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## Horseshoe kidney with retrocaval ureter: a case report

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### ABSTRACT

Horseshoe kidney and retrocaval ureter are uncommon congenital anomalies of the genitourinary system that are easily diagnosed by typical imaging features. Both anomalies presenting in one patient is a rare disease characterized by isthmus of horseshoe kidney between the abdominal aorta and inferior vena cava. The clinical diagnosis and treatment of horseshoe kidney with retrocaval ureter remain a challenge. Here, we reported a case of 10-year-old boy with the two anomalies who was preoperatively diagnosed by enhanced computed tomography scanning. The literatures on such combined anomalies are reviewed and the diagnostic evaluation and surgical management of this rare entity are discussed.

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## **Introduction:**

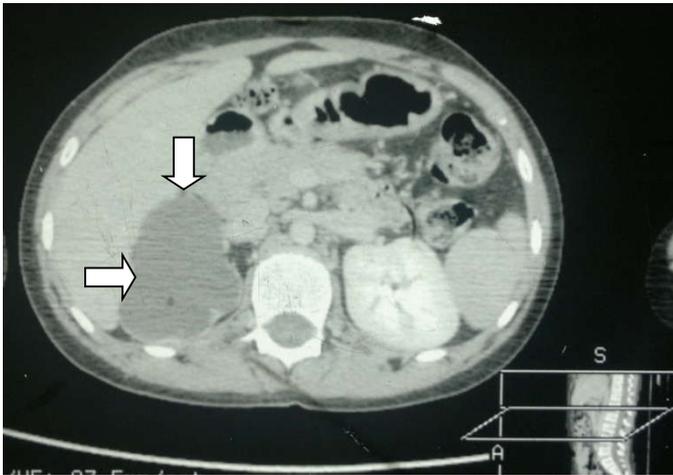
Horseshoe kidney with an incidence of 1 in 400 persons<sup>1</sup> and retrocaval ureter with an incidence of 1 in 1500 persons<sup>2</sup> are uncommon anomalies of the genitourinary system. However, the two entities coexisting in one patient seems so rare that only 10 relevant articles could be found on the PubMed database searched with keywords “horseshoe kidney” and “retrocaval ureter/circumcaval ureter”. When the two rare deformities coexist in one patient, it is easy to find horseshoe kidney; however, retrocaval ureter on the right side is difficult to diagnose. We present a case in which the diagnosis of both anomalies in one patient was made by preoperative enhanced computed tomography (CT) scanning with confirmation by surgical exploration.

**Case Report:** a 10-year-old boy presented with intermittent vague right flank pain associated with anorexia along the previous 1 year. He had a previous occasional attacks of urinary tract infection. Physical examination

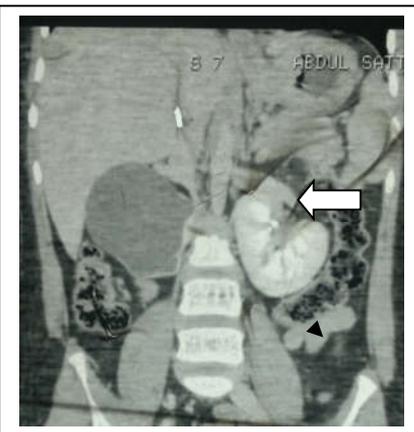
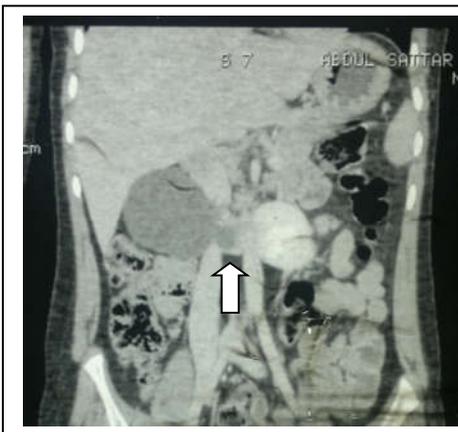
revealed a soft abdomen and no percussion pain on both kidneys. Renal function and other hematological findings were within normal limits. Urologic ultrasonography demonstrated a gross hydronephrotic right kidney with very thin cortex which appeared just a sac-like structure while the left kidney appeared larger than it was expected for this age and somewhat at a lower position in the abdomen. Intravenous urography (IVU) examination showed a non-visualized right kidney even in a delayed film and a mild compensatory enlargement of the left kidney with distortion of its lower pole calices, medially deviated left upper ureter and a normally looked urinary bladder.

CT-scan showed a horseshoe kidney with right severe hydronephrosis and diminished cortical paranchymal cover with absence of contrast excretion (Figure -1 A&B). The isthmus situated dorsal to inferior vena cava (IVC) and ventral to the aorta (Figure-2). A horseshoe kidney coexisting with right

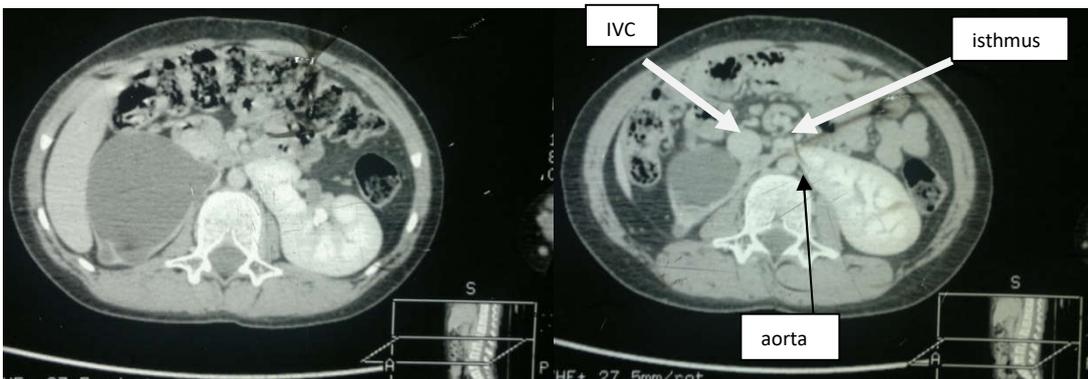
retrocaval ureter was thus identified.



**Figure (1 A):** enhanced CT-scan showing right sided severe hydronephrosis and diminished cortical parenchymal cover with absence of contrast excretion.



**Figure (1 B):** enhanced coronal section CT-scan showing contrast excretion in the left kidney with medially deviated lower poles & isthmus in midline.



**Figure (2):** enhanced CT-scan showing the isthmus situated dorsal to inferior vena cava and ventral to the abdominal aorta in cross-sectional films.

etroperitoneal approach. Horseshoe kidney concurrent with retrocaval ureter was confirmed through the operation. The special entity was isthmus of the horseshoe located between the abdominal aorta and IVC. Division of the isthmus and right heminephrectomy were performed. The patient tolerated the procedure well with no intraoperative and postoperative complications. At repeated follow-up visits, urinalysis and urinary ultrasound revealed no evidence of any abnormal finding.

**Discussion:** horseshoe kidney is the most common congenital deformity among renal fusion anomalies, with a male-to-female ratio of 2:1. This entity may be diagnosed at any age ranging from the fetus to the elderly patient. In 95% of horseshoe kidney,<sup>1</sup> the fusion occurs at the lower poles with an isthmus that usually lies anterior to the great vessels at the level of the third to fifth lumbar vertebra. Many horseshoe kidneys are asymptomatic and found

incidentally. However, they are prone to a variety of complications such as stone disease, ureteropelvic junction obstruction, trauma, infection, and a variety of benign and malignant tumors.<sup>3</sup>

Retrocaval ureter with a three-to-four-fold male predominance is also a rare congenital anomaly. Its etiology is presumed abnormal persistence of the subcardinal vein in embryologic development of the IVC resulting from the major portion of the infrarenal IVC being formed from the subcardinal vein that lies ventral to the ureter. It almost always occurs predominantly on the right side and usually becomes symptomatic in the third or fourth decade. In the majority of patients symptoms are due to ureteral obstruction and resulting hydronephrosis.<sup>2</sup>

Horseshoe kidney and retrocaval ureter are easily diagnosed by typical imaging features. IVU shows characteristic appearance of horseshoe kidney with abnormal renal axis,

incomplete inward rotation of the renal pelvis. Ultrasound examination shows an abnormal renal rotation and an isthmus; the diagnosis is easy to make in children and in nonobese patients. However, if the isthmus consists of a thin fibrous band or in adults with a large body habitus, ultrasound examination is insensitive and very operator dependent. Contrast-enhanced CT and magnetic resonance imaging (MRI) are good choices for the assessment of a horseshoe kidney because they show the isthmus and allow evaluation of potential complications. Retrocaval ureter can be divided into two types:<sup>4</sup> type 1, the more common form, has S-shaped or fishhook appearance accompanied by severe or moderate hydronephrosis with extreme medial deviation of the middle ureteral segment usually across the midline at L3 level; type 2, on the other hand, has sickle-shaped appearance with mild hydronephrosis and less medial deviation of the ureter. IVU, CTU, and MR-urography (MRU) can provide good appearance. When the

two rare deformities coexist in one patient, it is easy to find horseshoe kidney; however, retrocaval ureter on the right side is difficult to diagnose. Only two of 10 reported cases<sup>5,6</sup> were identified by preoperative CT; in the others the retrocaval ureter was found during exploratory operation of horseshoe kidney. In our case the horseshoe kidney was easily identified on CT-scan. However, an unusual feature was that the isthmus being between the IVC and aorta, and the right ureter was not found through IVU and even CTscan in the excretory phase (CTU) because of the non-functioning right kidney that interfered with ureteric filling by contrast medium.

Heminephrectomy in a horseshoe kidney can be done by open or laparoscopic approach. However, the blood supply to horseshoe kidneys may arise from the aorta, iliac arteries, and inferior mesenteric artery;<sup>7</sup> variability adds complexity to laparoscopic surgery. In our patient, we performed open surgical approaches, and successfully achieved right

heminephrectomy without damage of the aberrant vasculature.

**Conclusion:** horseshoe kidney concurrent with retrocaval ureter is a rare anomaly. If the isthmus of horseshoe kidney is between the abdominal aorta and IVC, it is likely that circumcaval ureter coexists.

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