Case Report

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Urachal Adenocarcinoma: A Case Report with 4-Year Follow-up

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Abstract

The urachus is an embryological remnant that lies between the bladder and umbilicus as a fibrous band that attaches the cloaca to the allantois in the intrauterine period. Urachal adenocarcinoma is a rare tumor and more aggressive in behavior than primary bladder tumors. In this case report, we present a 37 years old female patient with urachal adenocarcinoma in whom transurethral resection of the bladder revealed a signet ring adenocarcinoma of the bladder with invasion of the muscularis propria. The patient had left manubrium sterni and bone marrow infiltration. Partial cystectomy and sternal resection were performed, followed by radiotherapy and chemotherapy. There was no recurrence or new metastasis at the 4-year follow-up. In conclusion, a combination of surgery, chemotherapy, and radiotherapy is a suitable approach for the treatment of oligo-metastatic urachal carcinoma.
Keywords: Urachal carcinoma, adenocarcinoma, bladder tumor

Introduction

The urachus is an embryological remnant that lies between the bladder and umbilicus as a fibrous band, which attaches the cloaca to the allantois in the intrauterine period (1). Urachal adenocarcinoma is a rare tumor that accounts for 0.07-0.34% of all bladder tumors and has mucinous, enteric, signet ring cell, mixed, and unclassifiable histopathological types (2). The most common presenting symptom of urachal carcinoma is hematuria, and it rarely presents with irritative voiding symptoms, suprapubic mass, or pain. These tumors are mostly located within the muscular layer of the bladder dome but can lead to ulceration when reaching the mucosal layer and can invade through the Retzius space or anterior abdominal wall. On average, 11-13% of patients are reported as metastatic at disease presentation (3). Urachal adenocarcinomas are more aggressive in behavior compared with primary bladder adenocarcinomas, and the 5-year survival rate is 11-55% (4). Urachal carcinomas are rare but aggressive tumors with an incidence of approximately 1 case per million per year (3). Because of its rarity, treatment modalities and prognosis of urachal neoplasm are not well known, and prospective trials limited to this disease are lacking. Treatment is different for localized or metastatic disease. In this case report, we discuss the diagnosis, treatment, and follow-up of a patient with metastatic urachal adenocarcinoma in light of the current literature.

Case Report

Informed consent was obtained from the patient for the publication of the case presentation. In 2017, a 37-year-old female patient presented with irritative voiding symptoms, hematuria, and white particles in urine. The patient did not have any systemic disorders or a history of smoking or surgery other than cesarean section. Hemogram, routine blood biochemistry, and urinalysis were within normal limits. Ultrasonography revealed a 5 cm solid, vascular mass protruding into the bladder lumen from the bladder dome. Cystoscopy revealed an atypical mass lesion located at the bladder dome, back wall, and the base of the bladder, which had leukoplakia and edematous appearance. Transurethral resection of the bladder was performed concomitantly and macroscopically, and it appeared that the muscular layer was involved. Histopathological examination revealed a signet ring adenocarcinoma of the bladder with the invasion of the muscularis propria. The primary bladder, colon, or urachus origin could not be

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determined. CK20, CK7, CDX2, CD15, and beta-catenin were positive and carcinoembryonic antigen (CEA) was negative in immunohistochemistry. Thoracic computed tomography (CT) was normal. Abdominal CT revealed thickening and irregularity of the bladder fundus where the peritoneum was attached and mild wall thickening and omental contamination on the antrum of the stomach, which was suspicious for a primary stomach tumor. There was a residual mass in the bladder and lytic lesion in the left manubrium sterni on positron emission tomography (PET), but no uptake in the lymph nodes. An exophytic tumoral mass on the anterosuperior portion of the bladder, which was 5 cm in diameter and had a cystic component in its anterior part, was observed by magnetic resonance imaging (MRI) (Figure 1). Thoracic MRI revealed bone marrow infiltration suspicious for metastasis on the left manubrium sterni. Upper gastrointestinal endoscopy and colonoscopy were normal. A biopsy of the sternal lesion showed adenocarcinoma metastasis. A partial cystectomy was performed on July 2017. The mass extending from the urachal region to the bladder dome was resected with a 5 mm safety margin. Frozen biopsies were performed from all suspicious regions. Final pathology revealed signet ring mucinous adenocarcinoma of urachal origin with intact surgical margins (Figure 2). Stereotactic external beam radiotherapy was administered in 2 fractions 10 Gy on August 2017. FOLFOX (folinic acid + 5-fluorouracil + oxaliplatin) chemotherapy was administered for 9 cycles. After the completion of these therapies, sternal resection and thoracic reconstruction were performed. Sternal pathology revealed acellular mucinous islands. The patient was followed up with thoracic and abdominal MRI every 3 months without decreasing frequency. There was no recurrence or new metastasis at the 4-year follow-up (Figure 3).

Discussion

Urachus is an embryological remnant that lies between the fetal urinary bladder and allantois, and urachal tumors mostly spread to the bladder dome, umbilicus, and Retzius space (5). Urachal tumors are rare and mostly seen in the 5th and 6th decades showing male predominance (1). Most urachal tumors are adenocarcinomas. Rarely, sarcomas, squamous cell cancer, and urothelial carcinomas can be observed (6).

The diagnosis of urachal carcinoma is made by cystoscopic observation of the tumoral lesion located in the bladder dome and a resection revealing adenocarcinoma. CT and MRI are important for supporting the diagnosis and local staging of the tumor (1). A solid or cystic mass lesion that shows small calcifications and is located in the bladder midline is pathognomonic for urachal tumors (6). PET-CT can be used to detect distant metastases (7). Most commonly, regional lymph nodes, omentum, liver, lung, and bone metastases are observed (8). In line with the literature, we managed to diagnose the midline solid lesion in the bladder both radiographically and cystoscopically and confirmed the diagnosis by histopathological examination. The distant metastasis evaluation was performed by PET-CT and verified by MRI. Most urachal adenocarcinomas are positive for CDX2, CK20, and to a lesser extent CK7 and beta-catenin (9). CEA, CA-125, and CA-19.9 are used for the diagnosis and follow-up

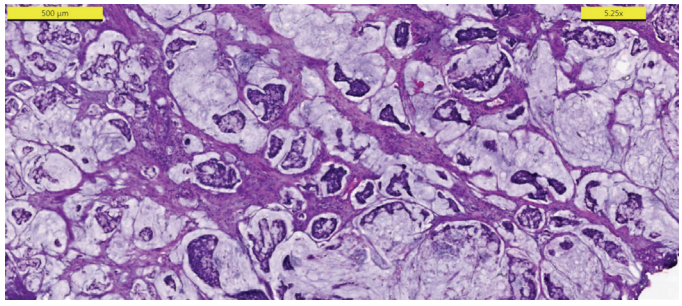

Figure 2. Microscopic appearance of urachal adenocarcinoma from a partial cystectomy specimen

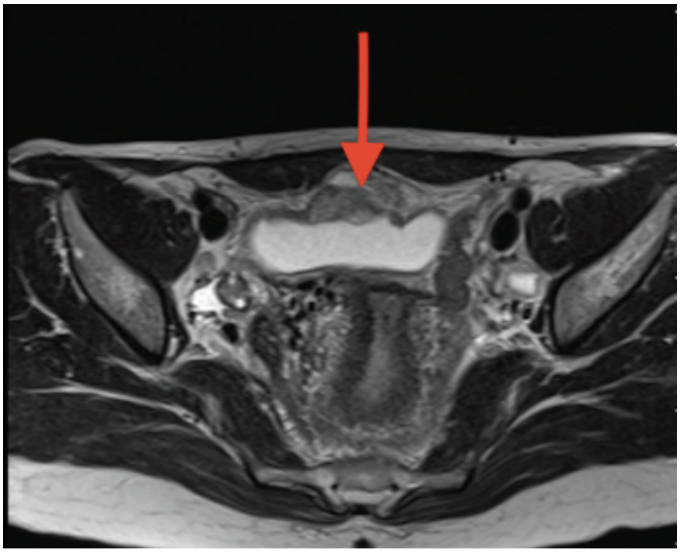

Figure 1. MRI showing a 5x6 cm diameter mass, which originates from the anterosuperior part of the bladder

MRI: Magnetic resonance imaging

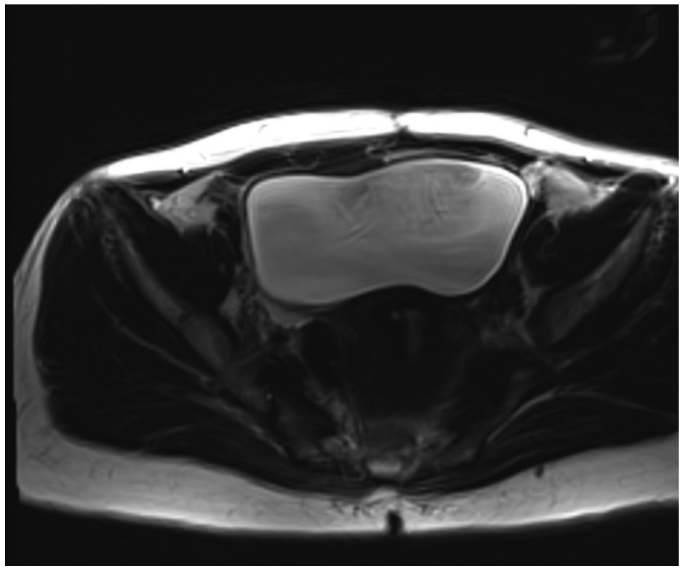

Figure 3. MRI shows that the bladder is intact after 4 year follow-up after combination treatment

MRI: Magnetic resonance imaging

of peritoneal carcinomatosis (10). CK20, CK7, CDX2, CD15, and beta-catenin were positive and CEA was negative in our patient. If necessary, colonoscopy, upper gastrointestinal tract endoscopy, mammography, or coloposcopy are recommended for primary tumor investigation (1,6). In our patient, upper gastrointestinal tract endoscopy and colonoscopy was negative for a tumoral lesion.

Stage is the most important indicator of prognosis in urachal carcinomas (11). Tumor-node-metastasis staging criteria for urothelial carcinoma of the bladder are irrelevant for urachal carcinoma because urachal carcinoma may primarily arise from outside of the bladder, secondarily grow into the bladder, and in some cases invade through the bladder (1). The Sheldon staging system is used for staging urachal carcinoma (Table 1) (6).

A suspicious lesion for metastasis was detected by PET-CT in our patient, and MRI and sternal biopsy verified the metastasis. Because of the presence of distant metastasis, our patient was classified as stage IVB according to the Sheldon staging system.

Treatment of localized urachal adenocarcinoma includes partial or radical cystectomy with urachal and umbilical resection and bilateral pelvic lymphadenectomy (12). In lymph-node-positive patients, lymphadenectomy is recommended after chemotherapy and lymph node regression (1). For stages III and IV, chemotherapy is the standard recommendation; however, for oligo-metastatic disease, local resection and metastasectomy can be performed (10). Treatment outcomes differ between different chemotherapy protocols, and there is no standard chemotherapy protocol for urachal carcinoma (1). It has been shown that the FOLFOX protocol, which shows a partial and complete response in metastatic disease, is suitable for urachal adenocarcinoma (13). Metastasectomy is recommended after chemotherapy with regression of metastatic lesions (1). Literature on the efficiency of radiotherapy in urachal carcinoma is limited, and its effects on the disease are not yet well known (10). Radiotherapy for metastatic lesions may prolong survival (14). In our patient, frozen section examination showed intact surgical margins during partial cystectomy; therefore, umbilicus resection was omitted. After radiotherapy to the sternal metastasis, FOLFOX was administered and metastasectomy was performed.

Urachal carcinoma is a rare and aggressive malignancy, and there is limited evidence for its diagnosis and treatment protocols. Stage and surgical margin status are important for local recurrence and survival after partial cystectomy (14). For metastatic urachan carcinoma, the 5-year survival rate is less than 50% (6). Chemotherapy and surgical interventions treat the disease with variable success rates, and with chemotherapy 1-year-survival is increased statistically significantly (15). After treatment, we followed up our patient with a 3-month interval thoracic and abdominal MRI, and with a 4-year follow-up, there was no local or distant recurrence. In conclusion, a combination of surgery, chemotherapy, and radiotherapy is a suitable approach for the treatment of oligo-metastatic urachal carcinoma.

Table 1. Sheldon staging system for urachal carcinoma

Stage I: Limited to urachal mucosa
Stage II: Limited to urachus
Stage III: Local dissemination
IIIA: To bladder
IIIB: To anterior abdominal wall
IIIC: To peritoneum
IIID: To viscera other than bladder
Stage IV: Metastatic disease
IVA: Regional lymph node
IVB: Distant metastasis

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Ethics

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Authorship Contributions

Surgical and Medical Practices: M.T.E., Concept: O.K., Design: M.T.E., Data Collection or Processing: B.H., Analysis or Interpretation: B.H., Literature Search: M.V.K., Writing: M.V.K.

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