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A Rare Case of Nephroureterectomy of Crossed Fused Renal Ectopia with Urothelial Carcinoma

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Abstract: ***Introduction: Crossed fused renal ectopia (CFRE) is a congenital anomaly, where one of the kidneys crosses the midline and located at the other side. Urothelial carcinoma in CFRE is an exceedingly rare clinical finding. Case Report: A 57 years old male who presented with pain abdomen and on evaluation, he was diagnosed with CFRE with tumour and stones in CT Scan. PET CT Scan showed crossed fused renal ectopia on the right side in the right iliac fossa with multiple calculi in fused left kidney. Metabolically active lesion in extrarenal pelvis of fused left kidney likely ?TCC. Metabolically active enlarged para-aortic, aortocaval and left iliac nodes with stage pT3,N2,Mx. The chosen treatment was nephrectomy of the left moiety with ureterectomy, bladder cuff excision and lymph node dissection. The final pathology revealed urothelial carcinoma with squamous differentiation, involving pelvicalyceal system and ureter, infiltrating into the sinus fat. Para-aortocaval and left common iliac lymph nodes showed metastasis. Conclusion: Our case highlights that nephroureterctomy with preservation of normal functioning moiety seems to be excellent option for urothelial carcinoma in a CFRE .***

Keywords: Crossed fused renal ectopia, Nephroureterectomy, Urothelial carcinoma, Squamous differentiation

Abbreviations: Crossed fused renal ectopia (CFRE), Transitional cell carcinoma (TCC)

1. Introduction

Crossed fused renal ectopia is a rare congenital anomaly in which both kidneys are located and fused on the same side of the body, with a high incidence of stone formation and urinary tract infection. The prevalence of the crossed renal ectopia with fusion was estimated to be 1 in 1000 live births [1]. Horseshoe kidneys characterize the most common renal abnormality of fusion, with an overall incidence of 0.25%, with a male predominance. Similarly, cross-fused renal ectopia also has a male predominance of 3:2 and is the second most common fusion abnormality [2]. The left kidney is usually the crossed and fused with the right kidney in most cases, between the inferior pole of the orthotropic kidney and superior pole of ectopic kidney [3].

It is very challenging to perform surgery in these patients due to atypical vasculature of both moieties. We report a case of left to right crossed ectopia harboring renal tumour managed by nephroureterectomy.

2. Case Presentation

A 57 years old male who presented with pain abdomen and on evaluation, he was diagnosed with CFRE with tumour and stones in CT Scan done elsewhere (Fig 1A & 1B). His medical history consisted of hypertension. He did not have any prior abdominal procedures. There were no family history of malignancy or similar congenital anomalies. Physical examination was unremarkable and routine investigation showed his serum creatinine of 2 mg/dl. He was referred to our center for further evaluation and management. 18 F-FDG whole body PET CT Scan showed crossed fused renal ectopia on the right side in the right iliac fossa with multiple calculi in fused left kidney (Fig 2A & Fig 2B). Left pelvi-ureteric junction calculi with upstream gross hydronephrosis of the left fused kidney. Metabolically active heterogenously enhancing lesion (3.6 x 2 cm in size) in the extrarenal pelvis of fused left kidney extending to the pelvi-ureteric junction likely primary malignancy of the collecting system ? transitional cell carcinoma (TCC). Few caculi in the ureter with transition in calibre. No focal lesions in urinary bladder. Metabolically active enlarged para-aortic, aortocaval and left iliac nodes suggestive of nodal metastasis. No evidence of distant metastasis with stage pT3,N2,Mx. DMSA scan showed right kidney moderate cortical functioning with left kidney cortex thinned out (Fig 1C). The background activity was elevated suggestive of overall impaired parenchymal extraction of tracer. Both kidneys are ectopically visualized in the lower abdomen as fused ectopic kidneys with prominent cortex. The right kidney shows moderate cortical functioning mass while the left kidney cortex is thinnned out with a large central photon deficient ‘cold’ area. Since the kidneys are fused together in composite planes the split function could not be assessed.

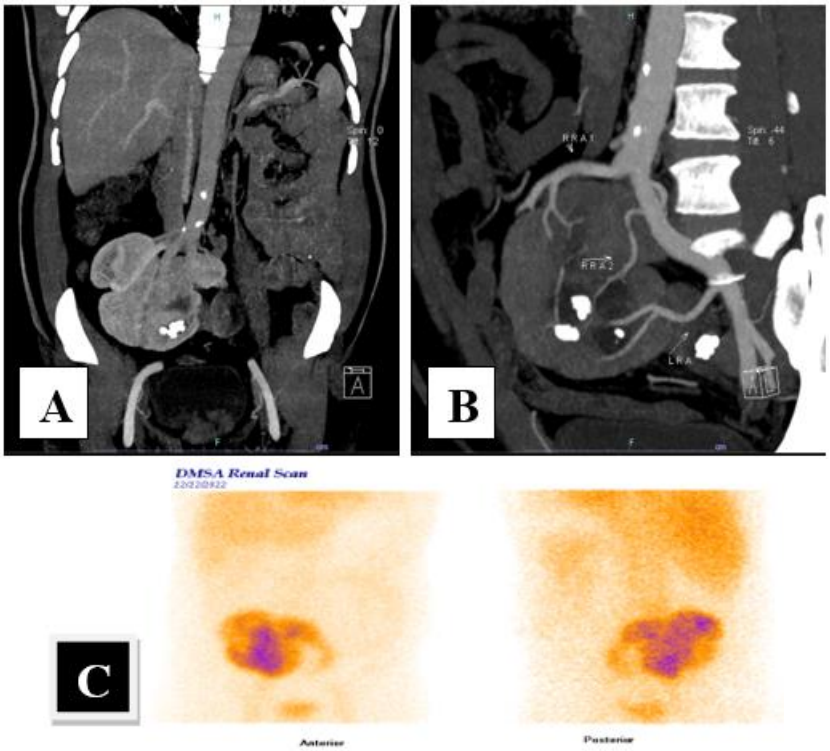

Figure 1: Preoperative CT of the abdomen coronal (A) and sagittal showing arterial supply (B) showed evidence of crossed fused renal ectopia on the right side. DMSA scan showing CFRE (C).

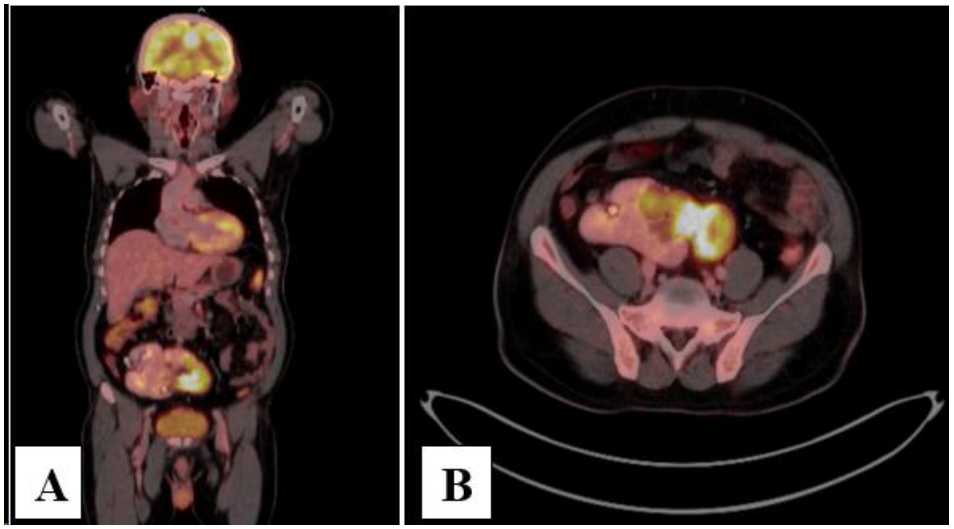

Figure 2: Preoperative PET CT of the abdomen coronal (A) and axial (B) showing evidence of crossed fused renal ectopia on the right side.

The chosen treatment was nephrectomy of the left moiety with ureterectomy, bladder cuff excision and lymph node dissection (Fig 3A & Fig 3B). Cystoscopy was performed to check the urinary bladder and ureteral orifices, where no abnormalities were detected, with placement of left ureteric catheter. A large midline incision was made to expose the fused kidney. Two arteries supplying the left renal moiety noted arising from the aorta directly and one more artery noted arising from the left common iliac artery. Left ureter was excised along with the bladder cuff, en bloc with specimen.. The left moiety then separated from the right moiety by sharp dissection. Lymph node dissections of paraaortic, right aortocaval and left common iliac were done. With careful consideration of the renal vasculature, the operation was completed without complications.

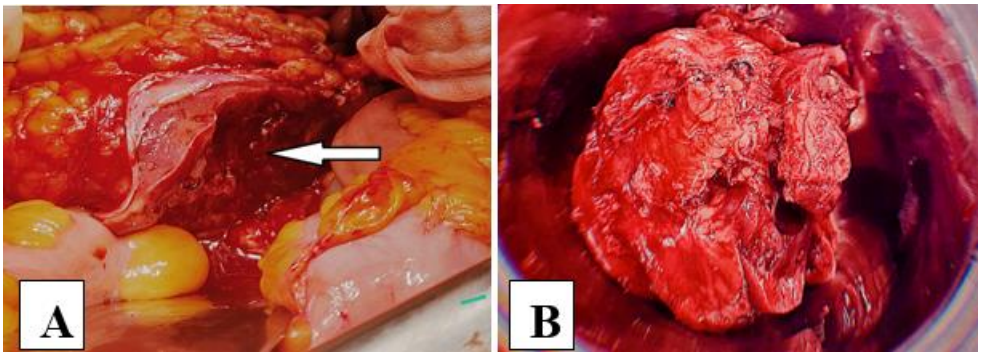

Figure 3: Intraoperative images showing the cut surface of the normal moiety after excision of the diseased moiety (A). The specimen (B)

Postoperatively, the patient recovered well despite the size and complexity of the surgery. The final pathology revealed urothelial carcinoma with squamous differentiation, involving pelvicalyceal system and ureter, infiltrating into the sinus fat (Fig 4A & 4B). Para-aortocaval and left common iliac lymph nodes showed metastasis. Renal resection margin, inter-aortocaval nodes, left distal ureter and bladder cuff were free of neoplasm.

Immunohistochemistry results were consistent with urothelial carcinoma with squamous differentiation (Fig 4C & 4D). Adjuvant chemotherapy was not recommended due to the elevated serum creatinine level. Postoperatively after 6 weeks, he received 45 Gy /25 fractions of radiotherapy to postoperative bed, paraaortic nodes and common iliac nodes. His serum creatinine is 3.4 mg/dl after 14 months postoperatively.

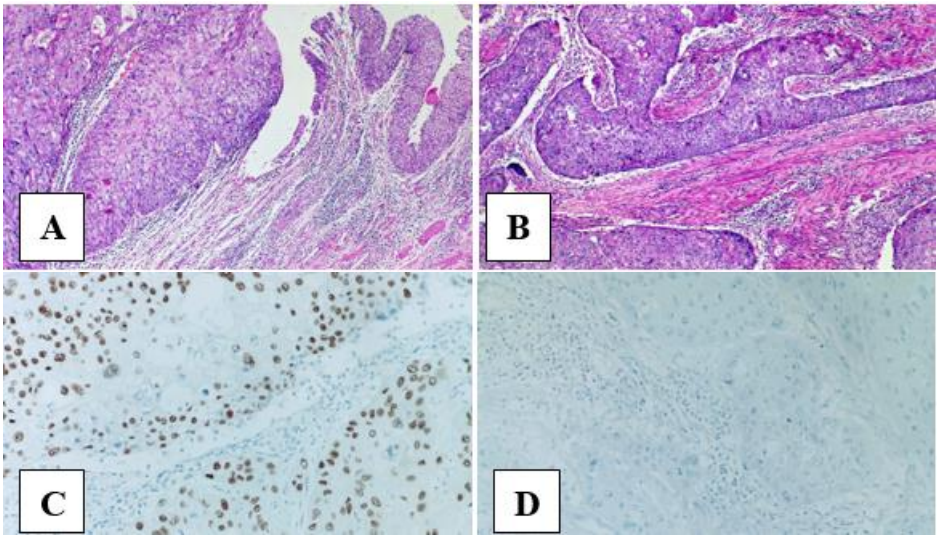

Figure 4: H&E, Urothelial carcinoma (200X) with adjacent urothelial lining showing transitional areas (A). H&E, Urothelial carcinoma (200X) infiltrating as large nests with squamous differentiation (B). IHC (400X) P40 diffuse strong nuclear positivity in neoplastic cells (C). IHC (400X) Uroplakin negative in neoplastic cells (D).

3. Discussion

Crossed fused renal ectopia is thought to result from the abnormal development of the ureteric bud and metanephric blastema during the fourth to eighth weeks of gestation. Wilmer, in 1938, first categorized the fusion anomalies of the kidney and McDonald and McClellan, in 1957, included crossed ectopia with fusion, crossed ectopia without fusion, solitary crossed ectopia and bilateral crossed ectopia in a modified classification. These abnormalities are clinically significant because approximately half the patients manifest complications e.g., hydronephrosis, infections and nephrolithiasis.[4]-[7]. The ectopic kidney is usually located inferior to the normal kidney with fusion. Crossed-fused renal ectopia is a rare congenital abnormal development of the urinary system. There are 6 subtypes of the anomaly:

1) Inferior ectopia, in which the ectopic kidney lies inferior to the normal kidney;
2) Superior ectopia, in which the upper pole of the normal kidney fuses with the lower pole of the ectopic kidney;
3) Sigmoid or S-shaped;
4) Lump or pancake;
5) L-shaped; and
6) Disc. [8],[9].

Zhuo Yin et al, however, reported a new subtype of crossed-fused renal ectopia named the “Y” type, in which the ureters of the kidneys are fused [10]. It has never been satisfactorily explained about how these kidneys are drawn to the opposite side of the kidney. One theory linked that entity to abnormal development of the ureteric bud and the metanephric blastema during early gestational age. Therefore, both kidneys are fused into a single mass, giving rise to two separate and distinct ureters with normally located ureteral orifices in the urinary bladder. Most of these patients usually have complications, such as hydronephrosis, nephrolithiasis, infection and rarely malignancy [11]. RCC is most

frequently associated tumor with fusion anomalies. Meanwhile, the prevalence of malignancy in kidneys with congenital anomalies is comparable to those with normal kidney, with similar prognostic parameters [12]. Careful consideration should be taken in approaching such cases due to the variability in vasculature and drainage of the renal units. Preoperative imaging for surgical planning, with contrast enhanced renal angiography and delayed urogram, is of utmost importance to delineate potential complex renal vascular supply and collecting system anatomy [13],[14].

Urothelial carcinoma of the kidney is also relatively uncommon, as upper urinary tract lesions account for only 5%-7% of all urothelial carcinomas [15]. The defining features of urothelial carcinomas include multiplicity and a high incidence of recurrence [16]. Due to these characteristics, the recommended treatment for urothelial carcinoma of the kidney is nephroureterectomy with excision of a bladder cuff. Moreover, given the likelihood of invasion into the bladder, regular cystoscopic follow-up is encouraged [17].

To our knowledge, only three cases of upper tract urothelial carcinoma in an individual with CFRE have been reported in the literature. The first reported case is of a patient with left-to-right CFRE and invasive urothelial carcinoma of the kidney. Open nephroureterectomy with bladder cuff resection was performed, and the pathology identified a high-grade T1 carcinoma of the ureter [18]. The second case is also of upper tract urothelial carcinoma in an individual with left-to-right CFRE. However, laparoscopic surgery was performed and the pathology revealed a low-grade T1 carcinoma [19]. The third case is of urothelial carcinoma in a right-to-left CFRE. It is the first reported case of urothelial carcinoma in a right-to-left CFRE [20]. Based on this review of the literature, we present the fouth reported case of urothelial carcinoma in CFRE as an entity and third case of urothelial carcinoma in a left-to-right CFRE, as well as the most advanced case of upper tract urothelial carcinoma in a patient with CFRE. It is also the third instance in which an open nephroureterectomy was performed to treat upper tract urothelial carcinoma in a patient with CFRE.

4. Conclusion

Urothelial carcinoma in crossed fused renal ectopia represents a rare of rarity entity. Nephroureterectomy with preservation of normal-functioning moiety seems to be an excellent option in a case of CFRE with urothelial carcinoma, despite the complexity of the surgery. We believe that a careful preoperative planning and meticulous delineation of renal vasculature are mandatory prior to surgery for preservation of the uninvolved renal unit and to avoid unpredicted anatomy. In addition, a comprehensive evaluation of the patient's preoperative general condition is important for the prevention of perioperative complications and also for the preservation of postoperative renal function.

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