CASE REPORT

Incidentally Detected Right-to-Left Crossed-Fused Renal Ectopia: A Case Report with Anatomical and Radiological Correlations

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Abstract

Crossed fused renal ectopia is an uncommon congenital renal anomaly in which both kidneys lie on the same side and are fused. In many patients, it causes no symptoms and is discovered as an incidental finding. We report a patient in whom the diagnosis was made during the investigation of lower abdominal pain.

A 36-year-old woman presented with a 3-day history of lower abdominal pain. Clinical examination and routine laboratory investigations were normal. Abdominal ultrasonography showed that the right kidney was not in its normal position or pelvis and raised the possibility of crossed fused renal ectopia. CT urography demonstrated a right-to-left crossed fused kidney, with the right kidney fused to the lower pole of the normally positioned left kidney. Both kidneys enhanced normally. There was no hydronephrosis, hydroureter, urinary calculus or any other urinary tract abnormality. The two ureters followed a normal course and entered the urinary bladder separately. Her abdominal pain improved with oral analgesics. The renal anomaly was unrelated to her symptoms and required no treatment.

Crossed fused renal ectopia is usually an incidental finding and can be suspected on ultrasonography. CT urography confirms the diagnosis and provides a clear demonstration of the renal parenchyma, collecting system, ureters and vascular anatomy, allowing an accurate identification and helping with future surgical planning when required.

Keywords: crossed fused renal ectopia; CT urography; congenital renal anomaly

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Introduction

Renal ectopic anomalies present with either a crossed fused state or a crossed unfused state. Out of these two anomalies, crossed-fused renal ectopia (CFRE) is the most prevalent, accounting for 1: 1000 live births, and occurs more frequently in males than females (1,2). Crossed fused renal ectopia is a rare congenital anomaly in which one side of the kidney is fused with the other side of the kidney while the two ureters open separately into the usual position at the trigone of the bladder (1, 3, 4). This condition is due to abnormal renal ascent and fusion during intrauterine development. However, its exact underlying pathogenesis remains unclear (3-5).

Most patients are asymptomatic, and the anomaly is often detected as an incidental finding during imaging performed for some other unrelated condition (1, 6, 7). When symptomatic, patients may present with flank or abdominal pain, haematuria, fever, recurrent urinary tract infections, or symptoms related to obstruction of the pelvicalyceal system (5, 7-9). Patients with CFTR can also develop complications such as hydronephrosis, urinary tract infections, and pelvi-ureteric junction obstruction (2, 5, 7-9). Multidetector Computed Tomography Urography (CTU) has become the imaging modality of choice for accurately demonstrating the anatomy, collecting systems, vascular supply, and associated complications of renal fusion anomalies (1, 10).

We report a case of right-to-left crossed fused renal ectopia diagnosed incidentally in a 36-year-old woman undergoing CTU examination for lower abdominal pain.

Case Presentation

We report the case of a 36-year-old female who presented with a 3-day history of lower abdominal pain. There was no history of fever, dysuria, haematuria, or urinary frequency. The physical examination was unremarkable. Initial laboratory investigations, including a full blood count, a urine full report, and renal function tests, were within normal limits.

An abdominal ultrasound (USS) was performed to investigate the cause of her lower abdominal pain. USS revealed the absence of the right kidney at its normal anatomical position in the right renal fossa, and no ectopic right kidney was found in the pelvis. The left kidney was present in its normal position but was enlarged, measuring about 14.0 cm in bipolar length. Two separate renal hila were identified. An additional renal moiety was observed fused to the lower pole of the left kidney, suggestive of right-to-left crossed-fused renal ectopia (Figure 1). No renal calculi, hydronephrosis or hydroureter were observed in either renal moiety. An intrauterine contraceptive device (IUCD) was detected within the uterine cavity. No other significant abdominal or pelvic abnormality was identified.

The patient was referred to the Urology team and CTU was performed for further evaluation the following day. CTU revealed the diagnosis of crossed fused renal ectopia, right to left, with the right kidney on the left side fused to the lower pole of the orthotopic left kidney. Both kidneys showed homogeneous contrast enhancement without calculi, hydronephrosis or hydroureter. Both ureters were of normal course and opened separately in the urinary bladder at the usual sites (Figures 2a, 2b, 3, 4a and 4b). No other congenital urinary tract anomalies were identified.

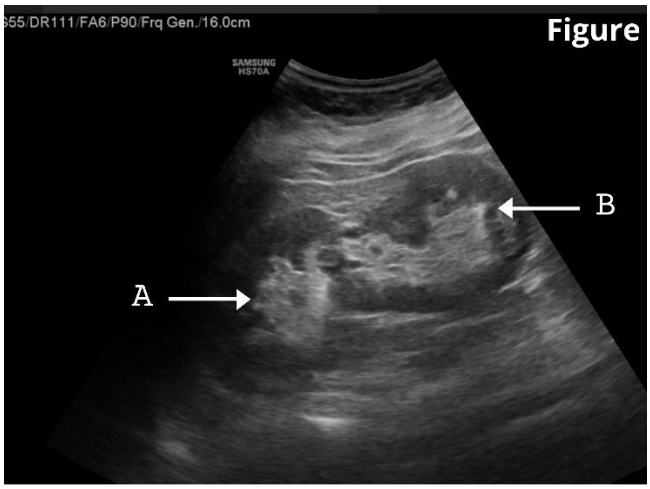


Figure 1: Ultrasonography of the abdomen demonstrating crossed fused renal ectopia. The fused renal mass is located on the left side. (A) Upper renal moiety. (B) Lower renal moiety. No hydronephrosis or renal calculi are identified.

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Her abdominal pain was relieved with oral analgesics and conservative management. As the crossed fused renal ectopia was an incidental finding and not related to her presenting complaint, no surgical or gynaecological intervention was required. She was advised to come to the urology clinic for follow-up if needed.

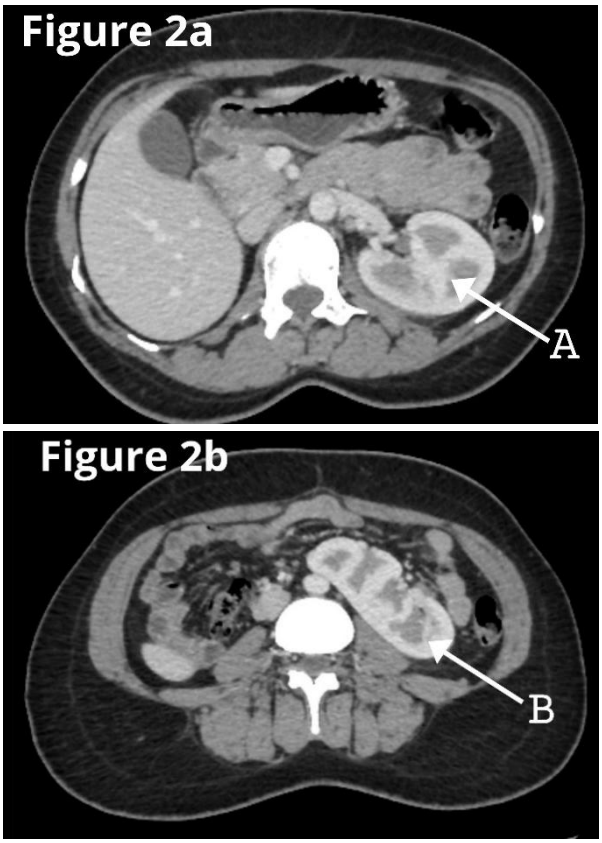


Figure 2: ***Axial venous-phase CT urography images demonstrating right-to-left crossed fused renal ectopia. Figure 2a: (A) Upper renal moiety. Figure 2b: (B) Lower renal moiety.***

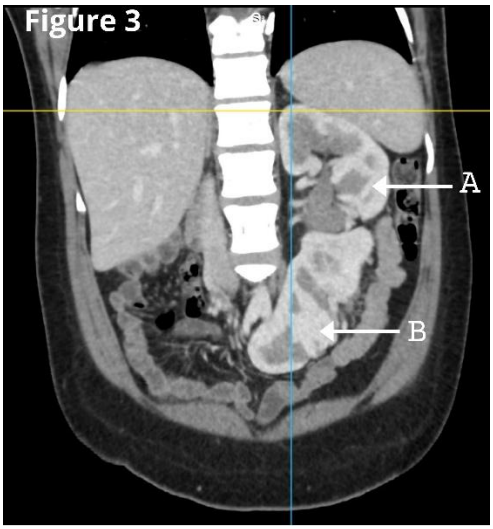
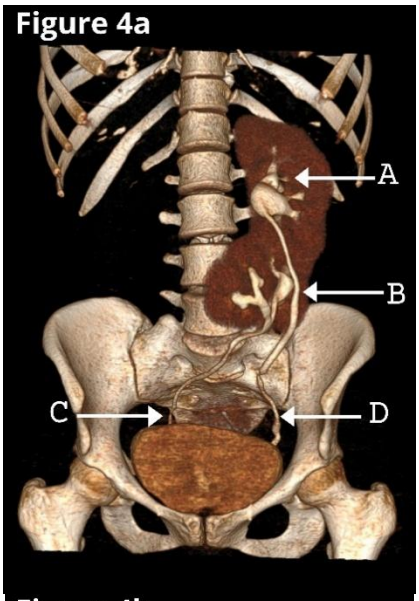


Figure 3: Coronal reconstructed venous-phase CT urography image demonstrating the fused left-sided renal mass in right-to-left crossed fused renal ectopia. (A) Upper renal moiety. (B) Lower renal moiety.



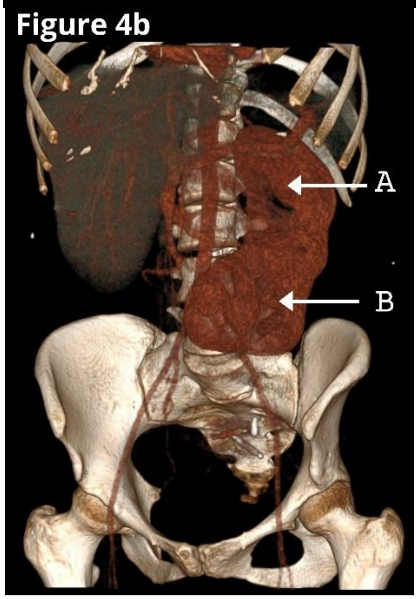


Figure 4: Three-dimensional volume-rendered CT urography images demonstrating right-to-left crossed fused renal ectopia.
Figure 4a: (A) Upper renal moiety. (B) Lower renal moiety. (C) Ureter of the lower renal moiety. (D) Ureter of the upper renal moiety.
Figure 4b: Three-dimensional volume-rendered CT urography demonstrating the vascular supply of the crossed fused renal ectopia. (A) Upper renal moiety. (B) Lower renal moiety.

Discussion

Crossed fused renal ectopia (CFRE) is a rare congenital anomaly in which both kidneys are located on the same side of the body, and their renal parenchyma is fused. The ureter of the ectopic kidney crosses the midline before entering the bladder at its usual contralateral vesicoureteric junction. Left-to-right ectopia is approximately three times more common than right-to-left ectopia (1, 7). In our patient, right-to-left crossed fused ectopia was present, with the superior pole of the right kidney fused to the inferior pole of the left kidney and two separate renal hila.

Renal fusion anomalies may be classified as partial or complete. Horseshoe kidney and CFRE are considered partial fusion anomalies, whereas the "cake kidney", also known as a fused pelvic kidney, represents complete fusion (1, 11). Horseshoe kidney is the most common renal fusion anomaly and is characterised by fusion of the lower poles, usually by an isthmus crossing the midline (1). In a cake kidney, the two renal masses are completely fused into a single pelvic mass, usually with two separate collecting systems and ureters (11). According to the McDonald and McClellan classification, CFRE can be divided into six anatomical variants: inferior crossed-fused ectopia, sigmoid (S-shaped) kidney, unilateral fused kidney, unilateral disc kidney, L-shaped kidney, and superior crossed-fused ectopia (7, 11) (Figure 5).

Generally, each kidney is supplied by a single renal artery arising from the abdominal aorta at the L1-L2 vertebral level just below the origin of the superior mesenteric artery. The right renal artery typically passes posterior to the inferior vena cava. In contrast, the blood supply to ectopic kidneys is highly variable and arises from the abdominal aorta, the aortic

bifurcation, the common iliac arteries, or the internal iliac arteries (1, 7).

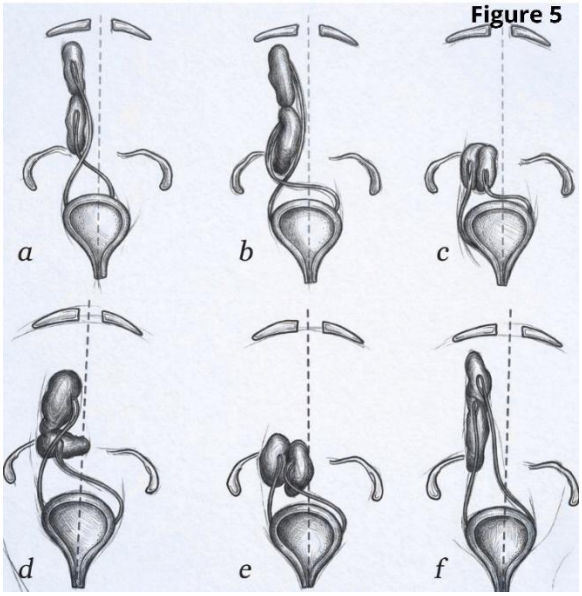


Figure 5: ***Diagrammatic illustration of the McDonald and McClellan classification of crossed fused renal ectopia (CFRE). (A) Inferior crossed fused ectopia. (B) Sigmoid (S-shaped) kidney. (C) Unilateral lump kidney. (D) L-shaped kidney. (E) Unilateral disc kidney. (F) Superior crossed fused ectopia. Illustration by K. Danansooriya, adapted from the original McDonald and McClellan classification***

Both the orthotopic kidney and the crossed-fused kidney have aberrant arterial supply (1, 12). Although the total number of arteries can vary from 1 to 6, most often there are 2-4 major arteries to each kidney. Accurate assessment of this vascular anatomy is particularly important when planning surgical or endovascular intervention.

Embryological development of the kidney system begins in the 4th week of gestation as a nonfunctional rudimentary system in the cervical region, the pronephros, which later regresses completely. This is followed by a temporary functioning system, the mesonephros, extending from the thoracic to the upper lumbar region. The mesonephros functions temporarily until the definitive kidney, the metanephros, develops during the 5th week of gestation (3, 4). The mesonephros contains the functional excretory units and its own collecting duct, called the Wolffian duct. Most of the Wolffian duct disappears except in males, where it gives rise to genital ducts (epididymis, vas deferens, and ejaculatory ducts). By the 5th week, the definitive kidney develops, with a unique dual origin from the ureteric bud and the metanephric blastema. The ureteric bud gives rise to the ureter, renal pelvis, calyces and collecting ducts, while the metanephric blastema forms the nephrons of the kidney (3, 4). Ascent of the kidneys from their pelvic position to definitive position at the L2 vertebral level occurs during the 8th week of gestation. During the ascent, the kidneys undergo 90° medial rotation, resulting in the anteriorly facing hilum moving medially (3, 4).

Although crossed renal ectopia was first described in 1654 by Pamarolus, the mechanism by which it forms remains unclear (6). Among the suggested theories, the most accepted are the mechanical theory and the ureteral theory (6, 13). According to the mechanical theory, the anomalous positioning of the umbilical arteries creates a physical barrier to the normal cranial ascent of the kidneys. Consequently, the metanephric blastema shifts along the pathway of least resistance, leading to migration to the contralateral side (13). According to the ureteric theory, the aberrant renal ureteric bud crosses the midline and induces the contralateral metanephric blastema for its nephrogenic differentiation (13). Consequently, the ipsilateral metanephric mesenchyme, deprived of inductive signalling, undergoes physiological regression. Malalignment and abnormal rotation of the

kidneys, genetic influences, and teratogenic factors are also considered to play a role (6).

Most patients with CFRE remain asymptomatic throughout life. The anomaly is usually detected incidentally during abdominal imaging or antenatal USS (6). When symptoms occur, they are usually related to associated complications rather than the anomaly itself (5, 11). Recurrent abdominal or flank pain is one of the most common presenting complaints, mainly in children and adolescents (5, 11). Other presentations include haematuria, dysuria, recurrent urinary tract infections, hypertension and, occasionally, a palpable abdominal mass (5–8, 11).

CFRE may be associated with a wide range of co-existing urogenital and systemic congenital anomalies. These genitourinary malformations affect both sexes and include vesicoureteral reflux ectopic urethral orifices, pelviureteral junction obstruction, hydronephrosis, ureteroceles, hypospadias, cryptorchidism, uterine and vaginal agenesis, such as Müllerian agenesis and Mayer-Rokitansky-Küster-Hauser syndrome (5–7, 11). In older patients, abnormal renal rotation and impaired urinary drainage predispose to urinary stasis, recurrent infection, and stone formation (8). Although not frequently reported, renal cell carcinoma is also associated with these renal fusion anomalies (14).

Furthermore, in paediatric patients, non-urological anomalies such as anorectal malformations, congenital heart disease and skeletal abnormalities. CFRE may also be associated with syndromic conditions such as VACTERL (10).

The structural overlap of anomalies in CFRE is due to the synchronised embryological development of the urinary and reproductive tracts within the shared intermediate mesoderm during the 4^{th}–8^{th} weeks of gestation. Due to this close anatomical proximity, a defect in localised development during this embryonic window can affect multiple organ systems at once. In our patient, none of these abnormalities were present.

USS is usually the first imaging modality, especially in children, due to its lack of radiation, lower cost, greater availability, and rapid results (5, 7, 11). When combined with Doppler technology, USS provides information on arterial supply and venous drainage (1). However, CT or MR angiography is often required for detailed vascular assessment before intervention (10).

Findings of USS include an empty renal fossa on one side and fused renal tissue on the opposite side. Associated hydronephrosis or renal calculi may also be detected (13).

A classic feature of ultrasound detected during CFRE diagnosis is the characteristic ‘notch’ between two fused kidneys, and in colour Doppler scans, a jet appearance is observed during the normal finding of bilateral urine flow to the bladder trigone (7).

CTU and MR urography (MRU) with advanced three-dimensional reconstruction are essential for defining urinary tract anatomy, vascular supply, and other associated genitourinary anomalies (1, 2, 10). CTU accurately demonstrates crossed fused renal ectopia by showing the malrotated fused kidneys and the characteristic course of the ureters, which descend before curving towards their normal openings in the urinary bladder (1).

CTU and MRU, combined with advanced three-dimensional reconstruction, are essential to diagnose urinary tract and genitourinary anomalies (1, 2, 6, 7). CTU is an extremely

useful modality to recognise cross-fused renal ectopia by demonstrating anterior or medial malrotation of both renal pelves and ureters that descend and diverge before emptying into the bladder (2, 6). Although CTU provides detailed visualisation of the renal parenchyma and vascular supply, its use is limited by higher radiation exposure, particularly in pediatric and pregnant populations. For these patients, MRU or renal pyelogram is preferred.

A micturating cystourethrogram (MCUG) is indicated when vesicoureteric reflux is clinically suspected, as USS cannot reliably identify the reflux. Dynamic renal scintigraphy using technetium-99m-labelled DTPA or MAG3 can assess differential renal function and urinary drainage of each renal moiety. Technetium-99m-DMSA imaging evaluates differential cortical function and detects renal cortical defects or scarring (11, 13). Intravenous urography (IVU) may demonstrate the collecting system and the characteristic crossing of the ureters, although it has largely been replaced by CTU and MRU.

Conclusion

Crossed fused renal ectopia is a rare congenital disorder that is commonly detected incidentally during medical imaging. However, some people may present with flank pain, haematuria, and recurrent urinary tract infections. As this condition may be associated with other genitourinary abnormalities and congenital syndromes such as VACTERL, early diagnosis and evaluation are essential. As demonstrated in this case, CTU is essential for accurately identifying the complex fusion of renal parenchyma, vascular anatomy, and aberrant collecting system. Radiological evaluations such as USS, CTU, MRU, and renal angiogram are crucial before any surgical intervention. In these patients, the functional aspect of the kidneys remains intact even though there is a structural disruption to the typical kidney architecture. Since the prognosis of our patient was good, surgical interventions were not indicated, and treatments are mainly focused on symptomatic relief and a follow-up plan.

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Conflicts of interest

The authors declare no conflict of interest.

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Availability of data and materials.

The data supporting the findings of this case report are available from the corresponding author upon reasonable request.

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