Malignancy in horseshoe kidneys: review and discussion of surgical implications

L. Fazio, MD, H. Razvi, MD, J. L. Chin, MD

Department of Urology, University of Western Ontario, London, Ontario, Canada

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Objective: *Horseshoe kidney is one of the most common congenital anomalies of renal structure. Not infrequently, surgical management of both benign and malignant disorders is required in patients with horseshoe kidney due to the susceptibility to certain conditions. The literature suggests a greater proclivity to certain renal tumors with this anomaly. We present three cases of malignancy in horseshoe kidneys. The unique technical challenges presented by these cases and the surgical approaches are discussed.*

Methods: *Three patients with tumors involving horseshoe kidneys are reviewed and their management discussed.*

Results: *Two patients were found to have renal cell carcinoma (RCC) and one had transitional carcinoma (TCC). Computed tomography (CT) and angiography were used in the work-up and preoperative planning of these cases. One patient with RCC received pre-operative renal artery embolization. Partial nephrectomy was performed in each patient with an aim at early vascular control of the tumors, identification of the collecting systems and ureters, as well as ensuring a 1 cm surgical margin. No patient required dialysis post-operatively. One patient died in the early post-operative period of a myocardial infarction; one patient developed brain metastases 18 months post-operatively, received palliative radiation and is alive 42 months after surgery; the other patient was free of disease for approximately 36 months but recently developed osseous metastases to her pelvis.*

Conclusions: *Techniques developed for partial nephrectomy may be used in the treatment of tumors in horseshoe kidneys. Survival is related to the grade and stage of disease.*

Key Words: horseshoe kidney, renal cell carcinoma, transitional cell carcinoma, surgical technique

Introduction

Horseshoe kidney (HK) is the most common abnormality of renal fusion with a reported incidence between 1/400 and 1/1800.[1-5] While the majority of patients with a horseshoe kidney are asymptomatic, the anomaly is associated with a heightened predisposition to several benign and malignant conditions. It is generally believed, that the overall risk of malignancy in HK is not increased.[4] However, the distribution of tumors involving HK varies when compared to malignancy in normal kidneys with a greater proportion of Wilms' tumor (WT) and transitional cell carcinoma (TCC) occurring in HK.[4,6,7]

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Address correspondence to Hassan Razvi, MD, St. Joseph's Health Centre, 268 Grosvenor Street, London, Ontario N6A 4V2 Canada

There is also an increased risk of primary renal carcinoid in this population.[2,8] While there is no increased risk of renal cell carcinoma (RCC) it remains the most common tumor of HK.[4] Due to their anatomy and frequently anomalous blood supply, surgical management of these tumors presents a special technical challenge. The following are three cases of malignancy in HK recently treated at our institution, with an emphasis placed on their operative management.

Methods

We present three cases of malignancy in horseshoe kidneys treated at our institution within the last three and a half years. The unique technical issues presented by these cases and the surgical approaches are discussed.

Results

Case one

A previously healthy, 51-year-old female presented with one episode of gross hematuria, diminished appetite and a 2 kg weight loss over a 3-month period. She denied flank or abdominal pain. On physical examination there was slight abdominal distention and a firm, non-pulsatile mass was palpable just beneath the umbilicus. Laboratory investigation revealed a hemoglobin of 84 g/L (normal range: 120 g/L-160 g/L) and serum creatinine was normal. Abdominal ultrasound suggested horseshoe kidney with a solid mass arising from the right moiety. Computerized tomography (CT) scan of the abdomen and pelvis showed an 8 cm inhomogeneously enhancing mass with irregular lobular outlines arising from the right component of the horseshoe system extending into the isthmus Figure 1. Renal angiogram revealed a single artery supplying the left renal moiety, a single artery supplying the majority of the right moiety and an artery draped over the renal mass Figure 2. All these vessels appeared to emanate from the aorta directly. Chest X-ray and bone scan were negative. The patient was counseled on the potential need for post-operative dialysis and was seen by the nephrology service pre-operatively.

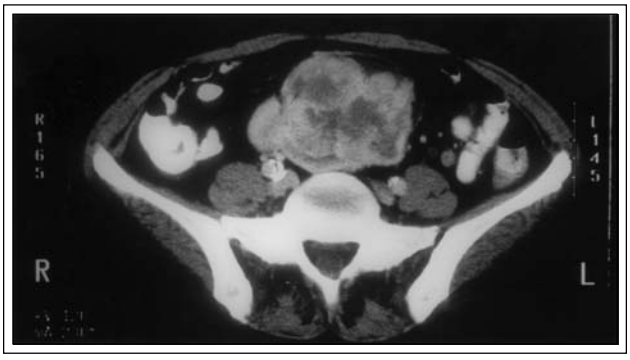

Figure 1. CT scan demonstrating 8 cm mass extending into isthmus of horseshoe kidney.

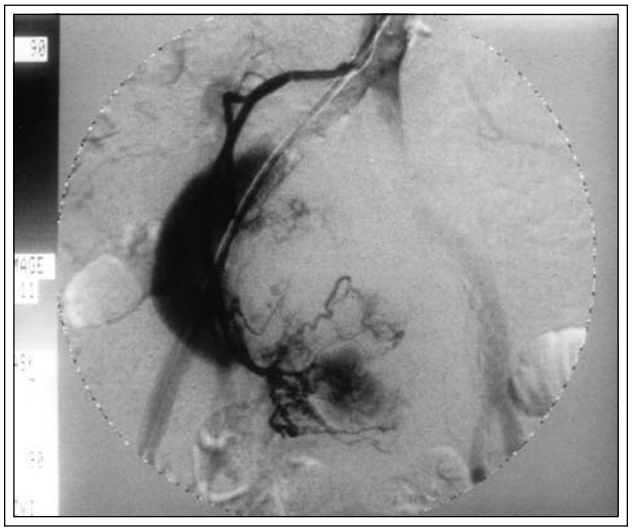

Figure 2. Artery draped over renal mass in case one into isthmus of horseshoe kidney.

The patient underwent right radical nephrectomy via a lower midline transperitoneal incision. The ureters were isolated. The isthmus was parenchymal of substantial thickness. It appeared technically feasible to control only the renal arteries to the tumor-containing right moiety and avoid clamping the left renal artery. The isthmus was divided on the left side with a 2 cm margin from the tumor mass. The collecting system was not entered. Large parenchymal vessels were oversewn with 3-0 chromic suture. The cut surface of the isthmus was re-approximated with 2-0 chromic vertical mattress sutures which were tied down over gelfoam plegets. The patient had an uneventful postoperative course. Dialysis was not required and the serum creatinine at discharge was 80 μmol/L (normal range 0 μmol/L-110 μmol/L). Histopathologic exam revealed a stage T3a grade III/IV clear cell variant of renal cell carcinoma. Resection margins were negative for malignancy. Eighteen months into her post-operative course she presented with headaches; CT scan of the head revealed metastatic disease involving the cerebellum. The patient responded to palliative

radiation. Subsequently she has been found to have pulmonary metastases and required further radiotherapy for her cerebral lesions. She is currently alive at 42 months follow-up.

Case two

A previously healthy, 72-year old woman presented with increased abdominal girth and abdominal discomfort. Abdominal ultrasound and CT scan showed a retroperitoneal mass Figure 3. A provisional diagnosis of sarcoma or lymphoma was made and she underwent laparotomy at another institution. Intraoperatively it was discovered that the mass involved the isthmus of a horseshoe kidney and most likely represented a renal cell carcinoma. Needle biopsy was performed only to confirm the diagnosis pending further investigations. Subsequent angiography demonstrated a separate artery supplying each of the right moiety, the left moiety and to the mass itself Figure 4. With respect to pre-operative considerations, vascular control was an issue. Special consideration was also given to the location of the tumor which meant controlling both moieties and possible involvement of the collecting system in the dissection.

With the vascular arrangement amenable to selective embolization, the mass was embolized via the percutaneous femoral arterial route 24 hours prior to repeat laparotomy. Cystoscopy and bilateral ureteral stent insertion was performed immediately prior to resection of her tumor with bilateral partial nephrectomies. The tumor, as expected, was supplied by a single vein and artery which were ligated. On the left side, the renal pelvis was noted to extend into the isthmus. The left renal pelvis was divided at the ureteropelvic junction (UPJ). The isthmus was isolated by clamping the normal parenchyma on each side with large Satinsky clamps and then transecting the parenchyma. The renal pelvis that opened on the left side was closed with a running 4-0 dexon suture. The cut edges of parenchyma were closed over with 2-0 chromic mattress sutures buttressed with autologous fat. Histopathologic examination revealed a stage T1, grade II/IV clear cell variant of renal cell carcinoma with negative resection margins. The patient had an uneventful postoperative course. She was disease-free up until 36 months follow-up. Recently, however, osseous metastases involving her pelvis have been discovered.

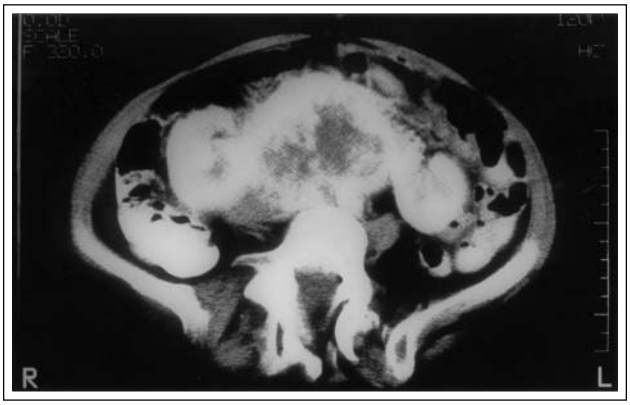

Figure 3. Retroperitoneal mass seen on CT scan in case two.

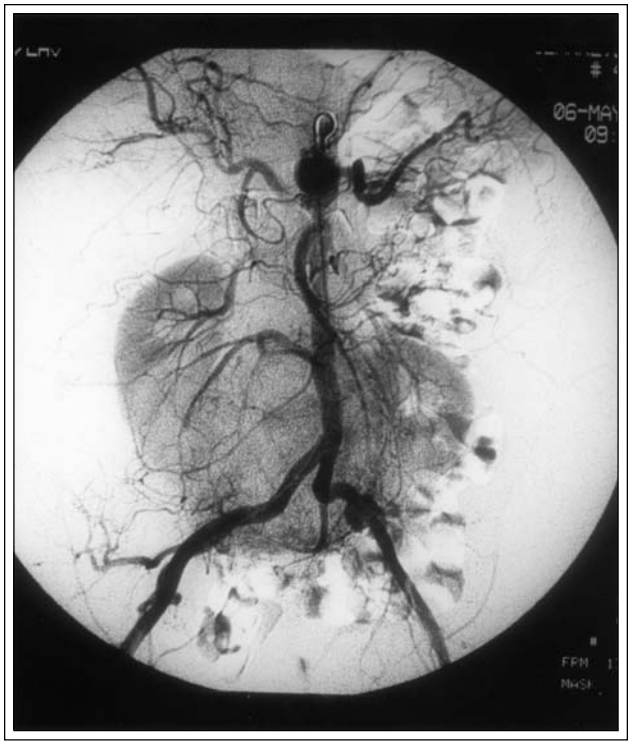

Figure 4. Arterial supply of tumor in case two.

Case three

A 74-year old man presented with painless gross hematuria. Past medical history was significant for myocardial infarction 3 years earlier, angina, diabetes, and a prior cerebrovascular accident. Intravenous pyelography (IVP) showed a horseshoe kidney with a calyceal diverticulum and a filling defect consistent with transitional cell carcinoma (TCC) in the upper pole of the left moiety Figure 5. IVP findings were confirmed on CT scan which also showed an isthmus with substantial parenchyma. The patient's serum creatinine was 120 μmol/L (normal range 62 μmol/L-120 μmol/L). CT scan of the chest and bone scan were negative. The main preoperative consideration was vascular control of

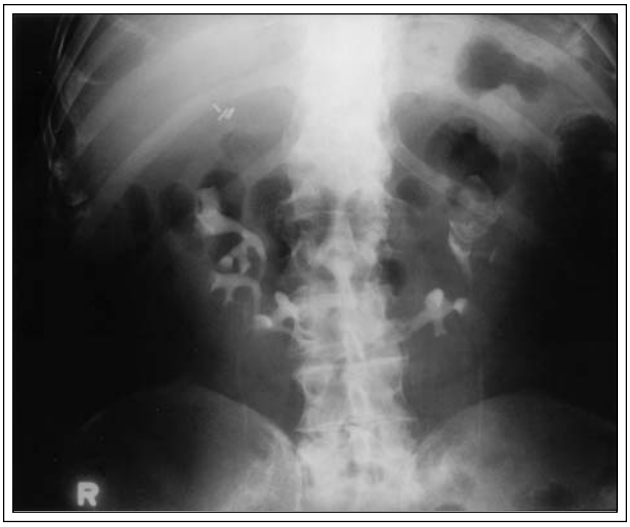

Figure 5. TCC involving upper pole of left moiety of horseshoe kidney.

the tumor. With the likely diagnosis of TCC, consideration had to be given to performing distal ureterectomy as well as dividing the isthmus and the related collecting system. Due to his significant comorbidities, preoperative internal medicine consultation was sought and despite significantly increased surgical risks the patient and family decided to proceed with surgery.

The patient underwent a left radical nephrectomy with distal ureterectomy and closure of the isthmus via a flank incision. The isthmus was noted to be 5 cm thick in its smallest diameter and was controlled with a large Satinsky clamp; it was transected and the cut surface was oversewn with 2-0 chromic sutures. The ureter was dissected down to its most distal portion and removed with a cuff of bladder. Unfortunately the patient suffered a massive myocardial infarct a few hours into his postoperative course. He did not recover from cardiogenic shock and died 5 days later. Histopathologic examination revealed a stage Ta, grade II/III transitional cell carcinoma involving the renal pelvis and proximal ureter of the left moieity with negative resection margins.

Discussion

The embryology of HK involves several theories. The abnormality appears to originate at approximately the fourth week of gestation,[9] prior to renal ascent. The theory of fusion dictates that a union occurs via median fusion of metanephric tissues; however this is likely applicable only for those cases of HK in which there is a fibrous isthmus.[1,10] For the 95% of cases in which there is a parenchymal isthmus,[7] it is postulated that the HK results from a teratogenic event:[2] the abnormal migration of cells from the posterior nephrogenic area. The horseshoe shape results as the embryo's trunk grows and the kidney is pulled cranially while the isthmus is restrained by the inferior mesenteric artery. This may also account for the frequently anomalous blood supply of these kidneys.[1,10] The abnormal anatomy not infrequently results in sub-optimal urinary drainage, leading to urinary stasis, calculi, recurrent urinary tract infections and chronic UPJ obstruction.[1,3,4,9,11-14]

An examination of the relative incidences of malignancy in normal and horseshoe kidneys Table 1 reveals the disparity between specific types of tumors; most notably the larger proportion of transitional cell carcinoma (TCC) and Wilms' tumor (WT) in HK. Renal cell carcinoma (RCC) is the most common tumor in both normal and horseshoe kidneys. Its incidence in HK is no greater than in the normal population.[4] The risk of TCC is three to four times greater in HK than in normal kidneys.[1,2] This increased risk is thought to be secondary to chronic urinary stasis, prolonging contact time between urinary carcinogens and urothelium, as well as the chronic noxious stimulation associated with infection and calculi.[7,15,16]

The incidence of WT is also increased in HK;[1-3,17]; the relative risk being 1.76-7.93, depending on the incidence of HK used in the calculation.[3] Since WT results from an abnormal proliferation of the metanephric blastema, it is possible that the teratogenic event that results in HK, i.e. the abnormal migration of metanephric tissues, may predispose to WT.[3] The relative risk of primary renal carcinoid in

TABLE 1.[6,7]

Tumor	Normal Kidneys	Horseshoe Kidneys
Renal cell carcinoma	83%	49.5%
Transitional cell carcinoma	7.7%	19.8%
Wilms tumor	5.6%	22.5%
Other (i.e. sarcoma)	3.0%	8.1%

HK is 82,[8] although this is an extremely rare tumor, with only 39 cases having ever been reported (seven have occurred in HK).

The presentation of malignancy in HK is similar to that in normal kidneys. However it can be difficult to image, as in our case two, in which a HK was only discovered intraoperatively, prior to which a non-contrast abdominal CT had been performed. In their 1985 study, the National Wilms' Tumor Group reported making a correct pre-operative diagnosis of HK only 46% of the time.[3] Furthermore, the isthmus of the HK may not be recognized due to the presence of abundant tumor.[17] The work-up is similar, involving standard imaging techniques such as abdominal CT scan and chest X-ray.[1,6,8,18,19] However, angiography is also recommended due to the presence of aberrant vasculature in 70% of cases.[3,6,9,19] Moreover, the blood supply to the kidney may depend entirely on the vessels to the isthmus.[13] If it is deemed possible that the entire renal blood supply will have to be controlled, with the ensuing risk of post-operative renal failure, a pre-operative consultation and advice regarding possible dialysis should be undertaken.

In terms of surgical issues, a midline transperitoneal approach provides the best exposure.[4,9,19] Control of the blood supply to the area to be resected is important considering the frequently anomalous vascular supply and the fact that the normal renal parenchyma may depend heavily on the tumor's blood supply. Selective ischemia to preserve the remaining renal tissue is an important principle of surgery in these patients.[1] If feasible, selective embolization of the tumor bearing area in the isthmus would be advisable. Regional hypothermia may be a strategy to preserve renal function while vessels are being controlled. Another consideration is the paucity of perinephric fat and poorly demarcated Gerota's fascia in these ectopically located kidneys which, theoretically, represents an increased risk of extracapsular invasion and local recurrence. In light of this, it is interesting to note, that although their resection margins were negative, both patients who have suffered recurrences had tumor involvement of the isthmus, where Gerota's fascia and perinephric fat would be thinnest.

Dissection of the isthmus is essential in providing access to regional lymph nodes and normalizing the course of the ureter.[1] An isthmectomy may be feasible if the tumor is confined to the isthmus,[9] as was performed in case two. Determining the extent of the collecting system is another important issue. This includes such aspects as pre-operative stenting of the ureters to fascilitate intraoperative identification; extension of the collecting system into the isthmus; and closure of the collecting system. When dealing with TCC, excision of the entire ureter obviously is advisable. Thus, preoperative stenting may fascilitate identification and dissection of a ureter with a possibly unpredictable and unusual course. Special attention has to be given to adequate resection margins in the case of TCC in one moiety of the renal pelvis if the pelvis extends into the isthmus. In order to minimize the risk of bleeding and urinary fistula formation, it is advisable to cover the exposed edge of resected parenchyma,[1,9,19] with either a peritoneal patch or with gelfoam soaked in topical thrombin.

There is little information about the long-term survival of these patients. RCC, TCC and WT survival appears to be independent of the presence of HK and is determined by grade and stage of disease.[12,20] On the other hand, primary renal carcinoid occurring in HK appears to follow a less aggressive course.[2,8]

Conclusions

In summary, there is an increased incidence of TCC, WT and primary renal carcinoid in HK. Pre-operative angiography is usually helpful in delineating aberrant blood supply and control of this blood supply is pivotal in excising tumors from HK and preserving renal function in the remaining healthy parenchyma. Other key surgical issues include dissection of the isthmus and coverage of the exposed, cut edge of remaining parenchyma. Employing the techniques developed for partial nephrectomy in morphologically normal kidneys, preservation of renal function and adequate surgical margins can be achieved in patients with tumor in horseshoe kidneys. There is, however, a theoretically increased risk of extracapsular extension due to a paucity of perinephric fat of horseshoe kidneys. Finally, the prognosis is related to the grade and stage of disease, and does not appear to be negatively influenced by the presence of a horseshoe anomaly. □

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