Clinical Case Report

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Right supernumerary kidney with urothelial carcinoma

A case report

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Abstract

Rationale: A supernumerary kidney is an extremely rare renal anomaly. Currently, <100 cases are reported in the literature. There are only 2 right unilateral supernumerary kidneys reported in the literature thus far, but no confirmed cases of urothelial carcinoma in supernumerary kidneys. We report a case of a right supernumerary with urothelial carcinoma, which is, to the best of our knowledge, reported for the first time.

Patient concerns: A 73-year-old female patient presented with intermittent, painless, whole course and gross hematuria for about 3 months. Her physical and laboratory examinations did not reveal any significant findings except positive occult blood in routine urine examination. Contrast-enhanced spiral computed tomography revealed a dysplastic supernumerary kidney under the normal right kidney.

Diagnoses: The ureteroscopy showed that the ureter was Y-shaped in the middle part. The medial ureter led to a normal kidney. The lateral ureter was just 2 cm and led to a small cavity in which there was a mass whose biopsy showed urothelial carcinoma. The patient was subsequently diagnosed with a right supernumerary kidney with urothelial carcinoma.

Intervention: Nephroureterectomy, including the right normal and supernumerary kidneys, and partial cystectomy by laparoscopy were performed after the ureteroscopy. The patient then received 6 cycles of gemcitabine and cisplatin regimen chemotherapy and regular intravesical epirubicin chemotherapy.

Outcomes: No recurrence or metastasis was found on follow-up computed tomography performed 13 months postoperatively.

Lessons: A supernumerary kidney is an extremely rare renal anomaly. Malignancy can occur in supernumerary kidneys.

Abbreviation: CT = computed tomography.

Keywords: congenital anomaly, supernumerary kidney, urothelial carcinoma

1. Introduction

A supernumerary kidney is an extremely rare renal anomaly. It is defined as the third kidney (in addition to the two independent kidneys), with a distinct collecting system, blood supply, and well-defined capsule.[1] Currently, <100 cases are reported in the literature, with the first case being reported in 1965.[2] The real incidence of supernumerary kidneys cannot be calculated because of its unusual appearance. There are only 2 right supernumerary kidneys reported in the literature thus far.[3,4] We report a case of a right supernumerary kidney with confirmed urothelial carcinoma, which is, to the best of our knowledge, being reported for the first time.

2. Case report

A 73-year-old female patient presented with intermittent, painless, whole course, and gross hematuria for approximately 3 months. The patient had a history of untreated hypertension. Her physical and laboratory examinations did not reveal any significant findings except positive occult blood in routine urine examination. Contrast-enhanced spiral computed tomography (CT) revealed a dysplastic supernumerary kidney measuring approximately $4.2 \times 5.0 \times 5.3$ cm in size under the normal right kidney (Fig. 1A–C), which had a separate arterial supply originating from the aorta (Fig. 1D).

The ureteroscopy showed that the right ureter was Y-shaped in the middle part (Fig. 2A). The medial ureter (green arrow) led to a normal kidney. The lateral ureter (yellow arrow) was just about 2

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The authors report no conflicts of interest.

All data generated or analyzed during this study are included in this published article [and its supplementary information files].

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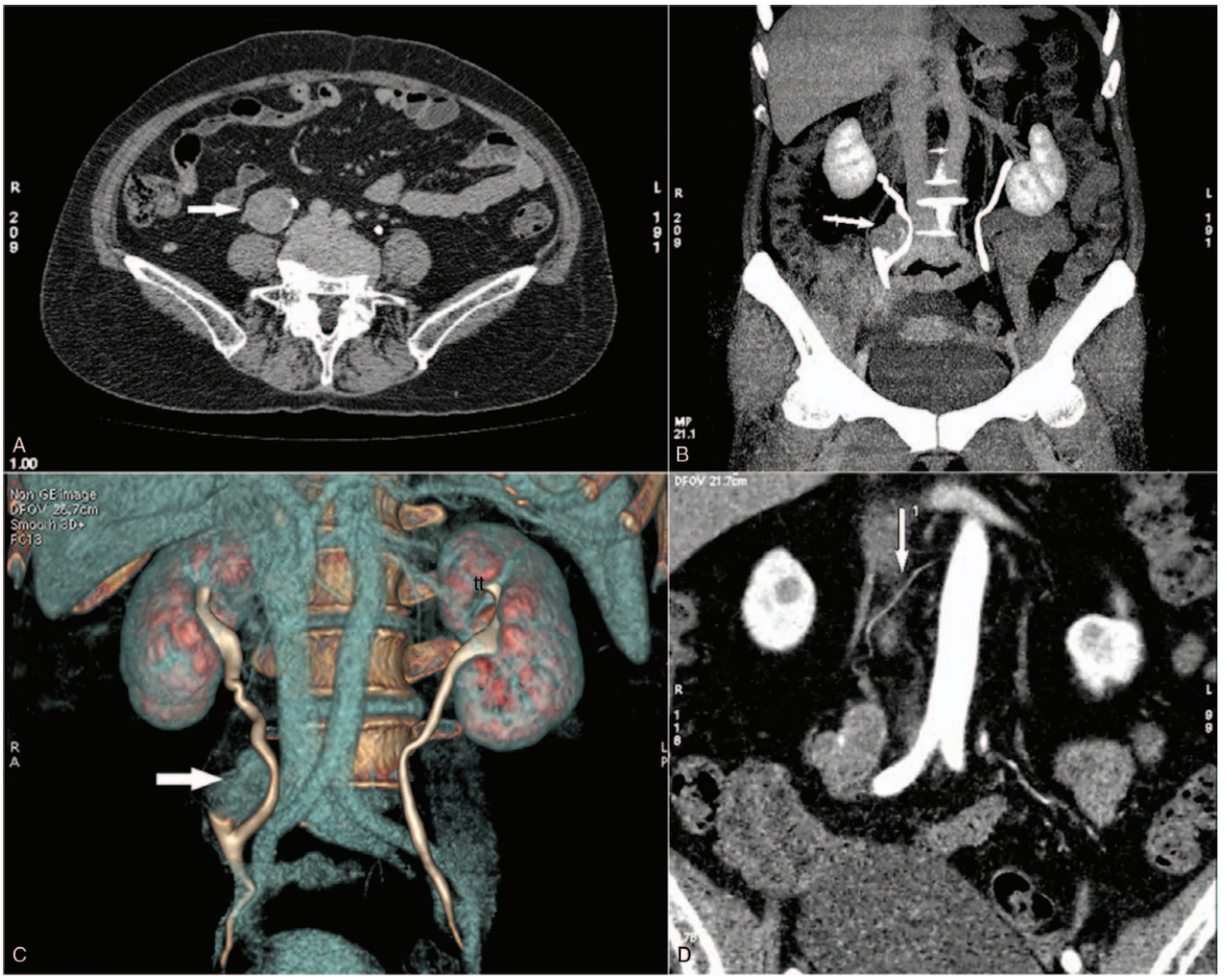

Figure 1. Computed tomography (CT) demonstrates a dysplastic supernumerary kidney (arrow) under the normal right kidney (A–C), and a separate arterial supply (arrow) originating from the aorta (D).

cm and led to a small cavity in which there was a mass (Fig. 2B), whose biopsy showed urothelial carcinoma. Nephroureterectomy, including the right normal and supernumerary kidneys, and partial cystectomy by laparoscopy were performed after the ureteroscopy. The postoperative specimen showed 2 branches of the right ureter and a tumor in the supernumerary kidney (Fig. 3). Postoperative pathology revealed that the tumor was a high-grade urothelial carcinoma (Fig. 4A). Glomerular and tubular structures were found in the supernumerary kidney (Fig. 4B). The patient then received 6 cycles of gemcitabine and cisplatin regimen chemotherapy and regular intravesical epirubicin chemotherapy.

No recurrence or metastasis was found on follow-up CT performed 13 months postoperatively. There was no discomfort except for hair loss and occasional nausea. The patient was satisfied with the timely and effective treatment.

3. Discussion

A supernumerary kidney is a very rare congenital anomaly of the urinary tract. Only <100 case reports can be found in the literature. It is usually smaller than a normal kidney in terms of size and function.[5] The supernumerary kidney can be either totally isolated from the ipsilateral kidney or attached to it through loose fibrous tissue.[5] The supernumerary kidney is thought to result from an abnormal division of the nephrogenic cord into 2 separate metanephric blastemas at the fifth to seventh week of gestation, and it may have partially or completely duplicated ureters.[6] Compared with a duplex kidney, a supernumerary kidney has a separate arterial supply originating from the aorta, venous drainage via the inferior vena cava, pelvicalyceal system, and distinct renal capsule.[3,7,8] A supernumerary kidney is usually present on the left side. Although there are several bilateral supernumerary kidney reports, it is extremely rare to have a right unilateral supernumerary kidney, with only 2 cases were reported thus far.[3,4,9]

Some supernumerary kidney-associated congenital anomalies include horseshoe kidney malformations, ureteral atresia, imperforate anus, vaginal atresia, ectopic ureter implantation, urethral duplication, coarctation of the aorta, and meningomyelocele.[10] Because of the hypoplastic nature of the involved renal element, urinary incontinence produced by ureteral ectopia from the supernumerary kidney is rarely seen.[9] These anomalies are often asymptomatic and usually go undiagnosed until the fourth

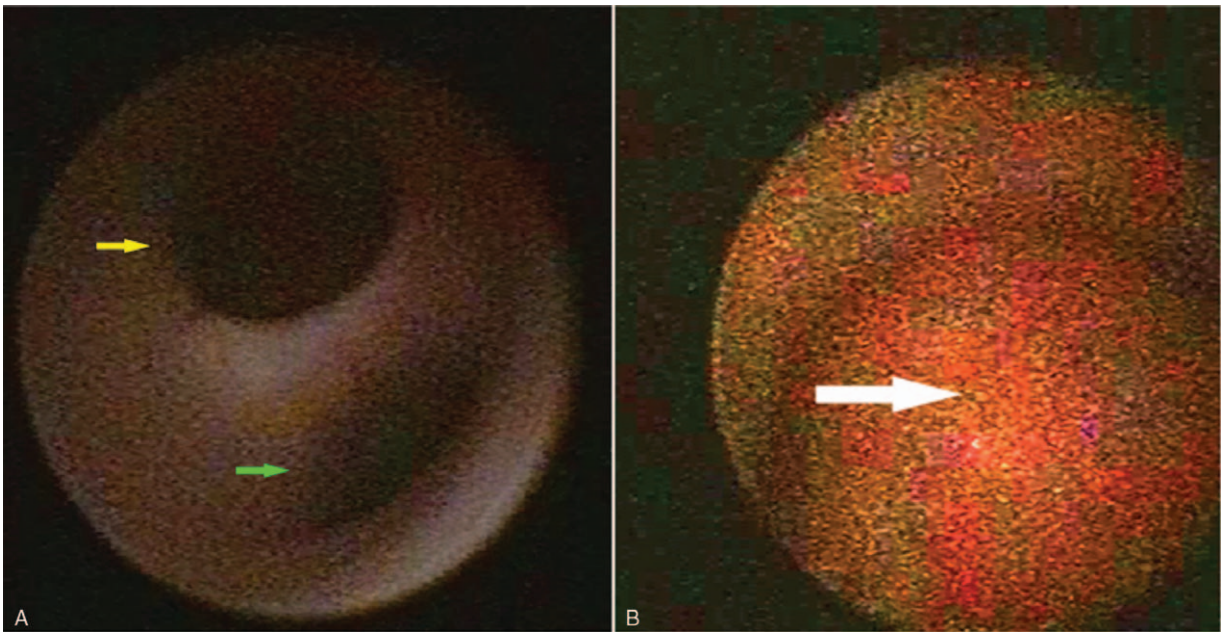

Figure 2. The ureteroscopy showing the right ureter with two branches in the middle part: the lateral ureter (yellow arrow) and medial ureter (green arrow) (A). The lateral ureter leads to a small cavity in which there was a mass (arrow) (B).

decade of life.[10] Abdominal discomfort or a palpable mass, hypertension, and fever may be the most common presenting symptoms.[3] A number of pathologic conditions, such as pyelonephritis, hydronephrosis, renal calculi, ureteropelvic junction obstruction, and benign and malignant neoplasms, may affect the supernumerary kidney.[4] Carlson reported that 2 carcinomas had been seen in conjunction with a supernumerary kidney in 51 cases, without a definite pathological type.[11] Exley and Hotchkiss reported a supernumerary kidney with clear cell carcinoma.[12] The present case is the first confirmed urothelial carcinoma in a supernumerary kidney. It should be considered that malignancy can occur in supernumerary kidneys.

A supernumerary kidney with urothelial carcinoma can be treated as a renal pelvic carcinoma, undergoing a nephroureterectomy including the supernumerary and ipsilateral normal kidney and partial cystectomy by laparoscopy. Intravesical chemotherapy and platinum-based chemotherapy can be beneficial.

To conclude, the present case is more interesting in 3 respects. First, this is the third right unilateral supernumerary kidney ever

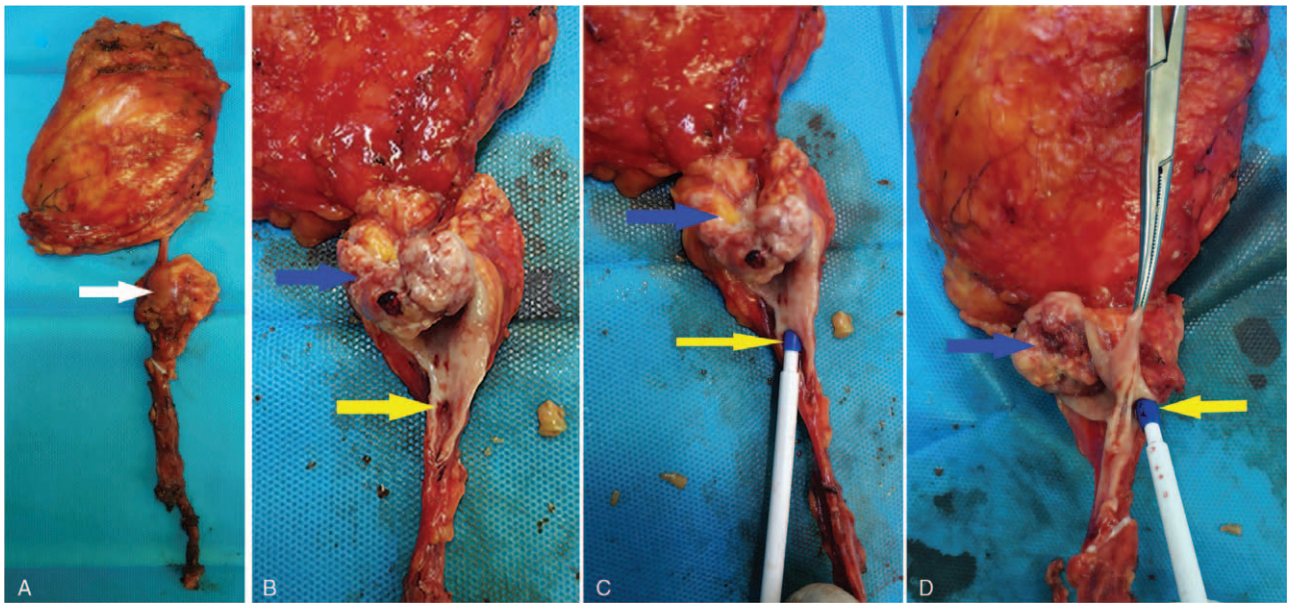

Figure 3. The postoperative specimen shows the supernumerary kidney (arrow) (A), tumor (blue arrow), and medial ureter leading to a normal kidney (yellow arrow) (B–D).

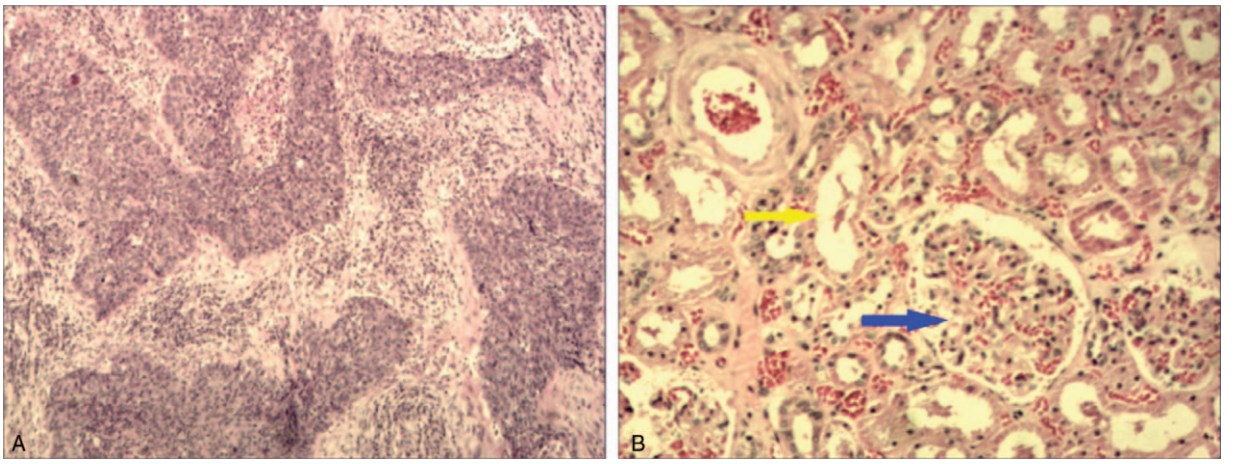

Figure 4. Postoperative pathology revealing high-grade urothelial carcinoma (hematoxylin and eosin staining, ×40) (A) and the glomerular (blue arrow) and tubular (yellow arrow) structures in the supernumerary kidney (hematoxylin and eosin staining, ×100) (B).

reported. Second, this is the first supernumerary kidney with confirmed urothelial carcinoma. Third, the combination of radical surgery and chemotherapy is safe and effective for this patient.

4. Consent for publication

Informed written consent was obtained from the patient for publication of this case report. The presented data are anonymized, and the risk of identification is minimal.

Author contributions

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Image recognition: Xudong Luo, Zhen Gao.
Writing – original draft: Xinghua Gao, Qingfei Xing.
Writing – review & editing: Longyang Zhang, Feng Guo.

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