

# RCC in Crossed Fused Ectopic Kidney: Rare of Rarest Entity

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**Abstract:** Primary renal cell carcinoma (RCC) in crossed fused renal ectopia represents a rare of rarest entity. Only nine cases were reported in the literature, including eight RCC and one transitional cell carcinoma. This report presents a case of a 75-years old male presented with incidentally discovered renal mass in a crossed fused ectopia. Careful preoperative planning and meticulous delineation of renal vasculature were performed to avoid unpredicted anatomy. Nephron-sparing surgery with preservation of the normal-functioning moiety was performed with uneventful postoperative course. These clinical, morphological and immune-histochemical features will be presented with a review of the current literature.

**Keywords:** RCC, Renal cell carcinoma, crossed fused ectopia, renal ectopia, Nephron sparing surgery

## 1. Introduction

Crossed fused renal ectopia is a markedly rare developmental anomaly with mild male predominance, where one of the kidneys crosses the midline and located at the other side, mostly fused with inferior ectopia. Association of renal malignancy with crossed ectopia is extremely rare, as well.<sup>1</sup> The exact incidence of crossed fused renal ectopia is not known, as most patients are asymptomatic. An estimated prevalence of 1:2000 live birth has been reported in autopsy series. Furthermore, the left kidney is usually the crossed and fused with the right kidney in most cases, between the inferior pole of the orthotopic kidney and superior pole of ectopic kidney.<sup>3</sup>

Surgery in these patients may be challenging due to atypical vasculature of both moieties. In this report, a patient with left to right crossed ectopia harboring renal tumor will be presented with a review of the current relevant literature.

Accompanying malignancies have been reported in only 9 cases in the literature. Among which 8 cases were RCC and 1 case was TCC. RCC is the most common type of neoplasm associated with this anomaly. Incidence and prognostic factors of RCC in ectopic kidney are similar to what has been described in the general population.<sup>4</sup> Due to unpredictable anatomy, preoperative imaging and ureteral stenting may be helpful in order to avoid significant surgical complications including haemorrhage and ureteral injury.

Contrast enhanced CT abdomen and CT angiogram can also provide additional information preoperatively.<sup>5</sup> While

several surgical approaches have been described for renal fusion abnormalities, transperitoneal approach provides the optimal exposure of the anomalous anatomy allowing for early vasculature control and improved opportunity for nephron sparing.<sup>6</sup>

## 2. Case Presentation

In this case, a 75 year old smoker male presented to a private clinic for evaluation of hematuria. Where on ultrasound he was incidentally diagnosed to have crossed fused left to right ectopic kidney with heterogenous multilobulated mass lesion in right kidney & was referred to our tertiary care center. He had a medical history of coronary artery disease with history of MI 4 years back with angioplasty of RCA. He did not have any prior abdominal procedures. He did not have any palpable masses on exam.

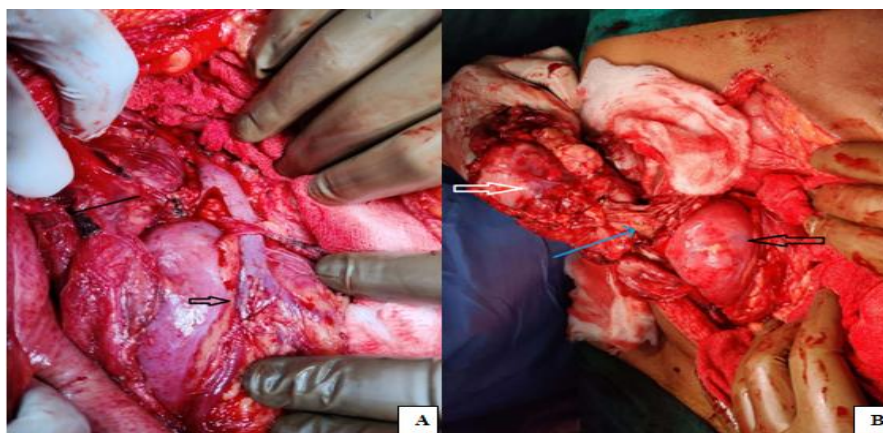
Following a contrast-enhanced CT scan, a right to left cross fused ectopic kidney was identified with the right kidney overlying the inferior vena cava (IVC) (Fig. 1). Fusion of the right kidney to the left was evident just lateral to the aorta (Fig. 2). Additionally, a 10.3 x 8.6 x 8.3cm enhancing mass on the superior and mid pole of right renal unit was seen with central necrosis concerning for a primary renal cell carcinoma (RCC) (Fig. 2). Renal angiography and delayed images demonstrated mild extension of tumour thrombus in the right renal vein not extending into the distal renal vein near confluence with IVC (Fig. 3). Imaging suggested no significant lymphadenopathy.



**Figure 1:** Preoperative contrast –enhanced CT of the abdomen :sagittal (A) and axial (C) section showed evidence of crossed fused renal ectopia on the right side, with single separate renal pedicles and collecting systems per each kidney. CT angiography delineating renal arteries of both renal moieties (B)

After a thorough discussion regarding his likely diagnosis and treatment options, the patient elected to undergo open right radical nephrectomy. A midline incision was made from xyphoid extending 6cm infraumbilically. The right-sided white line of Toldt was incised and the colon medialized. The duodenum was subsequently Kocherized. The right ectopic kidney was immediately visualized upon reflecting the small bowel mesentery. Dissection continued

to determine retroperitoneal anatomy of the right kidney, IVC, aorta, and vascular structures. The right-sided vessels including 2 arteries and 2 veins were identified, along with the right ureter, and marked with vessel loops. (fig 2.) The renal vein is then ligated at the confluence to IVC to include the tumour thrombus, renal artery were then ligated with 0 silk sutures and divided. The kidney and Gerota's fascia was subsequently freed of its attachments.

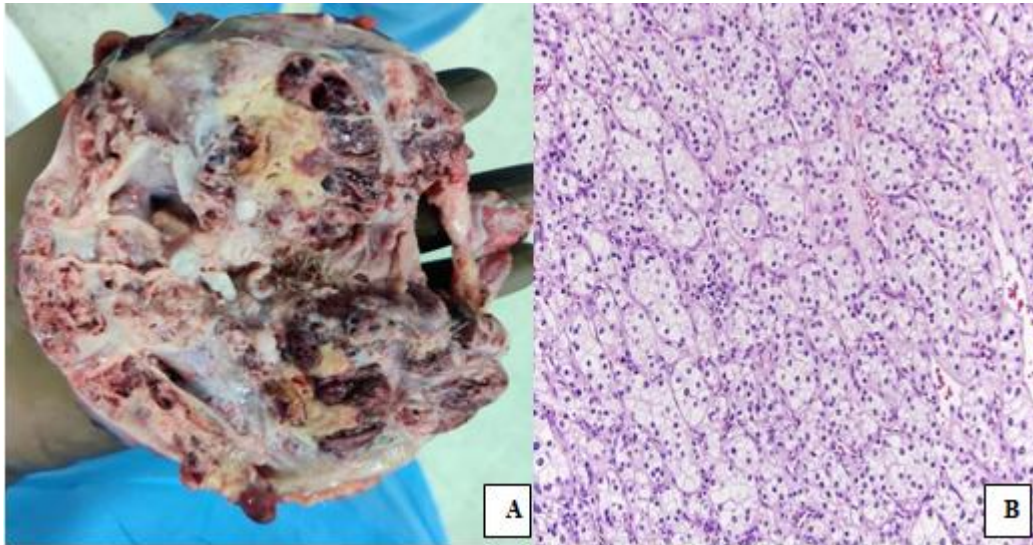


**Figure 2:** Operative images (A) arrow marked shows the vessels of lower moiety (B) black arrow points the lower ectopic moiety, white arrow points the upper moiety, blue line pointing on the fusion between both moiety.



The descending colon was mobilized after incision of the ipsilateral white line of Toldt to gain access to the left renal unit. A vascular clamp was used to clamp and compress the isthmus of the fused renal units and the right kidney was subsequently resected with a scalpel. Hemostasis was

obtained by individually suturing bleeding vessels from the resection site with 3-0 vicryl sutures. The renorrhaphy was completed using 2-0 chromic sutures with abgelpedgets. Finally, floseal was injected over the renorrhaphy.



**Figure 3:** (A) Gross appearance of excised Rt. renal moiety showing foci of central necrosis (B) Histopathology revealed clear renal cell carcinoma

On pathological review of the nephrectomy specimen, the final diagnosis was a pT2bNxMx clear cell renal cell carcinoma with no LVI and tumour limited within the renal capsule with necrosis & margins and ureter free from malignancy (Fig. 3). Post operative course was complicated by mild urinary extravasation which was successfully managed by the placement of double (DJ) catheters, it continued for 10 days, after which it started to decrease gradually and stopped completely. The tube drain was removed after two weeks, post operative ultrasound was normal. Otherwise, convalescence was uneventful, and the patient discharged home in a good general condition, with stable vital signs and normal kidney function. Serum creatinine at discharge was 1.08 mg/dl and hemoglobin level was 9.3 g/dl.

### 3. Discussion

Only nine cases of carcinoma in crossed fused renal ectopia were reported in the literature until 2020<sup>3-11</sup>, since the first case was presented in 1942. Embryologically, this anomaly occurs in the first trimester as a result of an abnormal development of the ureteric bud and metanephric blastema between 4 and 8 weeks' gestation. Mechanism involves arrest during kidney's migration, migration beyond normal limit, metanephric ectopia, contralateral metanephros induced by wandering ureteric bud or duplex Wolffian ducts<sup>1</sup>. While most patients are asymptomatic, ectopic kidneys carry increased risk of urinary calculus formation and hydronephrosis. Due to unpredictable anatomy, preoperative imaging and ureteral stenting may be helpful in order to avoid significant surgical complications including haemorrhage and ureteral injury.

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### 4. Conclusions

Crossed-fused renal ectopia with a renal mass represents an uncommon urological diagnosis. Renal cell carcinoma is the most frequent tumor type associated with fusion anomalies. It presents an operative challenge for the surgeon during preoperative and intraoperative management. The location of the kidney, its potential complex vascular supply and collecting system, provide surgical challenges to approach and ability to perform a partial nephrectomy.<sup>2</sup> Preoperative imaging with renal angiography and delayed images allow the surgeon to identify complex anatomy for surgical decision making. Whether a partial or radical nephrectomy is performed, a renorrhaphy will need to be accomplished to achieve hemostasis and repair the resection site.

### 5. Consent

Images and informations were obtained and shared with verbal and written consent by the patient.

### 6. Declarations of interest

None

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