CLINICAL CASE

Crossed renal ectopia with fusion and multiple renal calculi managed with nephrectomy through the anterior paramedian approach

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KEYWORDS
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Abstract  Congenital renal anomalies are not common. Crossed renal ectopia (CRE) is the second most frequent abnormality after horseshoe kidney. Its diagnosis is usually incidental in the third or fourth decade of life or when the patient presents with urinary tract infections, hematuria, lithiasis, or renal-ureteral colic. The aim of this article was to present the case of a patient with the classic symptoms of renal-ureteral colic who was diagnosed with CRE with an inferiorly fused, non-functioning kidney secondary to multiple renal calculi, and to describe the management with nephrectomy through the extraperitoneal anterior paramedian approach. A 39-year-old man presented with a classic case of renal-ureteral colic and during his evaluation multiple renal calculi outside of the normal renal topography were found. An abdominal computed tomography (CT) scan was done and CRE was diagnosed. The non-functioning inferiorly fused kidney was revealed in the contrast-enhanced and three-dimensionally reconstructed CT, and management with nephrectomy was decided upon. We believe that the extraperitoneal anterior paramedian approach provides good access for this type of congenital anomaly, given the anterior location of the renal unit.
Introduction

Crossed renal ectopia (CRE) with fusion represents 85% to 90% of the published cases and the most frequent variety is the unilateral fused kidney with inferior ectopia. CRE is defined as the position in the retroperitoneum different from the normal one, in which the kidney crosses the midline and is situated contralaterally to the side where its ureter normally inserts into the bladder. Diagnosis is incidental in the majority of cases and symptomatology is vague, simulating renal-ureteral colic in some cases. It is associated with urinary tract obstruction due to bridles or abnormal vascular supply to the ectopic kidney. It is also associated with other abnormalities such as vesicoureteral reflux, ureterocele, and anorectal malformation. Diagnosis is made through excretory urogram and contrast-enhanced three-dimensional computerized tomography (CT) and treatment is directed at the complications, more than at the anatomical anomaly, itself.

Case presentation

A 39-year-old man had illness onset 4 years earlier, with the clinical presentation of right renal-ureteral colic that was never managed by a physician. The patient self-medicated with anti-inflammatory agents for pain control. However, 7 months ago he presented with similar but more intense symptoms, to which were added fever, hematuria, and irritative urinary symptoms. He sought private medical attention and the physician ordered a simple abdominal x-ray and referred the patient to our hospital for management. He was first evaluated at the emergency room and was found to be in good general health. A complete blood count, blood chemistry, serum electrolytes, and urinalysis were ordered. The blood tests reported leukocytosis of 11,800, hemoglobin 12.2 g/dl, platelets 352,000, glucose 98, creatinine 0.8, and urea 17. The urinalysis showed a pH of 5.5, leukocyturia, erythrocyturia, and bacteriuria. The abdominal x-ray (fig. 1) revealed radiopaque images suggestive of stones measuring approximately 2 cm x 1.5 cm in the right lower quadrant above the iliac crest. An abdominal CT scan showed the absence of the left kidney and an enlarged right kidney with calcifications under the lower pole (fig. 2A); other views displayed an image that could correspond to an ectopic left kidney with multiple stones in its interior that was fused with the right kidney (figs. 2B and 2C). There was inadequate contrast material uptake by the fused ectopic kidney shown in the urotomographic reconstruction (fig. 3A).

The patient was evaluated together with the transplantation service. An angiotomography scan (fig. 3B) revealed a single artery and vein and inadequate vascular supply to the ectopic kidney. Nephrectomy through the extraperitoneal paramedian approach was performed on the ectopic kidney. It was carried out with no complications and the total surgery duration was one hour 10 minutes. Blood loss was 500 cc. Nine stones measuring 2.5 cm x 1.5 cm were extracted from the surgical specimen. The patient was released from the hospital after 72 hours.

Discussion

Lithiasis in kidneys that have some type of anatomical alteration is a particularly great challenge for the urologist, due to the fact that the abnormal anatomy prevents the use of the same disintegration or extraction access routes that are utilized in normal kidney units. Embryologically, the definitive kidney originates at the fifth week of intrauterine life and its development depends on the chemical interaction of the ureteric bud near its joining with the continuous mass of non-differentiated mesenchymal cells called the metanephric blastema. This union ascends from its pelvic position toward the ipsilateral renal fossa, turning inwards on its longitudinal axis until the definitive renal-ureteral unit is formed during the following 3...
weeks. Pelvic kidney, ectopic kidney, or renal malformation are explained by developmental defects in the migration stage.\textsuperscript{2,3} Normal fetal and embryonic development of the kidney can be altered by various factors that, in turn, are associated with other urinary tract malformations. A total of 35% to 40% of the congenital abnormalities are located in the genitourinary tract and 10% of all living beings are born with some type of urinary tract anomaly.\textsuperscript{2}

The ectopic kidney is defined as one that is congenitally in a position different from its usual location in the lumbar region due to a flaw in the process of its ascent, and that crosses the midline and becomes situated on the opposite side from where it normally connects to the bladder.\textsuperscript{2,4} CRE is the second most frequent anomaly with fusion after horseshoe kidney. The first case was published by Pamarolus in 1654 and in 1957 it was classified by McDonald and McClellan.\textsuperscript{2,3} Their classification is the one currently in use: crossed ectopia with fusion, which makes up 85% of the cases; inferior crossed ectopia without fusion; solitary crossed ectopia; and bilateral crossed ectopia. The fused varieties are divided into: unilateral fused kidney with inferior ectopia; sigmoid or S-shaped kidney; L-shaped kidney; disc kidney; and unilateral fused kidney with superior ectopia.\textsuperscript{5-8} Various theories have been proposed, but the precise mechanisms by which CRE occurs are not known.\textsuperscript{5} Among them are the mechanical theory, the ureteral theory, the theory of biochemical stimuli-induced migration, the teratogenic theory, and the theory of the abnormal rotation of the caudal end of the developing fetus.\textsuperscript{3,5}

CRE with fusion is a rare malformation with an incidence of 1:7,500;\textsuperscript{6} it is more frequent in men with a ratio of 1.4:1,\textsuperscript{7} and the left-to-right ratio is 3:1.\textsuperscript{3}

Its clinical presentation is asymptomatic in the majority of cases and it generally develops in the third or fourth decade of life.\textsuperscript{8} Because irrigation is different from the norm, kinking or compression of the urinary tract can cause ureteropelvic junction stricture, usually of the ectopic kidney, resulting in hydronephrosis with or without lithiasis in 9% of the cases,\textsuperscript{1,4,6} hematuria, non-specific abdominal pain, recurrent urinary infections, and renal-ureteral pain, among others.\textsuperscript{3}

Its diagnosis is based on intravenous urography; contrast-enhanced 3-dimensional CT is usually the best imaging technique for detailing the situation of the ectopic kidney.\textsuperscript{2,3,5,7,8} Treatment should be opportune and directed at the complications rather than at the congenital anomaly itself, and includes antibiotic prophylaxis, extracorporeal lithotripsy,
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Conclusions

Our patient presented with the classic symptoms of renal-ureteral colic and during his evaluation multiple renal calculi outside the normal kidney topography were found. Abdominal CT scan was done and CRE was diagnosed. Contrast-enhanced 3-dimensional reconstruction revealed a fused non-functioning kidney with inferior ectopia for which nephrectomy was performed. In our opinion, the extraperitoneal anterior paramedian approach is a good access route for this type of congenital anomaly, given the anterior situation of the renal unit.

Conflict of interest

The authors declare that there is no conflict of interest.

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References