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Case Report

A Rare Variation of Horse-Shoe Kidney it's Molecular and Embrylogical Basis – A Case Report

***Satyanarayana N*[1], *Victor Anand David A*[1], *Aswinprakash S*[1], *Arulmoli R*[1], *Sunitha P*[2]**

[1]*Anatomy Unit,* [2]*Physiology Unit, Faculty of Medicine, AIMST University, Smelling Bedong 08100, Kedah, Malaysia*

***Corresponding Author**

Dr Satyanarayana N

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Abstract: During routine dissection of a male cadaver in the anatomy unit, AIMST University, Malaysia, a solid mass was observed in the lower right and left lumbar region. Further fine dissection mass it was found to be a horseshoe shaped kidney. The both kidneys were found to be fused at the lower end by isthmus. The right kidney and left kidney are supplied by right and left renal arteries. Further, noticed that the accessory arteries arising from abdominal aorta and supplying to both kidney. The both lower poles of kidney connecting isthmus were supplied by a different artery taking origin from abdominal aorta. Ureters ran downwards anterior to isthmus. The inferior mesenteric artery ran over the mass of tissue joining the lower poles of two kidneys. The congenital malformation can be asymptomatic or can predispose the patient to ureteric obstruction, infection, hydronephrosis, malignancies and posing technical difficulties during renal surgeries, endovascular procedures, renal transplants.

Keywords: Horseshoe kidney; Accessory renal artery, Isthmus of kidney, Congenital malformation, Fibroblast growth factor (FGF2), Bone morphogenetic protein-7(BMP7).

INTRODUCTION

Congenital anomalies of the kidney and the urinary tract were very rare. Among the renal anomalies, there will be fusion of the lower poles or upper poles of kidneys in the earlier stages of embryo development. The fusion of poles of the kidney may be partial or total (Taghavi. *Et al*., 2016). The renal fusion anomalies result in the shape, position, ascent, rotation and blood supply of kidney (Bauer, S.B. 2002). Horse shoe kidney (HSK) comes under the anomalies of partial fusion (Taghavi. *et al*., 2016). The incidence of this anomaly is about 0.25% of the population, or 1 in 400 of the overall population. One third of individuals presenting the HSK were symptomless in their entire life and more commonly in males (Oktem, H. *et al*., 208; BASAR, H. *et al*., 1999). Horseshoe kidney is due to a connection of both the lower poles of the kidney. The fused part of the lower poles is called as isthmus, which is composed of Kidney parenchyma and fibrous tissue. The pelvic part of ureter is directed anteriorly and the calyxes face towards the. Ureters drawing from the anterior part were twisted together over the isthmus. This kind of abnormal anatomical pattern of renal pelvis and ureters over the isthmus will cause an obstruction. The abnormal vasculatures such as accessory renal arteries and other additional vessels extending to the isthmus may surround the ureters and leads to hindrance. The obstruction of urinary flow will lead to dilatation of the collecting system and results in hydronephrosis (Bauer, S. 2007). The retention of urine develops the formation of calculi in the horseshoe kidney. In common, this anomaly does not present any manifesting clinical features. The condition is rarely diagnosed during routine imaging studies (Cascio, S. *et al*., 2002). Some patients complain of abdominal pain but with an indefinite placement. 30% of the individuals with horse-shoe kidney are declared with urinary tract infection, 20–80% was affected by urolithiasis (Bauer, S. 2007). Patients with horseshoe kidney may have other urogenital, digestive, cardiorespiratory, musculoskeletal, or chromosomal anomalies (Cook, W. A., & Stephens, F. D. 1977; Boatman, D.L. *et al*., 1972). Presence of such renal fusion anomalies associated with difficulties and complications during abdominal aortic aneurysm, retroperitoneal and pelvic surgeries. Thorough understanding of their anatomical and radiological features will greatly aid in their surgical management and avoid complications (Babu, C. *et al*., 2015). The present study aims to explain about horseshoe kidney with related renal arterial variations recorded in a Malaysian cadaver. This article also discusses the embryogenesis of the above said anomaly.

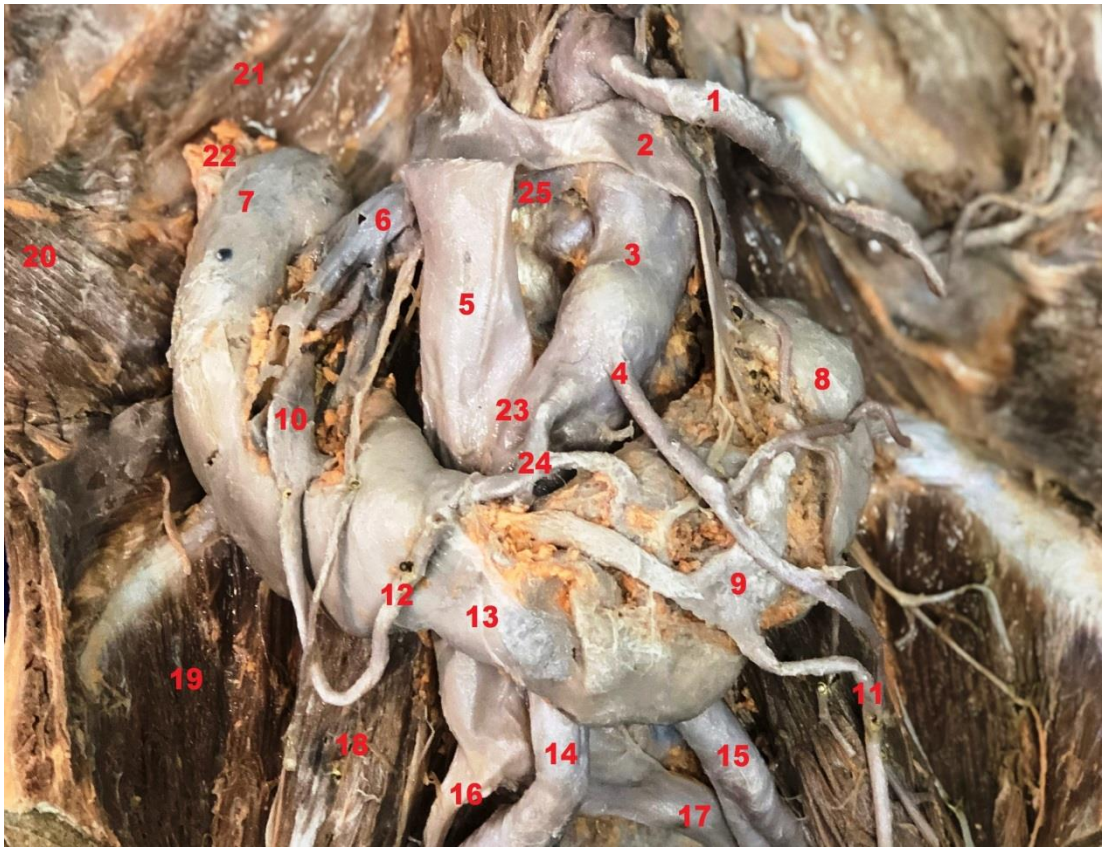

Figure 1: Horse shoe shaped kidney located at the lower lumbar region.

1.Superior mesenteric artery. 2. Left renal vein. 3. Abdominal aorta. 4. Inferior mesenteric artery. 5. Inferior vena cava. 6. Right renal vein. 7. Right kidney. 8. Left kidney. 9. Left renal pelvis. 10. Right renal pelvis. 11. Left ureter. 12. Right ureter. 13. Fused lower poles. 14. Right common iliac artery. 15. Left common iliac artery. 16. Right common iliac vein. 17. Left common iliac vein. 18. Right psoas major. 19. Right iliacus. 20. Transversus abdominis -Right. 21. Quadrates labarum- Right. 22. Right adrenal gland. 23. Accessory renal artery. 24. Accessory renal artery. 25. Right renal artery.

CASE REPORT

During meticulous routine dissection formalin fixed 70-year male cadaver in the anatomy unit, AIMST University, Malaysia, a mass was observed in the lower lumbar region. Further fine dissection of mass, it was found to be a horseshoe shape mass. The care full and fine dissection of the horseshoe mass was carried out. The horseshoe shaped mass was observed to be a horseshoe shaped kidney. The two parts representing right and left kidneys lower poles were joined by mass isthmus. The both kidneys renal pelvises were directed anteriorly. The Pelvis of both kidneys dilated and left renal pelvis bifid on renal pelvis starting from isthmus and other from hilum of left kidney observed. The ureters ran anteriorly downwards crossing the isthmus. It is observed to be a asymmetric midline HSK (**Figure 1**).

Arterial supply

The right and left renal arteries were found to lie at lateral side of the abdominal aorta, present below the superior mesenteric artery, the left being slightly higher than the right. The mass of the horseshoe shaped kidney isthmus part supplied by 2 accessory renal arteries arising from abdominal aorta. The 2 accessory renal arteries were observed to have an equal diameter compared to the corresponding right and left renal arteries. The coeliac trunk, superior and inferior mesenteric and testicular arteries showed no variations in position (**Figure 1)**.

Venous drainage

The horseshoe shaped kidney was drained by right and left renal veins which run anterior to the corresponding arteries. The left renal vein drained into the inferior vena cava after crossing from left to right side across the abdominal aorta. It was seen draining the left suprarenal vein superiorly and the left testicular vein inferiorly. It also received variable number of tributaries from the rest of left side of the HSK mass. A separate vein from the isthmus drained into the left renal vein. The right renal vein drained directly into the Inferior vena cava. The right testicular vein drained separately into the inferior vena cava (**Figure 1**).

DISCUSSION

The kidneys were normally in the retroperitoneal, located in the posterior abdominal

wall extending from the level of T-12 and L-3 vertebrae. During the developmental ascent both the kidneys comes to lie below the supra-renal glands (Kirkpatrick, J.J. 2018). HSK is the condition in which the ascent is caused by the inferior mesenteric artery, because of the central fusion between both the kidneys (Suwannakhan, A., & Meemon, K. 2016). This kind of condition is seen in 40% of cases, whereas in 20% of the cases, the fused kidney remains in the pelvis without further descent (Kirkpatrick, J.J. 2018). In some other cases the ascend of kidneys may be restricted by the renal artery (Suwannakhan, A., & Meemon, K. 2016). During the ascent of both kidneys, they rotate 90 degree of medial rotation (Mouriquand, P. *et al*., 1989; Türkvatan, A. *et al*., 2009). The rotation of the kidneys can be disturbed by the position of the ureters, which leads to urinary obstruction and drainage problems. In 70% of cases the arterial supply of both kidneys by a pair of renal arteries and 30% were found to be supplied by accessory renal arteries. In HSK, most of the cases were supplied by the branches emerging from the abdominal aorta or from the arteries. The type of arterial supply is depending up on the ascent the kidneys (Basso, L. S. *et al*., 2011; Natsis, K. *et al*., 2014).

Molecular basis of the kidney development

The ureteric bud epithelialization is due to the Wilms' tumor suppressor 1 ('WT1'transcription factor). The metanephric blastema mesenchyme produces the transcription factor 'WT1'. The 'WT1' regulates the Glial Derived Neurotrophic Factor (GDNF) and Hepatocyte Growth Factor (HCF) formation. The ureteric bud produces transcription factors PAX2 and WNT4, which helps in the differentiation of mesenchyme into epithelial cells and the formation of excretory tubules. The fibroblast growth factor (FGF2) and Bone morphogenetic protein (BMP7) that stimulates the proliferation of mesenchyme and WT1 expression (Shambharkar, S. B. *et al*., 2018; Cook, W. A., & Stephens, F. D. 1977; Garg, K. 2012).

Embryological basis of kidney development

During the development of the excretory system, there will be an interaction between the mesonephric duct and the metanephros. The metanephric blastema is formed in the fifth and sixth weeks of gestation and the lobulated kidneys ascends from the lesser pelvis reaches the posterior abdominal wall. Then they undergo a 90-degree axial rotation from horizontal to medial rotation (Taghavi, K. *et al*., 2016; Suwannakhan, A., & Meemon, K. 2016). During the process of kidney ascent from the pelvis the kidneys receive their blood supply from nearby blood vessel, initially median sacral, then common iliac, inferior mesenteric and finally the abdominal aorta. During ascent, if the kidneys comes into close contact result in horseshoe kidney or crossed renal ectopia (Bauer, S. 2007; Cook, W. A., & Stephens, F. D. 1977; Garg, K. 2012; Mouriquand, P. *et al*., 1989).

Mechanism of horse-shoe kidney

The growth of vertebral column and any abnormal forward bending of the spine results in the close approximation of both kidneys and union of lower poles leading to formation of horse-shoe kidney (O'Brien, J. *et al*., 2008). After the fusion of lower poles, the horse-shoe kidney ascends and reach the posterior abdominal wall, during this phase upper direction is restricted the inferior mesenteric artery due to the communication of lower poles. This may cause the absence of rotation of both the kidneys and anterior orientation of renal pelvis and ureters (Türkvatan, A. *et al*., 2009). According to the recent information the HSK is an anomaly caused by the teratogens (Tijerina., G.O. *et al*., 2009).

Among the 80% cases of horse-shoe kidneys the isthmus (fused lower poles) were not supplied by the branches emerging from renal arteries. (Roque, R. *et al*., 2007; Vaniya, V.H. 2004). The branches of the inferior mesenteric arteries supplying the isthmus have been reported (Yoshinaga, K. *et al*., 2002).

In the present case-report, three accessory renal arteries were observed, one directly emerging from the abdominal aorta, other two accessory renal arteries from the right inferior mesenteric artery and supplies the isthmus.

CONCLUSION

HSK manifests numerous anatomical variations. There are several mechanical and genetical factors closely relatable. The approximation of the renal blastema during ascent will play a significant role in the renal genesis. Based on the complete or incomplete ascent, the final position of the kidney that ranges from the pelvic, iliac fossa or lumbar region. The more renal blood vessels variations with respect to HSK were highly associated. Nephrologists must be aware of this type of rare variations with respect to the stenting of ureters, symphysiotomy, percutaneous nephrolithotomy and extra-corporeal shockwave lithotripsy. The present case might not only provide anatomical and surgical knowledge of the HSK but provide more interest in the embryological and molecular basis of its development.

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