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Case Report

Crossed nonfused renal ectopia with variant blood vessels: a rare congenital renal anomaly

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ABSTRACT

Crossed renal ectopia is a rare congenital anomaly, where one of the kidneys crosses the midline and lies opposite to the site of its normal ureteral insertion. Ninety percent of crossed ectopic kidneys are fused to their ipsilateral uncrossed kidney. Crossed renal ectopia without fusion is rare. We present the case of a 53-year-old male with an unusual incidental finding of crossed nonfused renal ectopia, with the left ectopic kidney lying anterior to the right kidney without fusion. The ectopic kidney had dual arterial supply: one from the aorta and another from the right renal artery. It also demonstrated dual venous drainage; a main left renal vein and an accessory renal vein. The main left renal vein joined the right renal vein to form a common renal vein before draining into the inferior vena cava. The accessory renal vein joined the left testicular and left lumbar veins to drain into the inferior vena cava. Multiple bilateral nonobstructing renal calculi were also noted. Although the patient was asymptomatic, the authors highlight potential complications related to the above-mentioned condition and the importance of identification of the findings.

Introduction

Crossed renal ectopia (CRE) is an uncommon congenital renal anomaly, where the ectopic kidney crosses the midline and is located contralateral to its normal position, and may be fused to the normal kidney or remains nonfused [1].The ureter of the ectopic kidney enters the bladder at its normal position. The blood supply to the kidneys in such cases shows many variations [2,3]. Although, it is often symptomless and found incidentally, it may be associated with urolithiasis, hydronephrosis, recurrent urinary tract infections, and vesicoureteral reflux. This article presents a case review of an incidental finding of crossed nonfused renal ectopia with anomalous blood vessels, in a male patient undergoing computed tomography (CT) scan for evaluation of a hepatic cyst.

Case report

A 53-year-old male, who was referred for evaluation of a hepatic cyst, underwent CT scan of the chest, abdomen, and

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pelvis. The scout radiograph showed absence of the normal renal outline on the left side, and an ectopic renal shadow was identified on the right side below the right kidney (Fig. 1). Pulmonary and hepatic hydatid cysts were noted along with cholelithiasis; however, the below-discussed findings regarding the kidneys were observed.

The right kidney was visualized in its normal position, but the left kidney was found on the right side, anterior to the right orthotopic kidney, and seen distinctly separate from it with no evidence of fusion (Fig. 2). The left kidney also appeared to be malrotated, with the hilum facing anteriorly. Multiple small nonobstructing calculi were noted in both kidneys. A small simple renal cyst measuring about 1.7 × 1.3 cm was also seen in the lower pole of the ectopic kidney. The ureter of the ectopic kidney descended on the right side for a short distance before crossing the midline, just below the bifurcation of the abdominal aorta and anterior to the right common iliac artery, to its normal position of insertion into the left side of the urinary bladder.

The arterial supply showed marked variation. The ectopic left kidney was noted to have a dual blood supply with the supplying arteries going through the renal capsule instead of the normal route through the renal hilum. The left renal artery originated from the aorta just lateral to the origin of the inferior mesenteric artery, ascending in a spiral manner and entering the left ectopic kidney in the lower pole by piercing the renal capsule (Fig. 3). The second artery, arising as a branch from the right renal artery about 4 cm from its origin, supplied the upper pole of the ectopic kidney (Fig. 4). The right kidney was supplied normally by the right renal artery originating from the aorta. Dual venous drainage of the ectopic kidney was also noted with one main left renal vein seen emerging from the renal hilum of the ectopic kidney and joining the right renal vein to form a short confluence before draining into the lateral aspect of the inferior vena cava (IVC) on the right side (Fig. 5). Another smaller accessory renal vein was seen emerging from the mid pole of the ectopic kidney, and joining the left testicular and left lumbar veins to drain into the IVC on the left side (Fig. 6).

There was also considerable variation in the drainage of the testicular veins. As mentioned above, the left testicular, left lumbar, and the left accessory renal veins joined to form a common trunk, which drained directly into the IVC (Fig. 6). The right testicular vein drained into the main left renal vein instead of the IVC (Fig. 7).

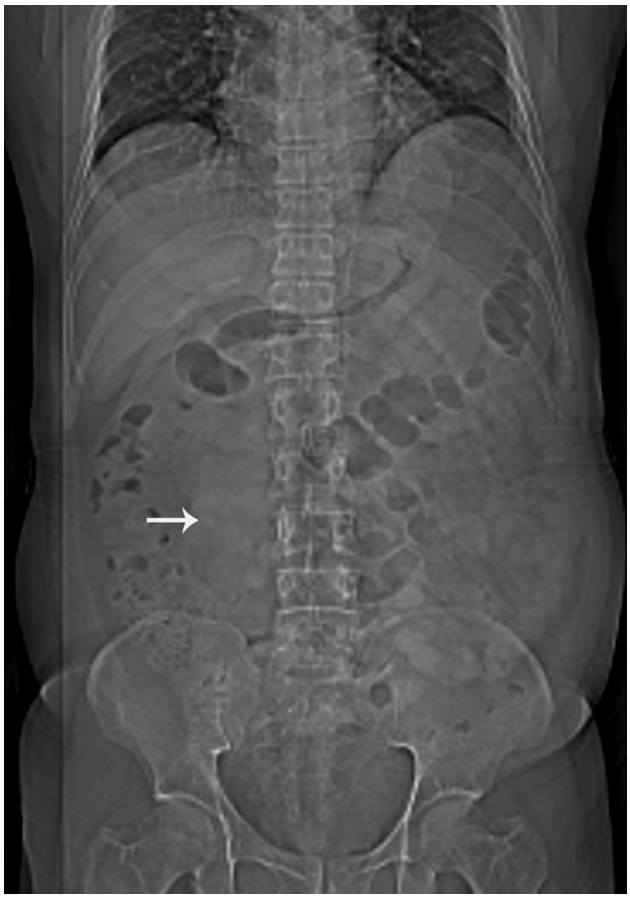

Fig. 1 – Scout radiograph showing absence of the left renal shadow and an ectopic renal shadow on the right side (arrow).

Discussion

CRE is a rare congenital anomaly in which a ureter crosses the midline from an ectopic kidney, which lies opposite of its normal location, to insert in its usual location in the urinary bladder. The autopsy incidence of CRE is reported as 1:7000. The incidence of nonfused CRE; however, has been reported to be 1 in 75,000 autopsies, which is about 10 times lower than that of the fused ectopia [3]. CRE is more common in men with a male to female ratio of 1.4:1 and is 2-3 times more frequent with the ectopic kidney on the right side than on the left side [4].The ectopic kidney is mostly found to be malrotated. CRE has been classified into 4 types by McDonald and McClellan [5]: *type A*, CRE with fusion; *type B*, CRE without fusion; *type C*, solitary CRE; and *type D*, bilaterally CRE.

In type A, the ectopic kidney crosses over to the opposite side and fuses with the normally located kidney. Commonly, the upper pole of the inferiorly positioned ectopic kidney fuses with the lower pole of the normally positioned kidney. The ureter of the ectopic kidney crosses the midline and enters the bladder in its usual position. It is further divided into the following subtypes: (1) unilateral fused inferior renal ectopia with the upper pole of the crossed ectopic kidney fusing with the lower pole of the orthotopic ipsilateral kidney; (2) sigmoid or S-shaped kidney in which the crossed kidney lies inferiorly with the renal pelvis facing laterally and the normally positioned kidney lies superiorly with the pelvis facing medially; (3) unilateral lump or cake kidney with fusion occurring over a wide margin, and both renal pelvises directed anteriorly; (4) L-shaped kidney in which the ectopic kidney lies inferiorly and transversely fusing with the lower pole of the normal kidney; (5) unilateral disc kidney in which the fusion occurs along the medial border of each pole; and (6) unilateral superior crossed fused ectopia type in which the lower pole of the superiorly positioned ectopic kidney fuses with the upper pole of the normally positioned kidney. It is the least common type.

In type B of CRE, the ectopic kidney crosses the midline, but each unit is separate and no fusion of renal parenchyma is

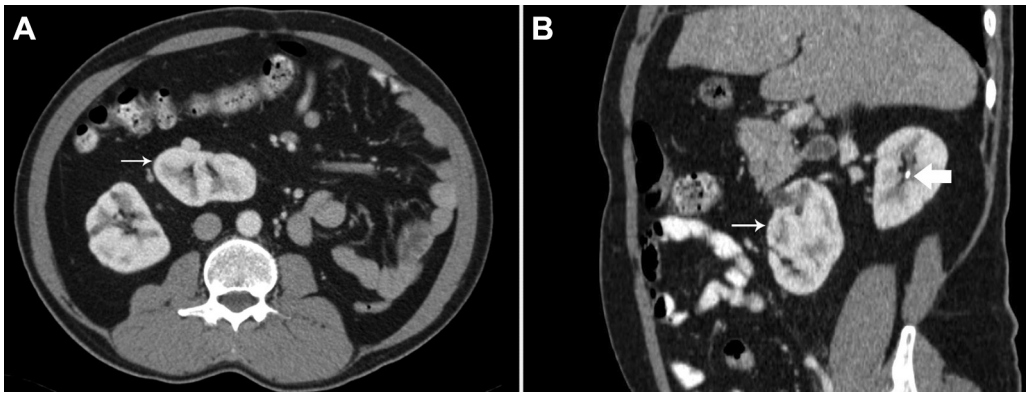

Fig. 2 – Contrast-enhanced computed tomography (CT) abdomen in the axial (A) and sagittal (B) views showing ectopic position of the left kidney (arrow) located anterior to the normally located right kidney with no fusion. A small calculus is also visible in the orthotopic right kidney (wide arrow).

seen. Type C demonstrates a solitary crossed kidney with unilateral renal agenesis. The ectopic ureter in solitary ectopia crosses the midline and enters the urinary bladder at its normal position. In type D, both kidneys are crossed with the left kidney located on the right and the right kidney located on the left. The ureters of both kidneys descend and cross the midline to enter the bladder in their usual locations [5]. Our patient demonstrated type B of CRE without fusion, which is rare with only a handful of cases reported in the literature to date [6].

Other types of renal migration anomalies are simple renal ectopia, which is characterized by the abnormal position of the kidney in the cranial or caudal direction, but at the same side of its origin. There are 4 different recognized types of simple renal ectopia: (1) pelvic or sacral kidney commonly located in the pelvis, but may also be seen opposite the sacrum or below the aortic bifurcation; (2) lumbar or iliac ectopic kidney located in the iliac fossa above the iliac vessels, but below the level of L2 and L3; (3) superior ectopic kidney found in the subdiaphragmatic position; and (4) thoracic kidney located above the diaphragm and in the chest [7]. The most common renal fusion anomaly is the horseshoe kidney in which the 2 renal masses are fused together in the midline. In most of the cases, the lower poles of both kidneys are jointed together across the midline and lying below the origin of the inferior mesenteric artery.

The normal embryological development of the kidney results from the interaction between the ureteric bud originating from the mesonephric duct, and the metanephric blastema during the fourth and fifth weeks of intrauterine life. In the beginning, the kidneys lie close to each other, with the hila facing anteriorly in the pelvis. Between the sixth and ninth weeks, the embryonic kidneys gradually ascend to the lumbar region along the posterior abdominal wall to their final position. During ascent, the kidneys also undergo a 90° axial rotation, so that the hila are eventually facing medially. Finally, the ureteric buds divide to form the pelvicalyceal system [8]. The embryological development of renal ectopia and other renal congenital variants is not fully understood, and many theories have been offered to explain it. Initially, during the ascent of the ureteral buds, the nephrogenic blastemas may get compressed between the umbilical arteries, which may lead to their fusion. If the metanephric masses are at the same level, then complete fusion results in a cake kidney, whereas fusion at one renal pole (usually the lower pole) results in a horseshoe kidney. It is thought that if the

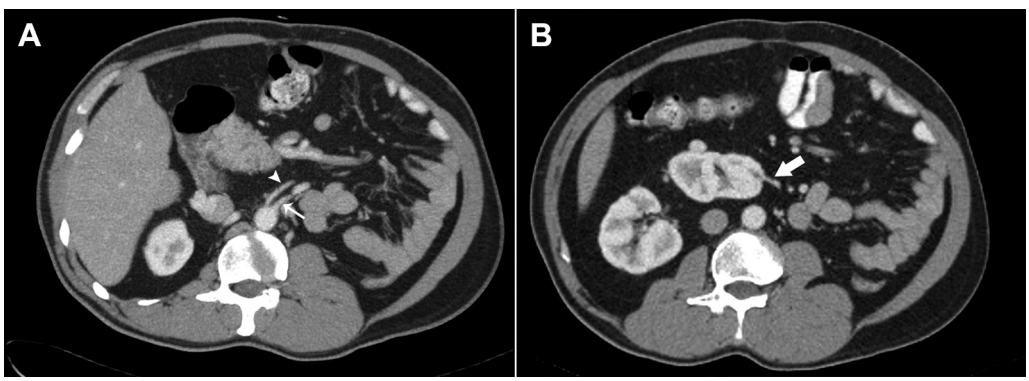

Fig. 3 – Axial views of contrast-enhanced CT abdomen. (A) Origin of the left renal artery (arrow) from the aorta lateral to the inferior mesenteric artery (arrowhead). (B) Left renal artery entering the lower pole of the ectopic kidney (wide arrow).

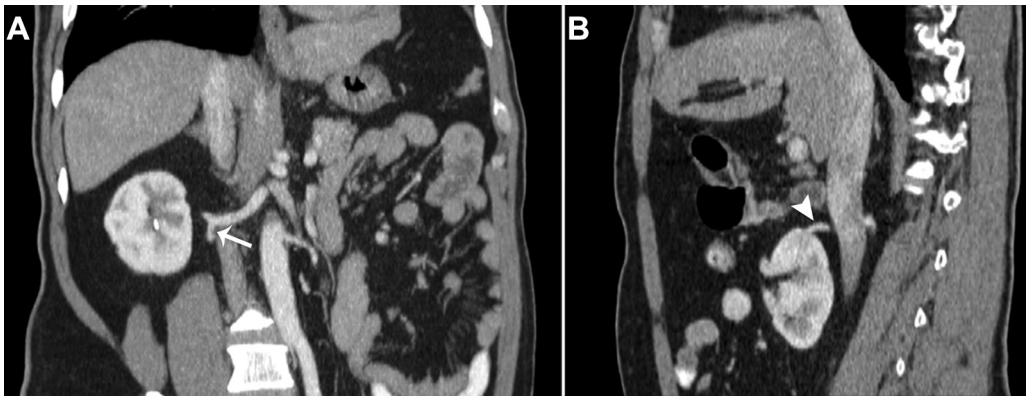

Fig. 4 – Coronal (A) and sagittal (B) views of contrast-enhanced CT abdomen showing the origin of the left renal artery (arrow) from the right renal artery and entering the upper pole of the ectopic kidney (arrowhead).

compression factor of the umbilical arteries persists at the start of the cranial migration in the presence of 2 unequal metanephric masses, the result will be crossed ectopia [9]. Horseshoe kidneys associated with parenchymatous isthmus are thought to be the result of a teratogenic event involving the abnormal migration of posterior nephrogenic cells [10]. The cranial migration of the cake and horseshoe kidneys is limited to the origin of the inferior mesenteric artery from the abdominal aorta. It is also postulated that abnormal rotation of the caudal end of the embryo prevents the ureteric bud from communicating with the ipsilateral metanephros, and it is attracted toward the closer contralateral side. In such cases, the migrated and the normally located ureteric buds induce the metanephric blastema twice to form 2 kidneys on one side resulting in crossed ectopia [11]. The nephrogenic tissue from the side that does not receive a ureteric bud will completely regress [12]. In very rare instances, the migrating kidney overshoots and ascends to a more cephalad location, resulting in superior ectopia or thoracic kidney [13]. During the cephalad migration, the kidneys derive their blood supply from neighboring vessels: initially median sacral, then common iliac and inferior mesenteric, and finally, the aorta. As the kidneys reach their final position, permanent renal arteries and veins develop. If migration is arrested at any point, then the temporary blood vessels become permanent, as a result, the ectopic kidneys usually contain many small vessels, which reflect the continuous changes in blood supply of the developing kidneys during the course of renal ascent [14].

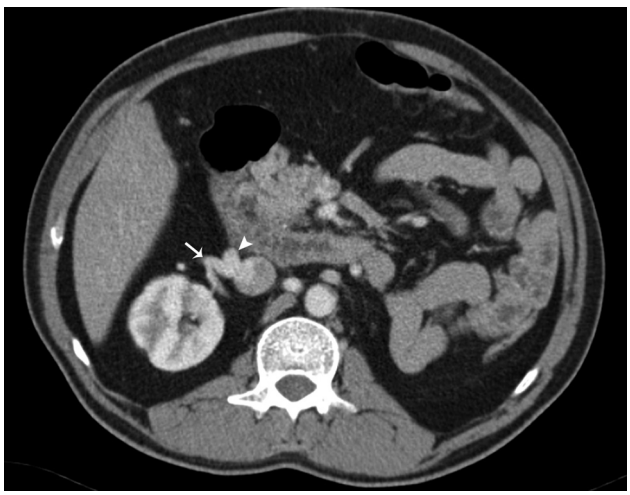

Fig. 5 – Contrast-enhanced CT abdomen in the axial view showing the right renal vein (arrow) and the main left renal vein (arrowhead) joining together to form a confluence and draining into the inferior vena cava (IVC).

Ultrasound is usually the initial modality that identifies the incidental finding of renal ectopia. It may demonstrate an empty renal fossa on one side and an ectopic kidney on either the contralateral side, subdiaphragmatic or pelvic in position. The characteristic ultrasound findings in crossed fused renal ectopia include an anterior and/or posterior notch with difference in orientation of the 2 collecting systems in the fused kidneys [15]. Horseshoe kidneys commonly demonstrate poor visualization of the lower borders of the kidneys on ultrasound. Renal ectopia may be suspected on plain abdominal radiograph, if the outline of either kidney is not seen in its expected location or if an abnormal mass is seen on a chest radiograph with an empty renal fossa on ultrasound, indicating a thoracic kidney. Intravenous urography often aids in providing anatomical details regarding the position of the kidneys and the course of the ureters. Micturating cystourethrography is recommended in cases presenting with infection to exclude vesicoureteral reflux. If obstruction is suspected on ultrasound, then drainage may be assessed using a technetium-99m mercaptoacetyltriglycine-3 renal scan. Radionuclide imaging may reveal a functioning isthmus in the case of horseshoe kidneys. CT or gadolinium-enhanced MR urography permits excellent definition of the exact anatomy in such cases.

CRE usually remains undiagnosed and may be found incidentally when a patient is being examined for other conditions, as was the case in our patient. If symptomatic, patients usually present with flank and lower abdominal pain, a palpable abdominal mass, hematuria, dysuria, and urinary tract infections [16]. The pathogenesis of renal ectopia is due to the lower positioning of the ectopic kidney, which is

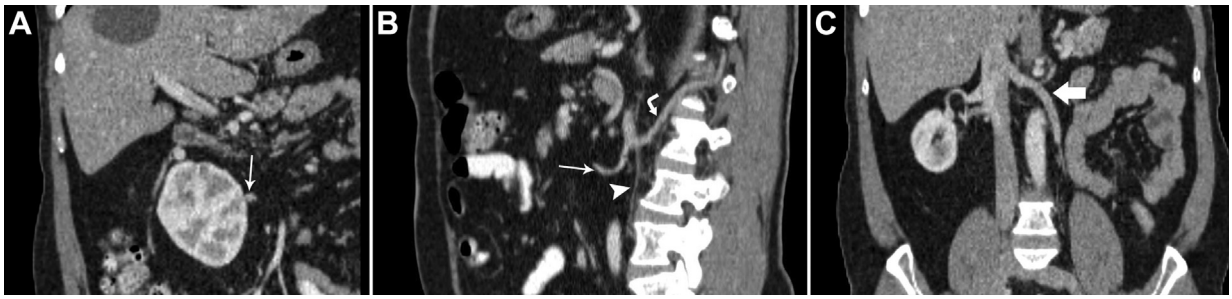

Fig. 6 – Coronal (A and C) and sagittal (B) views of contrast-enhanced CT abdomen showing the left accessory renal vein (arrow) joining the left testicular vein (arrowhead) and left lumbar vein (curved arrow) to form a common vein (wide arrow) draining into the IVC.

associated with a shorter ureter increasing the susceptibility to reflux and infection. Pelvi-ureteric junction obstruction is also noted, which is either due to a high insertion of the ureter on the renal pelvis, malrotation of the kidney, or an anomalous blood supply, which obstructs the collecting system [1]. The presence of an isthmus in horseshoe kidney also contributes to pelvi-ureteric junction obstruction. The most common complication noted in patients with crossed ectopia is vesicoureteral reflux, which may lead to deteriorating kidney function [17]. Other renal complications include hydronephrosis, infections, and nephrolithiasis. Obstruction resulting in urinary stasis and increased risk of infection has been thought to be the major contributing factor for nephrolithiasis. Urological abnormalities reported in children with renal fusion anomalies like horseshoe kidneys include ureteral duplication, ectopic ureter, ureterocele, and a retrocaval ureter [18]. The ectopic kidney, owing to its more anterior location in the lower abdomen or pelvis, and often located in a less-protected anatomical position in the retroperitoneum, may be more susceptible to blunt trauma [19]. There have also been reports of iatrogenic injury to the anomalous ureter during unrelated surgical procedures of the abdomen [6,20].

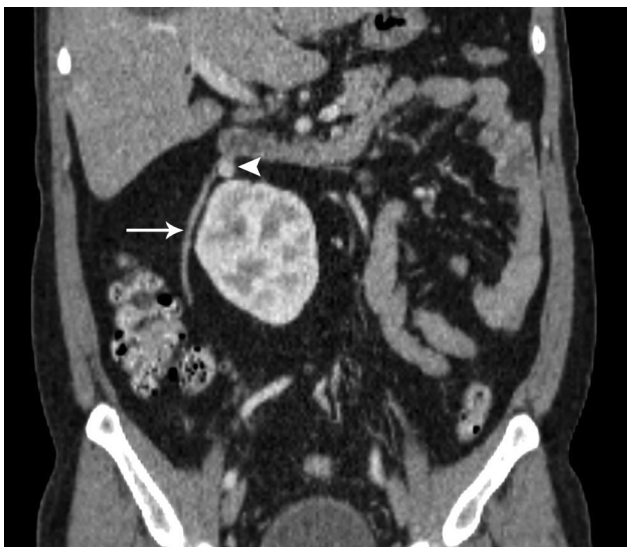

Fig. 7 – Coronal view of contrast-enhanced CT abdomen showing the right testicular vein (arrow) draining into the main left renal vein (arrowhead).

Other associated anomalies of renal ectopia include genital abnormalities in men like cryptorchidism and hypospadias [21]. In girls, genital anomalies such as agenesis of the uterus and vagina, for example, müllerian agenesis or unicornuate uterus have been associated with ectopic kidneys [22,23]. It can also be associated with other nonrenal anomalies (adrenal, cardiac, and skeletal abnormalities), and as a clinical feature in syndromes such as CHARGE syndrome (coloboma, heart disease, atresia choanae, retarded growth and development, genital hypoplasia, and ear anomalies) and VACTERL syndrome (vertebral, anal, cardiac, tracheal, esophageal, renal, and limb anomalies) [24,25]. Thoracic kidneys are often associated with congenital diaphragmatic hernia [26]. Horseshoe kidneys can be a feature of many fetal syndromes, such as Turner syndrome, Edwards syndrome and trisomy 9 [27,28]. Patients with horseshoe kidneys also appear to have an increased risk of Wilms tumor [29]. Primary renal cell carcinomas have also been reported in patients with crossed fused renal ectopia [30].

In conclusion, we reported a case of crossed nonfused renal ectopia with variant blood vessels, which was an incidental finding. Although multiple nonobstructing renal calculi were noted bilaterally, the patient did not complain of any urinary symptoms. In the absence of associated abnormalities, there is no specific treatment for CRE. Treatment is indicated in the presence of related pathologies like symptomatic nephrolithiasis and vesicoureteral reflux. Furthermore, preoperative recognition of the anomaly and its variant blood vessels is essential for any planned surgeries in the renal region. Hence, thorough assessment of the possible anomalies is necessary for proper diagnosis and management of these patients.

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