Partial and hemi-nephrectomy for renal malignancy in patients with horseshoe kidney

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Introduction: Horseshoe kidney is the most common congenital renal fusion anomaly, with an estimated incidence of 1.7 to 2.5 cases per 1000 live births. In these patients, nephron-sparing surgical management of renal tumors may be complicated by abnormal renal location, aberrant vasculature, and the presence of a renal isthmus. We present the largest known series of patients with renal malignancy in horseshoe kidneys managed by partial or hemi-nephrectomy with associated outcomes.

Materials and methods: A retrospective review of our institution's electronic medical record was conducted to identify consecutive cases over an 11 year period. Pediatric patients and those who underwent surgery for benign indications were excluded from analysis.

Results: Eight patients with horseshoe kidney who underwent partial or hemi-nephrectomy for renal malignancy were identified. Median tumor size was 6.0 cm (IQR 3.7 cm-9.5 cm). Six patients had clear cell renal cell carcinoma (RCC), 1 patient had papillary RCC, and 1 patient had a renal carcinoid tumor with concurrent adenocarcinoma. Median length of stay was 4 days (IQR 2-.5.5 days). Median perioperative change in eGFR was -6 mL/min/1.73m² (IQR -2.6-8.6 mL/min/1.73m²). One patient developed postoperative urine leak requiring percutaneous drainage and ureteral stent placement. Median follow up was 38.5 months, with a cancer-specific survival of 87.5% and an overall survival of 62.5%. **Conclusion:** Partial and hemi-nephrectomy for renal malignancy can safely be performed in patients with horseshoe kidney with acceptable operative and oncologic outcomes.

Key Words: congenital anomaly, renal cell carcinoma, partial nephrectomy, horseshoe kidney

Introduction

Horseshoe kidney is the most common congenital renal fusion anomaly, with an estimated incidence of 1.7 to 2.5 cases per 1000 live births.¹ Patients with a horseshoe kidney are thought to have the same risk of renal malignancy as the general population.² In patients with small renal tumors, partial nephrectomy is the reference standard and is associated with excellent rates of cancer-specific survival.³

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Address correspondence to Dr. Todd Yecies, Department of Urology, UPMC, 3471 Fifth Ave, Suite 700, Pittsburgh, PA 15213 USA However, the horseshoe kidney's abnormally low location, irregular vascular supply, anterior renal pelvis, and presence of the renal isthmus present challenges to nephron-sparing surgery. As such, surgical complications associated with altered renal anatomy have been reported.⁴ Though many patients with horseshoe kidney and renal malignancy would potentially benefit from a nephron-sparing approach, feasibility may be compromised due to the complexity of the operation in this patient population.

Partial nephrectomy in patients with horseshoe kidney has been described for renal malignancy via both open and laparoscopic approaches; however data regarding the feasibility of partial nephrectomy in the setting of horseshoe kidney is limited to individual cases or series of 3 or fewer patients.⁵⁻⁷ We report

ID	Age (yrs)	Sex	BMI (kg/m²)	CCI	Preop cr (mg/dL)	Preop eGFR (mL/min/1.73m²)	Laterality	
1	53	М	29.8	4	0.8	102	Left	
2	49	F	33.1	2	0.8	87	Left	
3	50	F	34.4	2	0.6	107	Right	
4	56	М	31.1	2	1.2	67	Right	
5	59	F	32.3	6	0.8	81	Left	
6	86	F	21.8	2	0.9	58	Right	
7	60	М	29.3	2	1.0	81	Left	
8	66	М	21.2	2	1.0	78	Right	

TABLE 1. Subject demographic information with pre and postoperative renal function

BMI = body mass index; CCI = Charlson Comorbidity Index; cr = serum creatinine; eGFR = estimated glomerular filtration rate; M = male; F = female

our results of an 11 year experience with partial nephrectomy and hemi-nephrectomy in patients with horseshoe kidney.

Materials and methods

A retrospective review of the electronic medical record of a single, multi-center, health system was performed. The inclusion criteria were patients with horseshoe kidney undergoing partial or hemi-nephrectomy for management of renal malignancy. Exclusion criteria included age < 18 years and surgery for benign indications. The MARS application was used to identify operative reports containing the words "horseshoe kidney" and "nephrectomy." The medical records of potential subjects were then reviewed by the primary author to ensure they met inclusion criteria.

Data collected included patient demographics, length of stay, comorbidities, tumor characteristics on preoperative imaging, final pathologic analysis, pre and postoperative renal function, treatment complications, and long term oncologic outcomes. Length of stay was determined from date of admission to date of discharge. Charlson Comorbidity Index was calculated upon review of patient history in the medical record.⁸ Glomerular filtration rate was estimate using the Chronic Kidney Disease Epidemiology Collaboration (CKD-EPI) creatinine equation.⁹ Descriptive statistics were performed using Stata/SE, version 14.0.

Results

We identified 22 patients with operative records that contained the terms "horseshoe kidney" and "nephrectomy." Of these, 8 were adult patients with

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horseshoe kidney who underwent partial or heminephrectomy for renal malignancy. Table 1 lists patient demographics and preoperative renal function. Patients had a median age of 58 years (IQR 52-63 years) and median Charlson Comorbidity Index of 2 (IQR 2-3). Median preoperative serum creatine and eGFR were 0.85 mg/dL (IQR 0.80-1.0 mg/dL) and 81 mL/min/1.73m² (IQR 72.5-94.5 mL/min/1.73m²). Representative imaging is demonstrated in Figure 1. All patients had computed tomography (CT) with contrast enhancement, including three patients who underwent CT-angiography. One patient underwent invasive angiography. Identified vascular anatomy



Figure 1. Representative CT images of described patients. **A)** 9.7 cm renal carcinoid tumor in patient #3 **B)** 9.2 cm clear cell RCC in patient #6 **C)** 7.5 cm clear cell RCC in patient #7 with atrophic contralateral moiety **D)** 4.6 cm clear cell RCC in patient #8.

ID	Approach	Operative time (min)	Discharge cr (mg/dL)	Discharge eGFR (mL/min)	∆ eGFR	LOS (days)	Tumor size (cm)	Pathology
1	Subcostal	92	0.8	102	0	2	1.5	Clear cell RCC
2	Subcostal	unknown	0.9	75	-12	2	4.2	Papillary RCC
3	Subcostal	75	0.6	107	0	2	9.7	Renal carcinoid tumor + adenocarcinoma
4	Hand- assisted	420	1.8	41	-26	4	9.8	Clear cell RCC
	laparoscopi	С						
5	Midline	354	1.1	55	-26	7	3.2	Clear cell RCC
6	Midline	102	0.9	58	0	5	9.2	Clear cell RCC
7	Midline	357	2.5	27	-54	7	7.5	Clear cell RCC
8	Midline	99	1.1	70	8	3	4.6	Clear cell RCC
cr = Creatinine; eGFR = estimated glomerular filtration rate; LOS = hospital length of stay; RCC = renal cell carcinoma								

TABLE 2.	Perioperative	features and	pathologi	c information	of the stud	y population
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ranged from two to five renal arteries feeding the two renal moieties.

Perioperative pathological data are summarized in Table 2. All patients underwent an open surgical approach through a midline or subcostal incision, except for one patient, who underwent hand-assisted laparoscopic partial nephrectomy. Median operative time was 102 minutes (IQR 92-357 minutes). Two patients required blood transfusion within the perioperative period. Median tumor size was 6.0 cm (IQR 3.7-9.5 cm). Six patients were diagnosed with clear cell RCC, one patient had papillary RCC, and one patient had a renal carcinoid tumor concurrent with moderately differentiated adenocarcinoma. All patients survived to discharge with a median length of stay of 4 days (IQR 2-5.5 days). Median serum creatinine and eGFR at the time of discharge were 1 mg/dL (IQR 0.9-1.5 mg/dL) and 64 mL/min/1.73m² (IQR 48-89 mL/min/1.73m²), respectively, with a median change of 0.1 mg/dL (IQR 0-0.38 mg/dL) and eGFR of -6 mL/min/1.73m² (IQR $-2.6-8.6 \text{ mL/min}/1.73 \text{m}^2$). The patient with the largest decline in eGFR (-54 mL/min/1.73m²) was known preoperatively to have an atrophic contralateral moiety. No patients required renal replacement therapy in the perioperative period.

One patient (#7) had a postoperative urine leak requiring re-intervention. He underwent concomitant radical retropubic prostatectomy and open left partial nephectomy for high risk prostate cancer and a 7.5 cm enhancing renal mass within a functionally-solitary left moiety of horseshoe kidney. His operation was technically difficult due to the presence of a second, lower pole renal artery and the anterior location of the renal pelvis and ureter. Urine leak resolved 6 weeks following percutaneous drainage and ureteral stent insertion. This patient's eGFR declined from 81 mL/min/1.73m² preoperatively to 27 mL/min/1.73m² postoperatively, stabilizing at 46 mL/min/1.73m² at 3 months.

Median follow up was 38.5 months. Only one patient (#4) progressed to metastatic disease. Time to progression was 13 months following surgery, and the patient died of his disease at 22 months despite the use of sunitinib after diagnosis of metastatic disease. Cancer-specific survival was 87.5%, and overall survival was 62.5% during the study period.

Discussion

These cases represent the largest series to date of partial or hemi-nephrectomy for renal malignancy in patients with horseshoe kidney and highlight that reasonable outcomes can be achieved despite the increased complexity of the operation. While the majority of patients in our series underwent open surgery, hand-assisted laparoscopic,¹⁰ pure laparoscopic,^{67,11} and retroperitoneoscopic^{12,13} approaches have been described.

Much of the technical complexity of partial nephrectomy in this patient population is due to the abnormal renal location and vascular anatomy. Approximately 95% of cases involve fusion of the lower pole, which prevents independent rotation and results in an anteromedial location of the renal pelvicies. Consequently, the ureters frequently course anterior to the isthmus.¹⁴ In this series, we frequently noted increased techincal complexity that resulted from this abberent renal anatomy. Specifically, reduced renal mobilization makes accessing posterior renal masses more difficult. In addition, access to lower pole tumors necessitated careful dissection of the anterior renal pelvis and ureter. To overcome the issue of reduced mobilization, others have suggested that posteriorly located tumors might best be approached by a retroperitoneoscopic approach.^{12,13}

The highly variable blood supply to a horseshoe kidney also increases the complexity of partial nephrectomy in these patients. A single renal artery to each moiety is estimated to occur in only 30% of cases.¹⁴ The renal isthmus will frequently have its own blood supply, with branches from the aorta, renal arteries, inferior mesenteric artery, and/or external iliac arteries. In our experience, arteries may arise anteriorly from the aorta either above or below the isthmus and brach to supply both moities. Thus, care must be taken if clamping to ensure that blood supply to the contralateral moiety is not affected. In our series, preoperative imaging including CT with IV contrast, CT angiography, and invasive angiography were found to be useful adjuncts to delineate renal vascular anatomy. We agree with the recommendation of others that CT angiography, prefeably with 3D volume rendering, is noninvasive and optimally defines the arterial supply and allows for appropriate preoperative planning.

Despite the increased complexity, all patients in this series underwent successful completion of tumor resection with negative margins and acceptable perioperative outcomes in terms of length of stay, transfusion requirement, and decline in renal function. As previously mentioned, the only identified surgical complication was following surgical resection of a 7.5 cm mass in a functionally-solitary renal moiety. In this case, urine leak resolved 6 weeks following percutaneous drainage and ureteral stent insertion.

While this study is limited by its retrospective nature and small sample size, we suspect that our methodology allowed us to identify a complete and consecutive series of pateints at our institution. Intraoperative blood loss was intended to be a study measure, but could not be consistently obtained due to insufficient documentation in older cases. Additionally, postoperative eGFR would ideally have been measured at a 3 or 6 month period, however inconsistency in follow up testing precluded this measure. Despite these limitations, our series supports the growing evidence that partial nephrectomy is feasible for the management of renal malignancy in patients with a horseshoe kidney and is associated with reasonable oncological outcomes.

Conclusion

Our case series demonstrate that surgical management of renal malignancy in horseshoe kidneys can be safely performed with acceptable perioperative and oncologic outcomes and emphasizes the technical complexity of partial nephrectomy in this patient population. $\hfill \Box$

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