CASE REPORT

Renal Cell Carcinoma in a Horseshoe Kidney

Luis Antonio R. Balajadia, MD; Michael Jonathan R. Latayan, MD, FPUA and Al-Widzrin M. Jupli, MD

Section of Urology, Department of Surgery, Southern Philippines Medical Center, Davao City

Horseshoe kidney is a rare form of congenital renal malformation. It occurs in 0.25 to 3% of the population and is usually asymptomatic. Occurrence of symptoms is usually related to infection, lithiasis, and rarely, malignancy. Presented is a case of a 62-year old male with a one-year history of occasional painless hematuria associated with epigastric discomfort. On physical examination, a palpable mass was noted on the right periumbilical area, 10cm x 10cm in size and was non-tender. CT-scan with IV contrast showed 10cm x 7cm x 12cm mass on the right side of a horseshoe kidney. He underwent Radical Nephrectomy, right with Isthmusectomy. Post-operative course was unremarkable. Histopathology result showed Renal Cell Cacrcinoma, clear cell type.. Although malignancy was present in an anomalous kidney, the prognosis is the same as with normal kidneys. To date, this is the first reported case of malignancy on a horseshoe kidney in the Philippines.

Key words: Horseshoe kidney, renal cell carcinoma, renal fusion anomaly

Introduction

A Horseshoe Kidney (HK) is a rare developmental anomaly that occurs in about 0.25% of the population and arises during the 4th and 6th week of embryonic development after the ureteral bud has entered the renal blastema.[1] In this anomaly, the poles of the 2 kidneys are fused, usually the lower poles. The polar fusion results in an isthmus of tissue (parenchymal or fibrous) between the 2 kidneys. The kidneys fuse in the pelvic cavity prior to ascending that leads to a large U-shaped kidney which is unable to ascend to the level of L1 because its ascent is arrested by the origin of the inferior mesenteric artery from the aorta at the level of L3 vertebra. This leaves the kidney in the hypogastric region.[8]

Alongside with the abnormality in its position, the horseshoe kidney calyces are normal in number but atypical in orientation as well as the ureter which may insert high on the renal pelvis and lie laterally. Its blood supply can be variable; there are frequently two or three arteries that supply each kidney from aorta and one or two arteries that supply the isthmus.[3]

Individuals having a horseshoe kidney are mostly asymptomatic but at risk to urinary stasis, infection, nephrolithiasis, and rarely malignancies. Fifty percent of the malignancies associated with this anomaly consist of adenocarcinoma.[6] Reported is a case of a renal cell carcinoma, clear cell type, in a horseshoe kidney of a 62-year-old male and its management.

The Case

A 63-year-old male, hypertensive but non-diabetic with no previous surgery, presented with a one-year history of occasional painless hematuria associated with intermittent epigastric discomfort one month prior to consult. There was no associated fever, dysuria nor weight loss. On physical examination, a palpable mass was noted on the right periumbilical area, 10cm x 10cm in size and was non-tender. Routine laboratory examinations were all normal except for the microscopic hematuria on urinalysis. Computed Tomography (CT) scan with IV contrast of the whole abdomen and pelvis showed an ill-defined heterogenous enhancing mass with areas of hypodense necrosis and calcific densities occupying the whole right kidney, measuring about 10cm x 7cm x 12cm. The tumor already extends beyond the Gerota's fascia and there is no thrombus noted on the vena cava and right renal vein. An enhancing renal parenchyma (Isthmus) connects the left and right kidney in the midline. (Figures 1 & 2).

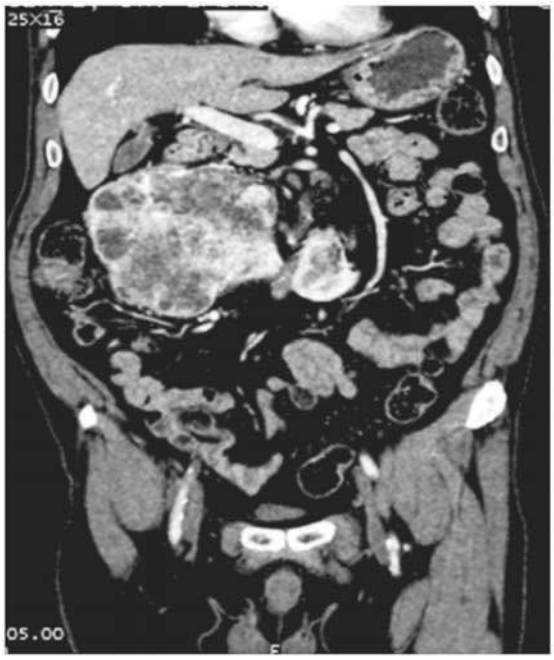

Figure 1. Contrast enhanced CT scan of the abdomen showing a horseshoe kidney, with a large mass on the right kidney displacing the surrounding tissue (Coronal view).

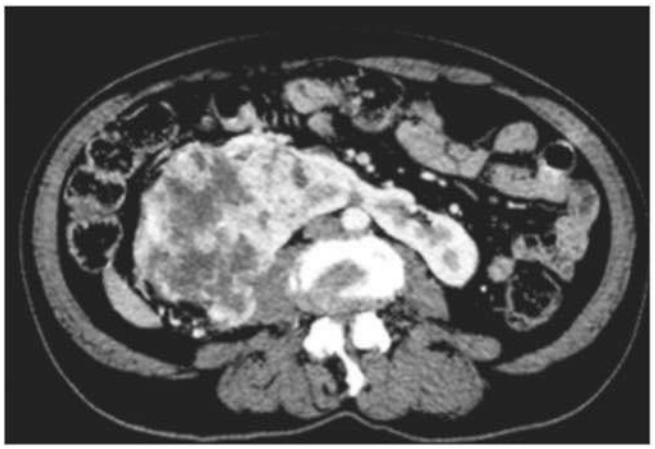

Figure 2. Cross section contrast enhanced CT scan of the abdomen, showing horseshoe kidney with right kidney mass anterior to the great vessels.

Presented with the salient features of the case, a malignant renal mass, most likely a renal cell carcinoma, was entertained. Differential diagnoses included pyelonephritis, angiomyolipoma, oncocytoma, and Wilms tumor. Acute or chronic pyelonephritis was considered because of the symptom of intermittent epigastric discomfort and hematuria but was ruled out because of the absence of fever and painful urination. Other renal tumors such as angiomyolipoma, oncocytoma and Wilms tumor were ruled in due to the presence of renal mass. Angiomyolipoma was later ruled out because of the absence of fat within the lesion visualized thru CT-scan. Wilms tumor was also ruled out because this entity usually presents during childhood. However, oncocytoma could not be ruled out because it mimis renal cell carcinoma on radiographic imaging and needs confirmation by biopsy.

After appropriate preoperative testing was completed and with a provisional diagnosis of renal cell carcinoma in a horseshoe kidney, the patient was scheduled for radical nephrectomy, right with isthmusectomy. A transperitoneal approach was done through a Chevron incision. The transverse colon was retracted inferiorly to expose the retroperitoneum. Intraoperatively, the tumor was noted on the right side of the kidney but there was no gross involvement of the isthmus and no palpable lymph nodes (Figure 3). After full

mobilization of the right side and proper identification and ligation of the right renal vessels and ureter (Figures 4 & 5), the isthmus was divided using electrocautery. Repair of the left side of the kidney was done using absorbable sutures ensuring no urine leak and good hemostasis. The procedure ended after placement of a Jackson Prat (JP) drain.

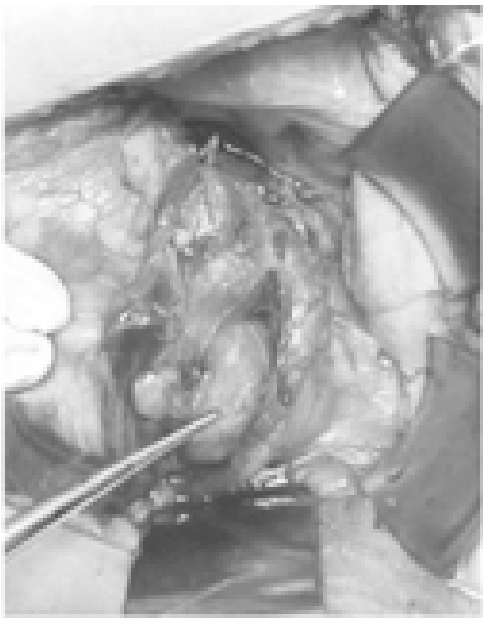

Figure 3. The isthmus with no nodes noted and right kidney fully mobilized with vascular variation.

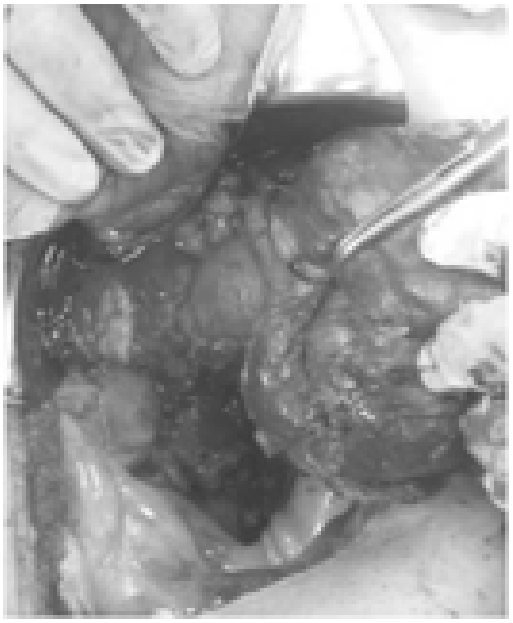

Figure 4. An intraoperative picture of the right kidney and the isthmus prior to division.

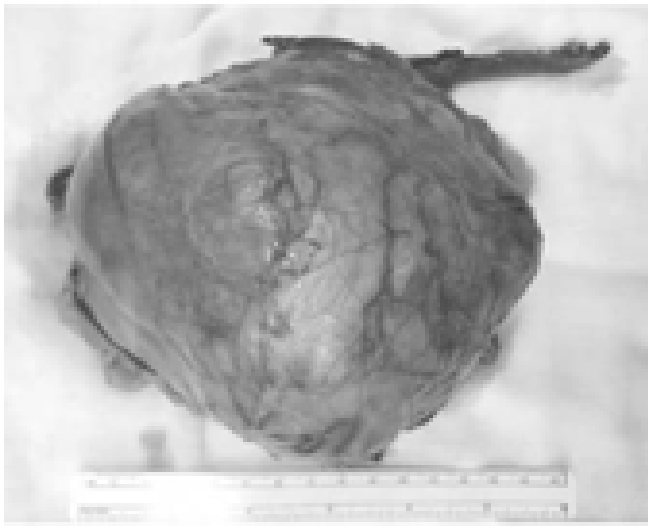

Figure 5. Gross appearance of the right mass involving the entire kidney.

Grossly, the specimen was 13cm x 7cm x 10cm in dimension and the tumor already extended towards the renal pelvis on cut section (Figure 6). Microscopic examination showed large cells with clear cytoplasmic appearance and sharply outlined boundaries consistent with renal cell carcinoma, clear cell type (Figure 7). The resection margin, renal vein, renal artery and ureter were all negative for invasion.

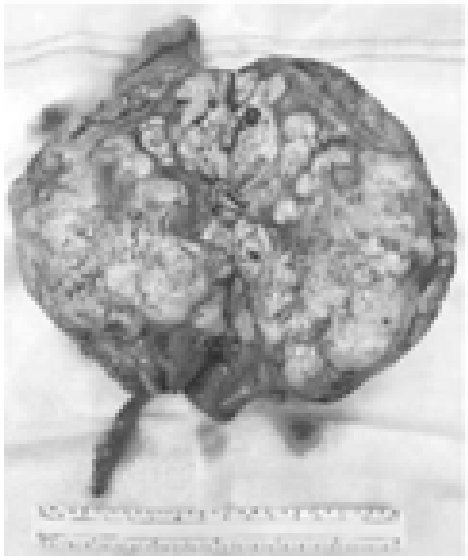

Figure 6. Cut-section, shows a large, solid, golden-yellow tumor, sharply separated from the surrounding tissues by a fibrous pseudocapsule. Note that the tumor already extends into the pelvis.

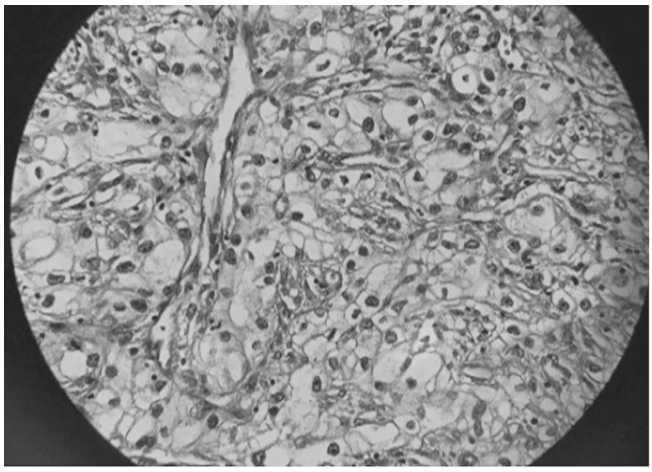

Figure 7. A diffuse pattern of growth with entrapment of glomerulus and an optically-clear cytoplasm and sharply outlined cell membrane.

Post-operatively, the patient was stable with unremarkable course in the ward. The patient was sent home on the 5th post operative day and was advised for surveillance CT scan after 6 months for possible recurrence on the contralateral side.

Discussion

The horseshoe kidney (HK) is an abnormal congenital fusion of the urinary tract, which combines three anatomic abnormalities such as the ectopia, malrotation, and vascular changes. The anomaly occurs in about 0.25% of the population. In most cases, the abnormality consists of two renal masses fused at their lower poles by a parenchymal or fibrous isthmus. In rare cases, the isthmus connects the upper poles and an inverted HK is created.[9] It was found out that the anomaly is at risk for infection, lithiasis, and rarely, malignancy.

As previously mentioned, adenocarcinoma comprises of about 50% of tumors arising in the horseshoe kidney, followed by transitional cell carcinoma and Wilms tumor.[6] Rubio Briones in 1998 reported 144 cases of tumorous pathology in horseshoe kidney, and another 43 cases were added by July 2006, of which 22 were renal cell carcinoma (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.[6] Urothelial carcinoma of the renal pelvis in a horseshoe kidney is relatively rare. In Japan, a report describes 35 cases of urothelial cell carcinoma presenting with hydronephrosis. This should alert clinicians of the possibility of malignancy is the presence of hydronephrosis on a horseshoe kidney.[14] The incidence of Wilms tumor was noted twice higher than was expected in the general population, perhaps related to abnormal migration of nephrogenic cells which form the isthmus and subsequently undergo malignant changes.[6] Furthermore, the National Wilms Tumor Study Group detected horseshoe kidney in 41 out of 8617 patients from 1969 to 1998 and reported the incidence as 0.48%.[10] Carcinoid tumor and sarcoma of the horseshoe kidney have also been reported, although occurring less frequently.

Although no genetic and racial determination is known, horseshoe kidney has been reported in identical twins and in siblings within the same family. The horseshoe kidney is found with even higher frequency in infants and children after autopsy examination, suggesting a higher risk of mortality possibly attributed to the severity of the associated abnormalities.[9] The tumor usually manifests itself by involving the isthmus even though it may be localized in any region of the kidney. In this case, the tumor involved the right side of the fused kidney. Bilateral tumors of the horseshoe kidney are quite rare as described in literatures and often pose health and therapeutic challenges.[11]

Majority of the cases of this anomaly were asymptomatic in presentation. If clinical manifestation does occur, it is usually attributed to the associated anomalies like hydronephrosis, infection, lithiasis and even tumor, with most patients being diagnosed incidentally by ultrasound, intravenous pyelography (IVP), or CT scan. Most authors in literature recommend a contrast enhanced CT scan, which is essential in the diagnosis and management. According to Jhobta, et al. 2003, pre-operative arteriography

will help in identifying any vascular anomalies that usually come with these cases but is not routinely recommended.

As always, success to the surgical procedure in this kind of case will rely on meticulous pre-operative planning. Either a midline abdominal incision or a subcostal incision can be done. The advantage of a midline and subcostal incision is that they provide adequate access to the entire horseshoe kidney and better vascular control. There is no role in doing a flank incision because horseshoe kidneys are located in the midline. In the present case, the attending physician opted for a bilateral subcostal incision (Chevron) due to the high position of the mass (epigastric), which provided adequate exposure of isthmus. Surgery will then proceed by careful isolation and ligation of the vessels. The isthmus, as a rule, has to be divided during the removal of the tumor from the horseshoe kidney. Not only does division of the isthmus provide access to the draining lymph nodes, it also helps normalize the course of the remaining ureter, thereby minimizing the potential complications.[1] In cases of transitional cell carcinoma of the pelvis, radical nephroureterectomy with excision of bladder cuff still remains the gold standard management, which can be approached thru open or laparoscopic surgery. The detection of the tumor in the bladder during cystoscopy prior to the resection would even worsen the prognosis and increase the risk of developing transitional epithelial cancer in the contralateral upper urinary tract. Hence, a very strict follow-up is required via cystoscopy every 3 months and computed tomography every 6 months since recurrence is highly anticipated.[10] For children with Wilms tumor in a horseshoe kidney, management is the same with that of a normal kidney, with chemotherapy and radiotherapy providing crucial role in its treatment.

The presence of horseshoe kidney does not alter prognosis of a malignant tumor arising in such a kidney. The problem, however, comes with the management that arise during and after surgery because of its anatomic variation. Therefore, prognosis is not affected by the anomaly and is dependent on tumor pathology and stage at diagnosis just as in normal kidneys.

Conclusion

Although horseshoe kidney is the most common renal fusion anomaly and clear cell carcinoma being the most common subtype of renal cell carcinoma, a combination of the two is rare. Moreover, malignancy in a horseshoe presents a dilemma to a urologist on a surgical point of view, as it usually gives anatomic variation, mostly emphasizing on its blood supply. Prognosis is not altered by such an anomaly, but will depend on tumor pathology and stage.

References

1. Kavousi L, Partin A, et al. Campbell-Walsh Urology; 11th edition; Part XV: Pediatric Urology, Anomalies of form and fusion, Horseshoe kidney; pp. 2993-6.

2. Jhobta R, Bawa AS, Attri AK and Kaushik R. Adenocarcinoma in horseshoe kidney; Yonsei Med J 2003; 44 (4): 744-6.

3. Isobe H, Takashima H, Higashi N, Murakami Y, Fujita K, Hanazawa K, Fujime M and Matsumto T. Primary carcinoid tumor in a horseshoe kidney; Int J Urol 2000; 7: 184-8.

4. Naveena S, Mrudula C, et al. Horseshoe kidney: a case report. Int J Res Med Sci 2013; 1(3): 304-30.

5. Ram N, Behera B, Rathi S, Trivedi S, Dwivedi US. Malignancy of a horseshoe kidney: A case series with a rare presentation; Uro Int J UIJ 5 (6) http://dx.doi.org/10.3834/uij.1944-5784.2012.12.15

6. Yaber-Gómez EK, Cortes-Arcos Y, González-Ruiz FG, et al. Clear cell carcinoma in horseshoe kidney: a case report and literature review; Urology Service. 2 Pathology Service Centro Médico Nacional 20 de Noviembre ISSSTE, Mexico City; Rev Mex Urol 2010; 70(2): 111-5.

7. Jones L, Reeves M, Wingo S, Babanoury A. Malignant tumor in a horseshoe kidney. Urol J (Tehran). 2007; 4: 46-8.

8. Qingfu Zhang, Jian Ming, Siyang Zhang3 and Xueshan Qiu. Primary micro neuroendocrine tumor arising in ahorseshoe kidney with cyst: report of a case and review of literature. Diagn Pathol 2012; 7: 126; http://www.diagnosticpathology.org/content/7/1/126

9. Rao EV, Battula SR. Horseshoe kidney: a review article. Int J Res Med Sci 2015; 3(11): 3004-7.

10. Natsis K, Piagkou M,Skotsimara A, Protogerou V, Tsitouridis I, Skandalakis P. Horseshoe kidney: a review of anatomy andpathology; Department of Anatomy, Medical School, Aristotle University of Thessaloniki, PlateiaIppodromiou 17, 54621 Thessaloniki, Greece; Surg Radiol Anat 2014; 36: 517-26.

11. Karakaya Y, Türk H, Yoldas M, Kulan S. Horseshoe kidney and concomitant urinary collector system tumor. A case report. J Surg 2016; 2(4): 133-5.

12. Petrovic M, Andrejovic V, Djurasic L, Starnenkovic V. Tumors of the horseshoe kidney - characteristics and literature review. 2012; UDK 616.61-007:616-006.04

13. Lai A, Marwaha RK, Narshiham KL Yadav K. Wilms tumor arising in a horseshoe kidney. PGIMER, Chandigarh, India; 1995; 32.

14. Kumar T, Jiwane A, Bhusare D, Kutumble R, Kulkarni BK. Wilms tumor in a horseshoe kidney 2001; 6.

15. Isobe H, Shiozawa S, Horie S. Urothelial carcinoma of the renal pelvis and ureter associated with hydronephrosis in a patient with horseshoe kidney: A case report. Juntendo Med J 2015.