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Renal cell carcinoma in a patient with crossed and fused renal ectopy: A case report

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A B S T R A C T

Crossed-fused renal ectopia (CFRE) is a rare congenital disease in which one of the kidneys with its ureter crosses the midline and fuses with the contralateral kidney. The association of this malformation with the presence of primary renal cell carcinoma (RCC) is even more anecdotal; there are only a few cases reported in the literature. We describe the case of a 62 year-old man with CFRE associated with renal cell carcinoma, which was successfully removed by retroperitoneoscopy, after careful preoperative study of blood supply and anatomical features.

1. Introduction

Crossed-fused renal ectopia is a congenital malformation with an estimated incidence of 1 in 2000 live births, being more common in men (3:1).[1] It represents the second cause of renal fusion malformation, after horseshoe kidney. It is characterized by the presence of an ectopic kidney that crosses the midline, merging with the contralateral orthotopic kidney. Because patients are mostly asymptomatic, it is usually diagnosed as an imaging finding. However, it can manifest as a palpable abdominal mass, or with symptoms derived from multiple associated complications, such as a higher rate of urolithiasis and recurrent urinary tract infection. Malignancy in CFRE is extremely rare[2]; there are less than 10 cases reported in the literature.[3] Surgery for tumor resection in these patients is a real challenge due to the anatomical alterations present, especially those related to the vasculature.

2. Case presentation

A 62-year-old man with a history of arterial hypertension and peripheral facial palsy, was studied with thorax abdominal pelvis CT with contrast after having a parenchymal brain hematoma in an atypical site. The image reported the presence of both kidneys in the flank and right iliac fossa, jointed by a band of parenchyma that fused the lower pole of the right kidney with the upper pole of the left one, compatible with fused crossed renal ectopia (Fig. 1- A). In the upper pole of the right unit, a partially exophytic hypervascular solid cortical mass was observed with wash out during the elimination phase, measuring 4.4 × 3.8 × 3.3 cm in the longitudinal, transverse and anteroposterior axes, respectively, consistent with renal cell carcinoma (RCC) (Fig. 1-B, Fig. 2). The lesion focally contacted the adipose tissue of the renal sinus, without contacting the perirenal fascia nor blood vessels. Two right renal arteries and a single left renal artery were observed, which originated distal to the inferior mesenteric artery. There was no evidence of metastatic involvement. RENAL Score was 7 points.

Partial nephrectomy was performed by retroperitoneoscopy, without complications. The biopsy confirmed renal cell carcinoma, clear cell variety, of 3.2 cm, Grade 1 WHO/ISUP Classification. No tumor necrosis, lymphovascular invasion, nor perirenal adipose tissue involvement was observed. The surgical margins were negative. At 6 months of follow-up, the patient is well without any complications.

3. Discussion

Of all cases of renal ectopy, in up to 90 % the ectopic kidney is fused with its contralateral.[4] Likewise, a higher prevalence has been described in males, and renal ectopy is more common from left to right, in a 3:1 ratio.[5] Regarding vascularization, it has been showed that in up to a quarter of the cases of CFRE the renal arteries originate from the upper abdominal aorta, while in the rest they originate from the lower abdominal aorta, or from the iliac arteries.[5] In our patient, there were two right renal arteries and one left, which originated distal to the origin of the inferior mesenteric artery.

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Usually, patients with CFRE are asymptomatic, constituting an imaging finding, as was the case with our patient. However, it can be accompanied by other malformations (mainly musculoskeletal and genitourinary) or by complications such as recurrent urinary tract infection, urolithiasis, hydronephrosis, and less commonly, associated with malignancy.[4] It should be noted that the prevalence of cancer in kidneys with some type of malformation is similar to that observed in the general population[5]; however, up to the date of publication of this article and to the best of our knowledge, fewer than ten cases of RCC have been reported in the literature.

Ultrasound findings suggestive of CFRE include the presence of an anterior or posterior notch in the evaluated kidney, along with the inability to visualize the contralateral kidney in its renal fossa. Additionally, it can be observed that the two collecting systems are oriented differently, contrary to what would be seen in the case of a kidney with a duplicated collecting system. A study revealing more detailed anatomical variations and associated malformations is recommended for clear characterization, especially when considering surgery in case of complications. The best images to evaluate CFRE are contrast-enhaced CT scan and MRI, both widely available. Other options are intravenous pyelography and angiography. Among the differential diagnoses to consider are horseshoe kidney, solitary kidney, solitary kidney with duplicated collecting system, or solitary kidney with a tumor.[4]

The management of these patients will primarily depend on the associated complications they have. In the case of association with malignancy, as in our patient's case, the surgical approach should be personalized based on the vascular alterations found in the preoperative study. Among the surgical approaches described in the literature for CFRE cases with RCC, transperitoneal partial nephrectomy is described as the most commonly used technique. However, in the presented case, retroperitoneoscopic partial nephrectomy was chosen for since the tumor was located at the posterior pole of the right kidney, which was difficult to mobilize as it was fused with the contralateral kidney, making tumor excision extremely complicated *via* the transperitoneal approach. For the installation of the ports, an incision was made 2 cm above the iliac crest at the level of the posterior axillary line, with expansion of the space using a surgical glove with physiological saline. Two 12 mm trocars were installed; one for the camera at the level of the posterior axillary line, and another for the right hand under the twelfth rib in the anterior axillary line. The left 5 mm trocar was then installed lateral to the psoas muscle (Fig. 3). The ectopic kidney was not specifically sought during surgery. Intraoperative images of the surgery are attached (Fig. 4-5). Estimated blood loss during surgery was 100 cc. No drainage was left, and postoperative creatinine was 0.9 mg/dL, without variations compared to preoperative creatinine. The patient was discharged after spending 48 hours hospitalized, without complications.

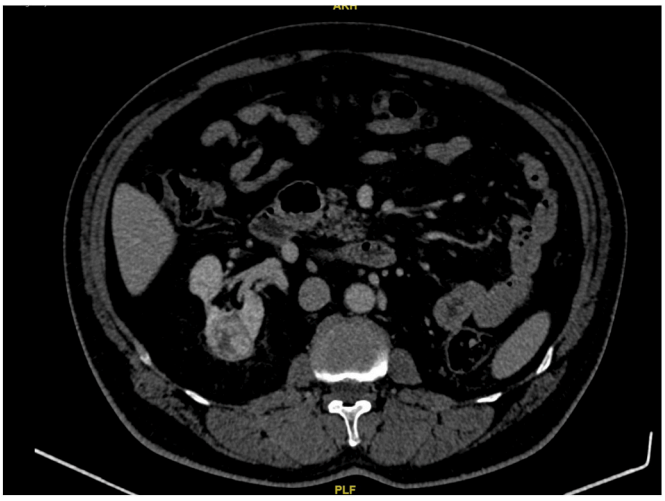

Fig. 2. Exophytic solid cortical mass in the upper pole of the right kidney.

To this date and to the best of our knowledge, this is the first CFRE with RCC operated using this technique.

4. Conclusion

Renal cell carcinoma in CFRE is a very rare entity, which represents a challenge for the practicing urologist. When presenting an altered anatomy, the decision of the surgical approach and the technique used is usually not easy, so a complete study with meticulous surgical planning is essential to avoid unexpected intraoperative findings that could lead to complications that are difficult to manage.

Consent

Informed consent was obtained from the patient for publication of this case report and its associated images.

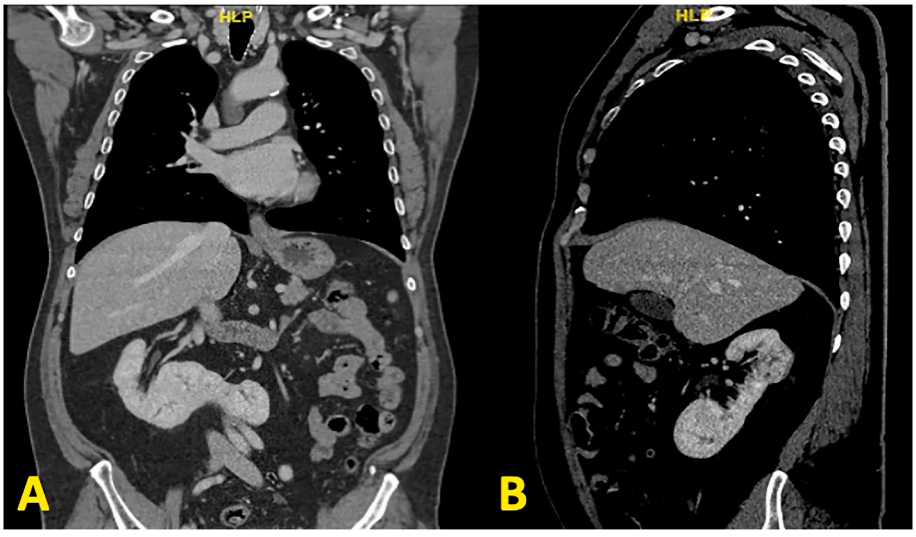

Fig. 1. A; Preoperative Abdominal Pelvic CT with contrast (A) Coronal view, showing the ectopic left kidney fused to the lower pole of the right kidney. (B) Sagital view, showing a renal mass in the upper pole of the right kidney.

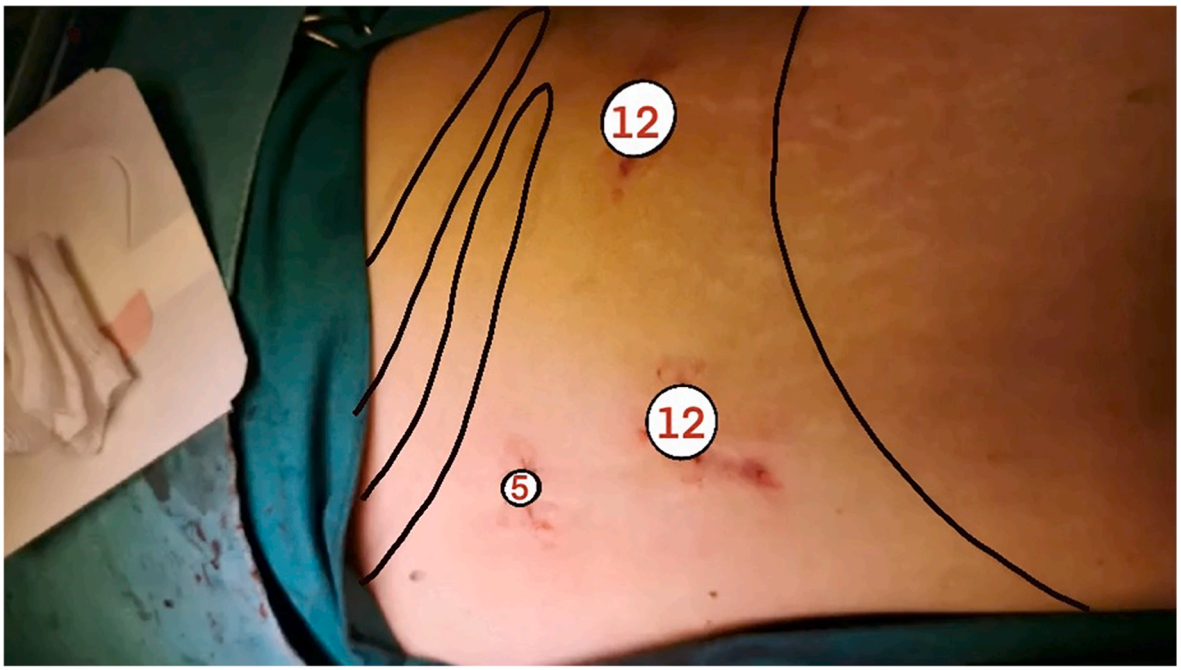

Fig. 3. Location of the ports, with their respective sizes in mm.

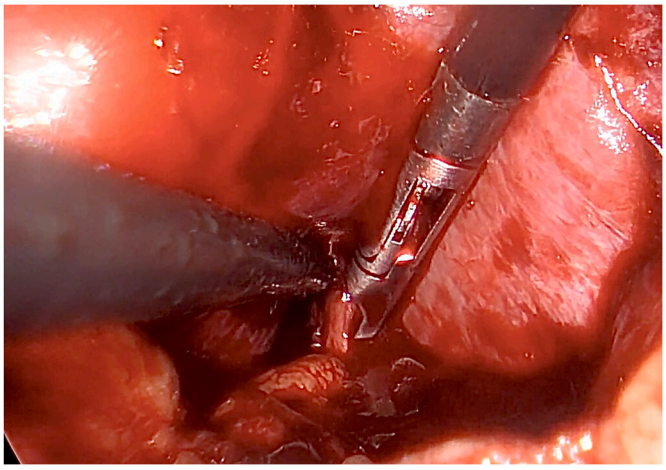

Fig. 4. Intraoperative picture; identification of the right main renal artery.

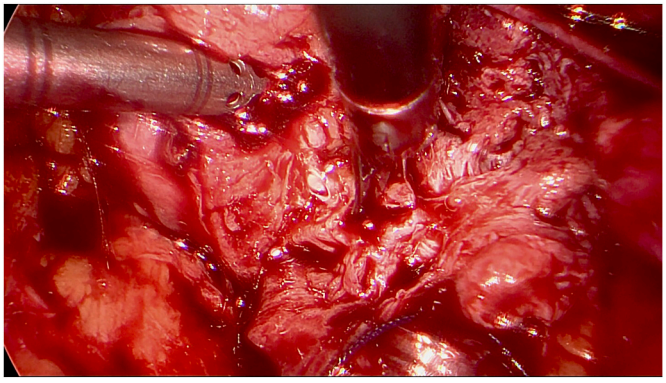

Fig. 5. Intraoperative picture during tumor dissection.

CRediT authorship contribution statement

Joan Cristóbal Bassa: Writing – review & editing, Project administration, Investigation. **Juanita Fernández Wenzel:** Writing – review & editing, Writing – original draft, Investigation. **Jorge Almonacid Grunert:** Resources, Formal analysis. **Pablo Aroca Siré:** Data curation. **Cristian Muñoz Fuentes:** Writing – original draft, Formal analysis, Data curation.

Declaration of competing interest

The authors declare that there are no conflicts of interest regarding the publication of this article.

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