Review Article

Horseshoe kidney: a review article

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ABSTRACT

Horseshoe kidney is a rare non-fatal congenital malformation of renal development. It usually remains asymptomatic and in many cases it is discovered incidentally. This anomaly is found twice as often in men than in women. The present report, horseshoe kidney was discovered in 62-year-old male cadaver during routine dissection. The inferior poles of the kidneys were fused to form a parenchymatous isthmus, resulting in a horseshoe kidney. The horseshoe kidney was located anterior to the abdominal aorta and the inferior vena cava at a level lower than the normal kidney. Both renal hila were directed anteriorly and the ureters which drained from each renal pelvis descended anterior to the isthmus to enter the urinary bladder normally. There were 3 renal arteries, 1 on the right and 2 on the left. The inferior vena cava was behind the isthmus and the lower pole of the right kidney. Two renal veins opened independently into the inferior vena cava. It is important to be aware of this renal anomaly in clinical practice, especially during renal surgeries, renal transplants, or surgical and endovascular procedures on the aorta.

Keywords: Horseshoe kidney, Abdominal aorta, Parenchymatous isthmus

INTRODUCTION

Horseshoe kidney is the most common renal fusion abnormality occurring in 1 in 500 persons. As with other fusion anomalies, it is more common in males.[1,2] The male to female ratio being 2:1. 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, this leads to a large U-shaped kidney[3] which is unable to ascend to the level of L1 because it's 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. The kidney is usually lower in position, at vertebra level (L1-L4) as in this case than a normal kidney (T12-L3) and anterior to the aorta and inferior vena cava. In at least one third of cases, the horseshoe kidney is not discovered until autopsy.[4]

Horseshoe kidney developmental anomaly occurs between 7 and 9 weeks of gestation. The intermediate mesoderm that gives rise to the metanephric blastema fails to separate. As the ureteric bud grows cranially, they come into contact with the fused nephrogenic cords and nephrogenesis proceeds. With growth of the embryo, the ascent of the kidney is arrested as the isthmus of the kidney makes contact with the origin of the inferior mesenteric artery. Normal posterior rotation of the kidney is prevented by the fusion resulting in the renal pelves becoming oriented anteriorly. Fusion usually occurs at the lower poles. Upper pole and mid fusion are rare.[3]

Even though the horse shoe kidney produces no symptoms, it is frequently concomitant with other vascular or genitourinary anomalies.[5-8] Therefore, it is important to be aware of these anomalies in clinical practice. Moreover, the horseshoe kidney can be transplanted en bloc or after division of the renal isthmus.[9,10] This report has described the anatomical

variations on the horse shoe kidney and discussed its anatomical and embryological significance.

HORSE SHOE KIDNEY

During a routine abdominal dissection carried out in the Department of Anatomy, Osmania Medical College, Hyderabad, an anatomical anomaly the horseshoe kidney was found in a single cadaver, that of a 62-year-old male. A description of the kidney its external appearance, dimensions and its blood supply were recorded with measurements, line drawings and photographs.
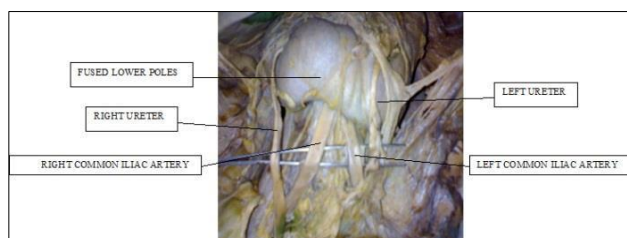
Figure 1: Horse shoe shaped kidney anomaly identified in 62 year old male.
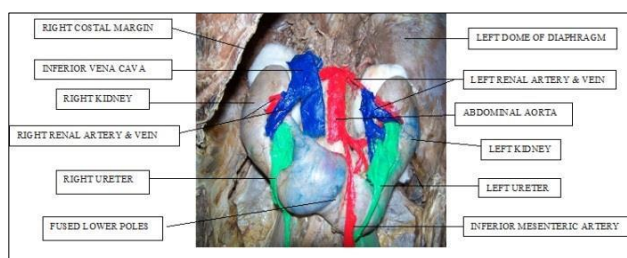
Figure 2: Horse shoe kidney with arteries, veins and ureters marked.

The right and left kidney were fused at their lower poles by a parenchymal isthmus located ventral to the abdominal aorta and formed a U-shaped structure with two unequal arms. The isthmus was oriented obliquely downwards and to the left (Figure 1 and 2). Inferior poles of the horse shoe kidney were at the level of the lower border of third lumbar vertebra on the right, and at the lower border of fourth lumbar vertebra on the left, whereas right and left superior poles were at the level of the middle of body of first lumbar vertebra and at the upper border of second lumbar vertebra respectively. The left side of the horse shoe kidney was slightly smaller in length and thickness than the right (Table 1). The superior pole of the right kidney was 35.6 mm lateral to the vertebral column while that of the left kidney was 47.3 mm lateral to the vertebral column. Their long axes were oriented inferomedially. A parenchymatous isthmus, at the level of the fourth lumbar vertebra, oriented obliquely downward and to the left, joined the inferior poles of the horse shoe kidney.

The horse shoe kidney was supplied by 3 renal arteries (Figure 2). The right renal artery originated directly from

the right side of the abdominal aorta, below the origin of the superior mesenteric artery, and it ran downwards posterior to the inferior vena cava and divided into four branches to supply the apical, upper anterior, middle and lower segments of the right part of the horse shoe kidney, respectively. There were two left renal arteries which originated from the left side of the aorta one below the origin of the superior mesenteric artery and the other just above the origin of the inferior mesenteric artery. The upper one divided into two branches, an anterior branch distributed to apical and upper segments, and the lower left renal artery divided into three branches which supplied the lower segment of the horse shoe kidney.

Table 1: Dimensions of the horse shoe kidney & ureters.

Structures	Dimensions	Right kidney (mm)	Left kidney (mm)
Horseshoe kidney	Length	114.6	112.5
	Width	47.1	48.1
	Thickness	33.5	33.1
Isthmus	Length	51.4	
	Width	37.1	
	Thickness	10.9	
Hilum	Length	19.1	22.4
	Width	10.7	15.1
Ureter	Diameter	5.6	6.1

The isthmus was located anterior to the abdominal aorta and the inferior vena cava, at the level of fourth and fifth lumbar vertebra. Both renal hila were oval in shape and opened anteriorly. The left hilum was larger than the right one (Table 1). There were four major calyces in each pelvis. A single ureter originated from each pelvis, which ran downwards obliquely on the anterior surface of the horse shoe kidney and crossed common iliac arteries to reach the urinary bladder as normal.

The inferior vena cava was located behind the isthmus and the lower pole of the right kidney. Two veins opened independently into the inferior vena cava at the level of L2-L3 vertebrae. A vein emerging from the middle segment of the right kidney, another formed by the union of three veins from the anterosuperior, middle and anteroinferior segments of the left kidney united at the level of L3 vertebra.

DISCUSSION

In the present case review, the horse shoe kidney is the result of an anomalous fusion of the inferior pole to form the parenchymatous isthmus. Its characteristics include a lower position (L1-L4 vertebrae), an isthmus at the level of the fourth lumbar vertebra, an anterior facing hilum, the ureters on the anterior surface of horse shoe kidney, and the abnormal blood vessels, appearance of which are similar to the previous studies.[17-20]

Embryogenesis

This developmental anomaly occurs between 7 and 9 weeks of gestation.[11] The intermediate mesoderm that gives rise to the metanephric blastema fails to separate. As the ureteric bud grows cranially, they come in contact with the fused nephrogenic cords and nephrogenesis proceeds (Figure 3). With growth of the embryo, the ascent of the kidney is arrested as the isthmus of the kidney makes contact with the inferior mesenteric artery. Normal posterior rotation of the kidney is prevented by the fusion resulting in the renal pelves becoming oriented anteriorly.
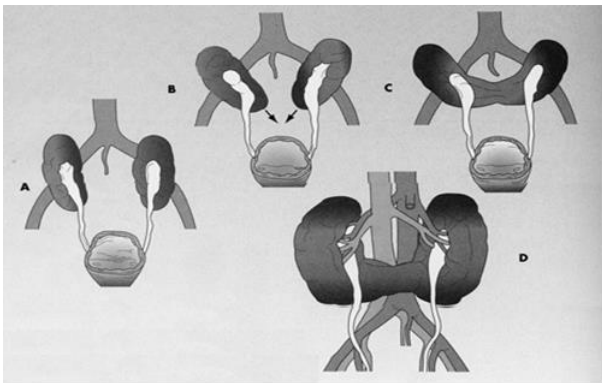
Figure 3: Showing embryogenesis of horse shoe kidney.

Fusion usually occurs at the lower poles. Upper pole and mid fusion are rare. However, nephrogenic cells alone cannot give rise to a horse shoe kidney, so the ureteric bud induction is also essential.[12] Concerning blood supply, the horse shoe kidney shows renal artery anomalies,[6,9] some of which may have an anomalous origin related to embryologic development.[13] The present case showed vascular anomaly with two left renal arteries.

Most (90%) of the cases of horse shoe kidney are asymptomatic. Wilson and Azmy[14] however, reported 15 out of 20 children with horseshoe kidney presenting with symptoms and 9 requiring surgery. In another review of 30 cases twice as many patients were male; 22 cases presented with abdominolumbar pain, 12 with hematuria and 2 with pyuria.

Horseshoe kidneys are sometimes at a greater risk than normal kidneys for obstruction, usually at the uretero pelvic junction, as well as for vesico ureteral reflux, infection, urolithiasis and malignancy. Because the pelves are both extrarenal, they appear more patulous than usual even if not obstructed. Hydronephrosis is caused by ureteropelvic junction obstruction, which may be due to the high origin of the ureter at the renal pelvis, crossing of the ureter over the isthmus, and ureteric entrapment by aberrant vessels, renal stones and infection.[21,22] Horseshoe kidney has the incidence of concomitant vascular or genitourinary anomalies, vesicoureteric reflux, hypospadias and undescended testis.

Horseshoe kidney is sometimes associated with Turner's syndrome (occurring in as many as 7%), trisomy 18 and trisomy 9. It occurs more commonly in patients with trisomy 18, and neural tube defects. 1/3rd of patients with horseshoe kidneys have other abnormalities including those of the genito-urinary, gastrointestinal, respiratory, and skeletal systems. Malignancies like Wilms' tumor (the relative risks of each tumor are increased four fold), Transitional cell carcinoma (the relative risks of each tumor are increased two fold), Carcinoid tumor (the relative risks of each tumor are increased 62 fold).[23-27]

The presence of horse shoe kidney is technically demanding during renal surgeries, renal transplants, or surgical and endovascular procedures on the aorta because of the anomalous complexity of the kidney, its collecting system, and renal blood vessels.[6,9,10] Hence its morphological structure and variations are important factors to be considered. The bulky isthmus located anterior to the abdominal aorta and it's bifurcation, in the horse shoe kidney can cause considerable difficulty in medical and surgical management.

CONCLUSION

Horseshoe kidney is a congenital malformation often asymptomatic throughout life but at some times may predispose the patient to numerous complications including pelvi ureteric obstruction, hydronephrosis, renal stones, infection, malignancies and loss of renal function. Urologists encounter technically difficult cases that are not responsive to standard operative procedures, affecting the patient's quality of life as well as the long-term functional protection of the horseshoe kidney.

This review demonstrates that the knowledge of such anomalies as described here is very important in planning and conducting surgical procedures.

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Cite this article as: Ephraim Vikram Rao K, Battula SR. Horseshoe kidney: a review article. Int J Res Med Sci 2015;3:3004-7.