

Case Report

Collecting Duct Carcinoma with Sarcomatoid Differentiation in S-Shaped, Crossed-Fused Renal Ectopic Kidney: A First Case Report and Review of the Literature

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Abstract

A 63-year-old male patient with hematuria and flank pain was admitted. Radiological evaluation revealed S-shaped, left-to-right cross-renal ectopia with a renal mass localized in the upper pole of the right kidney. Three-dimensional computed tomography (3D-CT) showed that the orthotopic moiety had 1 artery and 2 veins, and the ectopic moiety had 1 artery and 1 vein. A nephron-sparing partial nephrectomy was performed. The pathological diagnosis was collecting duct carcinoma with sarcomatoid differentiation. Using 3D-CT, we were able to achieve the appropriate pre-operative surgical planning for this challenging case. To our knowledge, this is the first case report of collecting duct carcinoma with sarcomatoid differentiation in a cross-fused renal ectopic kidney.

Keywords: Cancer, kidney neoplasm, nephron-sparing surgery

Cross-renal ectopia and fusion is a rare condition occurring in approximately 1 in 1000 live births.^[1] To best of our knowledge, to date, only fourteen cases have been reported in the literature. The incidence of renal malignancies in cross-ectopic kidney is no greater than that in normal population.^[2] Because the frequency of vascular variations is more common than normal population, surgical approach may become challenging. In this case report, we present a patient with collecting duct carcinoma with sarcomatoid differentiation in S-shaped, left-to-right cross-fused ectopic kidney.

Case Report

A 63-year-old male patient with the complaint of right lumbar pain and hematuria for a year was admitted. The pa-

tient had no family history of congenital anomalies. He is a smoker for 25 years and diabetic. He underwent an opened right pyelolithotomy by right flank incision for kidney stone 4 years ago. On laboratory examination, blood urea nitrogen was 48 mg/dL and serum creatinine level was 1.72 mg/dL; other values were normal. Ultrasonographic evaluation revealed 52×46-mm irregular, heterogeneous, hypoechoic renal mass on the upper side of the right kidney and 7-mm kidney stone in the middle renal calix. Because of high serum creatinine level, non-contrast-enhancement abdominal computed tomography (CT) was performed, which revealed malrotated right kidney and grade 2 pelvicaliectasia. Left kidney was on the right side and was fused with the lower pole of the right kidney. Millimeter-sized renal calculi were observed in the middle calyces. 46×28-

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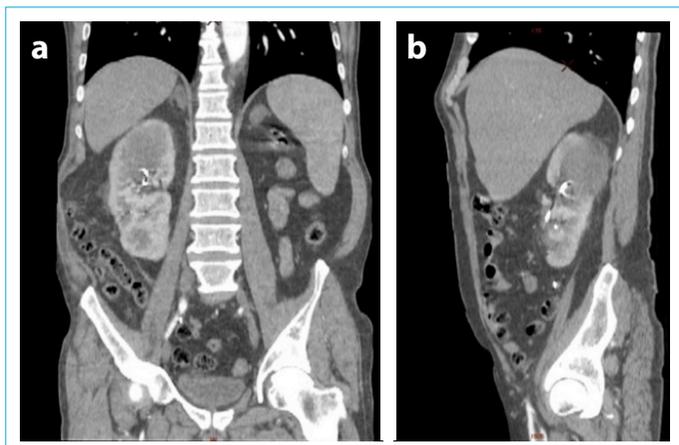


Figure 1 (a, b). Coronal (a) and sagittal (b) section imaging of kidney on computed tomography demonstrate renal mass in upper pole of the orthotopic moiety.

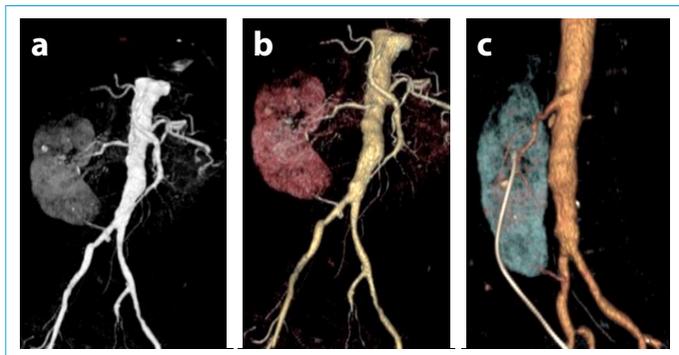


Figure 2 (a-c). Three-dimensional computed tomography demonstrates the arteries of both orthotopic and ectopic moiety.

mm sized, iso-hypodense, nodular, irregular renal mass was observed extending upper and middle lateral pole of right kidney. 26×16-mm lymph node was observed on para-aortic and aortico-caval region and was considered as metastasis of primary tumor (Fig. 1). Therefore, three-dimensional (3D-CT) was performed to demonstrate vascular anatomy. On 3D-CT, arterial supply of orthotopic moiety was arising from anteromedial part of the abdominal aorta from one artery and divided into three branches. Venous drainage of orthotopic moiety was draining into the inferior vena cava with two veins. In addition, arterial supply of ectopic moiety was arising from the antero-medial surface of the right common iliac artery from one artery and crossing common iliac artery anteriorly through the ectopic moiety. Venous drainage of the ectopic moiety was draining into the common iliac vein with one vein as well (Fig. 2). No distant metastasis was detected. Because of the patient's increased serum creatinine level, cystoscopy and ureterorenoscopy were performed. No abnormality was observed in the bladder and collecting system. Hematoma

was observed in the renal pelvis and double-J catheter was used to prevent urinary obstruction. On ureterorenoscopic evaluation, it was observed that the right and left kidney's renal pelvis were fused and continued with only one ureter. After the patient's serum creatinine level decreased to 1.7 ng/dL, open nephron-sparing partial nephrectomy was performed by midline incision. During the operation, one artery and two venous drainage were observed for the right kidney and one artery and one vein were observed for the left kidney. Left kidney was S-shaped and fused with lower pole of the right kidney. Only one ureter and fused renal pelvises were observed (Fig. 3).

Pathological Evaluation:

Gross Evaluation: A partial nephrectomy specimen with dimensions 9×7 cm, 5×6 cm. Sections revealed a solid, irregularly circumscribed, grey-yellow colored mass measuring 6.5×5 cm, with occasional areas of hemorrhage. Adjacent to the renal capsule, a yellow colored necrotic area with 3-cm diameter was observed.

Microscopic Evaluation: On hematoxylin and eosin-stained slides of the mass, sarcomatoid infiltration with occasional irregular tubule formation in an inflammatory desmoplastic stroma was observed. The cells forming the infiltration had eosinophilic or clear cytoplasm with prominent pleomorphism and nuclear atypia. The cytoplasmic borders of the cells were mostly indistinct. Large areas of

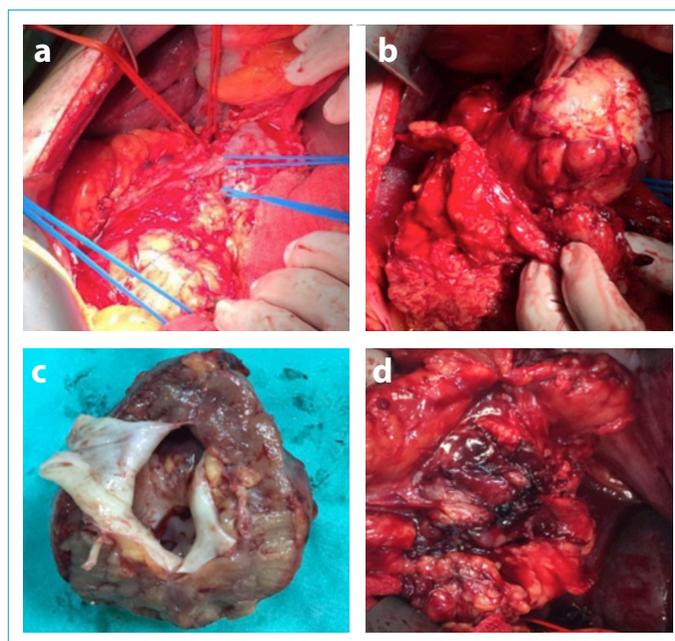


Figure 3 (a-d). Operative appearance of kidney. (a) Red tapes show arteries and blue tapes show veins. (b) Preparation of renal mass also observed in upper pole of orthotopic moiety. (c) View of surgical specimen. (d) View of orthotopic moiety after excision of mass.

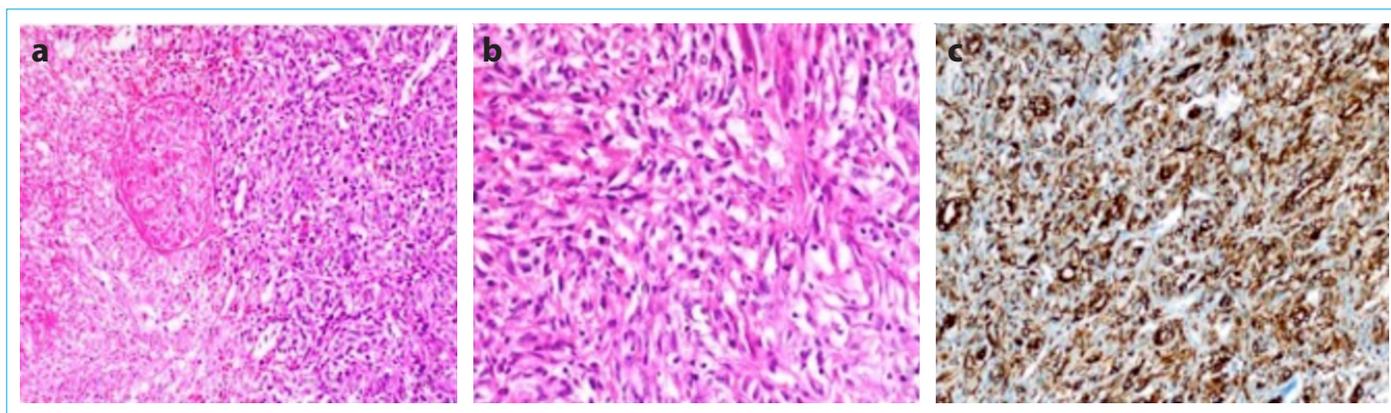


Figure 4 (a, c). Pathological view of surgical specimen. **(a)** Carcinoma zone was observed with wide necrosis and adjacent poor tubular structure (200×) **(b)** Sarcomatoid infiltration zone with increased mitotic index and prominent pleomorphism (400×) **(c)** Tumor cells were diffuse positive for CK7 (200×).

necrosis and increased mitotic activity were observed. Immunohistochemically, tumor cells were positive for pancytokeratin, CK 7, CD 117, and vimentin, whereas negative for p63, RCC, CK5/6, and CK20. According to the morphologic and immunohistochemical findings, a diagnosis of collecting duct carcinoma with sarcomatoid differentiation was made (Fig. 4).

Postoperative serum creatinine level of the patient was 3.5 ng/dL and potassium level of the patient was 4.2 mmol/L. Hemodialysis was not required postoperatively, and the patient was discharged without any complications on postoperative day 6 by comments of nephrologist. Because of the pathological features, the patient consulted an oncologist for chemotherapy. However, chemotherapy could not be administered because of low performance status of patient. Distant metastasis to lungs and brain occurred at postoperative month 4, and the patient died at postoperative month 6 because of distant metastasis.

Discussion

Cross-fused ectopic kidney is a rare renal abnormality. Ninety percent of crossed ectopic kidneys are fused with their ipsilateral mate. The ureter of the orthotopic kidney usually enters the bladder on the ipsilateral side and the ureter of the ectopic kidney crosses the midline and enters the bladder on the contralateral side. Its incidence ranges between 0.05% and 0.1% in the population. The crossed ectopic kidney is supplied by one or more branches from the aorta or common iliac artery. The normal kidney frequently has an anomalous blood supply, with multiple renal arteries originating from various levels of abdominal aorta.^[1] Regardless of the type of ectopia and fusion, vascular supplies of both ectopic and orthotopic kidney are variable.^[2] Cross ectopia is three times more common the left side than on the right side.^[3] Unless it is complicated, patient is frequently

asymptomatic. In the literature, few cases associated with malignancy have been reported.^[4] According to vascular varieties, surgical management may become challenging, and careful preoperative planning using imaging techniques, such as 3D-CT, magnetic resonance (MR) imaging, or CT/MR angiography are advised.^[5] Renal cell carcinoma is the most commonly reported pathological diagnosis for crossed ectopic kidney. In the literature, upper urinary system urothelial carcinoma has been reported in only one case. Collecting duct carcinoma is rare and comprises less than 1% of renal epithelial from the epithelium of Bellini's ducts, in the distal portion of the nephron. It is essential to differentiate this tumor from other renal tumors because of its aggressive behavior and early regional and distant spread.^[6] In the present case, we revealed collecting duct carcinoma with sarcomatoid differentiation. To best of our knowledge, this is the first case in the literature.

Surgical management of renal mass associated with renal abnormality contains complete resection of ectopic kidney if the mass is localized in ectopic moiety or excision of tumor.^[7] In the present case, because of increased serum creatinine level and localization of tumor, nephron-sparing surgery was considered and partial nephrectomy was performed. Preoperatively, 3D-CT was performed to reveal vascular anatomy. One arteria and two vein were observed for orthotopic moiety and one arteria and one vein was observed for ectopic moiety.

Conclusion

In terms of vascular anatomy, imaging result and operative appearance were compatible. We suggest that careful preoperative imaging is essential for surgical management in such cases. Thanks to developing imaging technology, favorable images can be obtained for appropriate preoperative surgical planning.

Disclosures

Peer-review: Externally peer-reviewed.

Conflict of Interest: None declared.

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