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# Primary renal cell carcinoma in crossed fused ectopia: Nephron sparing surgery for a rare of rarity entity

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ABSTRACT

Primary renal cell carcinoma (RCC) in crossed fused renal ectopia represents a rare of rarity entity. Only eight cases were reported in the literature, including seven RCC and one transitional cell carcinoma. This report presents a case of a 39-years old female 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.

## Introduction

Crossed fused renal ectopia is a markedly rare congenital anomaly, 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.[1] 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, with females are less frequently affected. 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.[2]

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.

### *Case presentation*

A 39- years old non-smoker female was referred to our tertiary care center after detected incidentally to have a right renal mass in a crossed-fused renal ectopia. Her medical and surgical history were irrelevant apart from a long-standing umbilical hernia, since 2005. There were no constitutional symptoms, no family history of malignancy or similar congenital anomalies. Physical examination was unremarkable and routine laboratory investigations were within normal range, as well. Serum creatinine was 0.76 mg/dl, while hemoglobin level was 10.3 g/dl. Contrast-enhanced CT of the abdomen and pelvis showed evidence of crossed fused renal ectopia on the right side, with single separate renal pedicles and collecting systems per each kidney, as confirmed with CT angiography (Fig. 1). A well-defined hypervascular mass originating from the lower pole of the left kidney was observed, measuring 4.6x4x4.7 cm, with no hydronephrosis. There were neither evidence of tumor thrombus in the venous drainage system, nor lymphadenopathy or radiological signs of distant metastasis.

Prior to surgery, flexible cystoscopy was performed to check the urinary bladder and ureteral orifices, where no abnormalities were detected, with placement of bilateral ureteral stents. The patient underwent trans-peritoneal partial nephrectomy with preservation of the ureter and renal pelvis of the affected kidney on October 2018 (Fig. 2A). Postoperative urinary leakage continued for 10 days, after which it stared to decrease gradually and stopped completely. The tube drain was removed after two weeks, where contrast-enhanced CT scan demonstrated normal function of the remaining part of the affected kidney (Fig. 2-B). Otherwise, convalescence was uneventful, and the patient discharged home in a good general condition, with stable vital signs and

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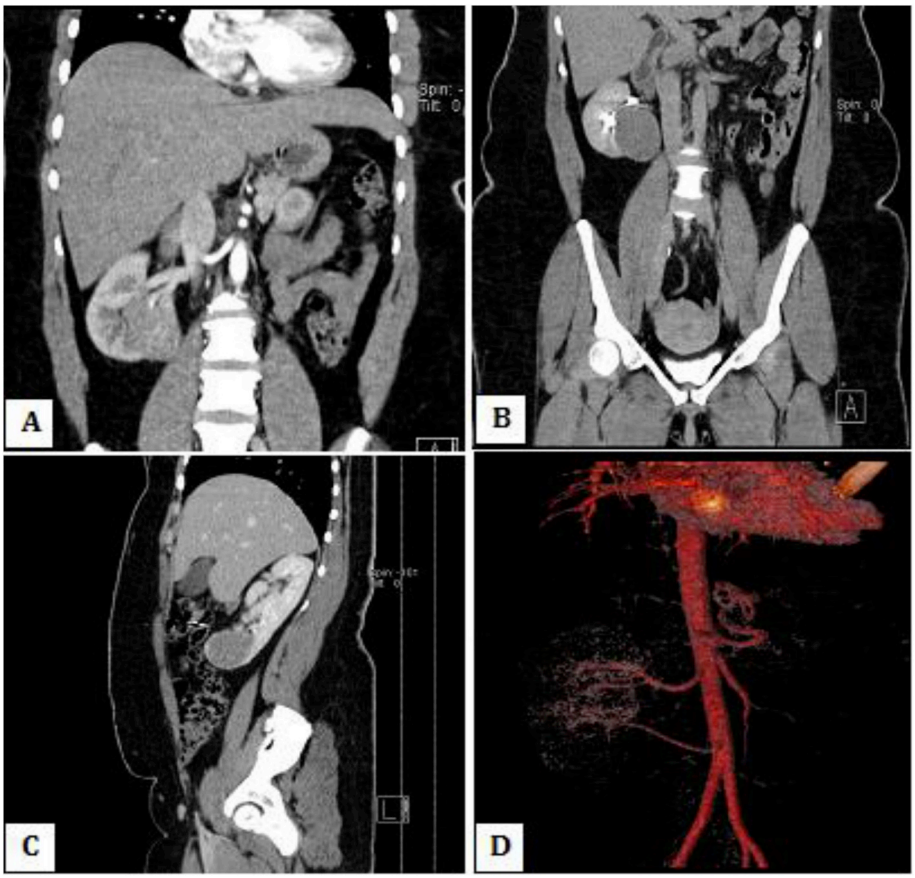


**Fig. 1.** Preoperative contrast-enhanced CT of the abdomen: coronal (A&B) and sagittal (C) sections showed evidence of crossed fused renal ectopia on the right side, with single separate renal pedicles and collecting systems per each kidney. CT angiography delineating renal arteries of both renal moieties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

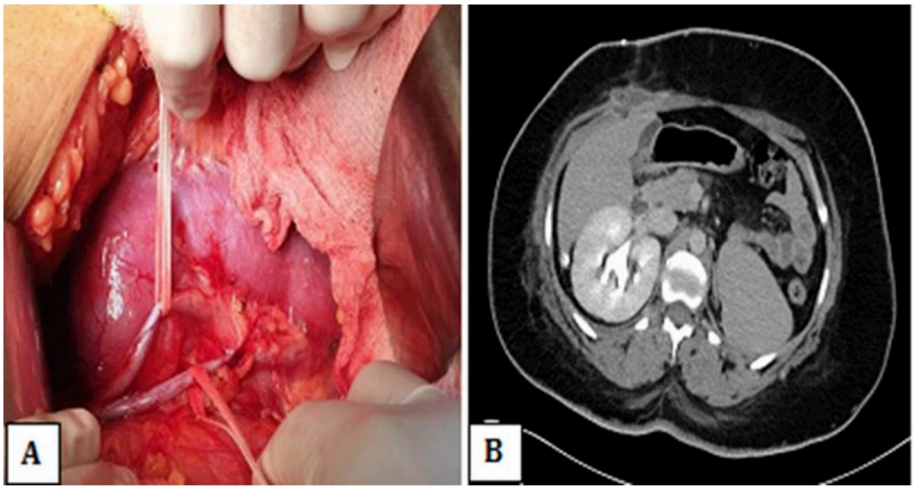


**Fig. 2.** Intraoperative exploration of ipsilateral two ureters (A) and renal postoperative axial (B) contrast-enhanced CT of the abdomen showed normal function of the remaining part of the affected kidney.

normal kidney function. Serum creatinine at discharge was 0.76 mg/dl and hemoglobin level was 9.7 g/dl.

Histopathology revealed unifocal chromophobe RCC, 6x 4 × 3.5 cm in size lower pole mass, which was yellowish in color, friable, with hemorrhagic areas and pathologic stage of pT1b, pNx, pMx, with no identified sarcomatoid or rhabdoid features (Fig. 3A–C). Immunohistochemistry showed CK7 to have strong diffuse positive (Fig. 3D), CD10 was weak multifocal positive, and CD34 did not highlight any venous invasion, while both C-kit and Vimentin were negative. Follow-up after 6- month showed normal kidney function and complete blood count. Contrast-enhanced CT study revealed no local recurrence or distant metastasis.

## Discussion

How these kidneys are drawn to the opposite side of the body has

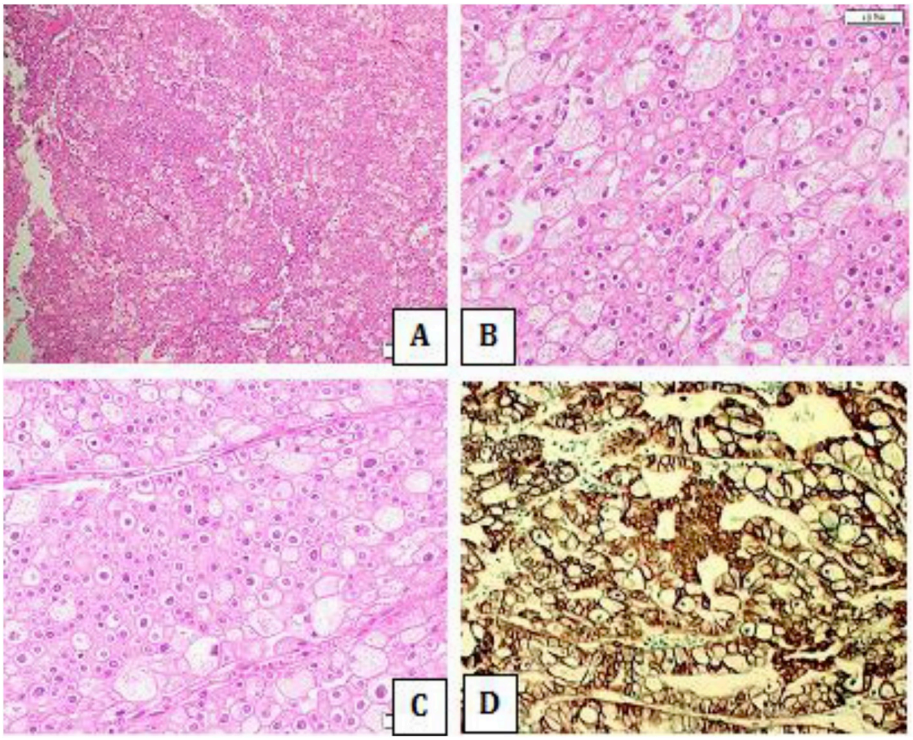


**Fig. 3.** Chromophore RCC (4X): Solid growth and nests of tumor cells composed of polygonal cells with "hard" or distinct cell borders. Granular and transparent cells are intermixed (A). High power (20X) showed mixture of cells composed of small cells with solid, slightly granular eosinophilic cytoplasm and cells having perinuclear halo/translucent zone in a background of pale, flocculent but not clear cytoplasm. Nuclei are irregular, wrinkled and angulated with perinuclear halos (B &C). Immunohistochemistry showed CK-7 positivity as strong and diffuse membranous (20X) (D).

never been satisfactorily explained. 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.[3]

Most of these patients usually have complications, such as hydronephrosis, nephrolithiasis, infection and rarely malignancy.[3] 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.[4]

Only eight cases of carcinoma in crossed fused renal ectopia were reported in the literature until 2017, since the first case was presented in 1942. A variety of surgical approaches were performed, where trasnperitoneal nephrectomy has been the slandered of care. Complete nephrectomy of renal moieties versus excision of the mass with preservation of normal functioning moiety depended on the clinical presentation and associated pathology. Four decades ago, *Gerber and associates* reported spread of malignancy to the normal moiety. The authors excised the tumor within the affected kidney with subsequent auto-transplantation of the residual kidney.[1] Only one case of renal malignancy in crossed fused ectopia has been managed with laparoscopic approach.[3] Intraoperative ultrasound was performed to determine the extent of tumor, which was excised and removed through a lower midline incision, with uneventful postoperative course.[5]

In the present index case, certain factors encourage nephron-sparing surgery, including young patient age, small tumor size, location of the mass in a favorable site, in addition to the availability of computed angiography, which is necessary before intervention.

## Conclusion

RCC in crossed fused renal ectopia represents a rare of rarity entity. Nephron-sparing surgery with preservation of normal-functioning moiety seems to be an excellent option in young patients with localized or small-sized mass. However, 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.

**CONSENT FORM:** A written consent was obtained from the patient for publication of this case report and accompanying images.

## Conflicts of interest

No potential conflicts of interest were disclosed.

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