Crossed Fused Renal Ectopia with Transitional Cell Carcinoma of Kidney: A Rare Association

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ABSTRACT

Crossed fused renal ectopia is a rare congenital fusion anomaly of the kidneys where one kidney is fused with the contralateral kidney. Most of the cases are asymptomatic and present as an incidental finding secondary to obstruction, stones, and tumours. Crossed fused kidneys do not need separation, and treatment is directed towards the cause of symptoms.

We hereby present a case of right-to-left crossed fused renal ectopia with transitional cell carcinoma of the kidney and renal calculi, which was managed by radical nephroureterectomy. This patient presented with lumbar pain, hematuria, and repeated transfusions due to low haemoglobin. Unlike all types of crossed-fused renal ectopia, this patient's ureters were lying on the same side and were fused at the distal end. The main objective of this case report is to describe the rare association of crossed-fused renal ectopia with transitional cell carcinoma and renal stones and to describe the treatment plan for the case.

Key Words: *Crossed fused renal ectopia, Transitional cell carcinoma, Renal calculi.*

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INTRODUCTION

Crossed-fused renal ectopia (CFRE) is a rare congenital fusion anomaly and presents as an incidental finding. Patients are often presented with associated conditions, including pelvic ureteric junction obstruction, stones, and tumours.[1] Stasis results in recurrent infections with the consequences of stones; both infection and stones result in epithelial changes with malignant transformation. Transitional cell carcinoma (TCC) of CFRE is a rare occurrence. The standard treatment is nephroureterectomy with bladder cuff excision.

We hereby present a case of right-to-left CFRE with TCC of the kidney and renal calculi which was managed by radical nephroureterectomy without bladder cuff excision.

CASE REPORT

A 70-year male with no known comorbidities and no known addiction presented with gross painless hematuria associated with clots for four months. Hematuria was significant and required multiple blood transfusions during the past two months. His lowest reported haemoglobin was 5.4 g/dl. His baseline serum creatinine was 1.6 mg/dl. He has had multiple urine tests, which showed significant hematuria with >20/hpf of red blood cells.

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He underwent a non-contrast CT (CT-KUB) which showed a right-to-left CFRE with multiple stones and soft tissue mass in the lower kidney (Figure 1A). The CT urogram showed CFRE on the left side and soft tissue mass with contrast enhancement arising from the lower kidney and multiple stones (Figure 1B). Two ureters were traced until the pelvic brim, after which they joined and opened as a single ureter in the bladder (Figure 1C).

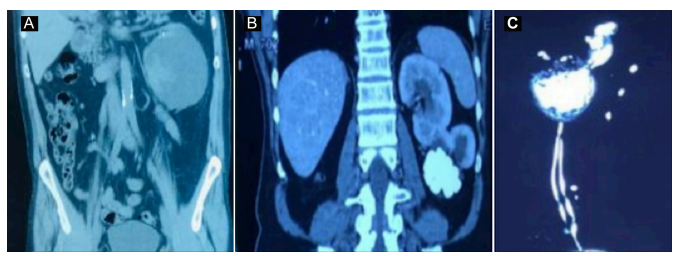

Figure 1: (A) CT KUB (coronal image) showing hydronephrotic lower kidney with soft tissue mass. (B) Contrast-enhanced CT abdomen showing multiple stones in the pelvis. (C) 3-D reconstruction CT showing both ureters joining together to form a single ureter near the bladder.

After the initial preoperative evaluation, the patient underwent a cystoscopy, followed by radical nephroureterectomy. Cystoscopy showed normal bladder anatomy with an absent right ureteric orifice and a normally placed left ureteric orifice. A stent was placed; however, retrograde pyelography was not performed at the time of stent placement.

The position of the patient was changed to right lateral. The left flank incision was made. Dissection was done until the renal hilum and cleavage plane of both kidneys were identified (Figure 2A). Intraoperatively, there was a crossed-fused ectopic lower kidney, and the upper kidney had a separate vascular supply and a separate ureter (Figure 2B). The stent was found to be in the lower pelvicalyceal system (the one with the

tumour). Both kidneys were separated without opening the pelvicalyceal system of the upper kidney (Figure 2C). Renorrhaphy of the upper kidney was done after separating both kidneys, and a stent was placed.

He made an unremarkable recovery and was discharged on the fourth postoperative day. His drain had insignificant output daily, and both the catheter and drain were removed before discharge. Intravesical mitomycin 40 mg in 50 ml of normal saline was instilled in a standard technique on the third postoperative day. Histopathology showed papillary urothelial carcinoma with a micropapillary component (at least 10%) and divergent differentiation (squamous).

Subsequently, he was planned for DJ removal, and retrograde pyelography was done. Cystoscopy findings confirmed an absent right ureteric orifice and a single normally placed left ureteric orifice, with the distal end of the DJ stent coming from it. Retrograde pyelography confirmed a single ureter on the left side draining the upper kidney with a few mm portion of nephrectomized ureter attaching the single distal ureter (Figure 2D). Postoperatively, the patient's creatinine is in the range of 2–3 mg/dl, while the recently reported creatinine is 2.7 mg/dl. We are planning this patient for close follow-up and surveillance because no bladder cuff was taken during radical nephro-ureterectomy.

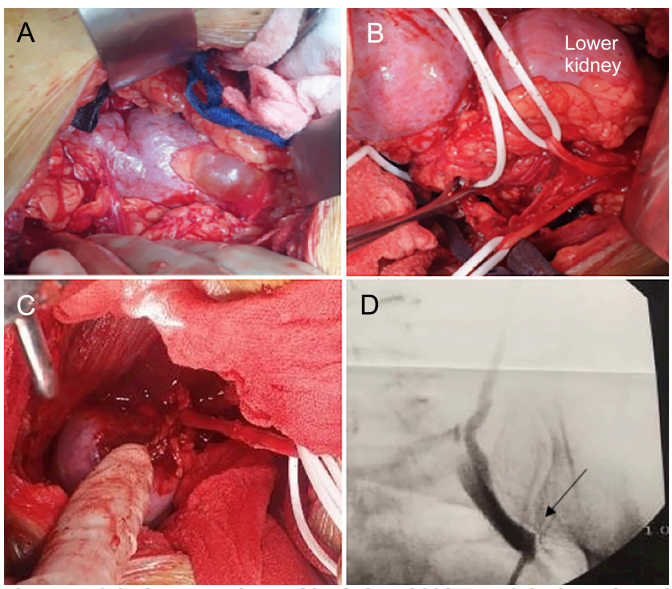

Figure 2: (A) Cleavage plane of both fused kidneys. (B) Dissection of hilum, both ureters taken in sling. (C) Upper kidney after removing the lower with the ureter in the sling. (D) Confluence of both ureters, distal few mm of resected ureter.

DISCUSSION

CFRE is a rare congenital anomaly. It is the second most common fusion anomaly of the kidneys after horse-shoe kidneys. The abnormality has a 3:2 predominance in males. The reported incidence is 1 in 2,000 to 1 in 7,500 at autopsy. It results from aberrant formation and midline crossing of the metanephric blastema and ureteric bud. CFRE is believed to develop from the fourth to eighth weeks of gestation.[1]

Six anatomical variations of CFRE have been described in the literature. Namely, 1: Inferior CFRE 2: Sigmoid kidney 3: Lump kidney 4: Disc kidney 5: L-shaped kidney 6: Superior CFRE.[2] Type 1 is the most common, while type 6 is the least frequent. Left-to-right CFRE is three times more frequent than right-to-left.[3]

The majority of CFRE cases have an incidental diagnosis. The symptoms are secondary to hydronephrosis, stones, pelvic ureteric junction obstruction, and tumours. Recurrent urinary tract infections are the most common presentation in children. There are no specific guidelines for the management of CFRE.[4] The fused renal units do not need to be separated. The treatment is guided toward the associated problems that lead to either a symptom or the deterioration of the upper tracts, e.g., a pelvic ureteric junction obstruction would require a pyeloplasty or nephrectomy for renal cell carcinoma (RCC) or TCC.[5] The CFRE has decreased renal function as compared to the contralateral kidney, which gives an idea that the remaining upper kidney in our case is ectopic because the patient has persistently raised creatinine postoperatively. In approximately 75% of cases, the renal arteries originate from the inferior abdominal aorta or iliac arteries.[6]

RCC, TCC, and Wilms tumours of the CFRE have been reported and managed by resection of the involved renal unit.[7] TCC of CFRE is rare. Distortion of the pelvicalyceal system leads to urinary stasis, which can be tumourigenic in some cases. The standard management for invasive high-grade upper-tract TCC remains nephroureterectomy. Since more than 90% of ectopic kidneys are fused, surgical management becomes difficult in relation to variant vascular origin and a complex anatomy, necessitating dissection within the renal parenchyma and likewise nephron-sparing surgery. The role of preoperative renal angiography and CT angiography has been described in the literature for the identification of the vascular anatomy and origin of renal arteries, which is essential in planning surgical dissection. Although separation and excision of a fused ectopic kidney can encounter surgical difficulty, the primary goal of cancer control should not be compromised.[8]

The embryological pathogenesis of CFRE has not been clearly explained. It was postulated that if the ureteric bud fails to fuse with the ipsilateral metanephric bud due to any cause, for example, rotation and overbending of the caudal end of the embryo, causing the ureter to turn more closer to the contralateral side, the migrated ureteric bud induces the metanephric blastema twice to form two kidneys on the same side.[9]However, this explanation failed to justify the cause of both ureters lying on the same side.

To our knowledge, three cases of CFRE with TCC have been reported in the literature. Two of them were left-to-right; one was managed by open nephroureterectomy, and the second was managed laparoscopically. Only one case of the right-to-left CFRE with upper TCC and one case with single ureter has been

reported. Our case is unique in the sense that both ureters were lying on the same side, which is a variant of the six types of CFRE described so far.

PATIENT'S CONSENT:
Informed consent has been obtained from the patient to publish the case.

COMPETING INTEREST:
The authors declared no competing interest.

AUTHORS' CONTRIBUTION:
AS: Conceptualising, literature review, and manuscript writing.
MHA: Proofreading and review of the manuscript.
All authors approved the final version of the manuscript to be published.

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