

Case Report

Renal pelvis urothelial carcinoma of the upper moiety in complete right renal duplex: a case report

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Abstract: Urothelial carcinoma (UC) originated from renal pelvis is the common tumor of the urinary system, however, neoplasia of the renal pelvis in duplex kidneys is extremely rare, especially in the complete renal and ureteral duplex cases. We present the first case of renal pelvis UC of the upper moiety in a complete right renal duplex. This male patient has bilateral complete renal and ureteral duplex. To the best of our knowledge, this is the first reported case of renal pelvis UC in a complete renal duplex system. After this experience we feel that the diagnosis of renal pelvis UC in duplex kidneys is not so easy, and once the diagnosis is determined, the whole renal duplex units and bladder cuff or ectopic orifice should be excised radically.

Keywords: Congenital malformation, hydronephrosis, renal duplex, renal pelvis, urothelial carcinoma

Introduction

Duplications of the renal collecting system and ureter are the most common anomalies of the upper urinary system, with about 0.8% having a ureteral duplication [1]. Though the renal pelvis UC is not rare, neoplasia of the renal pelvis in duplex kidneys is extremely rare, especially in complete renal and ureteral duplex cases. This report presents a case in which UC originated from the upper portion of the right duplicated renal pelvis with hydronephrosis, and this patient has bilateral complete renal and ureteral duplex. After the diagnostic dilemma, total nephroureterectomy with bladder cuff excision surgery of both renal units was carried out and the final diagnosis was proved.

Case report

In September 2012, a 65-year-old male patient presented to our department with more than a month of recurrent gross hematuria and occasionally right lumbar aching pain. He had no episode of frequency, urgency, odynuria and fever. The patient had a smoking and drinking history for more than thirty years and no other medical history. Urinary ultrasound revealed bilateral complete renal and ureteral duplex with heavy hydronephrosis and an equivocal

2.5*2.0 cm medium to high echogenic mass in the upper portion of the right duplicated renal pelvis. Noncontrast computed tomography (CT) revealed a small irregular lesion of soft tissue density in the inner wall of the dilated renal pelvis with the value of about 44HU (**Figure 1A**). It seems that the patient had a space-occupying lesion in the right upper renal pelvis, but no other evidence was found to support the diagnosis by the following examinations.

Urine cytology and KUB was negative. The right upper kidney was non-visualized in the intravenous pyelography (IVP). Magnetic resonance urography (MRU) also showed bilateral complete renal and ureteral duplex with heavy hydronephrosis and hydroureterosis of the right upper kidney and ureter and without other positive findings (**Figure 1B**). Cystoscopy showed no special sign of hematuria, and during the examination, an ectopic opening was found in the posterior urethra, close to the right bladder neck. In order to directly observe the possible disease, ureteroscopy was done through the ectopic opening, the urine was cloudy and the ureter was dilated without apparent bleeding or other disease. Unfortunately, the ureter was too long and the ureteroscopy cannot reach the renal pelvis. Next, the result of the retrograde

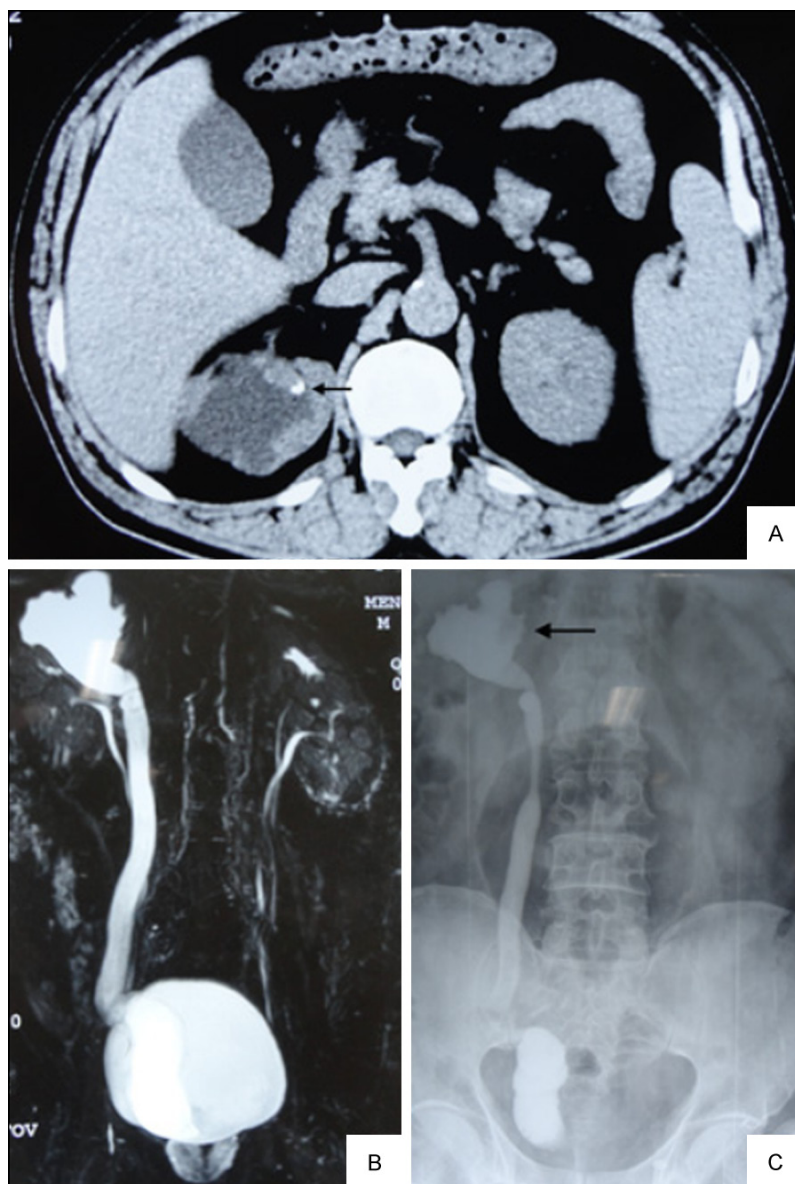


Figure 1. A: Noncontrast CT revealing a small irregular lesion of soft tissue density in the inner wall of the dilated renal pelvis with the value of about 44HU (black arrow); B: MRU showing bilateral complete renal and ureteral duplex without other positive findings; C: Retrograde pyelography showing a possible space-occupying lesion in the dilated renal pelvis (black arrow), however, most likely the filling defect was actually inflatable intestinal loop.

pyelography showed a possible space-occupying lesion in the dilated renal pelvis (**Figure 1C**). After joint consultation, nearly all the urologists and radiologists in our hospital were not sure about the imaging manifestation. They thought the filling defect was actually inflatable intestinal loop.

Summarily, though no conclusive evidence was found, this patient had gross hematuria and the bleeding most likely came from the possi-

ble space-occupying lesion in the abnormal right upper renal pelvis. According to common sense, the lesion was likely UC. However, the UC originated from the duplicated renal pelvis was so rare that we only retrieved one literature in which the authors just found one urothelial malignancy of 163 duplex patients [2]. After discussion and communication with the patient and his families, in order to solve the hematuria completely and reduce the possible complications, nephro-ureterectomy with bladder cuff excision surgery of both renal units was carried out by laparoscopic nephrectomy and lower abdominal incision. Gross specimen further confirmed the anomalies of complete renal and ureteral duplex and showed the typical small cauliflower-like tumor and multiple lichenoid lesions in the inner wall of the right upper renal pelvis (**Figure 2A, 2B**). Microscopic examination revealed that the tumor was poorly differentiated UC showing invasive growth features (**Figure 2C**). The patient is now under regular bladder perfusion with HCPT and intravenous chemotherapy with docetaxel and oxaliplatin.

Discussion

Many people with urinary congenital malformation remain asymptomatic, although multiple cases may present complications, often secondary to obstructive uropathy and urinary stasis (hydronephrosis, lithiasis, infections, etc). Moreover, these patients present a risk of having renal pelvis malignancy [3]. The incidence of squamous cell carcinoma in the renal pelvis of horseshoe kidneys is higher than that in nor-

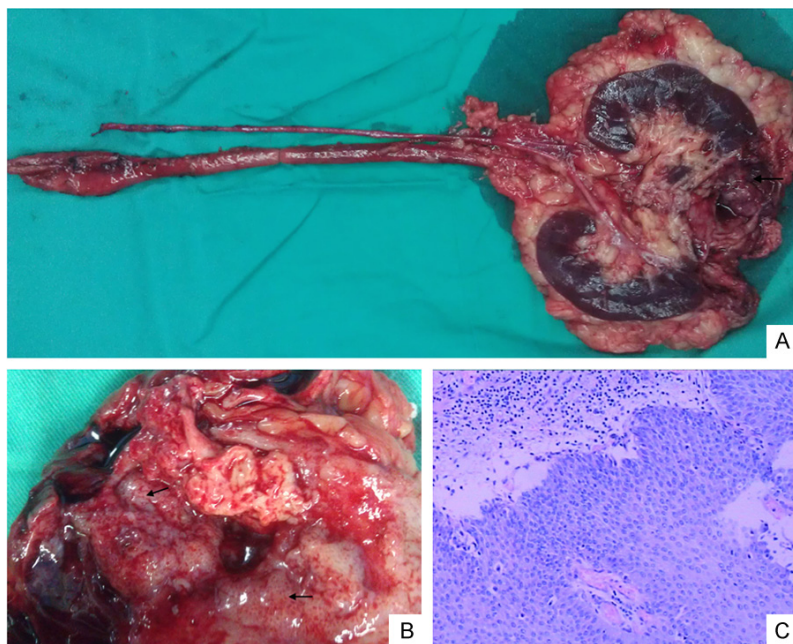


Figure 2. A: Gross specimen confirmed the anomalies of complete renal and ureteral duplex and showed the typical small cauliflower-like tumor and multiple lichenoid lesions in the inner wall of the right upper renal pelvis (black arrow); B: Local magnification (black arrows) of the lesion; C: Microscopic examination revealed that the tumor was poorly differentiated UC showing invasive growth features (hematoxylin-eosin $\times 200$).

mal kidneys. The increased occurrence of chronic infection, obstruction and stone formation may be instrumental in producing a higher incidence of renal pelvic tumors in patients with horseshoe kidney [4, 5]. However, neoplasia of the renal pelvis in duplex kidney is rarely seen. Chen [6] reported a case with sarcomatoid carcinoma of the renal pelvis in the upper hydronephrosis moiety of the complete left duplex kidney. UC originated from the duplicated renal pelvis was not reported in detail before, especially in the patient with bilateral complete renal and ureteral duplex. To our best knowledge, the present report is the first case. Presently, the development of renal pelvis UC of renal duplex is not understood, urinary stasis and long-time smoking may be instrumental in producing this kind of carcinoma. Therefore, it is important to stress the need for frequent monitoring or treating complex duplex kidney with hydronephrosis of either moiety in case of a risk of having neoplasia.

Usually, the diagnosis of renal pelvis UC is not difficult. But when the neoplasia is not so big and originates from a duplex kidney with heavy hydronephrosis, some diagnostic methods are

not useful, such as IVP, MRU and ureteroscopy. Otherwise, Urinary ultrasound and CT show some values. Some urologists may think the upper heminephroureterectomy is enough because the lower moiety of duplex renal system is normal. However, we believe the whole renal duplex units and bladder cuff or ectopic orifice should be excised radically, because the upper and lower moiety of duplex renal systems have common vessel traffic branches and the two ureters have the same origin. Furthermore, hemi-nephrectomy often damages collecting system and cause possible cancer cell implantation, so keeping the integrity of the collecting system is the rule of the surgery for UC of

the upper urinary tract. We think our experience of diagnosis and treatment for this rare case will be helpful to urologists. Of course, more cases are needed to improve the level of diagnosis and treatment for this kind of patients.

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Disclosure of conflict of interest

None.

Abbreviations

CT, computed tomography; IVP, intravenous pyelography; MRU, magnetic resonance urography; UC, urothelial carcinoma.

Renal pelvis UC of duplex kidney

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