Locally Invasive Primary Squamous Cell Carcinoma of the Left Ureter in a Patient with a Duplicated Inferior Vena Cava

Shaan A. Setia, Alexander K. Chow, and Christopher L. Coogan

PRESENTATION

A 56-year-old female presented to an outside institution for several days of abnormal uterine bleeding. At that time, she underwent a dilation and curettage and was discharged. Five days postoperatively, she presented to the emergency room with abdominal cramping, vaginal bleeding, hematuria, and left flank pain. Cross-sectional imaging was obtained which revealed left moderate hydronephrosis and a soft tissue midureteral mass (Figs. 1 and 2). The endometrial biopsy performed by gynecology resulted in only endometrial hyperplasia. Urology was later consulted and she underwent a diagnostic left ureteroscopy. A ureteral mass was identified endoscopically at the level of the iliac vessels. There was a small area of blanching necrosis but the mass did not have the classic papillary appearance of urothelial cell carcinoma. A biopsy was obtained and the stent was exchanged. Biopsy results were negative for malignancy.

She later presented to our institution for a second opinion regarding the source of her left renal obstruction. She was scheduled for repeat ureteroscopy and ureteral biopsy. A long (3-4 cm) segment of blanching necrotic obstructing mass was encountered and several biopsies were obtained (Fig. 3). Cytology and biopsy specimens were consistent with squamous cell carcinoma (SCC). The differential at this time included urothelial cell carcinoma with squamous cell differentiation or primary SCC of the ureter. She was then scheduled for an open left nephroureterectomy with a concomitant total abdominal hysterectomy and salpingo-oopherectomy by gynecology for intermittent abnormal uterine bleeding.

Pertinent History

Other pertinent medical history included hypertension, diabetes mellitus type II, and right carotid artery stenosis (on Clopidogrel). She also had a history of several intra-abdominal/pelvic surgeries including tubal ligation, cholecystectomy, and left laparoscopic oophorectomy which was complicated with postoperative hemorrhaging requiring return to the operating room and a prolonged ICU admission. She had no significant familial history of malignancy and she had no smoking history.

PHYSICAL EXAMINATION

She was obese with a body mass index of 32.5. Abdomen was soft, nondistended, and nontender with no palpable abdominal mass. Palpation did not elicit costovertebral angle tenderness. Lymphadenopathy was absent throughout.

LABORATORY

She had a creatinine of 0.96 and an estimated glomerular filtration rate of > 60. Remainder of laboratories was unremarkable.

Radiological Finding

See Figure 1 and 2. Chest imaging revealed no distant metastasis.

Intraoperative Findings

A midline incision was carried out from the xiphoid down to symphysis pubis. The retroperitoneum was exposed with medialization of the left colon. Gerota's fascia was opened between the kidney and the adrenal gland and the superior pole of the kidney was dissected free. Dissection was continued laterally and inferiorly until the ureter was identified. This was traced down to the level of her large left gonadal vein. Attention was turned to the renal hilum and the solitary renal artery and vein was identified and was taken en bloc with a Gastrointestinal Anastamosis (GIA) vascular stapler.

The proximal ureter was then mobilized cephalad and was found to be densely adherent to the left iliac artery and vein. At this time, the gynecology performed a total abdominal hysterectomy and salpingo-oopherectomy.

From the Rush University Medical Center, Division of Urology

Address correspondence to: Alexander Chow M.D., Department of Urology, Rush University Medical Center, Rush Professional Office Building, Suite 348, 1725 W Harrison St, Chicago, IL. E-mails: Alexander_k_chow@rush.edu; akch.umich@gmail.com

Submitted: April 29, 2019, accepted (with revisions): May 21, 2019

https://doi.org/10.1016/j.urology.2019.05.024
0090-4295

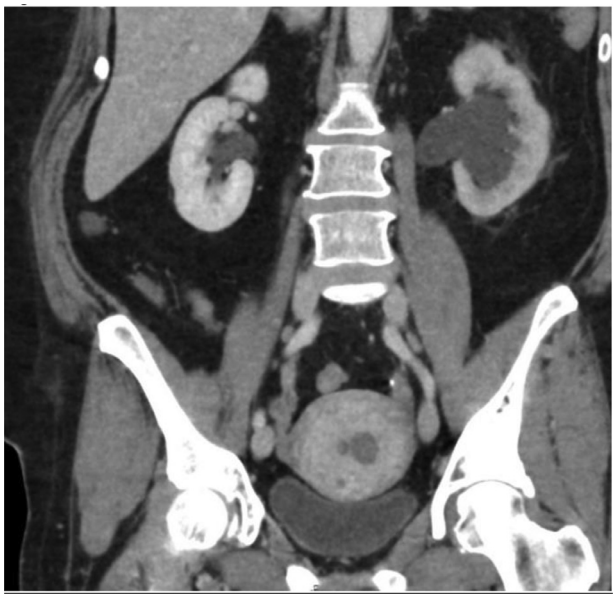

Figure 1. Shows a coronal cut from her original CT scan obtained 5 days after D&C. This reveals fluid within the uterus as expected and moderate hydronephrosis with mild perinephric stranding.

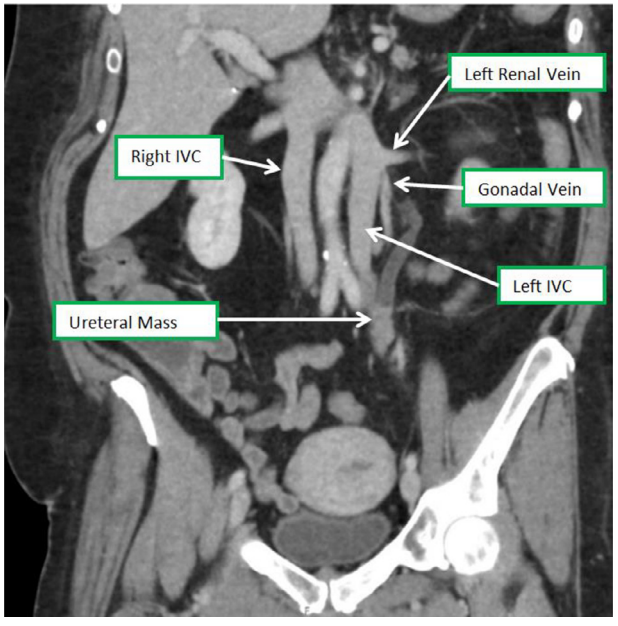

Figure 2. A duplicated inferior vena cava (IVC) is seen with a prominent left gonadal vein inserting at the confluence of the left IVC and left renal vein. A 3-4 cm solid midureteral mass is seen with locally invading the left gonadal artery and vein. (Color version available online.)

After completion of the total abdominal hysterectomy and salpingo-oopherectomy, the ureteral mass was carefully dissected off the iliac vessels which then released the kidney and ureter en bloc. The ureter distal to the iliac vessels was mobilized and the ureterectomy was completed with a transvesical excision of a bladder cuff. Both the anterior cystotomy and the bladder cuff defect were closed

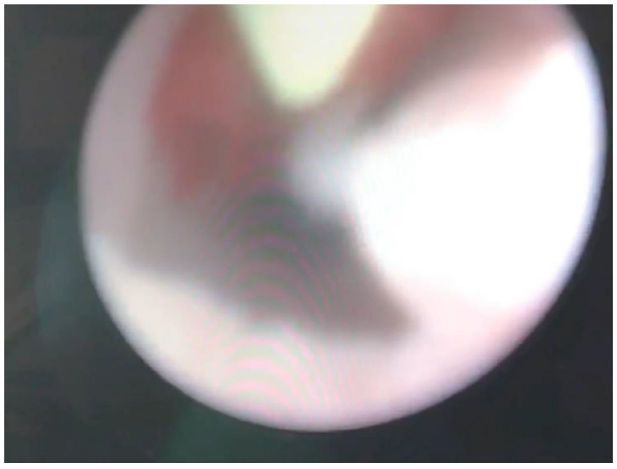

Figure 3. Ureteroscopy revealing the blanched necrotic mid ureteral mass. (Color version available online.)

in 2 water tight layers. Residual soft tissue on the left iliac artery and vein appeared concerning for malignancy and a frozen section was sent off from both vessels which returned as SCC (Fig. 4).

The iliac vein and iliac artery were skeletonized and proximal and distal vascular control was obtained. A wide excision of the left common iliac artery was performed and repaired with an interposed synthetic graft. A wide elliptical excision of the left common Inferior Vena Cava (IVC) was also performed. The left gonadal vein was conveniently dilated and was harvested and utilized as an onlay patch over the venous defect (Fig. 4).

A Jackson-Pratt drain was placed and the abdomen was closed. She was monitored in the ICU postoperatively and transitioned to the floor. She was eventually discharged in stable condition on post-op day 9.

PATHOLOGY

Pathology revealed moderately differentiated SCC with local periureteral soft tissue invasion. Vascular specimens revealed SCC invasion into the soft tissue and vessel muscle wall of the left iliac vein but not into the common iliac artery. All surgical pathology had negative margins. Final pathology: pT3Nx.

Hysterectomy specimen revealed benign inactive endometrium with no evidence of hyperplasia or malignancy.

CURRENT MANAGEMENT

The patient followed up with Medical Oncology 1 month after surgery and received a PET scan and MRI of the brain which revealed no evidence of distant metastasis. She was presented at our multidisciplinary tumor board and a recommendation was made for adjuvant gemcitabine and carboplatin. Chemotherapy was initiated 45 days postoperatively. She completed 2 cycles but unfortunately was unable to tolerate a third cycle due to carboplatin induced nephrotoxicity. After cessation of chemotherapy, her creatinine improved from 6.8 to 1.0.

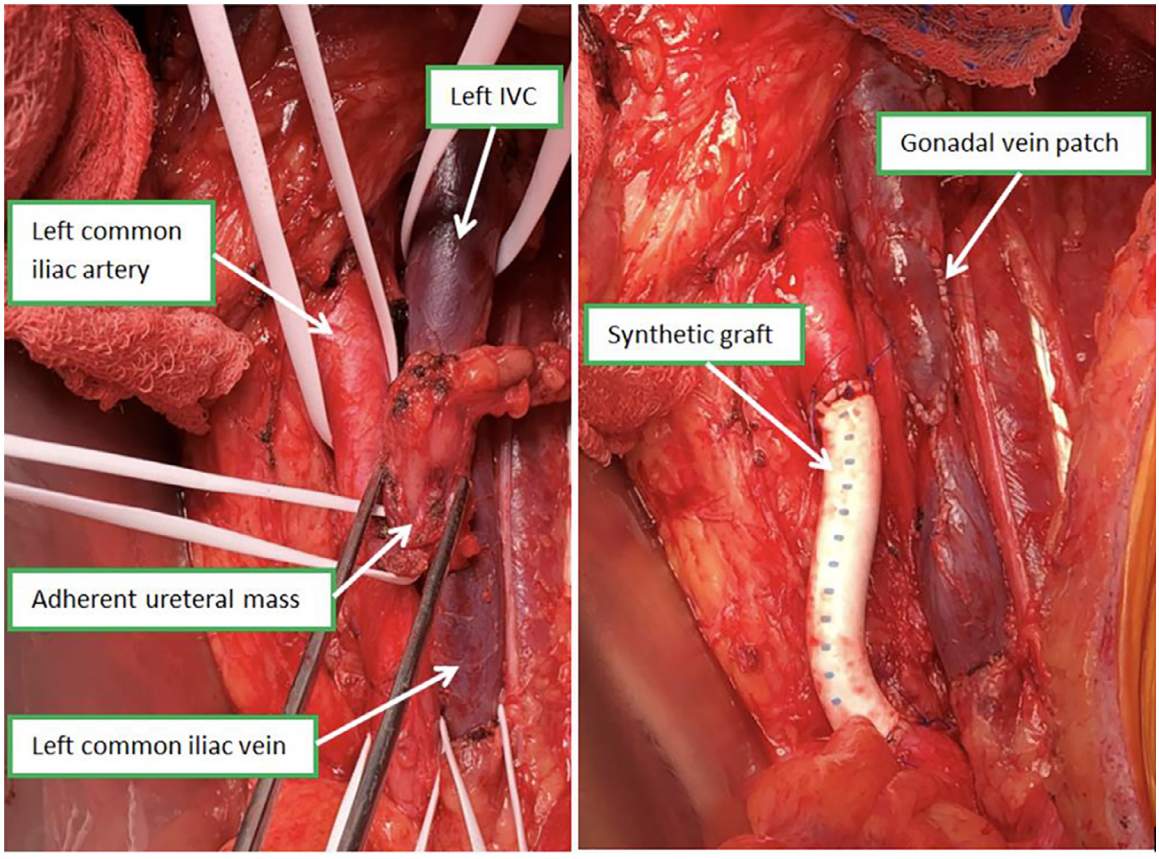

Figure 4. Surgical field before and after excision and grafting. (Color version available online.)

DISCUSSION BY CHRISTOPHER L COOGAN M.D.

Primary SCC of the upper urinary tract is a rare diagnosis with overall poor prognosis. It has an estimated incidence of anywhere between 1% and 8% of upper tract tumors with presentation often at later stages compared to UCC.[1] This patient had an abnormal presentation with abnormal uterine bleeding with abdominal pain, hematuria, and flank pain closely following a D&C.

Additionally, she had none of the commonly known risk factors of upper tract SCC such as history of recurrent nephrolithiasis, phenacetin abuse, or pelvic radiation. Most of the literature on these risk factors is based on relatively few case reports.[2-5] Additionally, these risk factors are all associated with chronic inflammation which may predispose to tumorigenesis.[6] This patient was also a nonsmoker and had no significant familial malignancy. Her surgical history was significant for a complicated left laparoscopic converted to open oophorectomy which may have induced a chronic retroperitoneal inflammatory reaction.

She was also noted to have a rare congenital anomaly of a duplicated inferior vena cava. The reported incidence of IVC duplication ranges between 0.4% and 4%.[7] The left-sided IVC continued as the left common iliac vein which had direct extension of the ureteral tumor through the vessel muscle wall.

There is relatively little literature on treatment of upper tract SCC due to its rarity. The largest published study by Holmang et al describes that most patients receive nephrectomy at presentation due to suspicion for renal cell carcinoma with relatively few undergoing completion ureterectomy at a later date.[1]

The decision to perform open nephroureterectomy was made due to the initial concern for urothelial cell carcinoma with squamous differentiation and the complexity of local vascular invasion. A robotic approach would prove to be difficult for oncological control and safe dissection for multiple reasons including the desmoplastic response of adjacent tissue, her vascular anomaly, and the need for vascular excision and repair.

Despite complete surgical resection, most patients have a poor prognosis with median survival of only 7 months after surgery.[1] Benefit of adjuvant chemotherapy or radiation therapy is thought to be limited with most studies showing poor outcomes despite aggressive management.[8,9]

SUMMARY AND CONCLUSION

This is a rare case of SCC of the upper urinary tract with locally advanced disease into anomalous vascular anatomy in a patient without obvious risk factors for this disease process. Presentation of upper tract SCC is often late and is associated with poor prognosis. Further investigation

into the management of this aggressive disease is needed to improve overall survival.

References

1. Holmäng S, Lele S, Johansson S. Squamous cell carcinoma of the renal pelvis and ureter: incidence, symptoms, treatment and outcome. *J Urol*. 2007;178:51–56.
2. Morita T, Izumi T, Shinohara N, et al. Squamous cell carcinoma of the ureter with marked leukocytosis producing granulocyte colony-stimulating factor. *Urologia Internationalis*. 1995;55:32–33.
3. Palmer C, Atty C, Sekosan, et al., et al. Squamous cell carcinoma of the renal pelvis. *Urology*. 2014;84:8–11.
4. Wagle D, Moore R, Murphy G. Squamous cell carcinoma of the renal pelvis. *J Urol*. 1974;111:453–455.
5. Li M, Cheung W. Squamous cell carcinoma of the renal pelvis. *J Urol*. 1987;138:269–271.
6. Multhoff G, Molls M, Radons J. Chronic inflammation in cancer development. *Front Immunol*. 2012;2:98.
7. Mayo J, Gray R, Louis E, et al. Anomalies of inferior vena cava. *Am J Roentgenol*. 1983;140:339–345.
8. Busby J, Brown G, Tamboli P, et al. Upper urinary tract tumors with nontransitional histology: a single-center experience. *Urology*. 2006;67:518–523.
9. Blacher E, Johnson D, Abdul-Karim F, et al. Squamous cell carcinoma of renal pelvis. *Urology*. 1985;25:124–126.