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A Horseshoe Shaped Kidney: A Case Report with Its Ontogeny, Phylogeny and Clinical Implications

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Authors' contributions

The work was done by all the authors in collaboration with each others. The horseshoe kidney was first observed by authors MS and SD during the dissection. They dissected the specimen and took clinical photographs under guidance of author Upasna. The first draft was prepared by authors Upasna and RKS searched the literature and corrected the first draft. Authors SK and RKS prepared the final draft. It was read and approved by all the authors.

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Case Report

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ABSTRACT

Aim: This study reviews the ontogeny, phylogeny and clinical importance of horseshoe kidney. In addition, the classifications in relation to variations in blood supply of horseshoe kidney have also been discussed.
Presentation: During routine dissection for undergraduate students, the gross anomaly of horseshoe kidney was found. The kidneys were fused at their lower pole by an isthmus. The isthmus was supplied by a separate artery emanating from the front of aorta and entering on its posterior surface. The pelvis was bifid on both sides.
Discussion: Horseshoe kidney is explainable owing to its ontogeny and phylogeny. Its detection is incidental during intravenous pyelography, routine ultrasound or CT scan performed for other reasons.
Conclusion: Variations in the blood supply of horseshoe kidney should be considered in any of the operative procedures related with the kidneys and preoperative digital substraction angiography should be performed.

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1. INTRODUCTION

Kidneys are common site of congenital anomalies, some of which may cause renal failure in middle age group. Horseshoe kidney (HSK) is one of the commonest renal malformation which combines three anatomical anomalies viz ectopia, malrotation and vascular changes. In most of the cases it consists of two renal masses fused at their lower pole by a parenchymal or fibrous isthmus. Rarely the isthmus may connect the upper poles and an inverted horseshoe kidney (HSK) is the result [1]. Such fusions may occur in approximately 1 of 400 individuals and 1 in 300 pyelograms [2,3] and 1:1000 necropsies [4]. As far as sexual dimorphism is concerned it is generally said to be twice more common in males as compared with females [1]. While Gupta et al. [5] are of the opinion that 7% of persons with Turner syndrome have HSK, Gleason and Krammar [6] and Yoshinaga et al. [1] denied any genetic or racial determinant of HSK. At the same time they added that it is found in identical twins and siblings within the same families.

Most of the workers are of the view that usually HSK produces no symptoms as there is normal development of both the collecting and excretory system. Also the ureters enter the urinary bladder normally [5]. So its detection is incidental during intravenous pylography, a routine ultrasound or a CT scan performed for other reasons [7]. Even its antenatal diagnosis may be made as early as first trimester of pregnancy by use of high frequency transvaginal sonography [8]. On the contrary Lallas et al. [9] opine that HSK is associated with propanisity to form uretropelvic junction (UPJ) obstruction in up to one third of cases. It is postulated that UPJ obstruction develops secondary to congenital stricture, high ureteral insertion and abnormal ureteral course over the isthmus, crossing the vessels supplying the isthmus or abnormal motility of the ureteropelvic junction segment [10]. A HSK was encountered in one of the dissection in our department which had several interesting variants. The senior authors who have done about 1000 dissections in total had never seen such a case so it is being reported here with special emphasis on its ontogeny, phylogeny and clinical implications.

2. CASE REPORT

During routine undergraduate dissection of about 50 years old male cadaver in the Department of Anatomy, Government Medical College, Patiala, the horseshoe kidney (HSK) was encountered. The two kidneys were fused at their lower pole by an isthmus crossing ventral to abdominal aorta (A) and inferior vena cava (IVC) forming a HSK. On the left side, the upper pole of kidney was at the level of lower border of T12 vertebra while on the right side it was at the level of lower border of L2 vertebra. The isthmus (IS) was horizontal with a superoinferior width 4.1 cm and located in front of abdominal aorta (A) and IVC between upper border of L3 and lower border of L4 vertebra. The inferior mesenteric artery (IMA) emanated just above the level of isthmus and coursed downwards across its anterior surface (Fig. 1).

2.1 Vascular System of HSK

2.1.1 On the right side

The HSK on the right side was supplied by a renal artery coming directly from the aorta at the level of L2 vertebra. This renal artery passed downwards and laterally behind the inferior vena cava and bifurcated, upper branch entering the upper pole of kidney and the lower branch entering the hilum (Fig. 2). Its length was 4.5 cm. The venous drainage was in the form of a single vein emerging from hilum going upwards and medially to drain into inferior venacava (Fig. 1).

2.1.2 On the left side

The HSK was supplied by a renal artery which came from abdominal aorta below the origin of superior mesenteric artery (Fig. 2). Its length was 2 cm. It was drained by three renal veins (RV) which were emerging from the hilum and joined to form a single vein draining into inferior vena cava. The left suprarenal vein was also joining the left main renal vein (Fig. 1).

2.1.3 Isthmus

The isthmus was horizontal connecting the lower poles of two kidneys, 4.5 cm wide and located in front of abdominal aorta and inferior venacava at the level of upper border of L3 and lower border of L4 vertebra. The inferior mesenteric artery was arising 1.5 cm above the upper border of isthmus and crossing it on its anterior side. Isthmus was supplied by a separate artery emanating from the front of aorta and entering on its posterior surface (Fig. 3). It was drained by two isthmal veins, right and left. The left isthmal

vein emerged on the anterior surface near its point of junction with the lower pole of left kidney, ascended to join lower left renal vein. The right isthmsl vein emerged from the anterior surface of the isthmus almost in the midline and coursed towards the right for a distance of 5cm and then turned superiorly at right angle and ran parallel to inferior vena cava to join the same near the upper pole of right kidney. During its course near the bend it received right testicular vein.

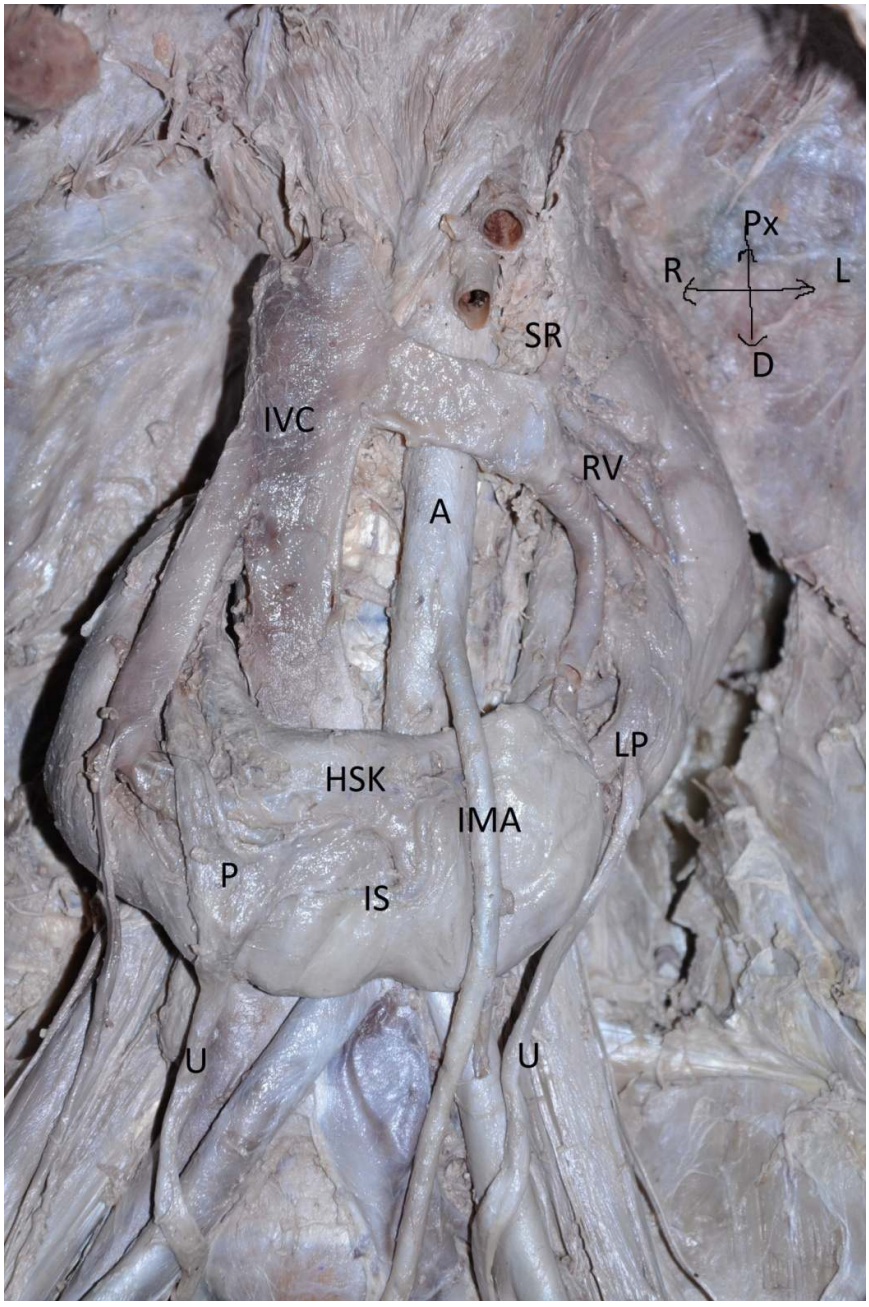

Fig. 1. *In situ* appearance of horseshoe kidney (HSK) showing venous drainage (RV), relationship to the ureters (U) and inferior mesenteric artery (IMA). Abdominal aorta (A), Isthmus (IS), suprarenal (SR), left pelvis (LP), right pelvis (P) and inferior vena cava (IVC) are seen

Fig. 2. Arterial and venous pattern of horseshoe kidney showing abdominal aorta (AA), renal artery (RA), renal vein (RV) and inferior mesenteric artery (IMA)

Fig. 3. Posterior view of horseshoe kidney showing isthmal artery (ISA) and right & left common iliac arteries (CI)

2.1.4 Pelvis and ureters

Two ureters (Right and Left) drained the HSK. The left ureter joined the pelvis near junction of left kidney with the isthmus. The pelvis was seen to be having two parts; one part was coming from the isthmus and one from the left kidney. The right ureter became continuous with the pelvis of right kidney on its anteromedial aspect. Here also the pelvis having two parts i.e. a bifid pelvis. Both ureters crossed the isthmus anteriorly, rest of their course was normal (Fig. 1).

3. DISCUSSION

Oktem et al. [11] classified the HSK into two types depending upon the morphological appearance of fusion. The U shaped HSK is formed by medial fusion of the kidney placed in symmetrical position or either side of vertebral spine. The L shaped HSK is asymmetrical and is formed as a result of lateral fusion between a horizontal and vertical kidney with an isthmus lateral to the midline. The presence specimen fits into the former U shaped HSK.

There is seen a great variation in the origin, number and size of arteries supplying each renal mass in HSK. The renal isthmus may have a separate blood supply. Boatman [7] classified the arterial supply of HSK in following types:

Type Ia: A single renal artery supplies the upper middle and lower renal segments on each side (one artery on right side and one on left side)
Type Ib: A single renal artery supplies upper and middle renal segments on each side while each lower segment is supplied by its own artery coming from abdominal aorta.
Type Ic: A single renal artery supplies upper and middle renal segments on each side (as in Type Ib). But the arteries to lower segment arise as a common trunk from abdominal aorta.
Type Id: Upper and middle renal segments are supplied by multiple renal arteries (more than one on each side) and lower renal segment by one renal artery on each side (arising from abdominal aorta).
Type Ie: Here in addition to type Id the isthmus is also supplied by arteries arising below the isthmus usually originating from abdominal aorta independentally of by a common trunk.
Type If: here the isthmus may be supplied by arteries coming from common iliac or internal iliac artery or middle sacral artery.

Natsis et al. [12] reviewed the work of earlier authors and found that type Ia is seen in 18%, Ib in 6%, Ic in 3%, Ib and Ic together in 17%, Id in 4%, Ie in 28% and If in 24%. In the present case upper and middle segments were supplied by single artery arising from abdominal aorta on each side while the isthmus was supplied by a single isthmal artery arising as a ventral branch of abdominal aorta and entering the posterior surface of isthmus. Thus it did not fit into any of the above six types.

Ruppert et al. [13] reported a classification according to variable arterial blood supply of HSK proposed by Eisendrath:

Type 1: One renal artery for each side of the HSK, 20% of cases.
Type 2: One renal artery for each side with an aortic branch to the isthmus, 30%.
Type 3: Two arteries for each side and one renal isthmal artery, 15%.
Type 4: Two arteries for each side with one or more arising from iliac arteries, including the isthmus branch, 15%.
Type 5: Multiple renal arteries originating from aorta and mesenteric and iliac arteries, 20%.

Depending upon this classification the present case of HSK falls into type 2 i.e. one renal artery for each side with an aortic branch to the isthmus which occurs in 30% cases (Maximum) of HSK. When the kidneys are malformed or misplaced than the postaxial arteries tend to persist and retain their primitive arrangements [14]. So the artery supplying isthmus from dorsal aspect can be a postaxial artery.

4. ONTOGENY

Two theories have been proposed for the ontogenesis of horse shoe kidney. According to the classical theory of mechanical fusion, during the metanephric stage i.e. 4^{th} week of gestation at 5-12 mm CRL stage, when the kidneys are still in the pelvis and are close to each other, their lower poles come in contact with each other and fuse in the midline forming a HSK with a fibrous isthmus. This fusion may be attributed to abnormal flexion or growth of the developing spine and pelvic organs that causes the fusion of nephrogenic blastemas of the immature kidneys which do not possess renal capsule. Normally during the 7^{th} and 8^{th} weeks the kidneys ascend and come out of the pelvis to the abdominal cavity and at the same time rotate so that the

anteriorly facing renal hilum turns medially [7,15]. On the other hand when HSK ascends, at the level of lower lumbar vertebrae, the inferior mesenteric artery (IMA) prevents the ascent and the kidney is trapped in the mid abdomen. The isthmus is further blocked underthe IMA and as a result HSK is malrotated and each renal pelvis remains anteriorly at a lower lumbar position [16].

It has also been proposed that HSK can result as a teratogenic event that involves the abnormal migration of posterior nephrogenic cells that form a parenchymal isthmus [7,15]. This could possibly explain the increased incidence of malignancies associated with HSK.

The formation of HSK usually occurs before the kidneys rotate on their long axis. So the hilum containing pelvis and ureters are usually anteriorly placed. These descend ventral to the isthmus. Rarely if the fusion occurs later after some rotation they are anteromedial. In the present case the renal pelvis of both the sides were directed anteromedial.

Collecting ducts of the permanent kidney develops from the ureteric bud, an outgrowth of the mesonephric duct close to its entrance to the cloaca. The bud penetrates the metanephric tissue, which is moulded over its distal end as a cap. Subsequently the bud dilates forming the primitive renal pelvis and splits into cranial and caudal portions, the future major calyces [17]. In the present case the cranial and caudal portions of the primitive renal pelvis seem to be extra renal in position. This anomaly could be due to slow development of metanephric tissue moulding over the primitive renal pelvis or a relatively rapid development of the ureteric bud leading to extra renal development of cranial and caudal portions of primitive renal pelvis.

5. PHYLOGENY

The functional pronephros is found in early larvae or unhatched embryo of most kinds of fish and amphibian, it degenerates as soon as the more posterior nephrostoms have given rise to a different type of kidney, the mesonephros. In frogs the kidneys are mesonephric in origin [18]. Example in lizard the adult kidneys are small, irregular and bilobed. Anterior broad lobes remain free while posterior narrow lobes become united forming a V shaped structure. The kidneys are attached dorsally and lie in the pelvic region [19]. In the present case, the kidneys were horseshoe shaped and located in lumbar region.

In mammals kidneys are metanephric in origin and occupy much anterior position in mammals as against the metanephric kidneys of reptiles and birds which remain on the posterior part of abdominal cavity. Lower end of the kidneys are also blended normally in some aquatic birds, as the Grebe and Coot, as also in many fishes [20]. Thus a horseshoe kidney in man which is normally seen in lower animals again supports the dictum “ontogeny repeats phylogeny”.

6. CLINICAL IMPLICATIONS

HSK is usually asymptomatic, symptoms of this anomaly are hydronephrosis, infection and lithiasis. Internal abdominal pain can be there in most cases. HSK is initially diagnosed by the ultrasonography or IVP or CT scan [5]. It should not be mistaken for a retroperitoneal mass. The relation between HSK and its arteries can be seen with the digital subtraction angiography. Multiple renal arteries require great skill from transplant surgeon to succeed a vital allograft and satisfactory haemostasis [12].

7. CONCLUSION

HSK affects 0.25% of population. 1/3rd of these are asymptomatic. Variations in the blood supply of HSK should be considered in any of the operative procedures related with the kidneys and pre-operative digital subtraction angiography should be performed.

CONSENT

It is not applicable.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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