

**HORSESHOE KIDNEY AND RENAL CELL CARCINOMA: A RARE ENTITY AND REVIEW ARTICLE***Manish Gupta<sup>1</sup>, Bharat Khadav<sup>2</sup>, H. L. Gupta<sup>3</sup>, T. C. Sadasukhi<sup>4</sup>, Amrit Pal Singh Gil<sup>5</sup>*<sup>1</sup>Associate Professor, Department of Urology, Mahatma Gandhi Medical College and Hospital, Jaipur.<sup>2</sup>Senior Resident, Department of Urology, Mahatma Gandhi Medical College and Hospital, Jaipur.<sup>3</sup>Associate Professor, Department of Urology, Mahatma Gandhi Medical College and Hospital, Jaipur.<sup>4</sup>Professor, Department of Urology, Mahatma Gandhi Medical College and Hospital, Jaipur.<sup>5</sup>Senior Resident, Department of Urology, Mahatma Gandhi Medical College and Hospital, Jaipur.**ABSTRACT****BACKGROUND**

Horseshoe Kidney is the most common congenital developmental anomaly associated with the Kidney. Adding to the rarity is the occurrence of Renal Cell Carcinoma in such a kidney. The incidence of a RCC in a Horseshoe kidney has not been substantially documented and hence makes it a challenge and interesting surgical exercise. We shall be discussing such a case here at length.

**KEYWORDS**

Horseshoe Kidney, Renal Cell Carcinoma, Radical Nephrectomy of the Right Moiety.

**HOW TO CITE THIS ARTICLE:** Gupta M, Khadav B, Gupta HL, et al. Horseshoe kidney and renal cell carcinoma: a rare entity and review article. J. Evid. Based Med. Healthc. 2018; 5(31), 2340-2343. DOI: 10.18410/jebmh/2018/482

**BACKGROUND**

Horseshoe kidney was first recognized in 1521 by De Capri in an autopsy; it affects about 1 in 400 live births.<sup>1</sup> These patients are usually diagnosed only on imaging, as most of these patients are asymptomatic. Renal Cell Carcinoma arising in these kidneys is a rare entity. Renal cell cancer (RCC) constitutes a group of tumours that is highly heterogeneous with respect to morphology and clinical behaviour. RCC is the most common malignant tumor arising in the kidney and accounts for 2% of all new cancers diagnosed world-wide. The incidence of Renal Cell Carcinoma in a Horseshoe Kidney suffers conflicting reports as to whether its more prevalent in Horseshoe kidneys compared to normal kidneys.<sup>2,3</sup> A proper pre-operative evaluation is very essential to minimize the risk for any intra-operative bewildering scenarios.

**REVIEW OF LITERATURE**

Jacopo Berengario da Carpi first described it in 1522 during an autopsy, but Botallo, in 1564, presented the first extensive description and illustration of a horseshoe kidney.<sup>4</sup>

Embryologically, the abnormality occurs between the fourth and sixth week of gestation after the ureteric bud has entered the renal blastema. There are several variations in the basic shape of the horseshoe kidney. In 95% of cases, the kidneys join at the lower pole, which occurs before the kidneys have rotated on their long axes. In a small subset, an isthmus connects both upper poles.<sup>5</sup>

Horseshoe kidneys are, in themselves, asymptomatic and thus they are usually identified incidentally. They are however prone to a number of complications as a result of poor drainage, which may lead to clinical presentation. These complications include:

- Hydronephrosis, secondary to pelviureteric junction obstruction.
- Renal Calculi.
- Increased susceptibility to trauma.
- Infection and pyeloureteritis cystica.
- Increased incidence of malignancy.
  - Wilms tumour.
  - Transitional Cell Carcinoma (TCC).
  - Renal carcinoid.
- (Renovascular) hypertension.<sup>6</sup>

Renal cell carcinoma that originates in a horseshoe kidney is an unusual entity. In tumor-bearing horseshoe kidneys, preoperative knowledge of the localization, extent and vascular supply of the neoplasm is indispensable for performing a complete resection of the tumorous focus without sacrificing more of the functioning renal tissue than is necessary

It has been stated that the occurrence of renal cell carcinoma in horseshoe kidneys is no higher than in non-fused kidneys, but that the incidence of transitional cell carcinoma in horseshoe kidneys is higher, and this is conceivably due to the presence of chronic urinary tract infections.<sup>7</sup>

The blood supply to the horseshoe kidney can be quite variable. In 30% of the cases, it consists of one renal artery for each kidney, but the blood supply may be atypical, with duplicate or even triplicate renal arteries supplying one or both kidneys. The isthmus and adjacent parenchymal masses may receive a branch from each main renal artery, or they may have their own arterial supply originating from the aorta either above or below the level of the isthmus. Not infrequently, this area is supplied by branches from the

*Financial or Other, Competing Interest: None.*

*Submission 26-06-2018, Peer Review 30-06-2018,*

*Acceptance 20-07-2018, Published 30-07-2018.*

*Corresponding Author:*

*Dr. Bharat Khadav,*

*Senior Resident, Department of Urology,*

*Mahatma Gandhi Medical College and Hospital, Jaipur.*

*E-mail: bharatkhadav@gmail.com*

*DOI: 10.18410/jebmh/2018/482*



inferior mesenteric artery, the common or external iliac arteries, or the sacral arteries.<sup>8</sup>

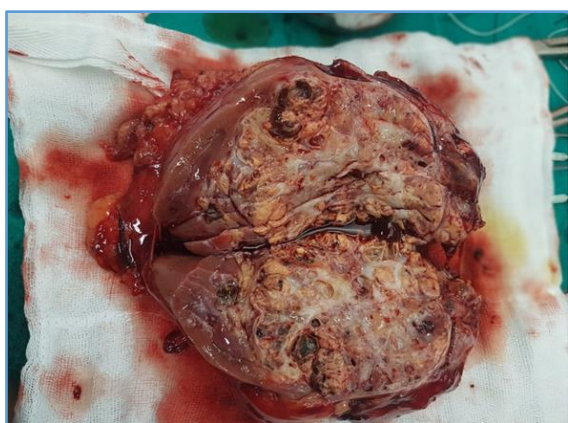
### Case History

A 54 years old male patient presented to the outpatient department with complaints of intermittent colicky right flank pain since 2 months, with sudden onset of haematuria since 5 days. There was no previous history of any medical or surgical illness. Basic biochemical and haematological investigations were negative.

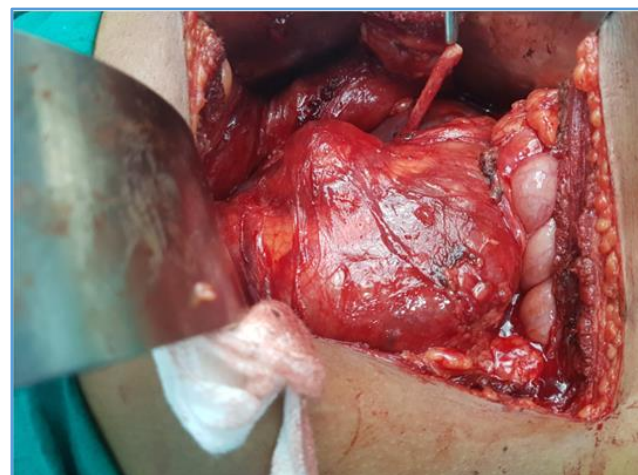
Ultrasound was suggestive of a Horseshoe Kidney with large 96\*98\*124 mm partially necrotic mass arising from right renal upper polar region. This warranted for further investigations and hence a Contrast enhanced CT with Renal Angiography was done that were confirmatory of a large ill-defined heterogeneous density lesion noted in upper and mid pole region of the right moiety of the Horseshoe Kidney. The lesion had intense heterogenous enhancement in arterial phase, which persisted in venous phase. It also indicated towards the exophytic component extending into the perinephric fat, with obliteration of fat planes between right lobe of liver and second part of duodenum.

After a thorough, pre-operative analysis the patient was taken up for a Radical Nephrectomy with Isthmusectomy of the Right moiety. With a Right Subcoastal Muscle cutting incision retroperitoneum was opened in layers. Intraoperative findings were suggestive of a 10\*10\*10 cm mass involving the entire right side of the isthmus. Kidney was mobilised, and total control of the vessels achieved before transacting the kidney with a clear margin of normal tissue over the isthmus. The transacted part was sutured over absorbable haemostatic gelatin sponge using 1-0 Polyvinyl sutures to achieve complete haemostasis. An abdominal drain was placed in-situ and closure done in layers. Post-operative stay was uneventful with the patient being subsequently discharged on post-operative day 5.

Histopathological examination was suggestive of Renal Cell Carcinoma Clear cell type with Nuclear Grade -2. Lymphovascular and Perineural invasion was not seen and resected margins of ureter and vessels were free from tumor invasion.



**Figure 1. Cross-sectional Image of the Excised Right Moiety with the Tumor**



**Figure 2. Right Renal Moiety with the Right Ligated Ureter**

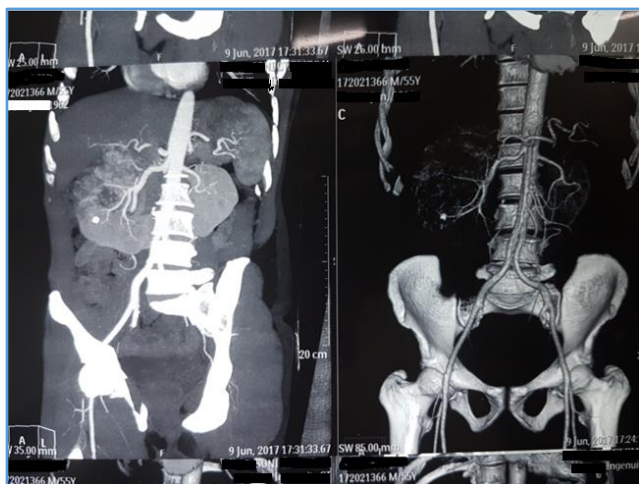


**Figure 3. The Isthmus of the Horseshoe Shape Kidney**

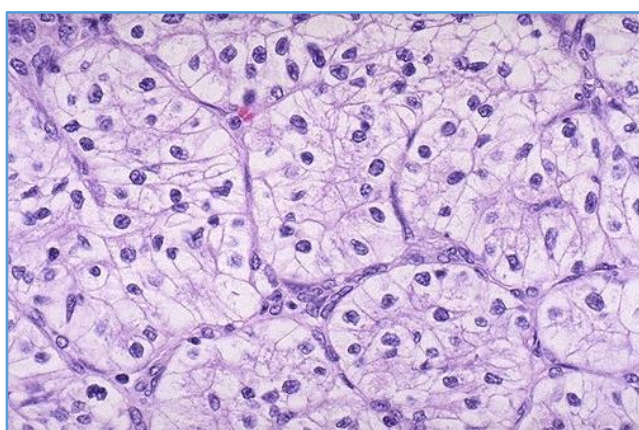


**Figure 4. CT Angiography Depicting the Horseshoe Kidney with the Mass in Right Moiety**





**Figure 5. CT Angiography Depicting the Horseshoe Kidney with the Mass in Right Moiety**



**Figure 6. Histopathological Depiction of the Clear Cell type of RCC**

## DISCUSSION

With less than 200 reported cases in history, Renal Cell Carcinoma in a Horseshoe Kidney is a rare manifestation.<sup>9,10</sup> Other tumours reported include Transitional Cell Carcinomas, Wilm's Tumours, Lymphomas, Carcinoid tumours and sarcomas.<sup>7,11</sup> The incidence of renal tumor in a horseshoe kidney is approximately 3 to 4 times greater than normal, and possibly the result of chronic obstruction, lithiasis, and infection.<sup>12</sup>

Cases have been reported in past 5 years from various surgeons and urologists with tumours affecting one or both entities along with the involvement of the isthmus. Kehinda Habeeb Tijani et al reported in 2016 a similar case where the intraoperative findings were of a 13.5 cm diameter tumour confined to the isthmus and inferior two-third of the shrunken right hemi-kidney. The tumour was most bulky on the left side of the isthmus.<sup>13</sup> Archit Gupta and Jagdish Gupta in 2017 published about a tumor that was seen in right mid and the lower pole of the kidney and was extending to the thick parenchymatous isthmus in 2017.<sup>14</sup> Michael Kongnyuy et al in 2015 reported of a tumor at the junction of the inferior pole of right renal component of the HSK and renal isthmus.<sup>15</sup> Anna Scavuzzo et al encountered a rare case of A 37 years old pregnant woman, gravida 4, para 3 With Renal Cell Carcinoma in a Horseshoe Kidney. The woman

underwent a right radical nephrectomy at 4 weeks after caesarean section.<sup>16</sup> Grygorenko et al reported in 2017 Partial nephrectomy of horseshoe kidney with renal cell carcinoma localized in the isthmus and the lower poles of both parts.<sup>17</sup>

These tumours present a radiological and surgical challenge with difficult diagnosis and a challenging course of surgery owing to the aberrant vascular supply. Hence, a thorough pre-operative analysis as done in our case with Computed Tomography and Renal Angiography is a must to prevent any unpleasant intraoperative surprises.<sup>18</sup> The availability of these investigations with our patient made the resection a lot more streamlined and reduced any unnecessary intraoperative blood loss.

In spite of the difficult surgical path the prognosis is independent of the anomaly and rather is only affected by the histopathology of the tumor. As in our case the histopathology report was a good prognostic indicator and patient was advised a regular follow-up.

## CONCLUSION

In conclusion, Renal Cell Carcinoma in a Horseshoe kidney is a rare entity but with appropriate investigations it can be confronted head on to yield favourable post-operative outcomes.

## REFERENCES

- [1] Mirzazadeh M, Richards KA. Complete duplication of collecting system in a horseshoe kidney presenting with recurrent urinary tract infections: report of an exceedingly rare congenital anomaly and review of literature. *Scientific World J* 2011;11:1591-1596.
- [2] Benidir T, Coelho de Castilho TJ, Cherubini GR, et al. Laparoscopic partial nephrectomy for renal cell carcinoma in a horseshoe kidney. *Can Urol Assoc J* 2014;8(11-12):E918-E920.
- [3] García AA, Burgos RM, Sánchez BC, et al. Horseshoes kidney isthmus carcinoma. A case report. *Actas Urologicas Espanolas* 2008;32(2):249-252.
- [4] Benjamin JA, Schullian DM. Observations on fused kidneys with horseshoe configuration: the contribution of Leonardo Botallo (1564). *J Hist Med Allied Sci* 1950;5(3):315-326.
- [5] Love L, Wasserman D. Massive unilateral non-functioning hydronephrosis in horseshoe kidney. *Clin Radiol* 1975;26(3):409-415.
- [6] Ichikawa T, Tanno K, Okochi T, et al. Evaluation of Renal Artery Anomalies Associated with Horseshoe Kidney Using CT Angiography. *Tokai J Exp Clin Med* 2015;40(1):16-21.
- [7] Buntley D. Malignancy associated with horseshoe kidney. *Urology* 1976;8(2):146-148.
- [8] Stuart BB. Anomalies of the upper urinary tract. In: Walsh PC, Retik AB, Vaughan ED, eds. *Campbell's urology*. Vol 3. 8<sup>th</sup> edn. Philadelphia: WB Saunders 2002:1885-1924.

- [9] Hohenfellner M, Schultz-Lampel D, Lampel A, et al. Tumor in the horseshoe kidney: clinical implications and review of embryogenesis. *J Urol* 1992;147(4):1098-1102.
- [10] Chibane A, Benatta M, Sallami S, et al. Neoplasia in horseshoe kidney: a diagnostic and therapeutic dilemma. *Tunis Med* 2015;93(5):332-334.
- [11] Hellstrom P, Ottelin J, Siniluoto T, et al. Renal cell carcinoma in horseshoe kidney associated with Turner syndrome and caval extension. *Urology* 1989;34(1):46-48.
- [12] Smith-Behn J, Memo R. Malignancy in horseshoe kidney. *Southern Med J* 1988;81(11):1451-1452.
- [13] Tijani KH, Ojewola RW, Orakwe DE, et al. Renal cell carcinoma in a horseshoe kidney: report of a rare disease. *Nigerian Postgraduate Medical Journal* 2016;23(4):232-234.
- [14] Gupta A, Gupta J. Renal cell carcinoma in horseshoe kidney. *J Med Res Prac* 2017;6(1):1-3.
- [15] Kongnyuy M, Martinez D, Park A, et al. A rare case of a renal cell carcinoma confined to the isthmus of a horseshoe kidney. *Case Reports in Urology* 2015;2015:1-3.
- [16] Scavuzzo A, Santana Rios Z, Diaz-Gomez C et al. Renal cell carcinoma in a pregnant woman with horseshoe kidney. *Urol Case Rep* 2017;13:58-60.
- [17] Grygorenko V, Zakordonets V, Danylets R, et al. Partial nephrectomy of horseshoe kidney with renal cell carcinoma localized in the isthmus and the lower poles of both parts: a case report. *Eur Urol Suppl* 2017;16(5):e2175.
- [18] Kim TH. Renal cell carcinoma in a horseshoe kidney and preoperative super-selective renal artery embolization: a case report. *Korean J Radiol* 2005;6(3):200-203.