Journal of Case Reports and Scientific Images

E-ISSN: 2708-0064
P-ISSN: 2708-0056
IJCRS 2024; 6(1): 24-27
www.allcasereports.com
Received: 02-01-2024
Accepted: 15-02-2024

Dr. Namdev Seth
Assistant Professor,
Department of Radiodiagnosis,
All India Institute of Medical Sciences, Gorakhpur,
Uttar Pradesh, India

Dr. Saumya Verma
BRD Medical College,
Gorakhpur, Uttar Pradesh,
India

Corresponding Author:
Dr. Namdev Seth
Assistant Professor,
Department of Radiodiagnosis,
All India Institute of Medical Sciences, Gorakhpur,
Uttar Pradesh, India

Clear cell renal cell carcinoma in crossed fused renal ectopia: A case report

Dr. Namdev Seth and Dr. Saumya Verma

DOI: https://doi.org/10.22271/27080056.2024.v6.i1a.75

Abstract
Crossed fused renal ectopia is an uncommon entity to be seen in clinical practice, owing to its silent clinical nature. Reports of renal malignancies in crossed fused ectopic kidneys have been made very rarely.
We are going to present the case of a 40-years old male patient, who came to our institute with the complaints of abdominal pain. His ultrasound scan showed a suspected mass lesion in right kidney. His left renal fossa was empty and left kidney was not visualized in expected ectopic areas. Further CT urography images show left to right crossed fused ectopia with an enhancing mass lesion in lower entity. Biopsy confirmed the diagnosis of clear cell renal cell carcinoma.
Crossed-fused renal ectopia (CRFE) with a renal malignancy is a very rare entity which presents a difficult task for the surgeon in surgical decision making, due to its location, complex vascular connections and collecting system. Hence, detailed imaging work-up with cross sectional and angiography imaging is necessary for surgeon to choose to best operative method, to avoid complications during and after surgery.

Keywords: Renal cell carcinoma, renal fusion anomalies, ectopic kidney, single kidney

Introduction
Crossed fused renal ectopia (CFRE) is rare congenital malformation of the urinary system, where both the kidneys are fused and located on same side of midline. Ureter of the crossed kidney opens into the urinary bladder on the contralateral side. The exact incidence of CFRE is unknown due to clinically silent nature of malformations; however, the estimated frequency is 1 in 1000–7500 [1, 2]. The symptoms of the crossed fused renal ectopia are mostly not obvious, and most Individuals are diagnosed only after they undergo investigations for other reasons. Incidence of renal cell carcinoma in crossed fused renal ectopia is further rare, with only 30 reports in PubMed journals during 1937-2018, with 11 of them being clear cell renal cell carcinoma [3]. Herein, we are going to report a rare case of clear cell renal cell carcinoma in crossed fused renal ectopia, along with review of relevant literature.

Case Report
A 40-years old male patient presented to our institute with the complaints of abdominal pain. His ultrasound scan was done at a peripheral clinic and reported a suspected mass lesion in right kidney along with empty left renal fossa and apparent non-visualization of left kidney. His medical and surgical histories were irrelevant, except the long-standing history of seizures. No history of diabetes and hypertension were present. Patient had never undergone any surgical intervention.
Physical examination showed tenderness in right lumbar region with no significant rigidity or guarding. No definite mass was palpable in abdomen. Rest of the examinations was unremarkable. The kidney function test and other routine lab investigations showed no significant abnormalities.
Subsequently patient presented in department of radio-diagnosis for evaluation of renal mass. Contrast-enhanced CT scan of abdomen and pelvis was performed. CT scan shows solitary kidney on right side with empty left renal fossa. Size of the visualized kidney was 13.1 x 6.2 cm. On careful assessment, two separate ureters were seen to arise from the right kidney which is seen to follow separate course on right and left side of midline. Ureter arising from upper pole was continuing as right ureter and the one arising from lower pole continuing as left ureter. Both ureters were terminated at vesico-ueretric junction on either side of midline - confirming the diagnosis of left to right inferior crossed fused renal ectopia (Figure 1).

Further assessment of CT images revealed a well-defined, nearly-spherical, iso- to hypodense (HU on non-contrast images ~ 30, as compared to normal renal parenchyma ~ 34) predominantly solid lesion at the lower pole of right kidney (Figure 2). Lesion measured 5.7 x 5.4 x 5.5 cm (AP x TR x CC) and showed significant enhancement (>20 HU) on post-contrast images with internal non-enhancing/minimally enhancing areas suggesting necrosis. Lesion was partially exophytic at anterior aspect with almost one-third appearing to be endophytic. Lesion was seen to extend to the inter-polar region (< 50% crossing the polar line) of right kidney. No e/o calcification/ visible fat within the lesion.

The lesion is seen in close proximity to the renal hilum abutting and causing mild displacement of the renal pelvis (no intervening fat plane) and the major calices. No any capsular breach or infiltration into adjacent structures seen.

Single renal artery is noted arising from abdominal aorta at the level of inferior endplate of L1 vertebrae, just inferolateral to origin of superior mesenteric artery at 11' O clock position (Figure 3). Renal artery is coursing posterior to the inferior vena cava. Single renal vein is noted, draining into inferior vena cava on right side at L1 vertebral level. Renal vessels at hilum appear normal - no any obvious invasion or thrombosis.

No e/o hydronephrosis is seen. Other abdominopelvic structures showed no significant abnormality.

Patient underwent USG-guided biopsy, which showed features suggestive of clear cell renal cell carcinoma (Figure 4).

Further course of patient is not available as patient opted to visit another center for surgical management.

Discussion

Crossed fused renal ectopia (CFRE) is a very infrequent congenital renal fusion anomaly characterized by fused bilateral kidneys, situated on one side of midline. The exact incidence of crossed fused ectopia is unknown, because of its asymptomatic nature. It has been reported more commonly in males. Reported male to female ratio in such cases is 3:2. It has also been reported that there are high chances of left kidney crossing the midline and fusing with right kidney than the vice versa [4]. Among renal fusion anomalies, horseshoe kidney is most frequent one, and is characterized by fusion of lower poles of both kidneys in midline. The midline tissue bridge is called isthmus. In crossed renal ectopia, one kidney completely crosses to the opposite side of midline. It can stay unfused or may get fused with the contralateral kidney, later is called crossed-fused renal ectopia. Ectopically located kidney, most commonly, fuses with lower pole of normally situated contralateral kidney.

The embryological basis of renal fusion anomalies is not completely understood. Many theories have been put forward over the years to explain the underlying mechanism of such fusion anomalies- (1) Mechanical theory says that upward migration of one of the kidneys is mechanically obstructed due to abnormally placed umbilical vessels (2) Ureteral theory – CRFE occurs due wandering of the ureteral bud to the contralateral side) (3) Teratogenic theory (4) Genetic theory (5) Theory of abnormal rotation of the caudal end of the fetus- this theory is supported by increased prevalence of spinal rotation anomalies in such cases. Another theory relates the abnormal development of the ureteric bud and the metanephric blastemal in early gestational age with renal fusion anomalies [1].

Patterns of renal fusion in such anomalies vary significantly. McDonald and McClellan [5] classified crossed fused renal ectopia in six anatomical variants. A- Inferior CRFE type, most common variant, in which, the upper pole of ectopically located kidney fuses with lower pole of normally placed kidney. B- S-shaped or sigmoid kidney where kidneys fuse like inferior CRFE pattern, however their hila face opposite to each other, making a S-shaped mass. C- Lump kidney - two kidneys fuses over a wide margin and ureter of ectopic kidney crosses the midline. D- Tandem or L-shaped kidney – horizontally placed ectopic kidney fuses with lower pole of normal kidney. E- Disc kidney - disc shaped mass is formed due to extensive fusion of two kidneys. F- Superior ectopia type- least frequent type, ectopic kidney is placed above the normally placed kidney.

Most of the patients with CRFE stay asymptomatic throughout the life; however fraction of them may present with complications, such as dilated pelvicalyceal system, calculi and pyelonephritis. Very rarely, CRFE may develop malignancy [4]. Renal cell carcinoma is most commonly associated neoplastic lesion with renal fusion anomalies. However, no significant difference has been reported in prevalence of neoplasm in fused kidneys in comparison to normal kidneys [6].

Typical management of renal cell carcinomas in patients with crossed fused renal ectopia is complete nephrectomy of both renal moieties [1]. However, in well-defined masses with accurate delineation of the renal vasculature through angiography may allow for complete excision of the affected renal moiety and at the same time, preserving the uninvolved kidney. Due to the uncertain and highly variable anatomy, careful pre-surgical planning with CT urography and angiography, ureteral stenting may be proved to be helpful [1]. Role of proper reporting of renal vasculature with other variant anatomy in the region helps in deciding the surgical management of such cases.

In approximately one-fourth of the CFRE cases, superior abdominal aorta gives origin to the renal arteries, whereas in rest of the cases, inferior abdominal aorta or the iliac arteries gives origin to renal arteries [7]. In our case, abdominal aorta was the originator of main renal artery.

Detailed understanding of the aberrant anatomy in CRFE cases is very important in pre-surgical decision making, to avoid possible complications during and after the interventions. Hence, preoperative imaging with renal urography and angiography images and accurate delineation of complex anatomy is critical on part of the radiologists.

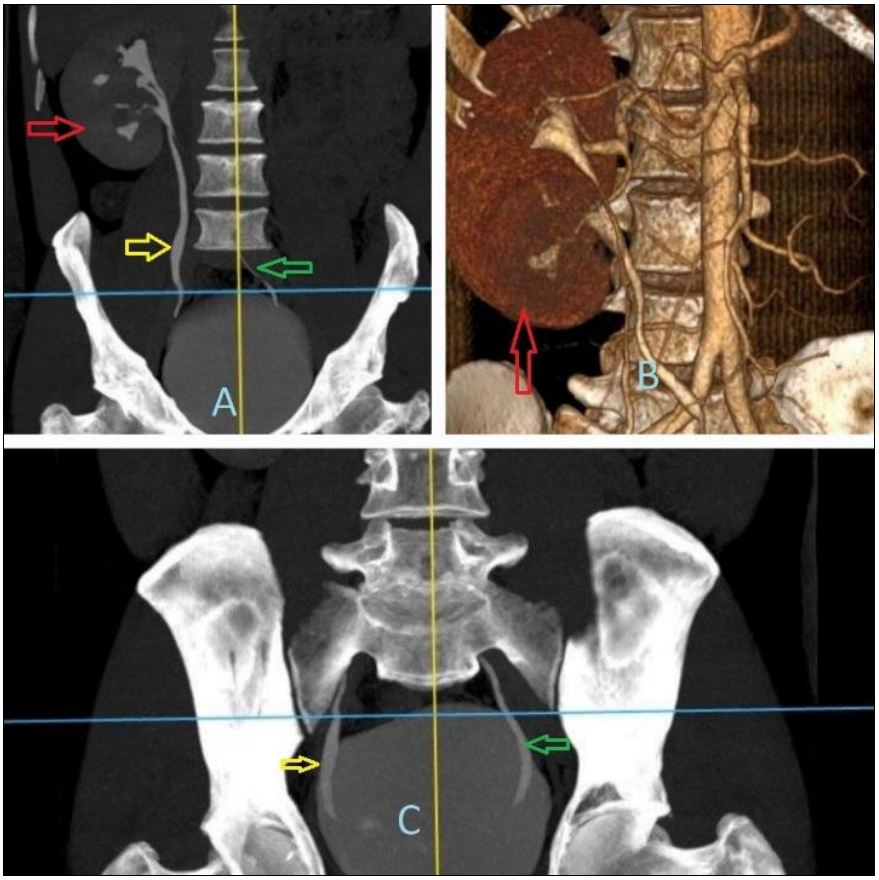

Fig 1: Maximum intensity projection (MIP) and VR images of CT Urography depicts single kidney (red arrow) on right, which is giving origin to two different ureters, coursing on right (yellow arrow) and left (green arrow) sides, to open on corresponding vesico-ureteric junctions- characteristics of crossed fused renal ectopia.

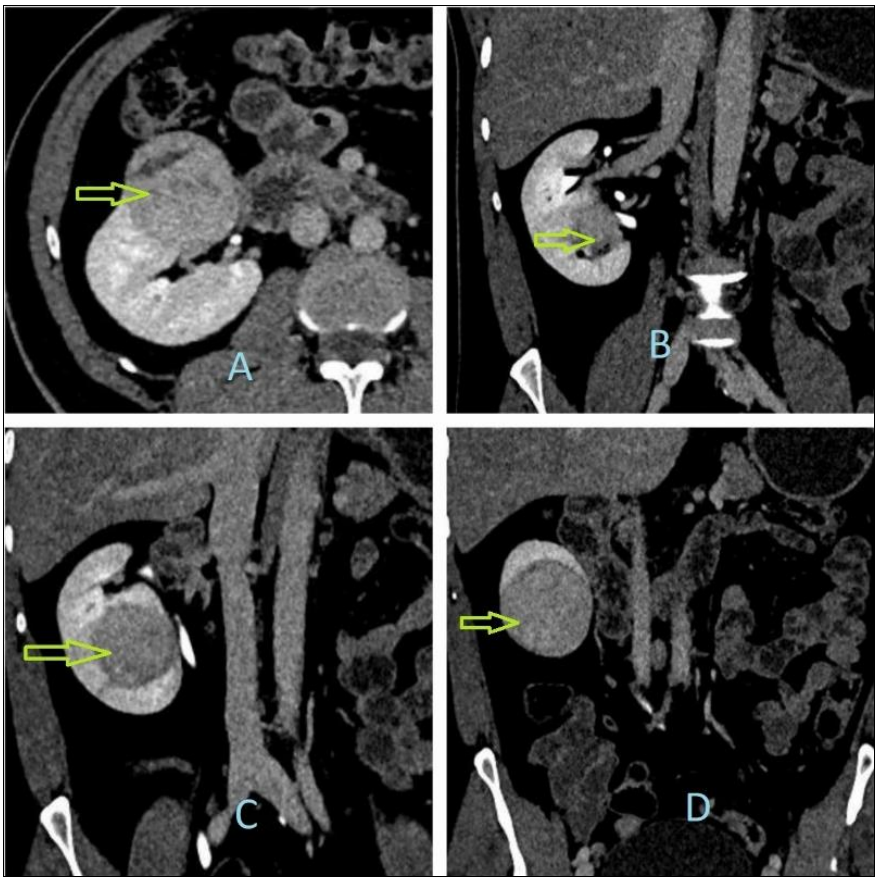

Fig 2: Contrast CT images of right kidney shows a well-defined, nearly spherical, iso- to hypodense, predominantly solid lesion (arrow) at the lower pole of right kidney with significant enhancement (>20 HU) on post-contrast images.

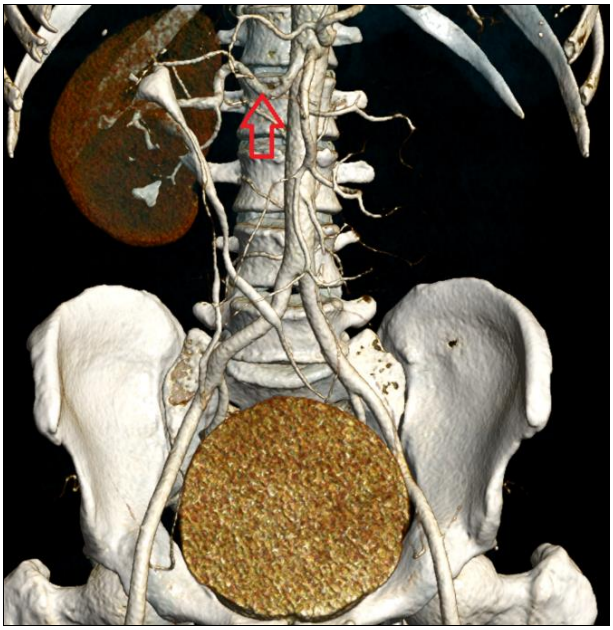

Fig 3: CT angiography image with VR reconstruction shows single renal artery (arrow) arising from abdominal aorta at the level of inferior endplate of L1 vertebrae

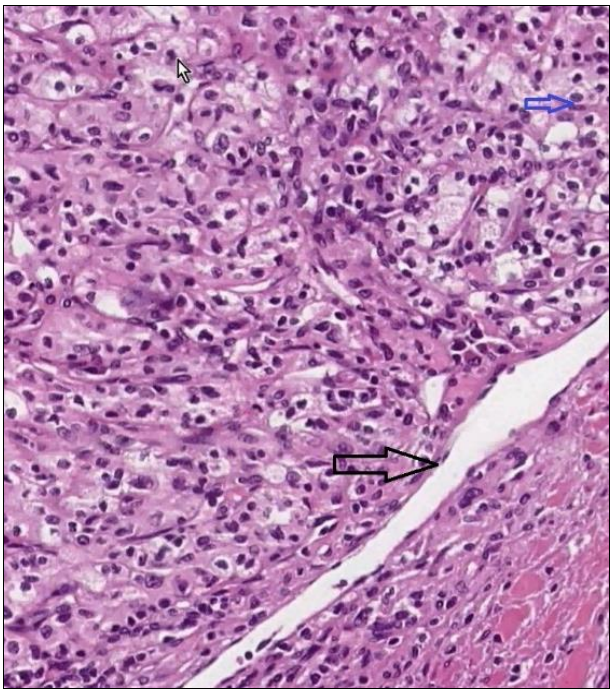

Fig 4: H&E stained section of biopsy sample shows tumour cells arranged in sheets and cord, traversed by small blood vessels (black arrow). Individual cells show centrally placed nucleus with abundant clear eosinophilic cytoplasm (blue arrow)- characteristics of clear cell renal cell carcinoma

Conclusion

Crossed-fused renal ectopia (CRFE) with a malignancy is regarded a very rare radiological diagnosis. It usually comes with a diagnostic challenge for the radiologists, and a surgical challenge for the urologists during intraoperative management, due to retroperitoneal location of the kidney, its complex arterial and venous connections as well as collecting system. Hence, Preoperative scanning with CT urography and angiography images allow the detailed reporting of complex and highly variable vascular connection and collecting system hence makes easier for surgeon in surgical decision making.

Conflict of Interest
Not available

Financial Support
Not available

List of Abbreviations

CRFE- Crossed fused renal ectopia
MIP- Maximum intensity projection
CT- Computed tomography
HU- Hounsfield unit
AP- Anteroposterior
TR- Transverse
CC- Craniocaudal

References

1. Stimac G, Dimanovski J, Ruzic B, Spajic B, Kraus O. Tumors in kidney fusion anomalies--report of five cases and review of the literature. Scandinavian Journal of Urology and Nephrology. 2004;38(6):485-489. DOI: 10.1080/00365590410018684
2. Solanki S, Bhatnagar V, Gupta AK, Kumar R. Crossed fused renal ectopia: Challenges in diagnosis and management. Journal of Indian Association of Pediatric Surgeons. 2013;18(1):7-10. DOI: 10.4103/0971-9261.107006
3. Cao Y, Zhang Y, Kang W, Suo N, Cui Z, Luo Y, et al. Crossed-fused renal ectopia with renal calculi: Two case reports and a review of the literature. Medicine (Baltimore). 2019;98(48):e18165. DOI: 10.1097/MD.0000000000018165
4. Mudoni A, Caccetta F, Caroppo M, *et al*. Crossed fused renal ectopia: case report and review of the literature. Journal of Ultrasound. 2017;20(4):333-337. DOI: 10.1007/s40477-017-0245-6.
5. McDonald JH, McClellan DS. Crossed renal ectopia. American Journal of Surgery. 1957;93(6):995-1002. DOI: 10.1016/0002-9610(57)90680-3
6. Akdogan L, Oguz AK, Ergun T, Ergun I. The Rarest of the Rare: Crossed Fused Renal Ectopia of the Superior Ectopia Type. Case Reports in Nephrology. 2015;2015:742419. DOI: 10.1155/2015/742419
7. Sharma V, Ramesh Babu CS, Gupta OP. Crossed fused renal ectopia multidetector computed tomography study. International Journal of Anatomy and Research. 2014;2(2):305-309. Available from: https://www.ijmhr.org/ijar_articles_vol2_2/IJAR-2014-426.pdf

How to Cite This Article
Seth N, Verma S. Clear cell renal cell carcinoma in crossed fused renal ectopia: A case report. Journal of Case Reports and Scientific Images. 2024;6(1):24-27.