

Rare Presentation of Crossed Fused Ectopia with Single Ureter and Ureteropelvic Junction Obstruction: A Case Report

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ABSTRACT

Crossed fused renal ectopia (CFRE) is a congenital anomaly in which one kidney is ectopically located on the opposite side of the midline and fused with its contralateral counterpart. However, CFRE with a single ureter draining both renal moieties is an exceptionally rare variant. We report on the case of a 12-year-old boy who presented with recurrent episodes of right flank pain. Ultrasonography and subsequent contrast-enhanced CT revealed that the left kidney was located on the right side, fused with the right kidney. Both renal pelvises were fused, forming a single pelvicalyceal system that drained through a solitary ureter entering the urinary bladder on the right side. This case highlights the characteristic imaging features of this unusual congenital anomaly, emphasizing the crucial role of CT in its accurate diagnosis and preoperative evaluation.

Keywords: Renal ectopia; CT scan; Radiology; Pakistan

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Introduction

Crossed fused renal ectopia (CFRE) is a congenital anomaly often detected incidentally on routine imaging, characterized by both kidneys located on the same side of the body and partially or completely fused¹. Typically, each kidney in CFRE is drained by its own ureter, which empties separately into the bladder. Rarely, however, CFRE is drained by a single ureter a highly uncommon variant with only a few cases reported in the literature². This anomaly is usually asymptomatic but may present with episodic flank pain, dysuria, hematuria, or other nonspecific symptoms when obstruction occurs³.

We report a rare case of crossed fused renal ectopia with a single ureter and UPJ obstruction.

Case Presentation

A 12-year-old boy was referred to the Radiology Department of a tertiary care hospital with recurrent right flank pain described as dull, colicky, and radiating toward the groin. He also reported intermittent, undocumented fever during these episodes but had not sought prior medical evaluation. At presentation, he was clinically stable, and general and systemic examinations were unremarkable. Hematological and urinary investigations were within normal

limits. There was no history of prior medical or psychiatric illness, and no family history of hereditary or congenital disorders. The patient belonged to a middle-class socioeconomic background. Ultrasound revealed the left kidney measuring $9.8 \times 3.8 \times 0.9$ cm in the left paraumbilical region, with its lower pole crossing the midline. Moderate hydronephrosis was noted up to the ureteropelvic junction (UPJ), with reduced cortical thickness. The right kidney was normal in size ($10.5 \times 3.8 \times 1.6$ cm), with its lower pole fused with the left kidney, initially suggesting a horseshoe kidney.

Contrast-enhanced CT scan demonstrated both kidneys positioned on the right side. The left kidney was malrotated, crossing the midline to fuse with the lower pole of the right kidney at the level of L3, with no intervening isthmus. The left kidney exhibited moderate hydronephrosis and hydropelvis (AP diameter 1.5 cm), with PUJ obstruction. The left renal pelvis was fused with the right renal hilum, which was oriented anterolaterally (L-Shaped fusion). Contrast pooling was normal in both kidneys; however, there was no excretion from the left kidney. Normal excretion was observed from the right ureter, which had an orthotopic insertion. The left ureter was not visualized or opacified, consistent with a single ureter draining both renal moieties. The urinary bladder was normal, with appropriate contrast opacification in the delayed phase.



Figure 1: Contrast enhanced axial image showing normal enhancement of renal parenchyma with crossing and fusion of lower pole with right kidney



Figure 2: 3D reconstruction of delayed contrast image showing crossed fused ectopia (L-Shaped).

Discussion

Crossed fused renal ectopia (CFRE) is a rare congenital anomaly, with an estimated incidence ranging from 1:1300 to 1:7500. Despite its rarity, advancements in imaging techniques have led to more frequent incidental diagnoses. As highlighted in the case reports by Jallouli et al. (2018) and Ahmad (2007), CFRE is characterized by the presence of both kidneys on the same side of the body, often fused. Our patient's case, where the left kidney was found on the right side, fused with the right kidney, aligns with this characteristic presentation^{4,5}.

While CFRE typically involves two ureters, each draining one kidney and inserting into the bladder separately, our case presented a particularly rare variant: a solitary ureter draining both fused renal moieties. Kumawat et al. (2020) also reported a similar, exceedingly rare instance of CFRE with a solitary ureter crossing the midline, further emphasizing the unusual nature of this anatomical variation⁶. The embryological basis of CFRE is

complex and not fully understood. Theories often involve abnormal migration and fusion of the ureteric bud and metanephric blastema during the 4th to 8th weeks of gestation⁴. In our patient, the non-visualization or non-opacification of the left ureter in conjunction with a single draining ureter on the right side suggests a unique deviation from typical ureteral development in CFRE.

Clinically, CFRE can be asymptomatic, as noted in the report by Sadiqi and Rasouly (2016)⁸, or it can present with complications such as hydronephrosis, urinary tract infections, or renal calculi, as seen in our patient with recurrent right flank pain. The presence of moderate hydronephrosis and a hydropelvis in the left kidney, along with pelviureteric junction (PUJ) obstruction, underscores the clinical significance of these anomalies. This aligns with findings from Dev et al. (2019)⁷ and Sadiqi and Rasouly (2016)⁸, who also reported CFRE cases complicated by UPJ obstruction and hydronephrosis.

Imaging plays a pivotal role in the diagnosis and detailed characterization of CFRE. Ultrasonography can initially suggest the diagnosis by revealing an empty renal fossa on one side and fused kidneys on the contralateral side⁶. However, contrast-enhanced CT (CECT) is crucial for a comprehensive evaluation, providing detailed anatomical information regarding renal fusion, malrotation, vascular supply, and the course and insertion of the ureters. This was vital in our case to confirm the diagnosis of crossed fused renal ectopia, identify the single draining ureter, and delineate the PUJ obstruction. The 3D reconstruction from CECT further enhanced our understanding of the complex anatomical arrangement. Previous studies similarly emphasize the importance of advanced imaging modalities like CT and MRI for accurate diagnosis and preoperative planning^{4,5}.

Management of CFRE is largely individualized, depending on the presence and severity of symptoms and associated pathologies. As reported in a study, asymptomatic cases may be managed

conservatively with regular follow-up⁴. However, symptomatic cases, particularly those with obstruction, often require intervention. The findings of PUJ obstruction in our case highlight the need for careful consideration of surgical options, to prevent further renal damage^{7,6}.

The rarity of CFRE with a solitary ureter draining both moieties necessitates thorough investigation and reporting of each case. These unique presentations contribute valuable insights into the spectrum of renal anomalies and aid in refining diagnostic and management strategies. Our case, consistent with the existing literature on CFRE but notable for its specific ureteral anomaly, further emphasizes the importance of advanced imaging for precise anatomical mapping and tailored patient care.

Thus, ultrasound serves as the initial or first-line imaging modality for the detection of abdominal anomalies due to its noninvasive nature, wide availability, and absence of ionizing radiation. It plays a crucial role in the early identification of pathologies, particularly in resource-limited settings⁹⁻¹¹. However, when further characterization or confirmation of findings is required, computed tomography (CT) becomes an essential next step. CT provides superior anatomic detail, higher spatial resolution, and cross-sectional imaging capability, making it invaluable for diagnosing and staging complex renal and abdominal conditions. This approach is not only widely adopted worldwide but is also routinely practiced in our region of Pakistan, where CT remains a cornerstone in the evaluation and management of challenging renal cases¹²⁻¹⁶. Thus, radiology continues to play a pivotal role in both the diagnosis and management of renal diseases, guiding clinical decisions and improving patient outcomes.

Conclusion

This case illustrates an exceptionally rare variant of crossed fused renal ectopia for which contrast-

enhanced CT-scan can play a pivotal role in identifying the precise anatomical configuration, fusion pattern, and drainage anomaly. Early detection of such anomalies can be improved through routine prenatal and pediatric ultrasonography with careful assessment of renal position and ureteric drainage patterns. Recognition of this rare congenital anomaly is essential to prevent misdiagnosis, guide appropriate intervention, and preserve renal function.

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