

Gross anatomy of the horseshoe kidney: a case report with brief review of literature

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SUMMARY

The various congenital malformations of the kidney are important as they may cause renal failure in middle-age groups. The horseshoe kidney (HSK) was originally regarded as a rare anatomical curiosity which was seen only on post-mortem. Horseshoe kidneys are congenital fusion anomalies with a prevalence of 1/400 to 1/800 and incidence of 0.25% in general population. HSK is more prevalent in men than women in a 2:1 relation.

During routine anatomical dissection in the formal course for undergraduate students of our college, the gross anomaly of a horseshoe kidney was found. The horseshoe kidney was arrested inferior to the inferior mesenteric artery in a middle-aged male cadaver. The kidneys were fused inferiorly by an isthmus of parenchymatous tissue, both the hila were wide and the right kidney was supplied by two and the left by three arteries. A review of the embryology and clinical importance of this rare anomaly are also discussed.

Key words: Horseshoe kidney – Gross anatomy – Congenital – Fusion anomaly

INTRODUCTION

The horseshoe kidney is one of the most frequent malformations of the urogenital tract with a prevalence of 1 in 400 to 1 in 800 (Kleta, 2000). HSK is a fusion anomaly of the kidneys with an incidence of 0.25% in general population being more preva-

lent in men than women in a 2:1 relation (Basso et al., 2011; Rubio et al., 1998).

The HSK was originally regarded as a rare anatomical curiosity which was seen only at the post mortem (Kilpatrick, 1967). In females with HSK, the physician should be alerted to the diagnosis of Turner syndrome (Kleta, 2000). With the aid of modern radiography (that is- ultrasonography, non-contrast CT, intravenous urography, and retrograde pyelography) the incidence of HSK was estimated at 1 in 200 to 400 individuals (Kilpatrick, 1967). Flower (2000) found HSK in 1 in 1000 necropsies, and states that HSK was more common in males. The reported incidence of this anomaly in Japan was 0.2-0.5% on anatomical dissections (Yoshinaga, 2002). Due to abnormal location of the kidney and ureter, it may be associated with other conditions like calculi formation, urinary infections, pelviureteric junction obstruction and tumoral pathology (Boatman, 1972).

In view of its rarity on necropsies, gross anatomy of horseshoe kidney in a 45-year-old male cadaver found during routine anatomical dissection for undergraduate students in our medical college was discussed.

CASE REPORT

During routine anatomical dissection in the formal course for undergraduate students of our college, the gross anomaly of horseshoe kidney was found in a 45-year-old male cadaver. Both the kidneys were fused at lower poles by an isthmus of parenchymatous tissue ventral to the abdominal aorta and inferior vena cava (IVC), forming the horseshoe kidney (Fig. 1). The parenchyma of the right

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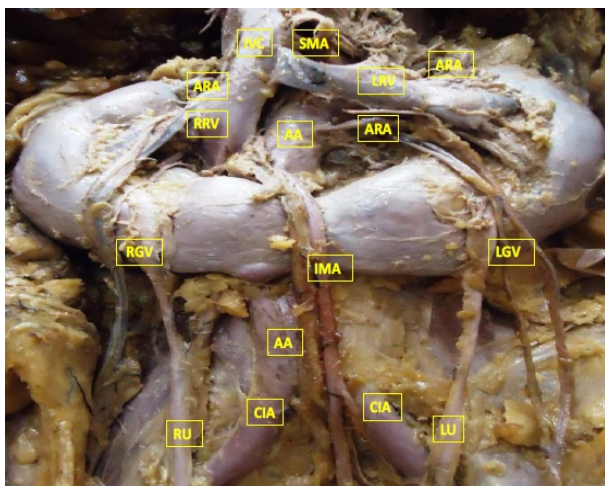


Fig. 1. In situ appearance of the reported horseshoe kidney. For abbreviations, see list at end of this paper.

kidney, the isthmus and the left kidney were normal. The kidney was retroperitoneal with a bridge at the of the L2 vertebral body level. Thereafter, the IVC and the aorta were separated, and the whole HSK was excised for further dissection and photography. The kidneys were fused inferiorly at their inferior poles just below the inferior mesenteric artery (Fig. 1). The ventral side of the kidney was convex while the dorsal side was flattened. The hila were wide and amorphous in both the kidneys. Relation of structures in hilum was normal on both sides as vein, artery and renal pelvis antero-posteriorly.

Vascular supply of the horseshoe kidney

Arterial supply - The right and left renal arteries were originated as usual in the lateral branches of the aorta just below the level of the superior mesenteric artery. The right kidney was supplied by two arteries, the right renal artery and one branch from it to the upper pole. The right renal artery had three short segmental divisions. The left kidney was supplied by three arteries, and the left renal artery has premature four segmental divisions. Two branches were directly from the abdominal aorta, one branch to the upper pole and another branch passing anteriorly to the hilum. This branch was passing anterior to the left renal vein (Fig. 3). There was no separate branch supplying the isthmus.

Venous drainage - Two tributary veins, superior and inferior, were originated from the right hilum to form the right renal vein that emptied into the lateral surface of the inferior vena cava. The right suprarenal vein was directly drained into IVC, but the right gonadal vein drained into right renal vein. The left kidney was drained by the left renal vein, which had four tributaries outside the hilum. The left suprarenal and gonadal veins were drained into the left renal vein as in normal situations (Fig.

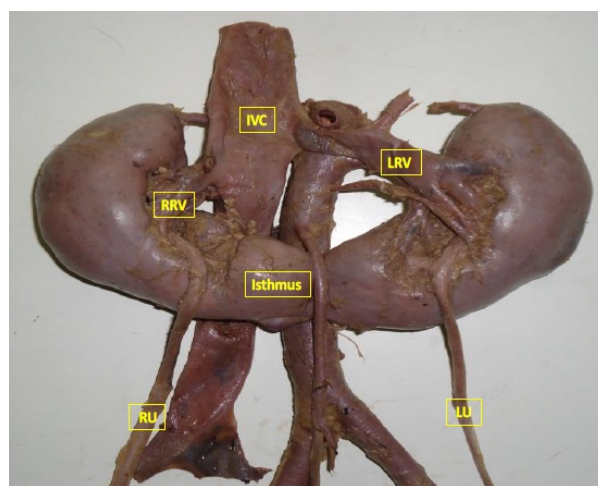


Fig. 2. Anterior view of the horseshoe kidney showing venous drainage.

2). There was no vein for isthmus.

Pelvis and ureter

The ureter on the right side emerged from the hilum on the medial side, and drained the right kidney exclusively. The left ureter drained the left kidney pelvis and the isthmus pelvis (Fig. 2). Both ureters were crossed anteriorly the psoas major muscles and the common iliac arteries, and inserted independently into the posterolateral angles of the urinary bladder (Fig. 1). The posterior view of HSK showed IVC and abdominal aorta which was shown in Fig. 4. There was no other gross variation in the cadaver.

DISCUSSION

The horseshoe kidney is defined as the fusion of the lower poles of both the kidneys ventral to the aorta. The fusion is called isthmus of HSK, consisting of parenchyma or fibrous tissue. HSK is the fusion anomaly that takes place as the result of a developmental defect occurring between 4th to 8th weeks of embryogenesis (Basso et al., 2011). This congenital anomaly resulted from an abnormal mediastinal fusion of the metanephric blastema, originally destined to result in anatomically normal right and left kidney (Basso et al., 2011). This abnormal fusion causes failure of ascent and rotation. Ascent is arrested by the inferior mesenteric artery, which arises anteriorly from the aorta just before its bifurcation (Basso et al., 2011). Therefore, the fused kidney lies more inferiorly in position compared with normal kidneys. Due to incomplete rotation, the renal pelvis is anterior to all the calyces (Gupta et al., 2007).

The cause of the fusion is not known, but it may be due to medial compression by the umbilical arteries (Gupta, 2007). The isthmus is usually composed of the parenchymal tissue and rarely

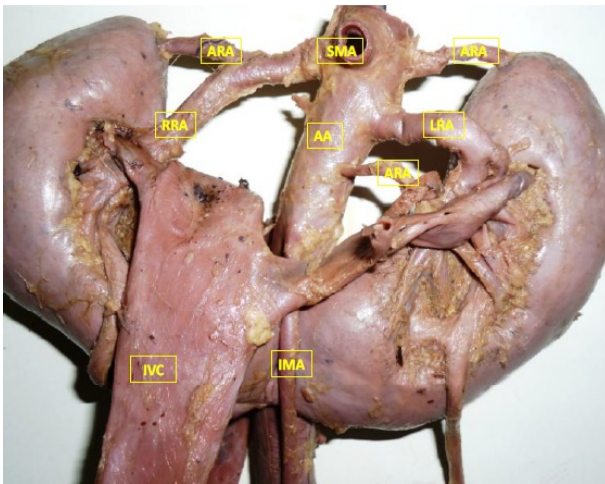


Fig. 3. Anterior view of the horseshoe kidney with reflected inferior vena cava to show arterial supply.

represented by a fibrous strand (Gupta, 2007). About one third of all HSKs have a normal pedicel blood supply, but in other two thirds an abnormal supply to the isthmus is seen (Gupta, 2007). There are two theories about embryogenesis of the HSK. The classical theory of mechanical fusion proposes that during organogenesis, when the lower poles of the kidney enter in contact and fuse in the midline, a HSK with a fibrous isthmus is formed. More recently, it is proposed that HSK is the result of the teratogenic event that involves abnormal migration of cells that form the isthmus, which is related to the parenchymal isthmus (Tijerina, 2009).

The incidence of kidney malformation with chromosomal anomalies is known. Thirty three percent of patients with Turner's syndrome presents with some renal malformation, and 7% of those patients have horseshoe kidney (Basso et al., 2011). Other congenital or aneuploidic anomalies are Edward syndrome, which has up to 20% cases of HSK. Rarely, non-aneuploidic anomalies such

as Fanconi anemia, Goltz syndrome and VACTERL association are noted with HSK (Basso et al., 2011).

Clinically, HSK is most frequently associated with calculi formation. Other manifestations are of urinary tract infections, pelviureteric junction obstruction, hydronephrosis, pyonephrosis and tumoral pathology like Wilms tumor, renal cell carcinoma or transitional cell carcinoma (Rubio et al., 1998; Gupta, 2007).

Technically, the term horseshoe kidney is reserved for cases in which most of each kidney lies on one side of the spine. This is the symmetric HSK (midline fusion). In asymmetric HSK (L shaped kidney), the fused part that is isthmus lies slightly lateral to midline (lateral fusion) (Sharma, 2013). HSK is generally differentiated from crossed fused renal ectopia, in which both fused kidneys lie on one side of the spine, and the ureter of the crossed fused kidney crosses the midline to enter the bladder (Sharma, 2013). HSK is usually incidental or postmortem finding, however for the diagnostic purposes ultrasonography, intravenous urography are usually first line investigation followed by computerized tomography or scintigraphy in cases of doubtful findings (Sharma, 2013). The majority of the HSKs are asymptomatic; hence in 25-30% of the cases no treatment is required. Surgery is necessary only when complications supervene as calculi formation, obstruction, infections or tumors (Kilpatrick, 1967).

Conclusion

In view of its unusual gross anatomy and rare incidence on necropsies, the case of horseshoe kidney in a 45-year-old male cadaver found during routine anatomical dissection was discussed. Clinicians must be aware of this rare anomaly, which may pose a diagnostic as well as interventional challenge.

ABBREVIATIONS FOR ALL FIGURES:

- AA - Abdominal aorta
- ARA - Accessory renal artery
- CIA - Common iliac artery
- IMA - Inferior mesenteric artery
- IVC - Inferior vena cava
- LGV - Left gonadal vein
- LRA - Left renal artery
- LRV - Left renal vein
- LU - Left ureter
- RGV - Right gonadal vein
- RRA - Right renal artery

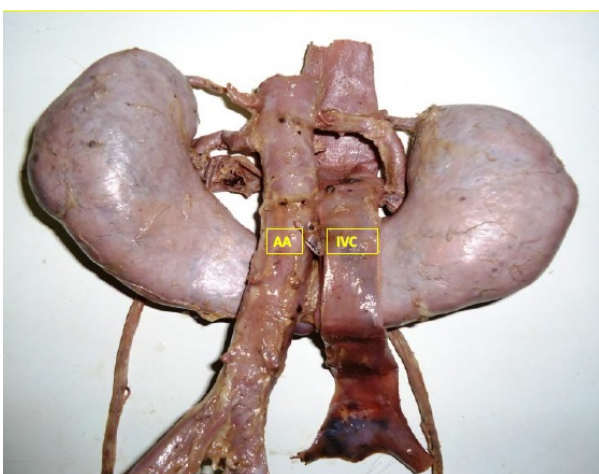


Fig. 4. Posterior view of the horseshoe kidney.

RRV - Right renal vein

RU - Right ureter

SMA - Superior mesenteric artery

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