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Case Report

Primary urachal adenocarcinoma: A case report

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Abstract
Primary urachal adenocarcinoma is an aggressive rare cancer that often presents at advanced stages with poor prognosis. We report this case of a 52-year-old patient with a stage-I (Mayo Clinic) primary urachal adenocarcinoma with good outcomes after surgery in a 2-year follow-up period. We analyze epidemiological, clinical and therapeutic features of this disease in the literature review.

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Introduction

Primary urachal adenocarcinoma is a rare and aggressive cancer. It often presents at an advanced stage and has a poor prognosis. We report the case of a 52-year-old patient with a stage I (Mayo Clinic) primary urachal adenocarcinoma with good outcomes after surgery with a follow-up of 2 years. We analyze epidemiological, clinical and therapeutic features of this disease in a literature review.

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Case report

Mr. M.A., a 52-year-old patient, had a history of epileptic disease treated by tegretol. He had a total intermittent hematuria and irritative urinary symptoms for a month. Clinical examination was normal. Ultrasound revealed an echogenic mass localized in the anterior wall of bladder. Hemoglobin was at 11.3 g/dl and renal function was normal. Urine was sterile at the culture.

Uroscan showed a 3 cm dense picture in the dome of the bladder, enhanced after injection of contrast product, which was typical for a urachal tumor (Fig. 1 and Fig. 2).

A rigid cystoscopy was performed under spinal anesthesia. It confirmed the presence of a solid tumor of 3–4 cm developed in the anterior wall of the bladder. Trigon, retrotrigon, lateral walls were normal. Ureteral meatus were free. Then, a transurethral resection was incompletely carried out.

Histology indicated a malignant tumor characterized by a glandular proliferation including well differentiated cells sometimes isolated sometimes grouped in polyadenoid clusters. These were covered by

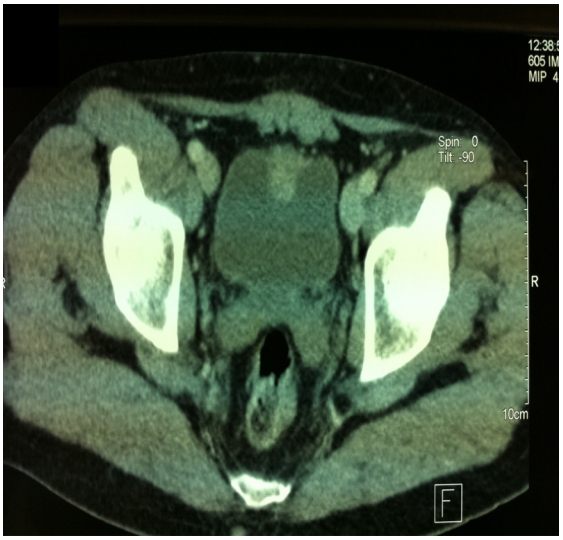

Figure 1 CT scan of the pelvis showing echogenic mass in the bladder dome.

pseudostratified cylindrical epithelium with cytonuclear atypia. The connective stroma was inflammatory. Bladder's muscle was invaded (Figs. 3 and 4).

Immunohistochemistry revealed positive marking for cytokeratin 7 and cytokeratin 20, but negative for β-catenin.

A primary enteroid urachal adenocarcinoma T2 was then concluded.

Prostate specific antigen (PSA) was at 0.66 ng/ml and Carcinoembryonic antigen (CEA) at 2.30 ng/ml (normal value). Colonoscopy has not found any colorectal tumor.

The computed tomography (CT) of chest, abdomen and pelvis showed neither regional nor distant metastasis.

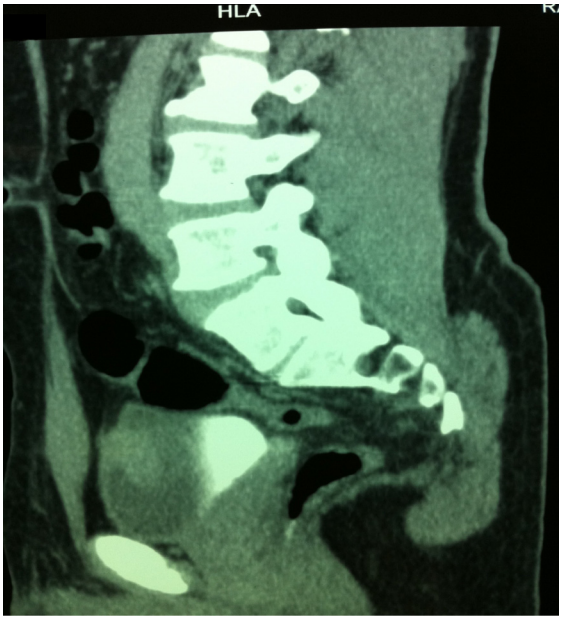

Figure 2 CT scan of the pelvis coronal view showing urachal tumor.

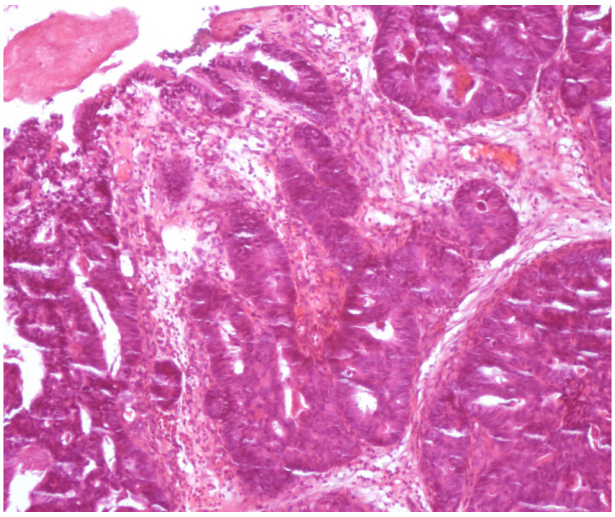

Figure 3 Histogram showing well differentiated adenocarcinoma with glandular proliferation.

A median laparotomy was performed. Perioperatively, a circumscribed and irregular mass of 5 cm × 4 cm originated from the urachus and extended to the dome of the bladder.

Partial cystectomy, with en bloc urachectomy up to the umbilicus, excision of the parietal peritoneum, and bilateral pelvic lymph node dissection, were performed.

Histology confirmed the diagnosis of enteroid adenocarcinoma T2 N0 with negative margins.

No adjuvant treatment was proposed.

The patient is still living free from disease after 2 years, as assessed by cystoscopy and CT of chest, abdomen and pelvis performed every 6 months.

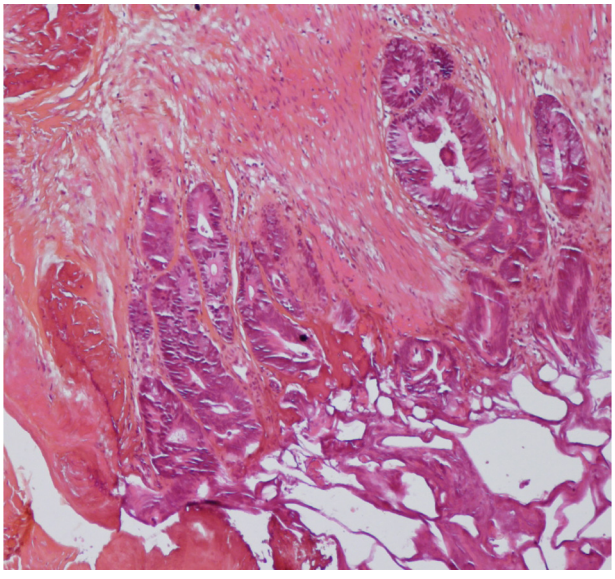

Figure 4 Histogram showing adenocarcinoma with infiltration of detrusor.

Discussion

The urachus is the embryologic remnant of allantois and the adjacent ventral cloaca. It is a tubular structure in which lumen becomes obliterated with the advancing age. But its patency with the urinary bladder may persist in a small proportion of adults [1].

Urachal tumors are rare and devastating cancers of the bladder which were first described by Hue and Jacquin in 1863. They account for only 0.5% of all bladder malignancies, and 20–40% of primary bladder adenocarcinomas [2–4].

Hematuria is the most common presenting symptom in about 90% of patients [5].

The MD Anderson Cancer Center (MDACC) suggested 5 criteria for the diagnosis of urachal cancers. These criteria include a midline location of the tumor; a sharp demarcation between the tumor and normal surface epithelium; an enteric histology; the absence of urothelial dysplasia, cystitis cystica or cystitis glandularis transitioning to the tumor; and the absence of a primary adenocarcinoma of another origin [6,7].

Wheeler and Hill in 1954 proposed 5 criteria: location in the bladder dome or anterior wall; invasion of the bladder wall from outside to inside; absence of cystitis cystica or cystitis glandularis; presence of embryonic remnants; absence of a primary adenocarcinoma of another origin [8]. All of these criteria were present in our reported case.

Immunohistochemistry may help in the distinction between primary and secondary adenocarcinoma.

In primary adenocarcinomas of the bladder, CK7 and CK20 are positive in contrast with colonic adenocarcinomas that express only CK20 [5]. A diffuse nuclear immunoreactivity for β-catenin would militate against the diagnosis of urachal adenocarcinoma [9].

Partial cystectomy with en bloc urachectomy up to the umbilicus is considered the gold standard for the treatment of urachal carcinoma when the disease is surgically resectable. Partial cystectomy is performed to ensure negative margins. En bloc resection of the urachal ligament and umbilicus is recommended because tumors can occur anywhere along the urachus, including at the umbilicus (7%) [4]. If the urachus is transected during surgery, spillage of the tumor containing fluid into the peritoneal cavity can increase the risk of relapse [10,11].

The open surgical approach is favored actually due to the lack of long-term data on either laparoscopic or robotic surgeries [12–14].

In 1984, Sheldon et al. [11] have proposed a system for clinical staging of urachal adenocarcinoma. In this system, early stage urachal cancers are localized to the urachal mucosa, whereas late stage disease involves local structures, like the bladder, abdominal wall or peritoneum, and metastases to regional lymph nodes or distant sites (Table 1). The Mayo clinic has suggested recently a more simplified system (Table 2) [10]. But none of them are validated.

There is currently no standard adjuvant or metastatic chemotherapy protocol for the treatment of urachal adenocarcinoma. The choice of protocols has been based largely on case reports and single institutional experiences. The results of the phase II trial of gemcitabine + cisplatin + 5-FU might further define a treatment standard for this disease [4].

Table 1 clinical staging system by Sheldon et al. [11].

Stage I Urachal cancer confined to urachal mucosa
Stage II Urachal cancer with invasion confined to urachus itself
Stage IIIA Local urachal cancer extension to bladder
Stage IIIB Local urachal cancer extension to abdominal wall
Stage IIIC Local urachal cancer extension to peritoneum
Srage IIID Local urachal cancer extension to viscera other than bladder
Stage IVA Metastatic urachal cancer to lymph nodes
Stage IVB Metastatic urachal cancer to distant sites

Table 2 clinical staging system by Mayo clinic [10].

Stage I Urachal cancer confined to the urachus and/or bladder
Stage II Urachal cancer extending beyond the muscular layer of the urachus and/or bladder
Stage III Urachal cancer infiltrating the regional lymph nodes
Stage IV Urachal cancer infiltrating the non-regional lymph nodes or other distant sites

Recent case reports show the benefit of combined chemotherapy in isolated cases of urachal cancers, most of them adenocarcinomas: the association of 5-FU, cisplatin or oxaliplatin, irinotecan and bevacizumab in different combinations demonstrated usually a partial and limited response [15–18].

Siefker-Radtke et al. [7] have reported a 46-month overall survival from diagnosis of 42 patients (including 7 with metastasis, and 35 with resectable disease). Forty percent of them survive for 5 years. Of the resected cases, 46% remain disease-free with a median follow-up of 31 months. Long-term survival was associated with negative surgical margins ($P = 0.004$) and absence of nodal involvement ($P = 0.01$).

Conflict of interest

There is no conflict of interest.

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