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Case Report

Urachal adenocarcinoma: a rare case report

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ABSTRACT

Urachal carcinoma is a rare and aggressive form of bladder cancer involving the urachus, a fibrous remnant of the allantois that extends from the bladder to the umbilicus. We report this case of a 49-year-old women with primary urachal adenocarcinoma treated with partial cystectomy who relapsed 5 years after surgery with lung metastases. This patient with unremarkable medical history presented with abdominal discomfort and a palpable pelvic mass. Follow-up imaging reveals a large mass on the dome of the bladder extending from the urachus. Subsequent ultrasound-guided biopsy result was suggestive of an urachal mucinous adenocarcinoma. The patient was treated surgically with a partial cystectomy.

Case report

A 49-year-old female patient with unexceptional past medical history presented to her primary care physician with complaints of a 12-month history of abdominal pain and an enlarging mass sensation along her previous cesarean section scar. On physical examination, a large anterior pelvic mass was palpable, firm, and nontender in the midline of suprapubic region slightly to the left. Other than increased frequency and nocturia, she did not complain of urgency, incontinence, pain with voiding, or hematuria.

Imaging of the abdomen was ordered to further assess the mass. Initial ultrasound (US) examination revealed a 15-cm mass localized to the dome of the urinary bladder (Figs. 1A and B). Subsequent magnetic resonance imaging (MRI) scan confirmed a mass measuring 14 × 8.5 × 7.3 cm arising from the left lower rectus abdominis muscle (Figs. 2-4). It extends anteriorly into the subcutaneous tissue and posteriorly imparts significant mass effect on the dome of the bladder. Contrast-enhanced computed tomography (CT) scan of the abdomen also confirms an enhancing mass lesion on the wall of the urinary bladder (Fig. 5). Given the imaging findings, the differential diagnosis at the time included: soft tissue sarcoma, dermatofibrosarcoma protuberans, and desmoid tumor. The patient underwent a cystoscopy, which detected the presence of a submucosal bulge at the urinary bladder dome in the expected area of the residual urachus, consistent with large urachal adenocarcinoma. An US-guided biopsy of the cystic mass showed significant histologic findings indicative of low grade mucinous adenocarcinoma. Colonoscopy at the time did not reveal evidence of primary cancer involving the colon. Also, CT scan of chest, abdomen, and pelvis in addition to positron emission tomography (PET) scan did not reveal regional nor distant metastasis at the time.

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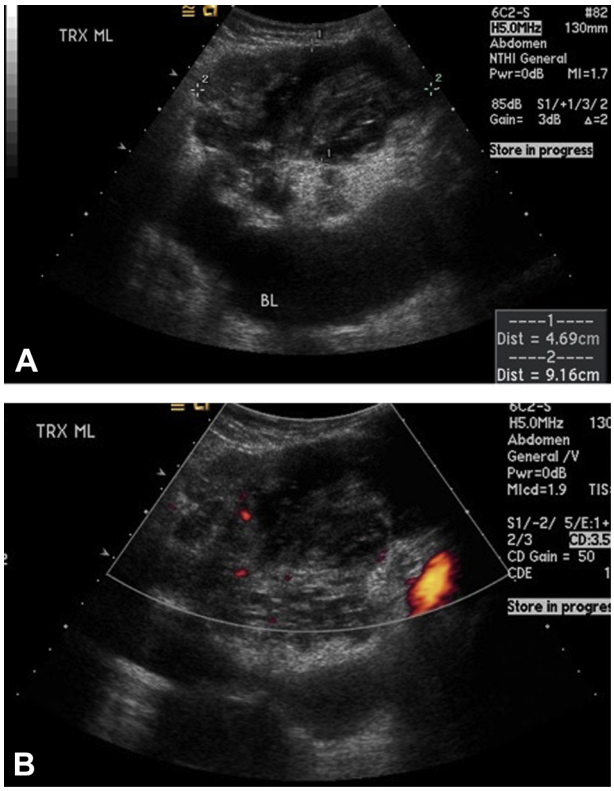

Fig. 1 – (A) Transabdominal ultrasound scan of the pelvis shows heterogeneous mass measuring 4.7 cm by 9 cm inseparable from the urinary bladder. (B) Transabdominal ultrasound scan of the pelvis showing heterogenous mass with internal vascular flow arrows.

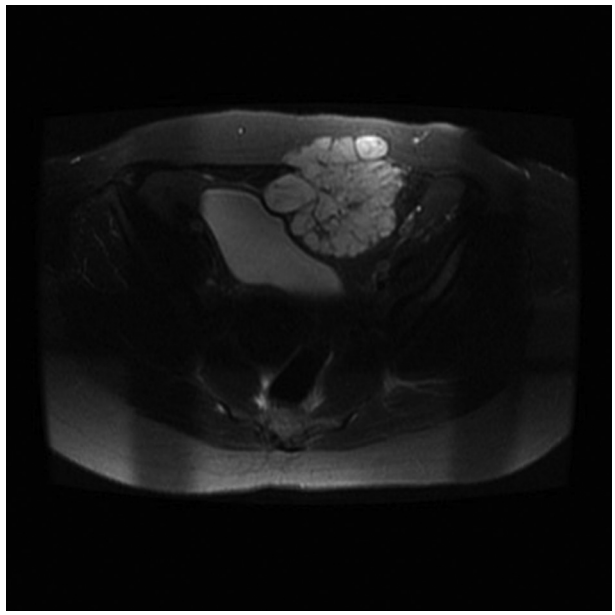

Fig. 2 – T2W axial magnetic resonance (MR) image through the pelvis showing multiseptated hyperintense mass exerting mass effect on the anterior lateral surface of the urinary bladder.

The tumor was removed surgically. The patient underwent a partial cystectomy, hysterectomy, bilateral salpingo-oophorectomy, ureterolysis, and left iliac node biopsy. Pathology confirmed the diagnosis of urachal adenocarcinoma with negative margins. No adjuvant chemotherapy treatment took place. After surgery, the patient is assessed by CT scan of chest, abdomen, and pelvis performed every 6 months (Fig. 6). Five years after her surgery, follow-up chest CT scan revealed multiple pulmonary metastasis.

Discussion

The urachus is a vestigial musculofibrous band of tissue located in the space of Retzius surrounded anteriorly by the transversalis fascia and posteriorly by the peritoneum [1]. During early embryonic development, the urachal canal connects the allantois to the early fetal bladder [2]. Following the descend of the bladder into the pelvis during the 4th month of fetal development, it is stretched until it becomes the median umbilical ligament that joins the umbilicus to the dome of the bladder. Although the tubular structure diminishes with advancing age, it persists in a small proportion of adults [3].

Urachal cancer was originally described by Hue and Jacquin in 1963. As a rare and devastating malignancy of the bladder, it accounts for an estimated 0.01% of all adult cancers, 0.5%-2.0% of all bladder malignancies, and 20%-40% of primary bladder adenocarcinomas [1,4–6]. The mean survival for a locally advanced or metastatic disease is between 12 and 24 months, and the 5-year survival rate is only 43% [7–9]. Late symptom presentation, propensity for early local invasion, and distal metastasis are 3 characteristics of urachal cancer that lead to its poor prognosis [9].

Because early urachal cancer is not accompanied with symptoms, patients often present at the time of diagnosis with higher stage and poor prognosis [7]. Only when invasion of the bladder takes place, patients would present with common symptoms such as irritative voiding, mucous-like discharge, and hematuria [10]. The strongest predictors of urachal malignancy are hematuria and age greater than 55 years [9]. As the predominant presenting symptom, hematuria occurs in 90% of patients and increases the risk of malignancy by 17-fold [7,9,11,12]. Abdominal symptoms such as umbilical pain and discharge have also been reported.

On rare occasions, urachal adenocarcinoma can metastasize to the ovaries. These metastases are similar to primary mucinous ovarian adenocarcinomas both macroscopically and microscopically [13]. Mucin stains are positive in 69% of urachal adenocarcinoma [4]. To differentiate primary ovarian tumors from secondary, immunohistochemistry panel consisting of CK7, CK20, CDX2, MUC2, 34βE12, and β-catenin can be used. Although this panel of biochemical markers can differentiate primary vs secondary ovarian tumors, it can also help in defining the secondary tumor [13].

Diagnosis of urachal cancers has been made easier by the MD Anderson Cancer Center (MDACC) criteria consisting of