An uncommon cause of gross hematuria: two cases of ureteroarterial fistula

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Ureteroarterial fistula (UAF) is a rare but potentially fatal cause of hematuria seen in patients with prolonged ureteral stenting in the setting of surgery or abdominal radiation. It presents as episodic, transfusion-dependent hematuria with formation of clots. There is no current consensus for the optimal way to diagnose or treat UAF. We report two cases of UAF that required repeated studies and provocative testing to confirm the diagnosis. Both were successfully managed by endovascular stenting. Clinicians must recognize the diagnostic difficulties involved and maintain a high index of suspicion for UAF in stented patients with intermittent, severe hematuria.

Key Words: hematuria, fistula, ureter, endovascular, stent

Introduction

Ureteroarterial fistula (UAF) is a rare cause of life-threatening hematuria that has increased in incidence since 1990.[1,2,7,8] Fistulae most commonly occur between the ureter and the common iliac or external iliac arteries leading to dramatic episodes of rapid blood loss requiring transfusion. Risk factors for the development of a UAF include a history of radical pelvic surgery or pelvic radiation and the presence of a chronic indwelling ureteral stent.[2,8] Because UAFs bleed intermittently, they can pose a significant diagnostic challenge. Initial studies, including retrograde pyelogram or angiography are frequently negative. Clinicians must maintain a high index of suspicion when risk factors are present to avoid significant delay in diagnosis and treatment. We present two cases recently treated for UAF at our institution, and review the existing literature.

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Case 1

CC is a 35-year-old female with history of mucinous cystadenocarcinoma of the ovary treated by pelvic exenteration and creation of an ileal conduit in 2010. Her course was complicated by bowel ischemia and bilateral ureteroenteric anastomotic strictures. For more than 2 years, she was managed with an indwelling 10-12Fr right ureteral stent and a left nephrostomy tube. Both were exchanged regularly in the interventional radiology suite. On presentation for routine tube exchanges, left antegrade pyelogram demonstrated a narrow opening of the distal left ureter and antegrade placement of left ureteral stent was attempted. During the procedure, a ureteral perforation with extravasation of contrast was noted. Ultimately, a wire was negotiated into the ileal conduit to permit balloon dilation of the strictured anastomosis and placement of an 8.5Fr ureteral stent. The patient was discharged to home

with bilateral ureteral stents and a left nephrostomy tube draining clear yellow urine.

She presented to an outside emergency department 2 days later with right flank pain and passage of a large volume of frank blood and clots from her ileal conduit. Hemoglobin was 8.6 gm/dL, down from 11.6 gm/dL 2 days earlier and creatinine was elevated to 2.1 mg/dL from a baseline of 1.4 mg/dL. She was transferred for IV fluid resuscitation and transfused with two units of packed red blood cells. Computed tomography of the abdomen and pelvis revealed high attenuation material consistent with blood filling the conduit and bilateral upper tracts and moderate right hydronephrosis. There was no evidence of a hematoma or fluid collection in the pelvis or retroperitoneum. On transfer, she was draining clear pink urine from her nephrostomy and conduit with no evidence of clots. She stabilized and urine continued to clear over the next 3 days. However, late on hospital day 4, she again passed a large volume of frank blood and clots from her stoma. Injury to the conduit was suspected and aortography with selective catheterization of the superior mesenteric artery was performed on hospital day 5. This demonstrated normal pelvic vasculature including jejunal vessels supplying the conduit with no evidence of pseudoaneurysm, fistula or active bleeding. Of note, the patient was again draining clear pink urine at the time of the study. The urine remained clear for 24 hours before she suffered another episode of frank bleeding. UAF remained high on the differential despite the prior negative study, and repeat angiography with removal of the indwelling stents was planned despite worsening renal insufficiency, likely exacerbated by IV contrast. She was prehydrated with 200 cc/h intravenous fluids and treated with acetylcysteine in an attempt to limit further renal deterioration.

At the time of the repeat study, the urine was again clear and pink. The initial aortogram showed no evidence of active bleeding. Wires were placed in the stents and the stents were removed. Angiography still demonstrated no evidence of active bleeding. Five French ureteral catheters were then introduced over the wires. Bilateral ureterograms demonstrated multiple filling defects consistent with clot. The ureters and conduit were irrigated with normal saline resulting in removal of a large amount of clot and immediate onset of brisk arterial bleeding. Contrast in the conduit revealed pulsatile swirling near the left ureter consistent with active bleeding. Immediate repeat angiography of the right common iliac artery finally demonstrated a right external iliac to left ureteral fistula, Figure 1. A 7 mm x 5 cm Viabahn vascular stent was advanced into the right external iliac artery and deployed to exclude the fistula. Post-stenting angiography confirmed absence of contrast extravasation, Figure 2. The remainder of her hospital course was uneventful. She was discharged home on hospital day 10 with clear yellow urine and was doing well with return to baseline renal function at last contact 2 months after discharge.

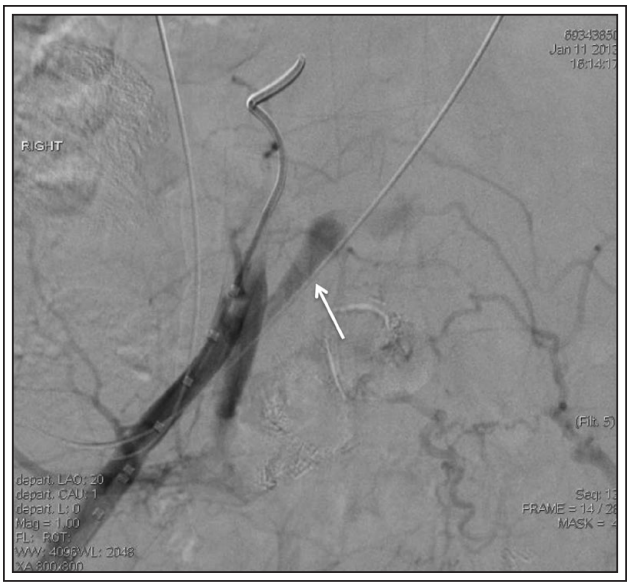

Figure 1. Right iliac arteriogram shows contrast extravasation from the right external iliac artery into left ureter (arrow). Endovascular stent is being advanced into the iliac artery.

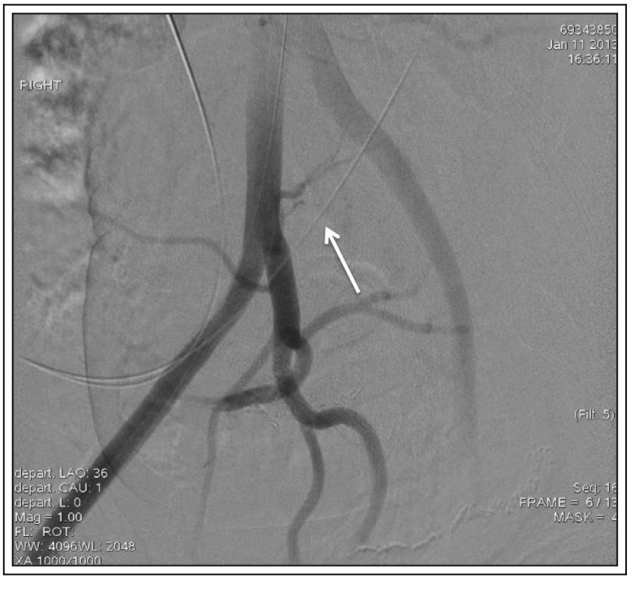

Figure 2. Following stent deployment, fistula has been excluded and the left ureter no longer fills with contrast (arrow).

Case 2

AC is a 36-year-old female with a history of cervical cancer treated with brachytherapy and external beam radiation to the pelvis. Her course was complicated by radiation colitis and retroperitoneal fibrosis causing bilateral ureteral obstruction necessitating chronic indwelling stents. Three months following placement of bilateral metallic double J stents, she presented complaining of recurrent urinary tract infections and several weeks of worsening hematuria with clots. Hemoglobin on admission was 8.1 gm/dL, four grams lower than 1 month earlier.

She was taken to the operating room for diagnostic cystoscopy and ureteroscopy. A large amount of clot was evacuated from the bladder. The posterior bladder was noted to be erythematous and friable and there was blood tinged efflux from the right ureteral orifice around the metallic stent. The stent was removed and right flexible ureteroscopy revealed clot filling the pelvis. After clearing the clot, inspection revealed slight ulceration at the ureteropelvic junction consistent with metallic stent irritation, but no abnormality or active bleeding was encountered in the kidney or ureter. Ureteroscopy was then performed on the left side, which also revealed significant clot burden but no ureteral or caliceal abnormalities. Bilateral non-metallic double-J stents were placed at the conclusion of the procedure.

On postoperative day 1, the patient complained of acute right foot pain and weakness. She was found to have only a palpable right femoral pulse, with absent distal pulses. Vascular surgery took her emergently to the operating room for thrombectomy of her right external iliac and popliteal arteries. Intraoperative angiogram revealed a 50% narrowing of her external iliac artery consistent with radiation arteritis. Postoperatively, she was maintained on heparin.

Over the course of the next week, gross hematuria with large clot burden continued, requiring frequent manual irrigation. On two occasions, approximately 3 days apart, she was noted to have a liter of frank blood in drainage bag that had collected over a period of less than 30 minutes. She required a total of eight units of packed red blood cells during this time. On hospital day 8, CT angiography was performed for persistent bleeding and the concern for possible AUF. This demonstrated an irregular contour of the right external iliac artery and a small blush of contrast extravasation where it crossed the right ureter, Figure 3. She was taken back to the operating room by vascular surgery and intraoperative angiogram confirmed ureteroarterial fistula from the right external iliac artery to the right ureter. A covered iCast 7 mm x 38 mm vascular stent was successfully deployed with resolution of extravasation. A right percutaneous nephrostomy tube was subsequently placed to allow removal of the indwelling right ureteral stent. The patient was discharged on clopidogrel and a 2 week course of oral antibiotics. At last contact, she is doing well with no recurrent hematuria.

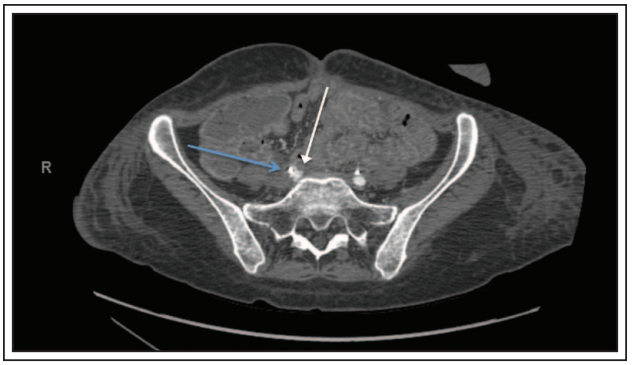

Figure 3. CT arteriogram shows contrast blush (arrow) where the right ureter (with indwelling stent, blue arrow) crosses the right common iliac artery.

Discussion

UAF are uncommon in urologic practice but can be the cause of life-threatening hematuria. The incidence of UAF appears to be increasing.[1] The most common presenting symptom is gross hematuria with clot passage which typically occurs intermittently, although persistent microscopic hematuria has also been described.[1-4] In a review of eight patients by Krambeck et al, 87.5% of patients had hematuria severe enough to require blood transfusion. The presence of prolonged ureteral stenting was common, with an average duration of 3.86 years of an indwelling stent prior to fistula development.[1] The exact pathophysiologic mechanism underlying the development of UAF is unclear, however several authors have postulated that the initial insult is ischemic injury to both the ureter and large blood vessels.[1-5] Compromise to the vaso vasorum, either from prior surgery or radiation, could lead to weakening and/or necrosis of major pelvic vessels.[2,7] When these vessels are adjacent to a fibrotic and fixed ureter, this pressure over time may lead to progressive weakening and fistula development.[2,5] The presence of an indwelling ureteral stent puts further pressure on the ureteral wall and may exacerbate this process.[1] In the case of CC, penetrating trauma from stent manipulation was likely an inciting factor.

Traditionally, open surgery was used to treat UAF, either via direct repair, embolization or vascular bypass.[6] However, these tended to be highly morbid

procedures given the uncertainty of diagnosis, technically demanding procedures and presence of comorbidities in this high risk population, making minimally invasive alternatives quite attractive. Kerns and colleagues first treated a UAF with an endovascular stent with immediate cessation of hemorrhage.[6] Subsequent experience with endovascular stenting for UAF has demonstrated good short term outcomes.[7]

Fox et al compared the long term outcome of endovascular versus open surgical repair.[8] A total of 19 patients with 20 ureteroarterial fistulae were studied with a mean follow up of 13 months. Endovascular techniques were initially attempted in 70% of cases, with two of these requiring subsequent open repair. This correlated to an 85% initial success rate with endovascular procedures. However, 36% of these required later revision either due to hemorrhage or limb ischemia. Of the patients initially treated with open repair, initial success was achieved in 67% of patients, with a 50% revision rate. Nephrectomy/renal loss was more common in the open repair group. Recurrent hemorrhage was more common in the open group (33% versus 14%), as were lower extremity complications (67% versus 50%). The authors concluded that there was no clear advantage to open or endovascular treatment of UAF, and recommended that endovascular techniques be first-line therapy in most cases of UAF due to their less-invasive nature. Regardless of treatment modality, patients appear to be at significant risk for recurrent hemorrhage or lower extremity complications.

The case studies above illustrate the difficulties in both diagnosis and management of UAF. Initial angiogram as well as CT scan in our first patient was negative for a fistula, which was only discovered later after continued transfusion dependent bleeding and provocative stent removal and irrigation. The second patient underwent a negative ureteroscopy, with confirmation of a fistula only on subsequent CT angiogram. This underscores the need for a high clinical suspicion of UAF as well as the shortcomings available imaging studies. Krambeck et al have reviewed the various diagnostic procedures for UAF, demonstrating low levels of sensitivity.[2] According to their analysis, cystoscopy can be used for diagnosis, however fistulae bleed intermittently, and bloody efflux may be absent at the time of the procedure. They did not recommend ureteroscopy due to the risk of exacerbating hemorrhage. Retrograde pyelogram was diagnostic in only 33% of studied cases and arteriography was positive in only 23%-41% of cases. This was improved to 63% by Krambeck et al using provocative angiography with simultaneous removal of the ureteral stent.

Both of our patients suffered secondary complications. CC developed acute kidney injury exacerbated by recurrent clot obstruction and repeated contrast loads. AC developed acute lower extremity ischemia following ureteroscopy requiring thrombectomy and revascularization. We postulate that mechanical stress from the ureteroscopy may have precipitated the formation of an arterial thrombus at the level of the fistula. These complications highlight the complexities of caring for patients with UAFs.

In both cases presented here, the decision was made not to replace a ureteral stent due to concern that it might increase the risk of infection of the vascular stent or predispose to future fistula formation. The risk of vascular stent infection in contact with the urinary tract is unknown. In the absence of data, both patients were treated with a prolonged course of antibiotics to decrease this risk.

In summary, our cases illustrate the diagnostic difficulty involved with identification of this life threatening urologic complication and the importance of maintaining a high index of suspicion in the face of negative studies. In both cases endovascular stenting proved immediately effective in managing bleeding from the UAF. Further studies will be required to determine the long term safety and efficacy of vascular stents in this setting. □

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