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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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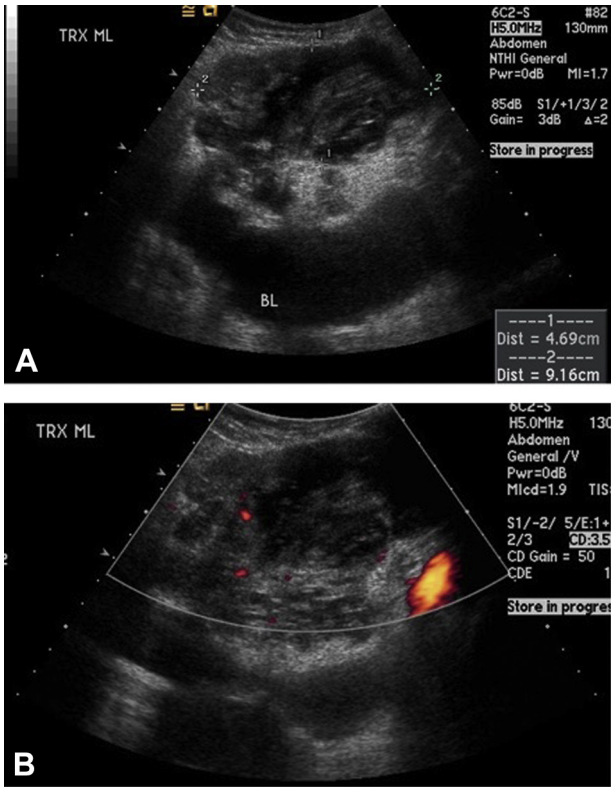

Fig. 1 – (A) Transabdominal ultrasound scan of the pelvis shows heterogeneous mass measuring 4.7 cm by 9 cm inseparable from the urinary bladder. (B) Transabdominal ultrasound scan of the pelvis showing heterogenous mass with internal vascular flow arrows.

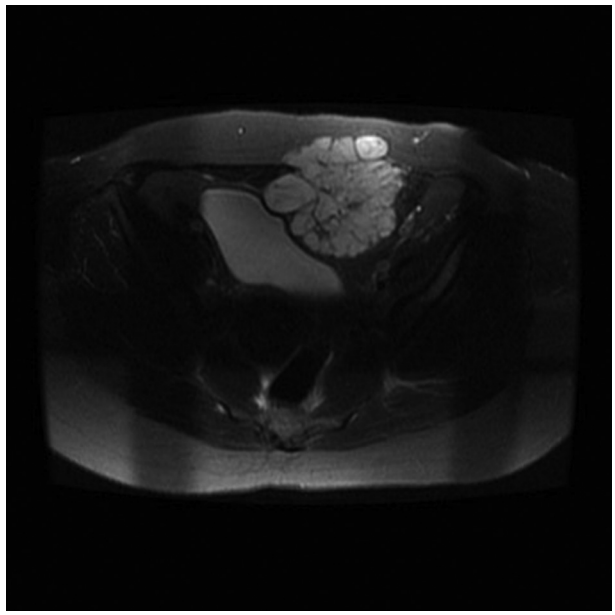

Fig. 2 – T2W axial magnetic resonance (MR) image through the pelvis showing multiseptated hyperintense mass exerting mass effect on the anterior lateral surface of the urinary bladder.

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

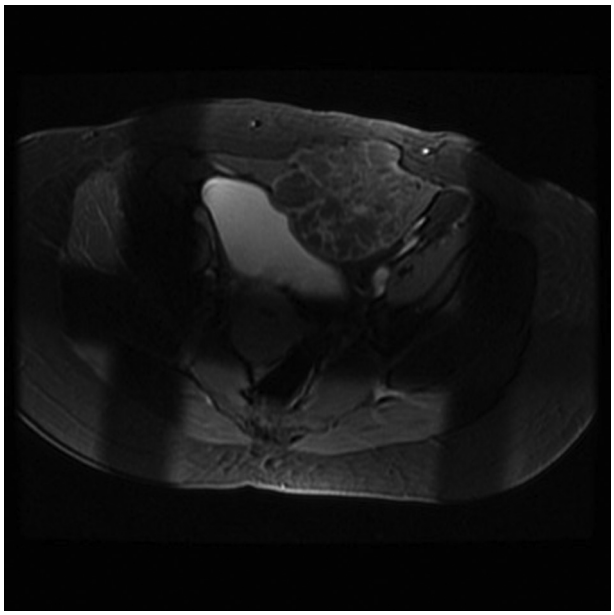

Fig. 3 – T1 FS postcontrast axial MRI image through the pelvis showing enhancing septations within the mass lesion anterior lateral to the urinary bladder.

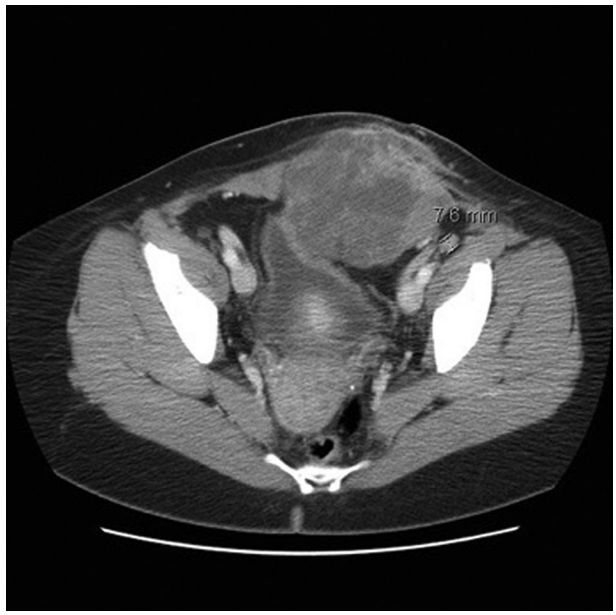

Fig. 5 – Axial image, contrast-enhanced CT scan of the abdomen and/or pelvis in the portal venous phase demonstrates heterogeneously enhancing mass lesion adjacent to the anterior lateral wall of the urinary bladder with associated left external iliac lymph node. There is a loss of fat plane with associated soft tissue stranding, highly suspicious for invasion into anterior abdominal wall.

2 main criteria and 4 supportive criteria [11]. The 2 main criteria consist of: midline location of the tumor and a sharp demarcation between the tumor and normal surface epithelium [10]. Supportive criteria include: an enteric histology; the absence of urothelial dysplasia; the absence of cystitis cystica; and the absence of a primary adenocarcinoma of another origin [8,10]. However, urothelial surface involvement and presence of cystitis cystica are not grounds for excluding urachal carcinomas from differential diagnosis [14].

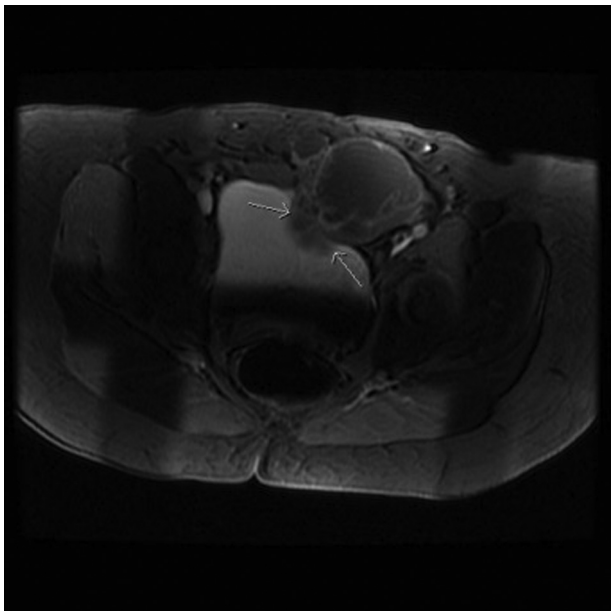

Fig. 4 – T1 FS postcontrast axial MRI image through the pelvis showing loss of tissue plane between the mass and urinary bladder suggesting bladder origin and/or invasion (arrows).

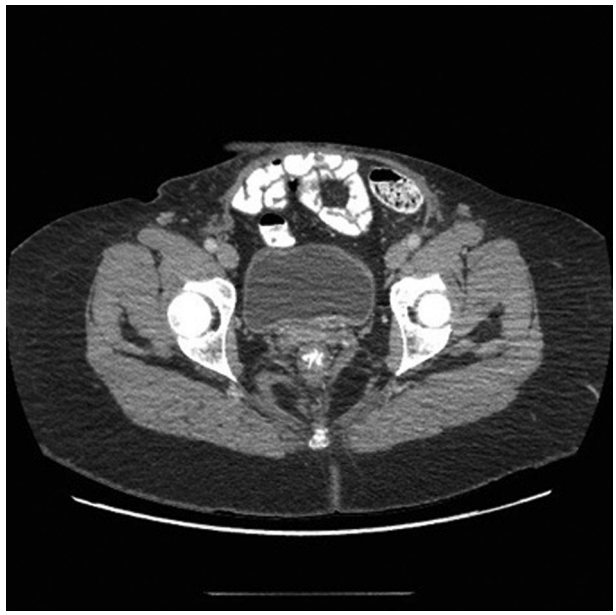

Fig. 6 – Contrast-enhanced CT scan of the abdomen and pelvis in the portal venous phase 4 years after surgical removal of the tumor. No evidence of local recurrence. Incisional hernia.

Table 1 – The urachal cancer staging system as defined by Sheldon et al in 1984.

Stage	Definition
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
Stage 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

Nonetheless, accurate diagnosis of urachal carcinoma is facilitated by a high degree of clinical suspicion and imaging correlation.

Standard imaging workup for urachal cancer includes US, CT scan, and/or MRI evaluation of the abdomen and pelvis. US is often the initial imaging modality. On US, the tumor is observed as a soft tissue mass, which may consist of heterogeneity and calcification. While nonspecific, internal vascularity can sometimes be seen with Doppler imaging. CT scan and MRI, on the other hand, are often used for local staging and evaluation of distant metastasis. On CT scan, in 84% of cases, the tumor is mixed solid and cystic [15], whereas in the reminder of the cases, the tumor appears solid. The cystic component commonly seen in these tumors is mucin. As a sensitive modality for detecting calcifications, peripheral calcification is also commonly seen in the CT scan. Regarding positioning, the bulk of the tumor can be seen outside the lumen of the bladder in 88% of the cases. To distinguish it from urothelial cancer, bladder wall invasion is seen in 92% of adenocarcinomas, and distant metastasis is found in 48% of the cases. On MRI, sagittal images are important to define the location of the tumor. Focal areas of high intensity on T2 sequence are produced by mucinous component, and are highly suggestive of adenocarcinoma. The solid component is isointense to soft tissue on T1 and enhances postcontrast administration. In addition, cystoscopy is also recommended. Diagnosis of urachal carcinoma is usually confirmed by cystoscopy and biopsy [9]. Immunohistochemistry may assist in the distinction between primary and secondary adenocarcinomas. Namely, primary adenocarcinomas of the bladder will be positive for both CK7 and CK20, whereas colonic adenocarcinomas express only CK20 [12].

One of the most significant predictors of urachal cancer prognosis is surgical margin status [16]. Hence, proper surgical intervention has proven critical to the survival of patients [9]. The gold standard surgical approach for the management of localized urachal cancer is an excision of the urachus, umbilicus, and partial cystectomy combined with bilateral pelvic lymphadenectomy [16]. While radical cystectomy has traditionally been done, partial cystectomy with en block urachectomy up to the umbilicus cures 70% of patients. Further resection of the urachal ligament and umbilicus is recommended to ensure negative margins as 7% of urachal cancer can occur at the umbilicus [4,9,17].

Tumor stage at presentation has been important in predicting outcome after surgery [16]. Three different staging systems of urachal cancer have been proposed, although they are yet to be validated: Sheldon, Mayo, and Ontario staging systems. Sheldon et al [4] Proposed a staging system involving localization of the tumor (Table 1). It classifies early stage urachal cancer as localized in the urachal mucosa, whereas late stage cancer involves the extraurachal structures: pT1—no invasion beyond the urachal mucosa; pT2—invasion confined to the urachus; pT3—local extension to the (A) bladder, (B) abdominal wall, and (C) viscera other than the bladder, and pT4—metastasis to (A) regional lymph nodes and (B) distant sites. A more simplified system has been proposed by Ashley et al [9]. The Ontario staging system is yet another simplified classification of urachal tumor involving 4 stages: confined to urachus (T1), confined to bladder (T2), Invading surrounding fat (T3), and extending to the peritoneum (T4) [5].

Currently, there are no standard protocol for the treatment of urachal adenocarcinoma with adjuvant chemotherapy. The role of chemotherapy and radiation therapy and its benefit to patient is yet unclear [7].

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