Upper tract urothelial carcinoma presenting at a bifurcation of a partially duplicated left ureter: A minimally invasive approach

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ABSTRACT
Urothelial carcinoma is the fourth most common solid organ malignancy. Rare cases arise from the upper urinary tract. A 78-year-old male presents with a chief complaint of hematuria, burning, urinary incontinence, and passing clots. The patient appeared to have a partial duplication of the left ureter. During the ureteroscopy, a mass was seen at the bifurcation of the partially duplicated left ureter. The mass in the ureter was classified as a T1 upper tract urothelial carcinoma. Minimally invasive endoscopic approaches were chosen to manage the patient and he presented to the operating room for laser ablation of the UTUC.

1. Introduction
Urothelial carcinoma is the fourth most common solid organ malignancy. Rare cases arise from the upper urinary tract. Lower urinary tract cancers (bladder and urethral) make up the majority (90–95%) of all urothelial carcinomas.1 The remaining cases (5–10%) arise from the upper urinary tract (calyces, infundibula, renal pelvis, ureters). This case highlights the unique presentation of a UTUC at the bifurcation of a partially duplicated ureter. Patients with an extensive genitourinary history should be considered for evaluation of this unique malignancy.

2. Case presentation
A 78-year-old male presents to the urology clinic with a chief complaint of hematuria, burning, urinary incontinence, and passing clots. The patient denied flank pain. The patient has a history of prostate cancer (T1cN0M0, 3 + 3+6, 3 + 4 = 7, 2/12 cores) that was biopsied and diagnosed in 2012 and treated with external beam radiation in 2013 (44 Tx). He presented to the clinic and was diagnosed with bladder cancer (CIS) that was treated with Bacillus Calmette-Guérin (BCG x 20 rounds) in 2015 and local transurethral resection of the bladder (TURBT). At time of initial workup a CT Urogram (CTU) was obtained. More recently, the patient presented to the emergency room for hematuria and clots. Chest X-ray done revealed a 2.2cm cavitory lesion in the right lower lobe of the lung in 2021 (Fig. 1). This was confirmed with follow-up CT scan of the chest.

The patient underwent a cystoscopy for his history and presenting symptoms. After cystoscopic visualization of the bladder, a retrograde pyelography was taken which showed mild hydronephrosis. A complex flexible ureteroscope was used to access the proximal ureter and kidney. The patient appeared to have a partial duplication of the left ureter (Fig. 2). During the ureteroscopy, a mass was seen at the bifurcation of the partially duplicated left ureter (Fig. 3). The scope was able to be passed proximal to the tumor. The patient underwent biopsies (x2) of the mass. The mass in the ureter was classified as a T1 upper tract urothelial carcinoma (UTUC). The specific location of this tumor at the bifurcation of the partial duplicate ureteral made this case unique. Minimally invasive endoscopic approaches were chosen to manage the patient due to low grade disease and cancer history. Once the treatment options of this cancer were discussed, the patient presented to the operating room for laser ablation of the UTUC. The patient required two additional laser ablative treatments for the cancer with a 90% treatment rate of the tumor. He will have another ureteroscopy in 3–6 months. If negative, then periodic retrograde pyelograms or maintenance ureteroscopies will be done.

3. Discussion
Although UTUC is a rare disease, there is important clinical significance among patients. Each year approximately 2 per 100,000 patients are diagnosed with UTUC in Western countries.2 Most of the UTUCs are made up of urothelial cancer (90%) and are most commonly located in
Risk factors for UTUC are similar to bladder cancer with some unique characteristics. The median age of diagnosis of UTUC is 70 years of age. The male-to-female ratio is around 2:1. Modifiable risk factors such as tobacco smoking and occupational exposure to aristolochic acid increases the risk of UTUC. Importantly, a prior history of bladder cancer places patients at higher risk of UTUC. UTUC most commonly presents with the complaint of hematuria. Some patients may also complain of flank pain initially.

An initial work-up of a patient with UTUC begins with a history and physical examination looking for risk factors. Lab work may reveal hematuria consistent with the patients presenting symptoms. Cystoscopy then assesses for the cause of the bleeding and is also done to check for concomitant bladder cancer. Retrograde pyelograms taken during cystoscopy may show filling defects at the location of the tumor. However, a CT urogram is preferred because of a higher sensitivity than retrograde pyelograms.

Treatment for UTUC is usually guided by tumor grade. Radical nephroureterectomy remains the gold standard of therapy. For low volume, low grade disease, such as the patient in this case, minimally invasive techniques are becoming more preferred. These nephron sparing therapies include endoscopic management, segmental resection, or intracavitary injections. Providers should recommend nephron sparing endoscopic therapies to decrease patient morbidity. Although not discussed here, lymph node dissection and chemotherapy do play a part in the management of UTUC and should be considered as well in treatment plans.

Duplicated ureters are found in approximately 1% of the population. They are often discovered incidentally on imaging. Excretory urography is diagnostic. These may have risk of future complications such as obstruction, urolithiasis, and vesicoureteral reflux. Although no screening methods exists, early detection and management should be considered.

The patient for this case presented with gross hematuria and was found to have an UTUC located at the duplication of a partial ureter. The choice to manage this patient with endoscopic ureteroscopy with holmium:YAG ablation was tolerated by the patient well. Minimally invasive approaches to this case were undertaken also due to his recent lung cancer diagnosis. When searching the literature, a few cases were found where UTUC cancer presented in a duplicated collecting system. Two cases examined patients with renal pelvic tumors and one case had a patient with a ureteral tumor, all three in a duplicated collecting system. The case of our patient presents another unique presenting of a UTUC in a partially duplicated ureter. However, its presentation at the
bifurcation has not been reported previously. With newer techniques allowing for nephron-sparing options, physicians now can consider less invasive treatment for local disease.

4. Conclusion

UTUC is a rare finding among patient populations. Management for local, low grade disease continues to be focused on minimally invasive treatments. For the patient in this case with T1 disease, the latter treatment is being used more commonly in this clinical situation. Anatomic abnormalities, such as duplicated ureters, exist in patients and providers should be aware of these. In this case, we presented a patient with UTUC at a bifurcation of a duplicated ureter, managed with minimally invasive endoscopic ablation with no evidence of complications or recurrence.

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