CASE REPORT

Horseshoe Kidney

Ewaldo Amirullah Hadi[1], Akhada Maulana*[2]

[1]1West Nusa Tenggara Province General Hospital, Mataram
[2]Division of Urology/Department of Surgery, Medical Faculty/Mataram University, West Nusa Tenggara Province General Hospital, Mataram
Coresponding Autho
*akhada_m@hotmail.com

ABSTRACT

Horseshoe kidneys are identified as having functioning renal masses present on both sides of the vertebral column fused together with ureters that remain uncrossed from the renal hilum to the urinary bladder. The anomaly is characterized by the fusion of the two distinct kidneys at the lower poles (in most of the cases), in the front of the body midline, through an isthmus of functional renal parenchyma or by fibrous tissue. Incidence of horseshoe kidney is 0.25% in general population and more frequent in male (male : female ratio = 2:1).

Keywords: horseshoe kidney, anatomy, embryology, diagnosis, treatment.

Introduction

Horseshoe kidney (HSK) is the most common congenital renal fusion anomaly and is characterised by three morphological anomalies: ectopia, malrotation, and changes in vascular supply. Jacopo Berengario da Carpi was the first person to describe this abnormality during autopsies in 1522. HSK usually consists of two renal masses fused at their lower poles by a parenchymal or fibrous isthmus.1

The isthmus connecting the two renal masses may be positioned in the midline or laterally resulting in asymmetric horseshoe kidney, 70% of which are left dominant, and consists of renal parenchyma in about 80% of cases with the remainder being composed of a fibrous band. In more than 90% of cases, fusion occurs at the lower pole, although fusion may occur at the upper pole in a minority of cases.2

Incidence

Horseshoe kidney is found more commonly in male with ratio to female is 2 : 1. It occurs in 0.25% of the population, or about 1 in 400 individuals. Reported incidence based on radiographic imaging has shown similar results, between 1 in 474 and 1 in 666 individuals.3

Embriology

Embryologically, kidney development is divided into 3 stages: pronephros, mesonephros, and metanephros. The pronephros stage is formed by 7-10 clusters of dense cells in the neck region. The first group forms the vestigial nephrotome, which disappears before the caudal nephrotome is formed. At the end of the 4th week, all signs of the pronephros system disappear. The mesonephros originates from intermediate mesoderm from the upper thoracic to upper lumbar segments. When the pronephric system regresses, the excretory ducts of the mesonephric appear to elongate to form the glomerulus (medial part) and Bowman's loop, both of which are called the mesonephric corpuscles (renal). Lateral part, the ducts converge on the mesonephric duct (wolfian

duct). In the middle of the second month, the medial part of the mesonephros forms the gonads and the ridge formed by these two organs is called the urogenital ridge. The caudal channel remains differentiated. Towards the end of the second month, the cranial portion has largely disappeared. The caudal part in the male remains and contributes to the sex system and in the female it disappears.4

At the time of the metanephrons phase, the metanephric blastema differentiates into a renal nephron, then its function and position become perfect. The metanephric blastema triggers the Wolfian duct and forms the ureteric bud (UB), which in turn fuses the ureter with the kidney. The normal kidney will ascend from the sacrum to the lumbar region which occurs in the six and nine weeks of pregnancy, the ascending kidney will receive a local vascular supply from the surrounding blood vessels.5

Anatomy

The kidneys are normally located in the retroperitoneum between the transverse processes of T12 and L3 with the left kidney slightly more superior than right. Horseshoe kidney is more commonly in a low position because the isthmus does not permit ascent beyond the inferior mesenteric artery. The isthmus of the kidney may not have a separate blood supply or is supplied by a single vessel from the aorta (in 65% of cases), from the common iliac, or inferior mesenteric arteries.1,6

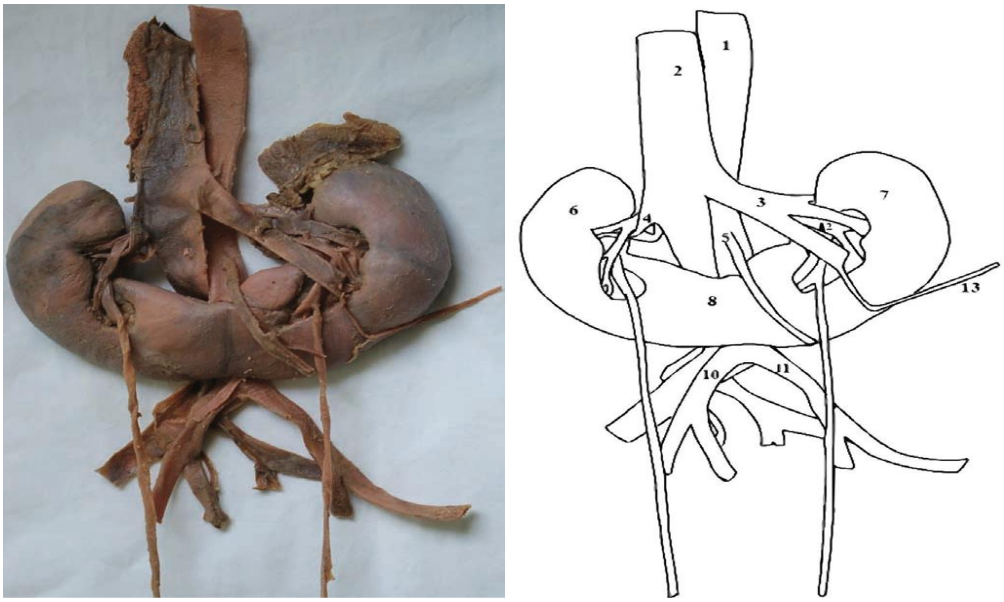

Figure 1 : Panoramic view of the anatomic sample. 1. Aorta; 2. Inferior vena cava; 3. Left renal artery; 4. Right renal artery; 5. Inferior mesenteric artery; 6. Right kidney; 7. Left kidney; 8. Isthmus; 9. Ureters; 10. Right common iliac artery; 11. Left common iliac artery; 12. Renal arteries for the isthmus; 13. left gonadal artery (reflected).6

Symptoms

Horseshoe kidneys are often asymptomatic, and so are often identified incidentally. Sometimes it is diagnosed after finding of a midline mass in the lower abdomen. In other patients the symptoms emerged due to renal obstruction, stones or infections. The

most common presenting symptoms in children are those related to urinary tract infection. About one third of the cases were found incidentally during radiological examination and only 60% of the patients were demonstrating complained symptoms.7

Evaluation

In recent years, ultrasonography has become the first imaging procedure performed in the evaluation of pathologic urinary tract conditions.8

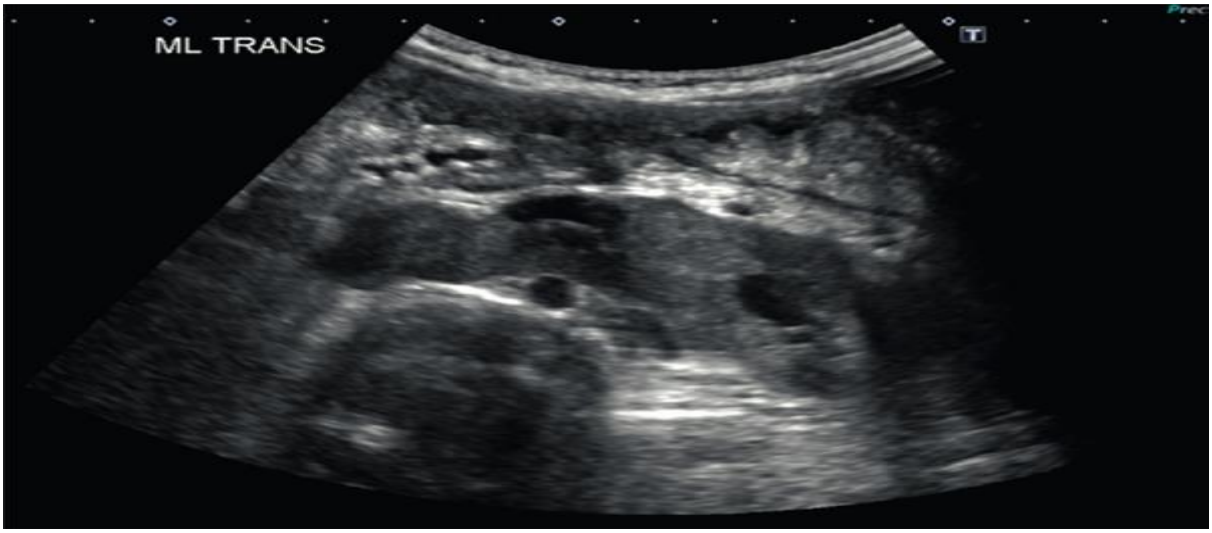

Figure 2 : Newborn with cloacal anomaly. Abdominal sonogram shows horseshoe kidney with mild dilation of the right and left collecting system. Note right side with anterior renal pelvis.3

CT scan are the best for demonstrating the anatomy and can detect accessory vasculature and surrounding structures.9

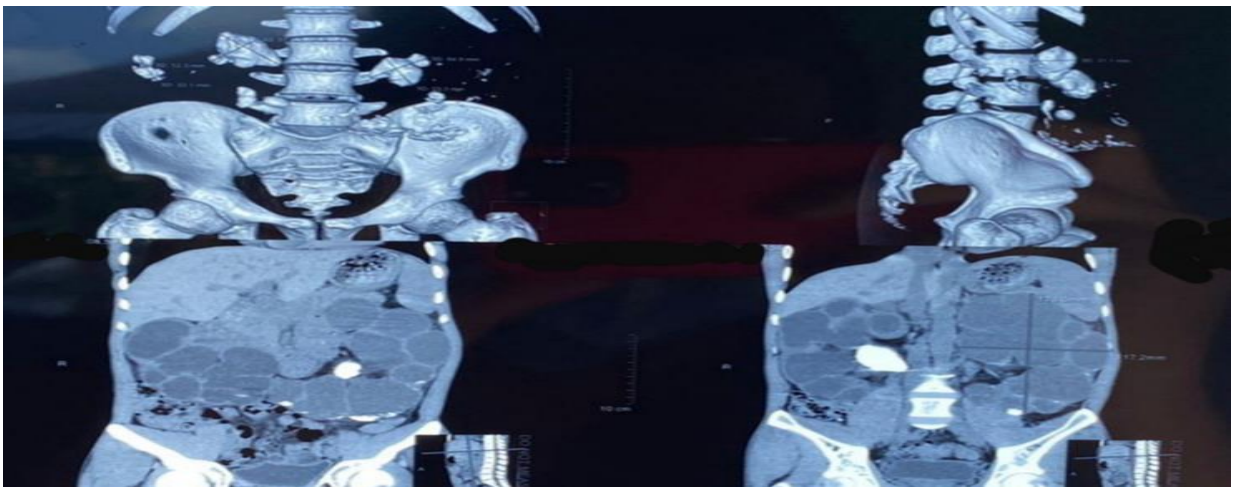

Figure 3: CT of an adult male with horse shoe kidney shows severe hydronephrosis and staghorn stones. Red arrow points to inferior isthmus of the kidneys, which separates left and right kidney. (Private collection)

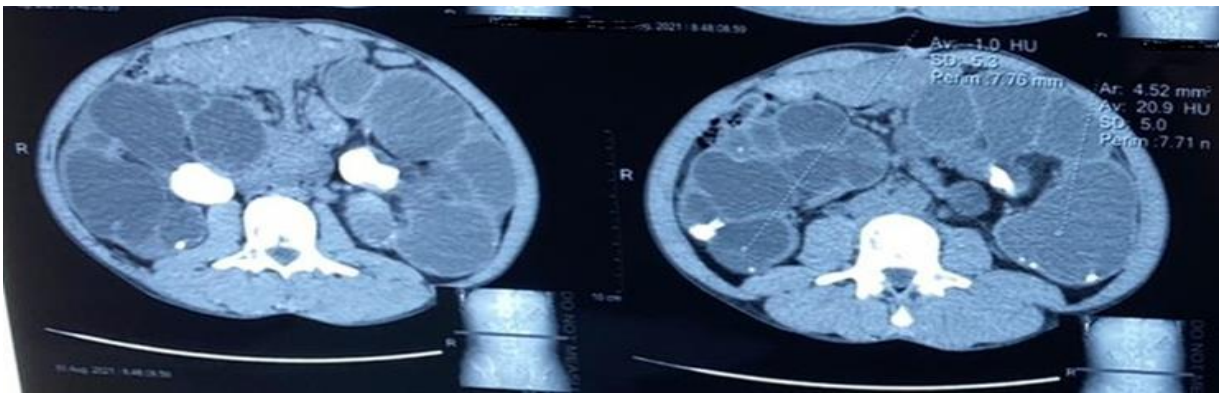

Figure 4: CT of an adult male with horse shoe kidney shows severe hydronephrosis and staghorn stones. Stone in renal-pyelum (red arrow) points to postero-medial, while in normal kidney stone points to antero-medial. (Private collection)

Treatment

Medical treatment may be necessary if complications are identified (renal stones or infections). PCNL is suggested as the first-line treatment method for staghorn stones in patients with Horseshoe Kidney. In 1973, Fletcher and Kettlewell reported the first PCNL in horseshoe kidney. From then on, percutaneous puncture of the Horseshoe Kidney has been found to be relatively safe because of favourable calyceal orientation and vascularity. PCNL showed a better stone-free rate (SFR) 81% to 87% than ESWL, and it was found that the risk of arterial bleeding did not increase in Horseshoe kidney compared with normal kidneys. Percutaneous puncture in horseshoe kidney is relatively safe, especially performed by passing through the superior poles. Janetschek and Kuanzel pointed out that the puncture should be made below the 12th rib on the posterior axillary line with caudad angle of puncture.10

Conclusion

Horseshoe kidney is the most common congenital renal fusion anomaly and is characterised by three morphological anomalies: ectopia, malrotation and changes in vascular supply. Horseshoe kidneys are often asymptomatic, and so are often identified incidentally. Sometimes it is diagnosed after discovery of a midline mass in the lower abdomen. CT scan are the best for demonstrating the anatomy and can detect accessory vasculature and surrounding structures. PCNL is suggested as the first-line treatment method for staghorn stones in patients with horseshoe kidney.

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