

Oncology

Lower and upper urinary tract urothelial carcinoma in crossed fused ectopic kidney



Dana Greenberg*, Yoav Avidor, Muhammad Majdoub, Asali Mohsin, Issac Kaver, Ronen Rub

Urology Department, Hillel Yaffe Medical Center, Hadera, 3810000, Israel

ARTICLE INFO

Keywords:

Urothelial carcinoma
Crossed fused renal ectopia
Crossed renal ectopia
Transitional cell carcinoma

ABSTRACT

Urothelial tumors in patients with anatomical abnormalities may pose significant challenges. Management follows the same principles which are employed in normal anatomy, however, thorough diagnostic investigation is warranted in order to delineate key anatomical landmarks. Meticulous pre-operative investigation should utilize every imaging modality which can assist the surgeons. We present a case of transitional cell carcinoma (TCC) in a crossed-fused kidney treated with nephro-ureterectomy. Only a handful of cases of TCC in CFRE have been reported. The case demonstrates the critical role of pre-operative anatomical studies and intra-operative identification of unique anatomy, which facilitate treatment and avoid complications.

1. Introduction

Crossed fused renal ectopia (CFRE) is a rare congenital renal malformation. In this condition, one kidney crosses the midline and may become fused to the contralateral kidney. This condition is mostly asymptomatic and usually diagnosed incidentally. We report a case of transitional cell carcinoma (TCC) in the ureter and kidney of a crossed ectopic kidney treated by nephroureterectomy and discuss the importance of pre-operative anatomical characterization in considering the surgical approach.

2. Case study

A 65-year-old man presented with painless macroscopic hematuria. Past medical history was remarkable for active smoking, ischemic heart disease and diabetes mellitus. Prior surgical history included coronary artery bypass graft, repair of penile fracture and segmental resection of left lung hamartoma. Physical examination was unremarkable. Blood workup indicated normal renal function. Urine cytology was suspicious for urothelial carcinoma (UC). Computed tomography urography (CTU) demonstrated a crossed fused left kidney to the right (Fig. 1). A mass within the lumen of the right renal pelvis was observed, alongside an irregularly thickened wall of the urinary bladder.

On *Trans-urethral* resection of bladder tumor (TURBT) a 2 cm papillary tumor located lateral to the right orifice was resected, consistent with pathology of T1 high-grade urothelial carcinoma

(HGUC). In re TURBT, no evidence of residual tumor was found. Diagnostic retrograde ureteroscopy detected a large papillary tumor that occupied the entire right renal pelvis, revealing pathology of TA low-grade UC (LGUC). To expedite surgical planning, CT angiography was done and carefully studied, which proved extremely beneficial in this case, demonstrating a lower-positioned left kidney on the right side of the body with its individual vessels and collecting system clearly delineated (Fig. 2).

The patient was admitted to right nephro-ureterectomy. A midline transabdominal incision was performed. The right retroperitoneum was exposed. After reflection of the large and small bowels, both kidneys were exposed with their arteries, veins and ureters. The right renal vessels and ureter were marked with blue vessel loops, the left renal elements were indicated with red vessel loops (Fig. 3).

After ligating and dividing the right renal vessels a clear line of demarcation was formed between the left and the right kidney. The right kidney was separated at the demarcation line and removed en block with the ureter and the bladder cuff which were managed in an extravesically manner (Fig. 4).

Postoperative laboratory tests revealed an elevation in creatinine levels to 1.6 mg/dL from a previously normal baseline, which was resolved after intravenous hydration of 2.0 liters. Additionally, a decrease in hemoglobin levels to 7.7 g/dL from a normal baseline was observed on the second postoperative day. The patient received two units of packed red blood cells, resulting in an increase in hemoglobin levels to 11.5 g/dL. Thereafter the patient's postoperative recovery was

* Corresponding author.

E-mail address: Dgreenberg91@gmail.com (D. Greenberg).

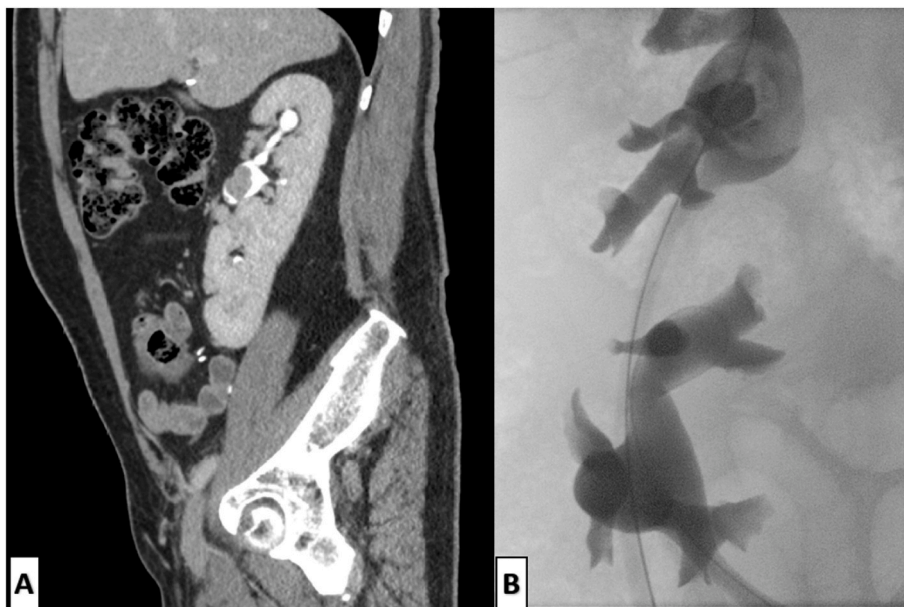


Fig. 1. (A) Computed tomography of the abdomen, sagittal view, demonstrating crossed fused kidneys, the upper portion representing the orthotopic right kidney with the left kidney fused to its inferior pole. (B) Retrograde pyelography demonstrating the collecting system of each kidney.

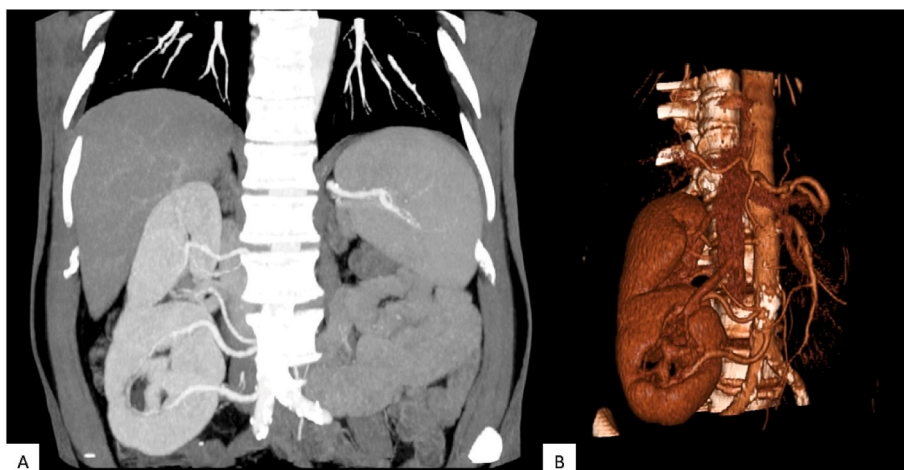


Fig. 2. (A) Computed tomography angiography and (B) 3D reconstruction of the renal vasculature.

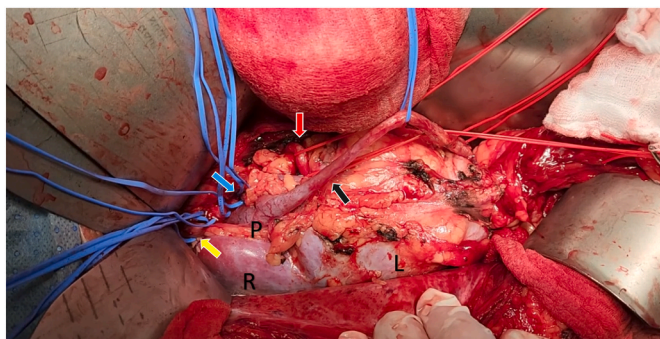


Fig. 3. Intraoperative view of the right kidney (R), left kidney (L), left renal arteries (red arrow), right renal arteries (blue arrow), right renal veins (yellow arrow), right renal pelvis (P), and right ureter (black arrow).

satisfactory.

Histopathology report of the tumor resected from the kidney showed a papillary LGUC with squamous differentiation, with tiny foci of early superficial invasion into the lamina propria without muscularis propria invasion. HGUC was found in some foci. The tumor involved the right renal pelvis and one of the calyces. There was no evidence of lympho-vascular or perineural invasion with tumor-free margins.

The patient received intra-vesical BCG (Bacillus Calmette-Guérin) immunotherapy, completing the treatment sessions nine months post-operatively. In subsequent follow-up sessions which included cystoscopy, CT scans, and magnetic resonance Imaging, no evidence of disease recurrence was detected. He continues to undergo follow-up at our outpatient onco-urological clinic to this day.

3. Discussion

Crossed renal ectopia (CRE) is a rare anomaly in which a kidney is displaced on the opposite side of the body. CRE accounts for the second most common renal fusion abnormality after horseshoe kidney and is more predominant in men with a ratio of 2:1. Approximately 90 % of

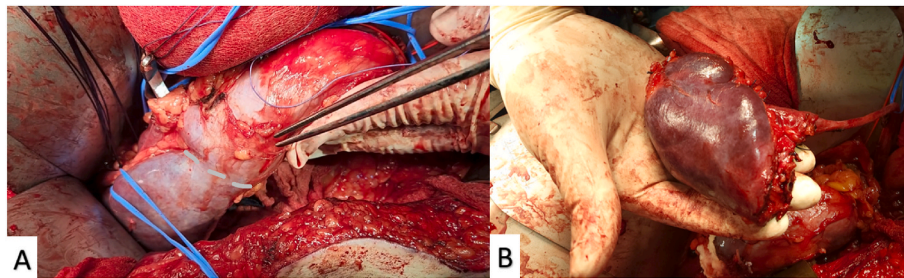


Fig. 4. (A) Intraoperative view showing the line of demarcation (grey dotted line) between fused kidneys after ligation of the right renal vessels. (B) the right kidney after separation together with its ureter.

crossed renal ectopia are fused¹ and estimated at 1 in 1000–7500 individuals.² Urothelial carcinomas (UC) are the sixth most common tumor in developed countries, out of which, upper tract UC (UTUC) accounts for only 5–10 %. 70.5 % of the patients are men and 53 % of them are smokers.³

Based on review of the literature, there have been only seven reported cases of space-occupying lesions in ectopic kidneys, including one renal cell carcinoma in CFRE,⁴ one TCC in CRE⁵ and five reported TCC in CFRE.^{6–10} To the best of our knowledge, this is the first reported case of synchronous upper and lower tract UC in CFRE treated with TURBT and nephro-ureterectomy. The uncommon nature and convoluted anatomy of such cases provides for a challenging diagnostic and management decision-making process. The surgical procedure is more challenging, mainly due to the complexity of the vascular system and merits careful advanced planning.

The treatment approaches in such rare cases vary from endoscopic to open approaches depending on the type and clinical stage of malignancy, site, co-morbidities and lastly the surgeon's experience. Cystourethroscopy is advised before surgery to detect any synchronous tumors. It is known that pelvic kidneys have an anomalous vascular supply and collecting system irrespective of the fusion type.⁵ Therefore, a fundamental yet crucial step is identifying and securing all renal arteries first before controlling renal veins, aiming to prevent kidney congestion and bleeding. Of note, while it is legitimate to treat the upper tract UC endoscopically, in this case, due to the variety of complexities and to rule out the presence of a high-grade tumor, we preferred an open approach.

4. Conclusion

This rare case of UC with CFRE highlights the importance of conducting a thorough investigation of the renal anatomy and vasculature prior to surgery, using the latest contemporary imaging techniques at the urologist's disposal. In this case, an open nephro-ureterectomy approach enabled optimal intra-operative anatomical and vascular characterization.

CRedit authorship contribution statement

Dana Greenberg: Writing – review & editing, Writing – original draft, Visualization, Validation, Resources, Project administration, Methodology, Investigation, Data curation. **Yoav Avidor:** Writing – review & editing, Visualization, Validation, Supervision, Resources,

Conceptualization. **Muhammad Majdoub:** Writing – review & editing, Supervision, Methodology, Data curation, Conceptualization. **Asali Mohsin:** Writing – review & editing, Writing – original draft, Methodology, Conceptualization. **Issac Kaver:** Writing – review & editing, Writing – original draft, Visualization, Validation, Supervision, Resources, Project administration, Methodology, Investigation, Data curation, Conceptualization. **Ronen Rub:** Writing – review & editing, Writing – original draft, Visualization, Validation, Supervision, Resources, Project administration, Methodology, Investigation, Data curation, Conceptualization.

Declaration of competing interest

No conflicts of interest.

References

- Partin AW, Dmochowski RR, Kavoussi LR, Peters CA, Wein AJ. In: Walsh Campbell, ed. *Wein Urology: 3-Volume Ser.* twelfth ed. Philadelphia, PA: Elsevier - Health Sciences Division; 2020:727–728.
- Glodny B, Petersen J, Hofmann KJ, et al. Kidney fusion anomalies revisited: clinical and radiological analysis of 209 cases of crossed fused ectopia and horseshoe kidney. *BJU Int [Internet]*. 2009;103(2):224–235. <https://doi.org/10.1111/j.1464-410X.2008.07912.x>. Available from.
- Rouprêt M, Seisen T, Birtle AJ, et al. European Association of Urology guidelines on upper urinary tract urothelial carcinoma: 2023 update. *Eur Urol*. 2023;84(1):49–64.
- Tsuboi I, Ogawa K, Yokoyama S, Araki A, Kadota K, Wada K. Open partial nephrectomy of a left-to-right crossed fused renal ectopia with clear cell renal cell carcinoma: case report and review of the literature. *Urol Case Rep [Internet]*. 2022;41(101964), 101964. <https://doi.org/10.1016/j.eucr.2021.101964>. Available from.
- Halalshah O, Ghawanmeh HM, Alshammari A, et al. Upper Tract urothelial carcinoma in ectopic pelvic kidney. *Urol Case Rep [Internet]*. 2017;11:42–43. <https://doi.org/10.1016/j.eucr.2016.11.026>. Available from.
- Figueiredo MS, Bruno PPV, Ivo de Sousa PN, et al. High-grade urothelial carcinoma in a pelvic ectopic kidney: a case report. *Biomed J Sci & Tech Res*. 2024;54(3). BJSTR. MS.ID.008543.
- Hearn J, Power RJ, MacDonald L, Johnston P, Organ M. Nephroureterectomy of right-to-left crossed fused renal ectopia with urothelial carcinoma [Internet] *Cureus*. 2020;12(6), e8544. <https://doi.org/10.7759/cureus.8544>. Available from.
- Miller DC, Kropp KA. Transitional cell carcinoma associated with crossed renal ectopia and previous cyclophosphamide therapy. *J Urol [Internet]*. 1992;148(4):1244–1246. [https://doi.org/10.1016/s0022-5347\(17\)36873-8](https://doi.org/10.1016/s0022-5347(17)36873-8). Available from.
- Gur U, Yossepowitch O, Baniel J. Transitional cell carcinoma in a fused crossed ectopic kidney [Internet] *Urology*. 2003;62(4):748. [https://doi.org/10.1016/s0090-4295\(03\)00577-6](https://doi.org/10.1016/s0090-4295(03)00577-6). Available from.
- Simforoosh N, Zare S, Mahmoudnejad N, Soltani MH. Laparoscopic radical heminephroureterectomy for management of urothelial tumor in a cross-fused ectopic kidney: first report. *Videoscopy [Internet]*. 2011;21(3). <https://doi.org/10.1089/vor.2010.0003>. Available from.