files. The patients included 16 women and 30 men, with a mean age of 53.4 years (range, 28-82 years). Forty patients had undergone cystectomy, and the remaining 6 had undergone transurethral bladder biopsy. Most tumors were located at the dome (n = 44); only 2 were located at both the dome and anterior wall. All tumors consisted of adenocarcinoma, including mucinous (n = 36), enteric (n = 7), not otherwise specified (n = 2), and signet ring cell (n = 1) types. Focal areas of signet ring cell features were present in 23 cases, but urothelial carcinoma in situ was not identified in any cases. The tumors invaded the muscularis propria (n = 8), perivesical adipose tissue (n = 27), and abdominal wall (n = 3). Twenty-five patients had died of cancer at a mean of 32 months (range, 12-74 months), and 21 patients were alive at a mean of 65 months (range, 7-230 months). The median cancer-specific survival time of urachal adenocarcinoma patients was 45 months, which was significantly longer than that of bladder urothelial carcinoma patients with similar-stage disease (P = .047). Patients' cancer-specific survival was associated with tumor stage according to the Sheldon, Mayo, and TNM staging systems. In conclusion, urachal carcinomas are predominantly composed of invasive adenocarcinomas, which commonly demonstrate mucinous features. Most tumors present at advanced stages but are still associated with a better survival rate than bladder urothelial carcinomas.

1. Introduction

The urachus is a vestigial structure of the allantois that generally loses its function after birth [1]. During embryogenesis, the allantois regresses to form the urachus, a tubular structure that connects the urinary bladder and the umbilicus.

☆ Competing interests: The authors have no conflicts of interest to disclose.
☆☆ Funding/Support: MD Anderson Cancer Center is supported in part by Cancer Center Support Grant CA16672 and Bladder SPORE grant P50CA91846 (Project 1 and Core C) to B. Czerniak from the National Institutes of Health.
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http://dx.doi.org/10.1016/j.humpath.2015.07.021
0046-8177/

After the third trimester, the urachus usually involutes as a fibrous cord, which coalesces with the obliterated umbilical arteries and forms the median urachal ligament. However, a urachal remnant, which is characterized by a tubular or cystic muscular structure that is lined by epithelium, can still be found in about a third of adults [2]. Although it is most commonly found at the bladder dome, the urachal remnant can also be found anywhere along the bladder midline, including the posterior and anterior walls [1]. It can be divided into 3 segments: intramucosal, intramuscular, and supravesical [3]. The urachal wall is usually composed of the urothelium, subepithelial connective tissue (or lamina propria), and muscularis propria layers. The urachal urothelial lining frequently undergoes metaplastic changes, mostly glandular metaplasia [4].

Carcinoma can arise from the urachal epithelial lining. Although the urachal remnant is generally lined with urothelium like the bladder, urachal carcinomas demonstrate pathologic and clinical features that are distinct from those of bladder carcinomas [5-12]. Although most bladder carcinomas are urothelial carcinoma, most urachal carcinomas are adenocarcinoma, which typically produces abundant extracellular mucin, and exhibit focal signet ring cell features. Most bladder urothelial carcinomas present as noninvasive papillary urothelial carcinomas and do not invade the muscular wall [13]. In contrast, most urachal adenocarcinomas mainly involve the muscularis propria and perivesical soft tissues, with a sharp demarcation from the bladder surface urothelium [5-12]. In urachal carcinomas, the bladder urothelium is usually intact or ulcerated and lacks papillary urothelial carcinoma or urothelial carcinoma in situ.

Urachal carcinomas are rare, accounting for less than 1% of all bladder cancers [5-7]. Although most urachal carcinomas are adenocarcinoma, urachal adenocarcinoma is far less common than is nonurachal adenocarcinoma in the bladder [8]. A previous study from our institution reported that urachal adenocarcinomas accounted for one-third of all adenocarcinomas of the bladder, but this figure may have been high because of the referral-based nature of our institution [14]. A recent multicenter study reported that urachal adenocarcinomas account for approximately 10% of bladder adenocarcinomas, which may reflect their true incidence in the general population [15]. Because of the rarity of urachal tumors, there have been limited studies of this malignancy reported in the medical literature; most studies have been single-case and small series reports. Here, we determined the pathologic and clinical features of urachal carcinoma in a large series of cases from a single institution.

2. Materials and methods

With the approval of the institutional review board, we retrospectively reviewed our surgical pathology report databases to identify all patients with urachal carcinoma who had been treated at The University of Texas MD Anderson Cancer Center (Houston, TX) from 1990 to 2010. The following diagnostic criteria were used to diagnose primary carcinoma of the urachus: (1) the tumor was located in the dome or midline of the bladder; (2) the tumor predominantly invaded the muscularis propria and perivesical soft tissues with either intact or ulcerated bladder epithelium; (3) there was a sharp demarcation between the tumor and the bladder surface urothelium; (4) there was no extensive cystitis glandularis or cystitis cystica, particularly atypical intestinal metaplasia, in the bladder wall; (5) urothelial carcinoma or urothelial carcinoma in situ was not found in the bladder; and (6) there was no primary adenocarcinoma of another organ.

Forty-six patients with urachal carcinoma were identified. All patients had undergone transurethral biopsy of the bladder, and 40 had undergone partial or radical cystectomy, including partial cystectomy with removal of the urachus and umbilicus (n = 14), partial cystectomy with removal of the urachus (n = 10), partial cystectomy (n = 9), cystoprostatectomy with removal of the urachus and umbilicus (n = 2), cystoprostatectomy (n = 2), cystectomy with removal of the urachus (n = 1), cystectomy with removal of the urachus and umbilicus (n = 1), and total pelvic exenteration with removal of the umbilicus (n = 1). The 6 patients who had not undergone cystectomy presented with metastases. The biopsy and cystectomy specimens from all 46 patients were routinely processed for histologic examination. Usually, more than 40 sections were taken from a cystectomy specimen, including the tumor (1 block per centimeter of the largest tumor dimension); resection margins (ureter, urethra, umbilicus, and soft tissue); and noncancerous bladder, medial ligament, and other associated organs. Three sections were typically taken from the noncancerous areas of the bladder dome and the median umbilical ligament, respectively. Hematoxylin-eosin–stained slides were used for morphologic analysis. The reviewed pathologic parameters included tumor size, location, histologic type, tumor stage, signet ring cell features, lymphovascular invasion, resection margin, urachal remnant, and other urothelial changes.

The following demographic and clinical information was obtained from patients' charts: age; sex; clinical presentation; cystoscopic findings; clinical stage; treatment; and clinical outcome, including metastasis. Clinical stages were evaluated using 3 different staging systems. The Sheldon staging system was defined as follows [5]: stage I, tumors were limited to the urachal mucosa; II, tumors invaded into but not beyond the urachal muscular layer; III, tumors extended to the bladder, abdominal wall, and other adjacent organs; and IV, tumors metastasized to the lymph nodes or other distant organs. The Mayo staging system was defined as follows [9]: stage I, tumors were confined to the urachus or bladder; II, tumors extended beyond the muscular layer of the urachus or bladder; III, tumors metastasized to the regional lymph nodes; and IV, tumors metastasized to nonregional lymph nodes or other distant sites. The TNM clinical staging system