Horseshoe Kidney and Renal Cell Carcinoma: A Rare Entity and Review Article

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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.


BACKGROUND
Horseshoe kidney was first recognized in 1521 by De Capri in an autopsy; it affects about 1 in 400 live births.1 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.2,3 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.4 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.5

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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.6

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.7

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
inferior mesenteric artery, the common or external iliac arteries, or the sacral arteries.8

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 heterogeneous 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.
underwent a right radical nephrectomy at 4 weeks after caesarean section.\textsuperscript{16} 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.\textsuperscript{17}

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.\textsuperscript{18} 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


