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Laparoscopic nephrectomy in crossed fused kidney ectopia with severe hydronephrosis in 2 year-old child

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ABSTRACT

Introduction: There are no specific recommendations for the surgical management of crossed fused renal ectopia (CFRE), but a laparoscopy has been proposed in a limited number of children. We report the case of a complex CFRE with severe hydronephrosis in a child treated by laparoscopic nephrectomy.
Case report: A 2-year-old boy was admitted for hip and abdominal pain. A CFRE was diagnosed. Laparoscopic nephrectomy was suggested. The right crossed ectopic kidney appeared fused at its upper pole to the lower pole of the left kidney. The left kidney was in total rotation postero superiorly The left ureter crossing over the ectopic dilated right pelvis deliniated the fusion line between both kidneys. The right ureter was dissected distally. The right artery and vein were dissected and the right kidney pelvis opened making the separation of the kidneys easier and so reducing the danger of damaging the lower pole of the left kidney. During the dissection an aberrant vein arising from the left iliac vein and draining from both kidneys was identified. The right vein was dissected preserving the left vein, and the nephrectomy completed.
Conclusions: Laparoscopy is a recommended option to treat CFRE, offering all advantages of this approach.

1. Introduction

Crossed renal ectopia (CRE) is a rare congenital anomaly where both kidneys are located on the same side of the spine, and, if the ectopic kidney is fused with the opposite kidney, it is defined as crossed fused renal ectopia (CFRE) [1-5]. Ninety percent of crossed ectopic kidneys are fused to the contralateral one [5]. The exact incidence of CFRE is not known because the majority of patients are asymptomatic and are often diagnosed incidentally. However, an estimated prevalence of 1:2000 to 1:7000 has been found in autopsy series [5]. Abdominal pain, hematuria, fever, urinary tract infection, hypertension, renal failure, a palpable abdominal mass or anorectal malformation [2,3,5,6] may be present in symptomatic patients.

There are no specific recommendations for the management of crossed fused renal ectopia. The surgical treatment is guided toward the associated problems that lead to either symptoms or the deterioration of kidney function. The laparoscopic approach has been reported as an acceptable option to treat this anomaly in a limited number of pediatric cases [2,3,6], with all the associated advantages of minimally invasive surgery.

We report a case of right CFRE (both kidneys found on the right side of the spine) with severe hydronephrosis in a 2 year-old child, who presented with addominal pain, and was successfully submitted to laparoscopic nephrectomy.

2. Case report

A 2-year-old boy was admitted to hospital for hip pain without any urinary symptoms. The patient had a prior perinatal diagnosis of hydronephrosis in an ectopic right kidney but was lost to follow up.

On clinical examination, the child presented right hip pain with very mild limitation of mobility as well as mild abdominal tenderness

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in the right lower quadrant, without any rebound tenderness. External genitalia were normal, testicles were in place and he had a satisfactory urinary stream. There were no signs of heart defect or skeletal abnormalities. Urinalysis, blood urea nitrogen and creatinine were all normal.

Kidney ultrasonography showed a crossed ectopic severly hydronephrotic right kidney. Uro-MRI confirmed the diagnosis with fusion bewteen the upper pole of the right crossed ectopic kidney and the lower pole of the left kidney (Figs. 1 and 2). MAG3 scintigraphy demonstrated only 4% right residual renal activity. Laparoscopic nephrectomy was advised.

The child was positioned in a supine position, slightly tilted to the left side. A 3 mm laparascope was inserted at the right hypocondrium. A 3 mm trocar was inserted at the left hypocondrium and a 5 mm trocar at the right iliac fossa. Another 3 mm trocar for traction was inserted at the right umbilical line. The mesoperitoneum covering the right crossed ectopic hydronephrotic kidney was dissected to uncover the renal pelvis. The right ureter was identified and retracted on 2/0 Ethibond thread introduced percutaneously. This exposure made it much simpler to identify the whole distal ureter as well as the pelvi-ureteric junction. The right crossed ectopic kidney appeared fused at its upper pole to the lower pole of the left kidney. The right renal vein and artery were identified.

The left kidney that was in total rotation postero superior which made the identification of the pelvi-ureteric junction very difficult. The ureter that was crossing over the dilated right kidney pelvis deliniates the fusion line between both kidneys. Intraoperative images are showed in Fig. 3.

A vessel loop passed through the 5 mm trocar was used to deliver traction to the right vessels to clearly identify the exact dissection line.

The right ureter was followed to its near insertion in the bladder and dissected using Thunderbeat®.

The right artery and vein were dissected using 5 mm Thunderbeat and ligasure. The right kidney pelvis was opened and the separation between the two kidneys was thus made easier and whilst also reducing the danger of damaging the lower left kidney pole. During the dissection, an aberrant vein coming from the iliac vein and draining from both kidneys was identified. The right vein was dissected by Thunderbeat whilst preserving the left vein. A small remnant of right kidney calyx was closed with 2/0 PDS endoloop.

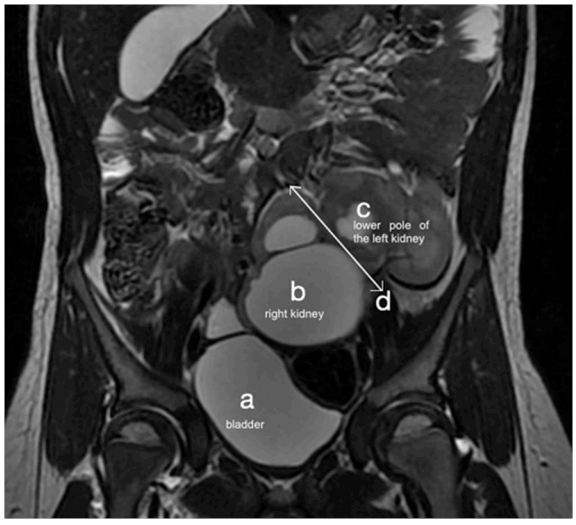

Fig. 1. Preoperative uro-MRI. The location of the bladder (a), right kidney pelvis (b) and left kidney (c) is showed. The line fusion between the upper pole of the right crossed ectopic kidney and the lower pole of the left kidney is also represented (d).

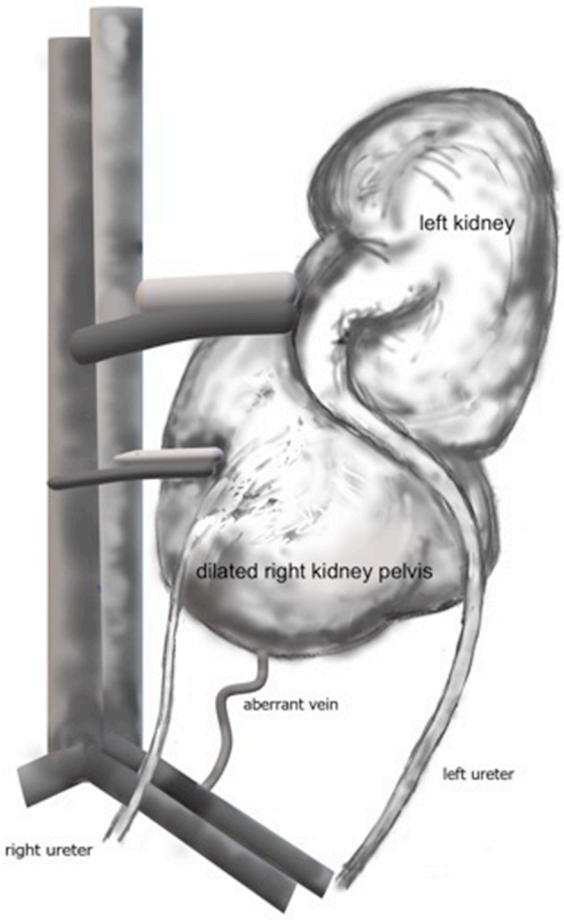

Fig. 2. Illustration of the renal malformation. The dilated right kidney pelvis appeared fused at its upper pole to the lower pole of the left kidney. The left kidney was in total rotation postero superior. The left ureter was crossing over the right kidney pelvis deliniating the fusion line between both kidneys (drawn by Dr. Salvatore Amoroso).

The entire kidney was removed by enlarging the 5 mm incision at the RIF to 1.5 cm. Control of hemostasis was then achieved. Closure of the wounds by glue and the RIF using 4/0 Vicryl and 5/0 Monocryl to skin.

There were no perioperative or post-operative complications. The child was discharged 48 h postoperatively.

3. Discussion

Crossed fused renal ectopia results from the abnormal development of the ureteric bud and metanephric blastema during the fourth to eighth weeks of gestation [7,8] Under normal conditions, the two masses of metanephrogenic tissue arise in the pelvis and ascend to their definitive position in the lumbar region, bilaterally [9]. During the formation of fused kidneys, the nephrogenic blastemas will be compressed between the umbilical arteries at the beginning of the cranial migration of the ureteral buds, and this can lead to their fusion. Fused kidneys are prevented from ascending and so remain in an ectopic pelvic position [9,10].

CFRE is the second most common congenital anomaly of the kidney and urinary tract after “horseshoe kidney”. Most patients with CFRE remain asymptomatic and are only incidentally encountered on autopsy or during routine medical imaging for unrelated disorders [1-5]. However, CFRE may become clinically significant because approximately half the patients manifest complications such as, hydronephrosis, infections and nephrolithiasis. We describe a pediatric

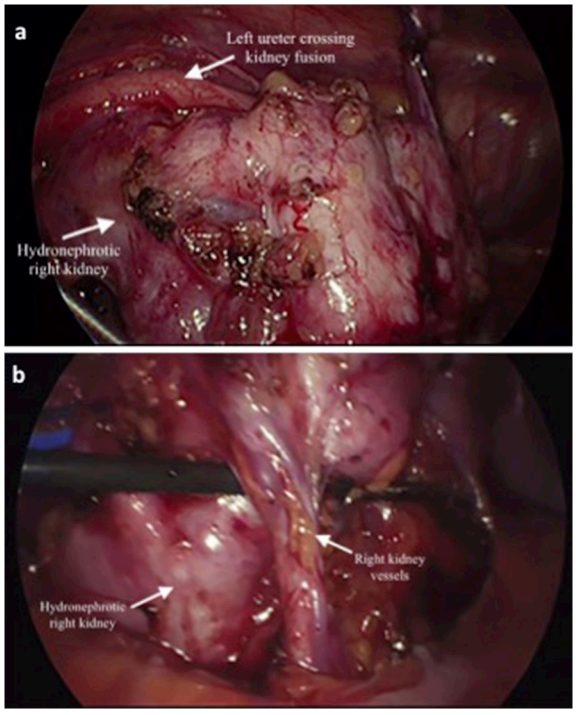

Fig. 3. Intraoperative images. Panel a: visualization of the left ureter crossing above the kidney fusion; Panel b: identification of the right kidney vessels.

case in which CFRE was associated with severe hydronephrosis with mild addominal pain, and was successfully treated by laparoscopic nephrectomy.

Ultrasonography is the primary imaging carried out in most cases for detecting congenital fusion anomalies. However, CT and MRI can give more precise anatomical configuration of the kidneys and their collecting system, and also detect any associated anomalies [11,12].

According to McDonald and McClellan's classification, six subtypes of CFRE are described: unilateral fused kidney inferior ectopia type, sigmoid or S-shaped kidney, lump kidney, L-shaped kidney, disc kidney and unilateral fused kidney superior ectopia type [13]. Vascular supply may be derived from a single renal artery (from the distal aorta or common iliac) and a single renal vein (draining into inferior vena cava or common iliac veins) [7,13].

In our case, CFRE type D was diagnosed. This type of malformation, in which the ectopic kidney is placed horizontally fusing with the lower pole of the normal kidney, shows abnormalities of position (ectopia), migration, rotation and vascular supply [7]. The presence of such a renal fusion anomaly and, in addition, an anomalous blood supply, poses difficulties and risks during interventional procedures. Therefore, the treatment of CFRE represents a potentially difficult surgical task. Angiography and MRU are indicated in CRFE prior to surgery as this will greatly aid in the surgical planning and management and help to avoid complications, especially when nephrectomy is indicated.

The laparoscopic approach is a possible option to treat CFRE. As reported by previous investigators, laparoscopic treatments have been successfully performed [14,15] in the pediatric age-group. As in our case, laparoscopic surgery offered the well-established benefits of good exposure, reduced blood loss, shorter operative time, and faster convalescence, versus a traditional open procedure. The minimally invasive approach also allows one to limit the risks of complications in the treatment of a complex malformation, with a good surgical result.

In conclusion, the laparoscopic approach appearsa very effective approach for the treatment of CFRE. The management of CFRE is individualised according to the underlying urological anomaly and vascular supply. The preservation of as much renal function as possible is important whenever possible; when the residual renal activity is not sufficient, nephrectomy is indicated. The laparoscopic approach offers the combination of good exposure with limited risk of complications, and all the other advantages of minimally invasive surgery.

Patient consent

Consent to publish the case report was not obtained. This report does not contain any personal information that could lead to the identification of the patient.

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Declaration of competing interest

The following authors have no financial disclosures: GN, ZN, SA, VC, GP.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.epsc.2019.101368.

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