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Pyeloureteral Jonction upon a Horseshoe Kidney

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Abstract **Case Report**

The horseshoe kidney is the most common renal fusion anomaly, affecting around 0.25% of the general population. We report the rare case of a 14-year-old child with a horseshoe kidney, admitted to hospital for chronic abdominal pain and burning micturition. Physical examination revealed umbilical tenderness. Abdominal ultrasound showed a horseshoe kidney associated with moderate bilateral hydronephrosis. CT scan revealed fusion of the kidneys at their lower poles and obstruction of the ureteropelvic junction (UPJ).

Keywords: Horseshoe kidney; Malformation Pyeloureteral junction; imaging.

INTRODUCTION

Horseshoe kidney is the most common of all renal fusion anomalies, occurring in approximately 0.25% of the general population [1]. It is usually asymptomatic, but can be associated with nephrolithiasis, ureteropelvic junction stenosis, renovascular hypertension, polycystic kidney, and polycystic liver [2]. Pyeloureteral jonction upon a horseshoe kidney remains a frequent congenital malformation [3]. This case report describes the incidental diagnosis of a horseshoe kidney with a single ureter.

CASE

14-year-old children was admitted to hospital complaining chronic vague abdominal pain with burning micturition. At physical examination, umbilical tenderness. An abdominal ultrasonography showed a horseshoe kidney associated with moderate bilateral hydronephrosis. Computerized tomography revealed kidneys fused by their lower poles (Figure 1) and ureteropelvic junction (UPJ) obstruction (Figure 2).

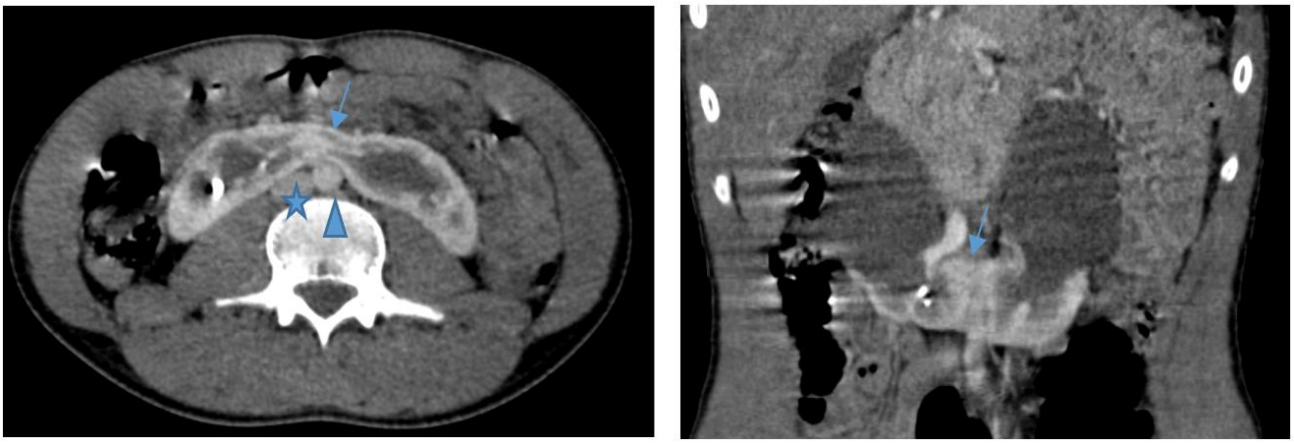

Figure 1: Computed tomography (CT): axial and coronal plans scan revealed kidneys fused by their lower poles (arrow) in front of the aorta (arrowhead) and inferior vena cava (asterisk)

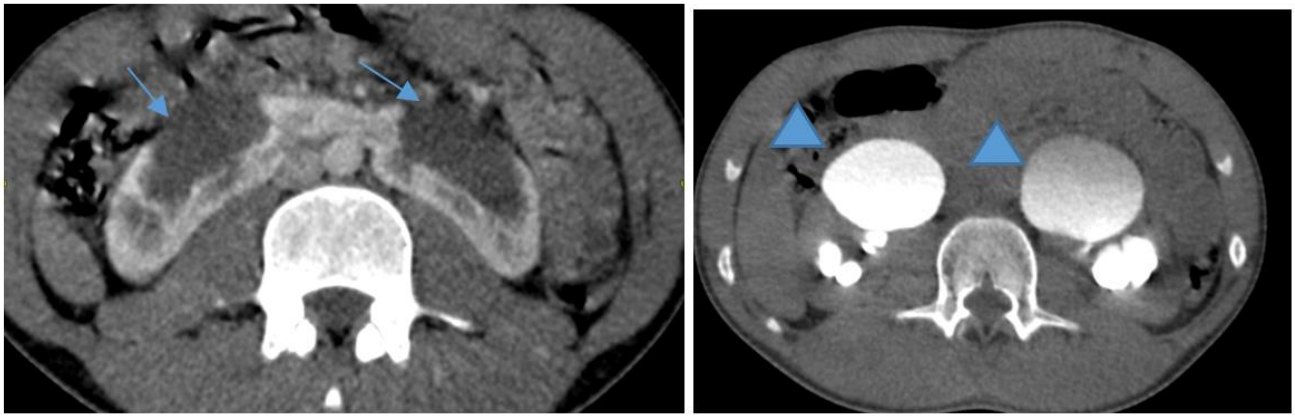

Figure 2: Axial and coronal computed tomography (CT): showing the two kidneys with pelvic dilatation(arrows) contrasting in late times (arrowhead) and normal caliber ureter suggesting stenosis of the ureteropelvic junction.

DISCUSSION

Horseshoe kidney (HSK) remains a frequent congenital malformation, occurring in approximately 0.25% of the general population and more frequently in men. HSK usually combines three distinct anomalies: renal ectopia, malrotation of the kidneys and renal vascular abnormalities [4]. In most cases the kidneys are connected by an isthmus located at their lower poles. Horseshoe kidney represents a defect of renal ascending combined with fusion defect. The theory of mechanical fusion suggests that between the week 4 and 6 of gestation the metanephric blastema fuses before the kidneys ascend and rotate. As the horseshoe kidney ascends, the isthmus is trapped under the inferior mesenteric artery (IMA), arresting further ascent and rotation, resulting in lower location of the kidneys with anteriorly facing pelvis. Approximately 98–99% of the horseshoe kidneys are located at or around the origin of IMA from the aorta and the rest in the pelvis [5]. HSK may be associated with other urinary tract and extrarenal abnormalities, including stenosis of the ureteropelvic junction, double pelvis, abnormalities of the urinary bladder, supernumerary kidneys [6]. HSK can be complicated by the formation of kidney stones, hydronephrosis (due to stenosis of the ureteropelvic junction, malrotation and/or malposition of the kidneys and ureters, obstruction by stones), urinary tract infections and tumors (Wilms' tumor – usually in the isthmus, transitional cell carcinoma, oncocytoma, carcinoid tumors [4,7].

In the adult population HSK is usually asymptomatic and can be discovered accidentally during imaging studies for other reason or for renal stones or urinary tract infections [8]. The ultrasound image is very typical – low-lying and malrotation of one or both kidneys, anterior position of renal pelvises with and without hydronephrosis/pyelectasia, boomerang-shaped kidneys with elongation, poorly-defined lower pole curving away from the transducer [8]. The isthmus may not be clearly seen on ultrasound investigation [8]. In addition, it is operator dependent and detection rate varies depending on operator skill and experience. Moreover, if unsuspected, the isthmus may mimic a midline mass on US [9]. CT is the modality of choice for evaluation of the horseshoe kidney and relation to surrounding structures. It also plays an important role in the evaluation of potential complications and surgical planning. MRI can be used with similar advantages and without the risk of radiation, however, some complications including stones and trauma are better evaluated with CT. Finally, computed tomography angiography (CTA) and magnetic resonance angiography (MRA) are both useful for the depiction of vascular anatomy of horseshoe kidney [10]. Traditionally intravenous urography/pyelography (IVU/IVP) has been performed for assessing for pelvi-ureteric junction obstruction. Ultrasound will often show a dilated renal pelvis with a collapsed proximal ureter. CT may show evidence of hydronephrosis +/- caliectasis with collapsed ureters. Useful for assessing crossing vessels at the pelvi-ureteric junction, especially when surgical intervention is planned [8, 10]. The typical imaging of PUJ obstruction features can be seen on intravenous urography (IVU) or CT excretory urogram and include dilated renal pelvis with normal caliber ureter. CT is a better imaging tool because it not only delineates the anatomy of horseshoe kidney but also its relation to surrounding structures, cortical thickness, and any associated vascular abnormalities. All the above information is of utmost importance if surgery or radiological intervention is planned [7, 10].

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