Oncology

Cross fused renal ectopia with associated renal cell carcinoma

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Introduction

Congenital malformations of the urinary tract are relatively common. As many malformations are relatively silent, the true incidence may not be truly known. The quoted incidence of malformation ranges from 3.3 to 11.1% and accounts for 50% of congenital abnormalities. Abnormalities in urinary tract development can include differences in development of the collecting system, presence or absence of renal units, ectopia, or fusion. 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. Congenital malformations of the urinary tract are relatively common. As many malformations are relatively silent, the true incidence may not be truly known. The quoted incidence of malformation ranges from 3.3 to 11.1% and accounts for 50% of congenital abnormalities. Abnormalities in urinary tract development can include differences in development of the collecting system, presence or absence of renal units, ectopia, or fusion. 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.

Case presentation

In this case, a 69 year old male presented to the clinic for evaluation of a 5.5cm right sided renal mass. He had previously been diagnosed with a solitary kidney, however, regular check up with an outside urologist yielded a retroperitoneal ultrasound concerning for a pancreatic lesion, and he was subsequently referred for a CT scan of the abdomen and pelvis. His medical history consisted of hypertension, pre-diabetes, and an unknown surgery in childhood that required adjuvant radiation to the neck. 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) and aorta at the L4-5 level (Fig. 1). Fusion of the right kidney to the left was evident just lateral to the aorta (Fig. 2). Additionally, a 5.5cm enhancing mass on the superior right renal unit was seen concerning for a primary renal cell carcinoma (RCC) (Fig. 2). Renal angiography and delayed images demonstrated complex vascular and ureteral collecting system anatomy (Fig. 3). Imaging was also significant for retroperitoneal and mesenteric lymphadenopathy, along hypodense hepatic lesions thought to represent hemangiomas.

After a thorough discussion regarding his likely diagnosis and treatment options, the patient elected to undergo open right radical nephrectomy and retroperitoneal lymph node dissection. 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. The renal vessels were then ligated with 2-0 vicryl sutures. The kidney and Gerota’s fascia was subsequently freed of its attachments.

The descending colon was mobilized after incision of the ipsilateral white line of Toldt to gain access to the left renal unit. A Fogarty 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. Bioglue was injected over the renorrhaphy. The transplant surgery team then performed an ultrasound of the liver due to concern for a liver lesion, which was ultimately negative for any concerning lesions.

On pathological review of the nephrectomy specimen, the final diagnosis was a T1b Furhman grade 2 clear cell renal cell carcinoma.

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Fig. 1. Right to left cross-fused ectopic kidney with overlying vasculature. Blue arrow marks aorta, orange arrows mark renal units. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

Fig. 2. Sagittal image showing fusion of right kidney to left, lateral to the aorta. Blue arrow depicts aorta. Orange arrow depicts inferior mesenteric artery traversing over isthmus. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

Fig. 3. Right renal artery and vein wrapping around the right renal mass. Middle image shows second right renal artery and vein entering the right renal hilum. Last image shows duplicated right collecting system with the blue arrows and the left ureter depicted with the orange arrow. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)
Discussion

Current literature on cross-fused renal ectopia consists of case reports.\(^3\)\(^4\) Its overall prevalence is estimated to be 1 in 900 live births.\(^5\) This congenital renal abnormality in conjunction with a renal mass is even more uncommon. (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.\(^7\) In our case, the right kidney demonstrated 2 sets of renal vessels with the inferior arising from the right common iliac vessels. Delayed phase imaging also identified a right duplicated ureteral pelvic system with early proximal confluence to form a right ureter. Furthermore, intraoperative exposure should allow access to both renal hilums, as ligation of ipsilateral vessels may not fully provide hemostasis as the contralateral kidney's vascular supply remains intact. In our case, we used a Fogarty vascular clamp to provide regional ischemia during renorraphy to maintain the contralateral renal unit's vascular perfusion. As this particular ectopic kidney was anatomically centered over the IVC and aorta, a transperitoneal approach was used to allow direct and immediate access to the retroperitoneum for appropriate vascular control. Our patient did well in the post-operative setting and was discharged home on postop day 4 with bowel function and tolerating a regular diet.

Conclusion

Crossed-fused renal ectopia with a renal mass represents an uncommon urological diagnosis. It presents an operative challenge for the surgeon during peroperative 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.\(^6\) 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 renorraphy will need to be accomplished to achieve hemostasis and repair the resection site.

Consent

Images were obtained through EMR appropriate means with verbal and written consent by the patient.

Declarations of interest

None.

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