Malignant Tumor in a Horseshoe Kidney

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Keywords: horseshoe kidney, kidney neoplasms, renal cell carcinoma, diagnosis

INTRODUCTION

Horseshoe kidney, first recognized at autopsy by DeCarpi in 1521, is a renal fusion anomaly found in about 0.25% of the population. Malignancies are associated with this anomaly and adenocarcinoma comprises about 50% of tumors arising in the horseshoe kidney. We report a case of renal cell carcinoma in a 66-year-old man and depict our experience in its management.

CASE REPORT

A 66-year-old Caucasian man presented with intermittent epigastric discomfort of several days’ duration. Review of systems was negative for cardiac or pulmonary symptoms and the patient denied any unexplained weight loss. He was afebrile and physical examination revealed no abnormalities. Basic hematological and biochemical investigations were unremarkable. Urinalysis, urine cytology, and urine cultures were negative. The patient’s Eastern Cooperative Oncology Group performance status was zero. Computed tomography (CT) of the abdomen and pelvis revealed benign hepatic cysts, a benign lesion in the pancreas, and a horseshoe kidney. To the right of the isthmus was a 5.4 × 4.4-cm enhancing mass. It measured 24 HU in the noncontrast phase, 75 HU in the corticomedullary phase, and 96 HU in the excretory phase. The CT angiography demonstrated unremarkable celiac axis and superior mesenteric artery origins and single renal arteries to the left and right portions of the kidney. Of note, the inferior mesenteric artery draped over the left side of the lesion and the right ureter passed close to its right side (Figures 1 and 2).

Figure 1. Corticomedullary phase of renal dedicated CT reconstruction. Asterisk indicates mass; solid white arrow, inferior mesenteric artery; and broken white arrow, right ureter.

Figure 2. Computed tomography reconstruction. Asterisk indicates mass and black arrow, inferior mesenteric artery.
After appropriate preoperative testing was completed, the patient was taken to the operating room for a right open partial nephrectomy of the right renal unit of the horseshoe kidney. A double-J right ureteral stent was placed cystoscopically. A midline abdominal incision was then made, the posterior peritoneum was incised from the ileocecal junction to the ligament of Treitz, and the horseshoe kidney was exposed. The right ureter was dissected off the mass and identified with a vessel loop. The right renal hilum was exposed, umbilical tapes were placed around the right renal artery and vein, and a Rumel tourniquet was prepared for the artery. The inferior mesenteric artery was dissected free of the mass as well (Figure 3). Intravenous mannitol was given and the kidney was immersed in ice slush. The renal capsule around the mass was scored with Bovie electrocautery and the tumor was then removed with several millimeters of normal parenchyma, while the right renal artery was occluded. The defect was closed over a Surgicel bolster with 0-0 chromic and fat bolsters. Excellent hemostasis was obtained. The total ischemic time was 13 minutes. A closed suction drain was placed and the wound was closed after copious irrigation. The patient was discharged home after a speedy recovery with a normal serum creatinine level, adequate pain control, and good ambulatory status.

Pathology revealed a grade 4 clear cell renal cell carcinoma with sarcomatoid features. Vascular invasion was present, but the surgical margins were negative for malignancy (Figure 4). The patient continued under diligent surveillance, and to date has shown no evidence of disease recurrence.

DISCUSSION

Horseshoe kidney is a renal fusion anomaly found in about 0.25% of the population. Virtually, every urologic disease process has been described in the horseshoe kidney, including malignancy. Adenocarcinoma comprises about 50% of tumors arising in the horseshoe kidney, followed by transitional cell carcinoma and Wilms tumor. While prognosis is dependant on the same factors as in nonfused kidneys, careful attention must be paid to highly variable vascular supply and anomalies of the collecting system when planning surgical intervention.

In a review of the literature up to 1998, Rubio Briones reported 144 cases of tumorous pathology in horseshoe kidney. We searched the literature and found 43 other cases by July 2006, 22 of which were RCC. Transitional cell carcinoma accounts for 28% to 40% of these malignancies, with an increased incidence over the general population, possibly related to an increase in calculus pathology, frequent urinary obstruction, and chronic infection leading to prolonged exposure to carcinogens in horseshoe kidneys. The incidence of Wilms tumor is twice as that expected in the general population, perhaps related to abnormal migration of nephrogenic cells which form the isthmus and subsequently undergo malignant changes. As mentioned, renal cell carcinoma is the most common neoplasm described in horseshoe kidneys, accounting for about 50% of...
cases, and occurs no more often than in the general population. Prognosis is unaffected by the anomaly and is dependant on tumor pathology and stage at diagnosis just as in normal kidneys. 

CONFLICT OF INTEREST
None declared.

REFERENCES