CASE REPORT

A Case Report on Rare Congenital Anomaly: Crossed Fused Renal Ectopia

Sonali Satpute, Lakshmi Kiruba N, Swapna Parate

Abstract

Congenital anomalies of the kidney and urinary tract are relatively rare and can present with a wide range of clinical manifestations, making diagnosis and management challenging. A rare developmental anomaly of urinary system L-shaped kidney with bilateral Double pelvis and bifid ureter located anterosuperior to the bifurcation of abdominal Aorta at the level of L4. Resulted as a consequence of abnormal renal ascent in embryogenesis due to migration and fusion of left kidney to the lower pole of the right kidney. Surgeons must remain vigilant for potential complications, including infections, obstructive issues leading to stone formation, and concurrent malformations in other organs that may increase the risk of malignancy. Additionally, ectopically located kidneys are prone to injury during pelvic surgeries, particularly those involving the aorta, due to their anomalous blood supply originating from the abdominal aortaht, and environmental factors, such as other nutrients and hormones.

Keywords: Bifid ureter, Congenital anomaly, L-shaped Kidney, Renal ectopia.

Journal of Research in Medical and Interpathy Sciences. 2(2);2024

Background

Kidney is an excretory organ located retroperitoneally on either side of vertebral column. Developed from the metanephric blastema. Very rarely both kidneys are seen in same side and usually fused called crossed-fussed renal ectopia. In this case we found a rare developmental anomaly of urinary system L-shaped kidney on right side with double pelvis and bifid ureter located anterosuperior to the bifurcation of abdominal Aorta at the level of L4 formed by fusion lower pole of left kidney with the lower pole of right kidney.[1]

Very few authors have reported this type of rare and unique congenital anomaly of kidney.

Case Description

During routine dissection practical for 1st MBBS in the Department of Anatomy, SMBT IMS & RC, Nashik, MS, India, A 58-year-old male cadavers was dissected to expose the abdominal organs. Noticed lower pole of left kidney fused with lower pole of right kidney Infront of bifurcation of aminal aorta at the level of L4. The right kidney presented with double renal pelvis, two renal arteries arising from right side of abdominal aorta and one renal vein opening into right side of Inferior vena cava. Lower pole of Left kidney completely migrated to right side seen across the major blood vessels of abdomen in the midline with two hilum one on either side of vertebral column.

Accessory renal artery arises from left side of abdominal aorta enters the hilum present at the upper pole of the left kidney and left renal artery arises from the right side of Abdominal aorta enters hilum present near the lower pole of the left kidney. Accessory Renal vein from the upper pole of left kidney joins with left supra renal vein to form a common vein and opens into Inferior vena cava. Left renal opens into right side of Inferior vena cava.

Thus, left kidney presents double pelvis and bifid ureter, both the ureter joins to form single ureter anterior to the left kidney in midline to open into the urinary bladder as shown in (Figure 1).

Discussion

Development of kidney begins in the 4th week of intrauterine life from intermediate mesoderm, and undergoes changes from pronephros, mesonephros, and metanephros. The permanent kidney develops in the sacral region around 5th week, metanephric blastema forms the excretory system and ureteric bud forms the collecting system. At 9th week kidney undergoes rotation 90 degree medially and ascends up to reach adult position.[2,3]

Crossed fused renal ectopia arises from aberrant ureteric bud development, fetal rotation, and malalignment.

Department of Anatomy, SMBT IMS RC, Dhamangoan, Nashik, Maharashtra, India

Corresponding Author: Lakshmi Kiruba N

(Email: lakshmikiruba1@gmail.com)

Conflict of Interest: None

Source of Funding: None

How to cite: Satpute S, Kiruba LN, Parate S. A Case Report on Rare Congenital Anomaly: Crossed Fused Renal Ectopia. Journal of Research in Medical and Interpathy Sciences. 2024;2(2):82–84

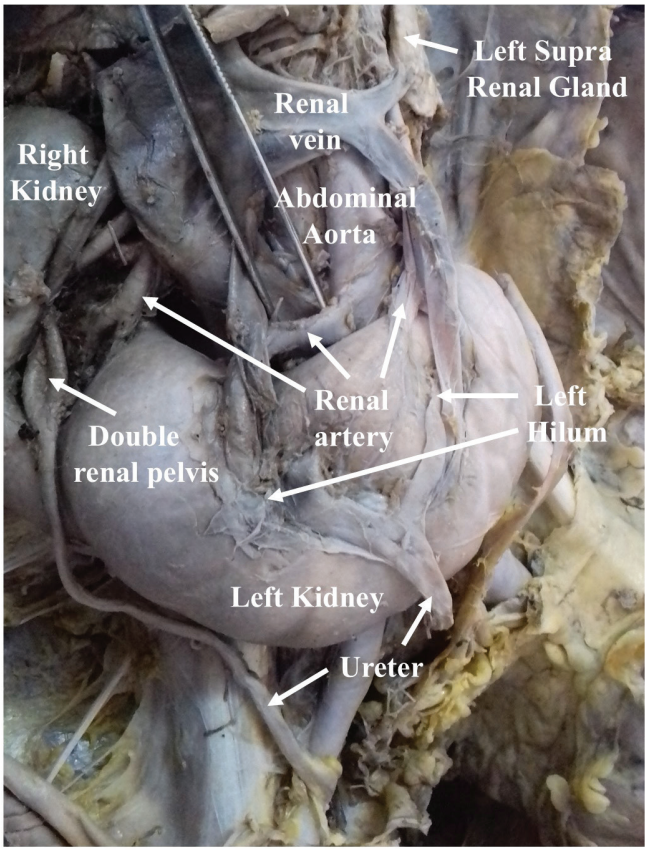

Figure 1: Crossed Fused Renal Ectopia

Renal fusion occurs during ascent and rotation, sometimes forming an L-shaped kidney. The ectopic kidney's ascent is limited by the inferior mesenteric artery, anchoring it against the L4 vertebra.[4,5]

Congenital kidney anomalies are relatively common, with renal fusion anomalies resulting from partial or complete fusion during embryonic development. These anomalies, including horseshoe kidney and crossed fused renal ectopia, often involve abnormalities in kidney position, migration, rotation, and vascular supply, and are more common in males.[6]

A retrospective autopsy study of 1,900 consecutive cases in the Indian population revealed a relatively low incidence of kidney anomalies compared to Western countries. The study identified nine cases (0.47%) of congenital renal anomalies, comprising: Horseshoe kidney (3 cases, 0.16%), Extrarenal calyces (2 cases, 0.11%), Renal ectopia with fusion (1 case), Trilobar kidney (1 case), Polycystic kidney (1 case), Unilateral renal agenesis (1 case) These findings suggest a lower prevalence of kidney anomalies in the Indian population compared to Western countries.[7]

In 1957 **James H McDonald** and **Don S McClellan**, American urologists classified renal ectopia into four types[8]

1. bilateral crossed renal ectopia without fusion
2. unilateral crossed renal ectopia
3. bilaterally crossed fused renal ectopia
4. crossed unfused renal ectopia

Further, he classified crossed fused renal ectopia into six subtypes according to the degree of fusion with normal kidney-

- Type A: the upper pole of the ectopic kidney fuses with the lower pole of the normal kidney.
- Type B: S-shaped kidney, the hilum of the ectopic kidney faces laterally and the normal kidney faces medially
- Type C: Both kidneys fuse over a wide margin with the ureter of the ectopic kidney crossing midline
- Type D: L-shaped kidney, in which the ectopic kidney is horizontally seen and fused with the lower pole of the normal kidney
- Type E: Extensive fusion of both kidneys forming a disc-shaped mass
- Type F: The ectopic kidney is placed above the normal kidney and fused with its upper pole.

In this case. we found Type D of crossed fused renal ectopia.

Vascular supply to the kidneys appears to be abnormal in all reported cases of renal ectopia. Arteries supplying either arise from the abdominal aorta or iliac artery, renal vein opens directly into inferior vena cava or indirectly through suprarenal vein.[9]

An ectopic kidney is more prone to injury during pelvic surgery, particularly in aortic procedures. Because the arterial supply arises from the abdominal aorta and its bifurcation, the entire kidney undergoes ischemic during proximal aortic cross-clamping.[10]

Yano *et al*. reported a rare case of crossed renal ectopia without fusion, associated with an abdominal aortic aneurysm. Due to the heightened risk of renal injury during surgical intervention, thorough preoperative evaluation is crucial. In patients with crossed renal ectopia, renal failure is a significant concern following vascular surgery. Therefore, the approach to renal preservation must be tailored to each patient's unique anatomical characteristics.[11]

Clinical Significance

Crossed fused renal ectopia is a very rare developmental genitourinary anomaly yet surgically it is important. As a surgeon, it is essential to understand the variations of the kidney with its vascular supply. When planning for any surgery related to abdomen and pelvis in patients with renal anomaly especially ectopic kidney care should be taken and appropriate sonography and radiological investigations to be done thoroughly to perform surgery successfully and to avoid any complications.

Acknowledgement

I would like to express my sincere gratitude to Dr. Swapna Parate, Professor and Head, Department of Anatomy, SMBT IMS RC, Dhamangoan, Nashik for her invaluable support and guidance.

References

1. Standring S, editor. Gray's Anatomy. 42nd ed. Philadelphia, PA: Elsevier; 2020.
2. Singh V. Text book of Clinical Embryology. 2nd ed. New Delhi: Elsevier; 2021.
3. Larsen WJ. Human Embryology. 6th ed. New York: Churchill Livingstone; 2020.
4. Shapiro E. Upper urinary tract anomalies and perinatal renal tumors. Clin Perinatol. 2014 Sep;41(3):679-94. doi: 10.1016/j.clp.2014.05.014. Epub 2014 Jul 18. PMID: 25155735.
5. Shapiro E, Bauer SB, Chow JS. Anomalies of the upper urinary tract. In: Wein AJ, Kavoussi LR, Novick AC, Partin AW, Peters CA, editors. Campbell-Walsh urology. 10th ed. Philadelphia: Saunders; 2012. p. 3140-5.
6. Babu CSR, Sharma V, Gupta OP (2015) Renal Fusion Anomalies: A Review of Surgical Anatomy. Anat Physiol 5: S5-001. doi: 10.4172/2161-0940.S5-001.
7. Gupta T, et al. Congenital Renal Anomalies in Indian Population. J Postgrad Med Educ Res 2017;51(1):12-16.
8. McDonald JH, McClellan DS. Crossed renal ectopia. Am J Surg 1957;93(6):995-1002. doi: 10.1016/0002-9610(57)90680-3 PMID: 13424850.
9. Simonds E, et al. Case Report of a Pelvic Crossed Fused Renal Ectopic Kidney. Kurume Med J 2019; 66:55-58.
10. Pasquali M, Sciascia N, D'Arcangelo Liviano G, La Manna G, Zompatori M. Pancake kidney: when it is not a problem. BJR Case Rep. 2018 Feb 16;4(3):20170117. doi: 10.1259/bjrcr.20170117. PMID: 31489215; PMCID: PMC6711276.
11. Yano H, Konagai N, Maeda M, Itoh M, Kuwabara A, Kudou T, Ishimaru S. Abdominal aortic aneurysm associated with crossed renal ectopia without fusion: case report and literature review. J Vasc Surg. 2003 May;37(5):1098-102. doi: 10.1067/mva.2003.184. PMID: 12756361.