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CASE REPORT

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Partial nephrectomy of a horseshoe kidney associated with renal cell carcinoma and ureteral stone: A clinical case report

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Key Clinical Message

Although anatomical and vascular abnormalities of the horseshoe kidney might be challenging, complete preoperative imaging evaluations and accurate organ-sparing surgical planning can lead to much lower complications.

Abstract

Horseshoe kidney (HK) is one of the most common renal fusion anomalies. Renal carcinoids are rarely reported in HK patients. Here, we described a rare case of advanced right renal cell carcinoma (RCC) along with proximal left ureter stone in a 41-year-old man who presented with a complaint of turbid urine. Early blood tests revealed a blood urea nitrogen of 44 mg/dL and serum creatinine of 1.35 mg/dL. The urine analysis showed microscopic hematuria (6–8 RBCs) and few calcium oxalate crystals. The imaging evaluations revealed an HK anomaly with a solid mass on the right side and a 4 mm stone in the proximal left ureter. The findings suggested RCC which was confirmed by histopathology examination. Consequently, the patient was scheduled for an organ-preserving open surgery of a right kidney tumor with concomitant left ureterolithotomy. The 16-month follow-up showed no urological complications, metastasis, or tumor proliferation. Although the anatomical and vascular abnormalities of HK might be challenging, organ-sparing surgical treatment should be considered in feasibly resectable tumors. Complete preoperative imaging evaluations to identify the characteristics of HK, as well as accurate surgical planning, can lead to much lower complications.

KEYWORDS

horseshoe kidney, organ-sparing surgery, partial nephrectomy, renal cell carcinoma, ureteral stone

1 | INTRODUCTION

Horseshoe kidney (HK) is the most common renal anomaly, with an incidence of 0.1%–0.25% of the population that often appears in males with chromosomal aneuploidies, including 20% of trisomy and 60% of Turner syndrome.[1,2] Although most HK patients are asymptomatic, urolithiasis and ureteropelvic junction (UPJ) obstruction

commonly occur due to anatomical variation of the HK, causing hydronephrosis.[3] Renal carcinoids have rarely been reported in HK patients. However, the risk of developing Wilms tumor in HK has been described as double.[4] Renal infection, obstruction, and stone disease in patients with HK may lead to renal pelvic tumors.[5] Currently, surgery including partial nephrectomy, is the standard treatment for renal neoplasms, but it is challenging for tumors developed in HK because of its complex anatomy and abnormal vascularization. Here, we describe a rare case of advanced right renal cell carcinoma (RCC) occurring in HK simultaneously with a proximal left ureteral stone.

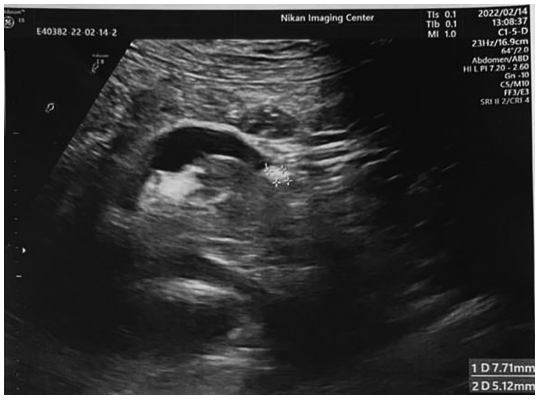

FIGURE 1 The kidney ultrasonography; left renal malrotation with pelvicalyceal fullness due to a stone in proximal left ureter.

2 | CASE PRESENTATION

A 41-year-old male was admitted to Mostafa Khomeini Hospital, Tehran, Iran, with a complaint of turbid urine. The patient did not complain of pain. His medical history included migraine, bipolar disorder, and hyperlipidemia. On examination, the patient was alert and oriented and his vital signs were normal. Initial blood tests revealed a blood urea nitrogen (BUN) 44 mg/dL and serum creatinine (SCr) 1.35 mg/dL. Urine analysis revealed yellow, semi-clear urine with a trace of blood, 0–2 WBCs, 6–8 RBCs, and a few calcium oxalate crystals. The urine culture was negative.

Furthermore, kidney ultrasonography was performed, which showed a well-defined round hetero-echoic solid mass (52 × 51 mm) with extrarenal extension in the middle pole of the right kidney (103 mm) with a pressure effect on the renal sinus. The left kidney (120 mm) appeared to be malrotated and located caudal to the normal anatomical site (Figure 1). Subsequently, abdominal and pelvic spiral computed tomography (CT) and abdominal magnetic resonance imaging (MRI) demonstrated an exophytic well-defined 50 × 53 × 48 mm mass lesion in the middle pole of the right kidney, with a mass effect on the pyelocaliceal system adjacent to the right liver lobe, suggesting renal cell carcinoma (RCC), along with a 4 mm stone in the proximal left ureter with 21 mm distance from the left ureteropelvic junction (UPJ) of the HK (Figures 2 and 3). The type of kidney stone was calcium oxalate. Until then, the patient was unaware of his kidney abnormalities and had not observed any symptoms of urological disorders.

Based on these findings, the patient was a candidate for organ-preserving open surgery for the right kidney tumor with concomitant left ureterolithotomy. Initially, renal angiography was performed to reveal an aberrant pattern of vascularization, which showed two arteries in the right kidney. We initiated our surgical approach via a midline abdominal incision. The ascending colon and ileum mesenterium were then dissected, which was essential for complete exposure to HK. The lower pole of the right kidney was occupied by a large tumor. Due to the large size of tumor, the renal ischemia was prepared by clamping the three arteries of right kidney using bulldog clamp technique. The polar resection technique completely excised the tumor and prepared the perfect visualization of the layers. Then, the visible vessels and collecting system were sutured (figure-of-eight stitch and running sutures, respectively). After unclamping, the renal defect was assessed for bleeding, and selective single-layer renorrhaphy was done. The ischemia time was 11 min. Fortunately, most of the right renal bulk tissue was successfully preserved. The upper left ureter was then explored and released. Once a ureteral stone was identified, ureterolithotomy was performed. A double-J ureteral stent was inserted and the ureter was sutured.

Pathological examination of the tumor (Figure 4) confirmed Grade 2 RCC with a clear cell subtype, 6.0 cm in diameter, and adhered to 0.5 cm surrounding tissue. The surgery lasted 215 min, with an estimated blood loss of 200 mL. The patient was discharged after 7 days of hospitalization. During the 16-month follow-up period, the patient reported no urological symptoms. Serial imaging studies after surgery showed no metastasis or growth of another tumor. His SCr level was stable at approximately 2 mg/dL.

3 | DISCUSSION

HK is one of the most common fusion anomalies of the kidneys, accounting for 0.15%–0.25% of renal developmental disorders in the general population.[6] Almost 30% of all HK are asymptomatic and are discovered accidentally by physical examination or imaging modalities such as CT and ultrasound during the management of other diseases.[7] Renal tumors of HK have been identified in

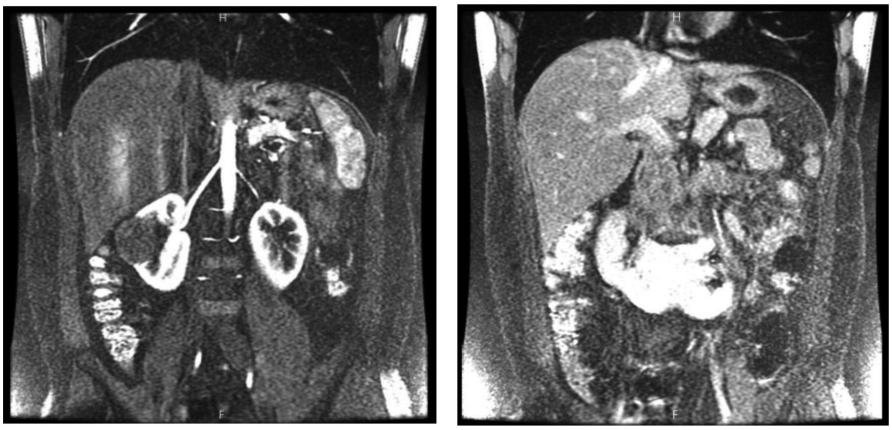

FIGURE 2 The magnetic resonance imaging revealed horseshoe kidney with exophytic mass lesion in the right middle pole, adjacent to right liver lobe.

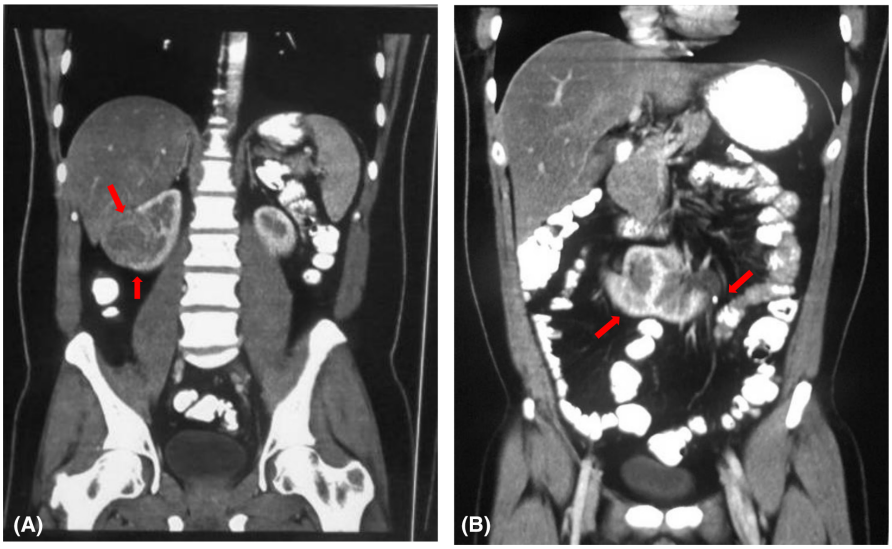

FIGURE 3 Abdomen and pelvis spiral CT scan with and without contrast; (A) A well-defined hypodense round shaped mass with heterogenous enhancement (53 × 51 mm) in middle pole of right kidney, near approximation with Segment VI of liver and pressure effect to renal sinus. (B) Horseshoe kidney and 4 mm stone in proximal left ureter with 21 mm distance from left UPJ.

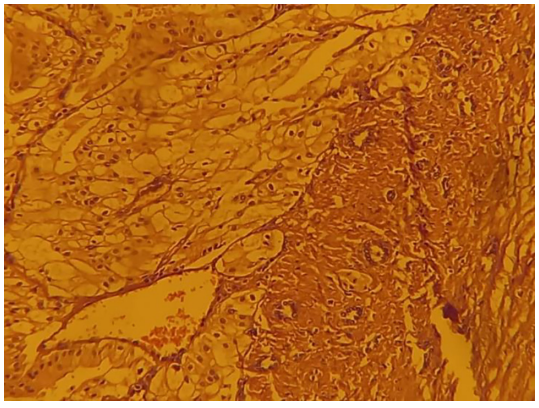

FIGURE 4 The microscopic examination of tumor, Van Gieson stain; showed the Grade 2 renal cell carcinoma with a clear cell subtype.

almost 12% of patients with these anomalies.[8] Although the risk of urothelial carcinoma is approximately four times higher in kidneys with fusion anomalies, the most commonly reported tumor is RCC, which is apparent in approximately 50% of patients.[8–11] Previous studies have reported flank pain, hematuria, and abdominal masses as the most common symptoms in patients with kidney tumors.[8,10] Similarly, our patient had hitherto unknown HK with microscopic hematuria that developed RCC confined to the kidney.

CT is the gold standard for identifying renal tumors. However, in the case of a solitary kidney, bilateral tumors, or in patients with HK, CT angiography, or MRI is also required because of its unusual anatomy.[12] Abnormal vascularization is a common finding in patients with fusion anomalies of the kidney. Graves[13] described six blood supply patterns to HK. However, nonconformity with Graves' classification has been identified, and arteries may be asymmetrically distributed.[14] Clodney et al., in a study of 209 cases with fusion anomalies, reported that the average number of arteries of the left and right kidneys were 1.9 and 2.4, respectively, and only 5% of patients with HK had one artery for each kidney. In this study, CT angiography revealed that accessory blood vessels (two arteries) supplied the right hemi-kidney of the

patient. However, we encountered three vessels when the HK was exposed.

Surgery is the best treatment for HK cancers with resectable cases. Since Vermooten presented the clinical and technical aspects of organ-sparing tumor resection in 1950,[15] many surgeons have developed techniques based on various surgical experiences. Three main factors resulting from the nature of the HK defect should be considered when deciding to use an organ-sparing surgical method: the anatomical position of the kidney, variations in renal vascularization, and different structures of the isthmus.[16] Partial nephrectomy for the treatment of HK tumors has been demonstrated to be a practical method. Considering surgical complications, including perioperative blood loss, urine leakage, peri- and postoperative renal function, and survival rate, partial nephrectomy has led to an excellent oncological and functional prognosis. Furthermore, in several case series, the average estimated blood loss was approximately 400 mL.[11] Our patient underwent a successful organ-preserving partial nephrectomy for a tumor with a diameter of 6 cm, and the estimated blood loss was 200 mL.

Partial nephrectomy of HK could be challenging due to complex anatomy and abnormal vasculature. In this case, although the angiography showed two supplier arteries, we faced three vessels when we were exposed to the HK. The uncertainty of the collecting system, the stone in the opposite ureter, and the lack of previous experience due to the rarity of such conditions, were other challenges of our patient's surgery.

4 | CONCLUSION

The current case report presents a case of incidentally diagnosed RCC simultaneously with a proximal left ureteral stone in a patient with HK who underwent complex organ-preserving open surgery (partial nephrectomy), resulting in a good prognosis during a 16-month follow-up. This case demonstrates that although anatomical and vascular abnormalities of HK might be challenging, organ-sparing surgical treatment should be considered in feasibly resectable tumors. Thorough preoperative imaging evaluations to identify the characteristics of HK, as well as accurate surgical planning, can lead to fewer complications.

AUTHOR CONTRIBUTIONS

Feraidoon Khayyamfar: Conceptualization; investigation; supervision; writing – review and editing. **Davood Dalil:** Data curation; project administration; writing – original draft; writing – review and editing. **Amirmahdi Khayyamfar:** Writing – review and editing.

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None.

CONFLICT OF INTEREST STATEMENT

The authors declare that there is no conflict of interest regarding the publication of this article.

DATA AVAILABILITY STATEMENT

The data used to support the findings of this study are available from the corresponding author upon request.

ETHICAL APPROVAL

The Research Ethics Committee of Shahed University approved all procedures performed in the current case report. The patient expressed his informed consent through a written form.

CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

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