



Endourology

Spontaneous Ureteral Urine Extravasation From Invasion of a High-grade Angiosarcoma

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ABSTRACT

A 69 year-old male with a past medical history of hypertension, diabetes, and atrial fibrillation presented to the Urology clinic with asymptomatic microscopic hematuria. His work up for hematuria included a negative cystoscopy and a computed tomography (CT) scan, which revealed what appeared to be a fluid collection around the left kidney with a perinephric infiltrative mass and two para-aortic enlarged lymph nodes.

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Introduction

Spontaneous ureteral urine extravasation is defined as perforation not induced by iatrogenic manipulation, external trauma, degenerative kidney disease, urography with external compression, or previous surgery. Fewer than 100 cases of spontaneous ureteral urine extravasation have been reported in the literature and are usually caused by stone disease. In this case report, we describe an unusual case of extrinsic invasion of a tumor directly into the ureter causing spontaneous ureteral urine extravasation.

Case presentation

A 69 year-old male with a past medical history of hypertension, diabetes, and atrial fibrillation presented to the Urology clinic with asymptomatic microscopic hematuria. His work up for hematuria included a negative cystoscopy and a computed tomography (CT) scan, which revealed what appeared to be a fluid collection around the left kidney with a perinephric infiltrative mass and two para-aortic enlarged lymph nodes (Fig. 1, left). A CT-guided percutaneous biopsy was obtained of the most posterior mass, which revealed atypical cells mixed with inflammatory cells, but no

malignancy. The patient elected to continue surveillance rather than surgical exploration.

Two weeks later, the patient presented to the hospital with increased abdominal distension with a physical examination consistent with ascites. He was noted to have mild tachycardia, tachypnea, and a white blood cell count of $16.8 \times 10^3/\text{mL}$. His physical examination was remarkable only for abdominal distension and diffuse mild tenderness. CT-guided paracentesis was performed with drainage of 14 L of fluid. Peritoneal fluid analysis revealed serum-ascites albumin gradient (SAAG) < 1.1 g/dL, 2803 nucleated cells and 149×10^3 red blood cells with 85% lymphocytes, protein 3.1 g/dL, fluid creatinine was 1.64 mg/dL, and no growth on aerobic culture. The liver appeared homogenous without evidence of cirrhosis and his liver function tests were normal. The differential diagnosis for ascites fluid includes spontaneous bacterial peritonitis, malignant ascites, and liver disease. Although the patient presented with signs of sepsis, there was no evidence of infection of the ascites fluid. Liver disease was unlikely given laboratory data and imaging. An SAAG of < 1.1 g/dL is more consistently seen with carcinomatosis, tuberculosis, pancreatitis, serositis, peritonitis, or nephrotic syndrome. Cytology from multiple repeated paracenteses never revealed malignant cells.

A CT scan of the abdomen and pelvis was repeated and was unchanged compared to prior imaging except for interval development of an irregular high density area around the left proximal ureter concerning for extravasated contrast-enhanced urine seen on delayed images (Fig. 1, right). Urology was consulted and the decision was made to proceed with a retrograde pyelogram to

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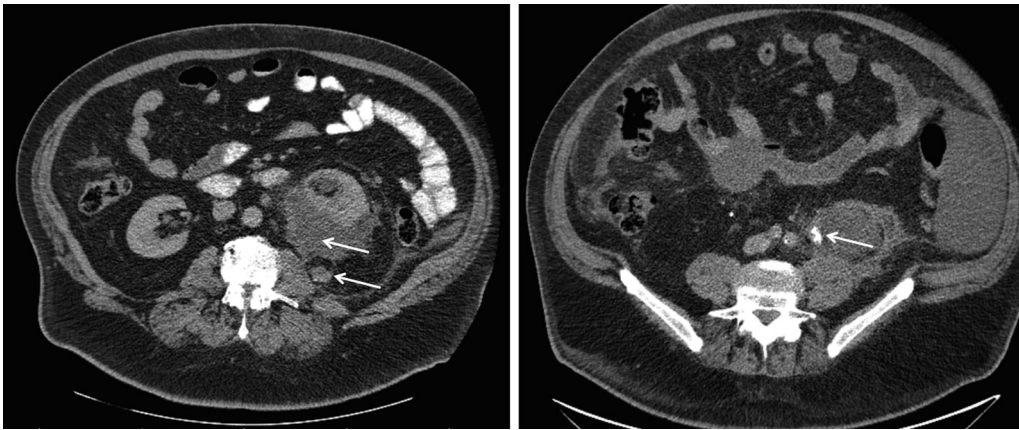


Figure 1. CT scan (left) demonstrates a crescentic fluid collection posteriorly and inferior to the left kidney (top arrow) measuring approximately 7.6×7.4 cm with extensive infiltrative changes extending inferiorly into the pelvis. There is a 2 cm nodule posterior to the kidney, which was biopsied (bottom arrow), and two enlarged para-aortic nodes (not seen on this image). CT scan with delayed images (right) demonstrated irregular high density noted around the proximal ureter, which may represent urine extravasation. The arrow indicates contrast-enhanced urine at the level of the left proximal ureter.

verify the presence of extravasation and placement of a ureteral stent if indicated. Retrograde pyelogram indeed revealed extravasation of contrast at the proximal ureter. More proximally, there was a ureteric irregularity and tortuosity of the upper ureter, and blunting of the calices (Fig. 2, left). Ureteroscopy was deferred due to the presence of extravasation and a left-sided double J ureteral

stent was placed. The patient ultimately refused a urethral foley catheter, and since bladder urine can reflux with a stent, a left nephrostomy tube was later placed for better urinary diversion. An anterograde nephrostogram at the time of nephrostomy tube placement redemonstrated proximal ureteral extravasation (Fig. 2, right).



Figure 2. Retrograde pyelogram (left) demonstrated moderate hydronephrosis of the left kidney (upper pole greater than the lower pole). There is a focal stricture identified within the proximal ureter. Additionally, there is subsequent contrast extravasation at the level of the proximal ureter. Anterograde nephrostogram (right) after ureteral stent and nephrostomy tube placement also demonstrates contrast extravasation at the left proximal ureter.

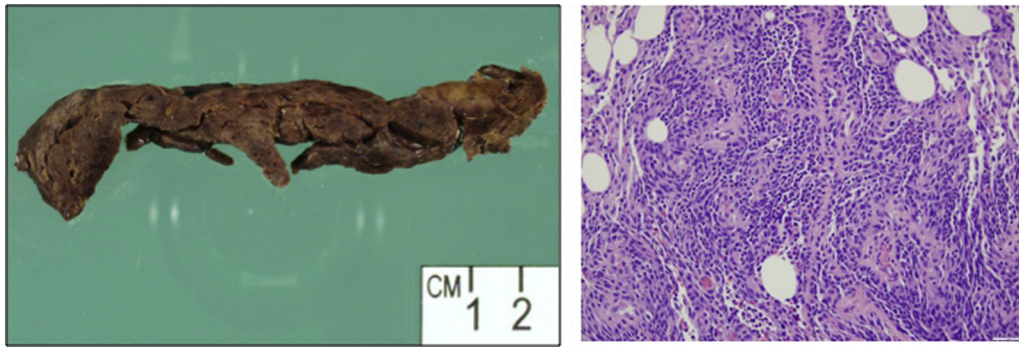


Figure 3. Gross photograph of a section of omentum showing diffuse hemorrhage and necrosis with scant normal adipose tissue present (left). Angiosarcoma involving the omentum (20 \times) (right). Sections of omentum showed diffuse involvement by a spindle cell neoplasm demonstrating both formation of vascular channels and solid areas of growth. The cells were positive for the vascular markers CD31 and CD34, consistent with angiosarcoma.

The patient continued to develop painful abdominal distension from ascites requiring repeated paracenteses with drainage of high volumes of bloody ascites (as much as 14 L at a time) and frequent blood transfusions. Surgical Oncology was consulted for malignancy work up. Tumor markers including AFP, CEA, and CA 19.9 were negative. Several days later the patient developed acute respiratory failure requiring intubation with increasing vasopressor requirements. An emergency laparotomy was performed revealing 9 L of bloody ascites fluid in the abdomen. The greater omentum showed gross evidence of necrosis with widespread areas of purplish discoloration and was thus resected. The left retroperitoneum was entered revealing a large thickened mass involving Gerota's fascia, extending along the lower pole of the kidney and upper ureter. The mass appeared necrotic and was biopsied.

On gross examination, the omentum appeared diffusely hemorrhagic and necrotic. The resected portion of the perinephric lesion consisted of largely necrotic-appearing tissue (Fig. 3, left). On microscopic examination, the omentum was diffusely involved by a spindle cell proliferation with prominent formation of vascular channels as well as solid areas of growth in a background of necrosis and tissue hemorrhage (Fig. 3, right). Mitoses were frequent. Immunohistochemical stains demonstrated that the tumor cells were diffusely strongly positive for CD31 and focally positive for CD34. They were negative for CKit, DOG1 and keratins AE1/3 and cam5.2. These findings are most consistent with an angiosarcoma. Only a small amount of biopsy material was available to evaluate the perirenal mass, but demonstrated totally necrotic tissue.

After return from the operating room, the patient continued to have poor hemodynamics and increased vasopressor requirements. The following day, the patient expired, 3 months after his initial presentation of microscopic hematuria.

Discussion

Here we describe an unusual case of spontaneous ureteral urine extravasation caused by an invasive angiosarcoma in a patient

initially presenting with microscopic hematuria. Spontaneous ureteral urine extravasation is a rare occurrence and is defined as perforation not induced by iatrogenic manipulation, external trauma, degenerative kidney disease, urography with external compression, or previous surgery.¹ The most common causes of spontaneous ureteral urine extravasation are stone disease (56%) and stricture (16.7%), and tumor (5.6%).² Cases of spontaneous ureteral extravasation involving tumors are almost always involving tumors of urothelial origin with intrinsic tissue disruption or obstruction.^{3–5}

Conclusion

To our knowledge, this is the first reported case of ureteral urine extravasation caused by extrinsic tumor invasion. The primary source of the angiosarcoma is unclear. We conclude that when spontaneous ureteral urine extravasation is found in the absence of urolithiasis or distal obstruction, malignancy should be considered in the differential diagnosis, particularly in the setting of suspicious radiographic and clinical findings.

Conflict of interest

None.

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