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Ureteric Fusion – An Unusual Presentation of Crossed Fused Renal Ectopia

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ABSTRACT

Background: *Crossed fused renal ectopia is a rare congenital anomaly where the kidneys are fused and located on the same side of the midline. The ureter of the ectopic kidney crosses the midline and enters the urinary bladder on the opposite side. The purpose of this case report is to show the rare but important variation of this anomaly and to highlight its embryological and clinical significance.*

Case: *A 58-year-old female presented with upper abdominal pain. CECT abdomen showed choledocholithiasis with an incidental presence of right-sided crossed fused renal ectopia with ureteric fusion.*

Conclusion: *Careful assessment of an incidental finding on imaging is also very important for overall prognosis besides searching for the actual etiology behind the clinical manifestations. Any congenital anomaly or normal variant is important to document. This prepares both the patient and the treating clinician for any future endeavour/eventuality.*

Keywords: *renal ectopia; crossed fused renal ectopia; ureteric fusion*

INTRODUCTION

When a kidney is located on the side opposite to that of its ureteric insertion into the bladder, it is defined as crossed renal ectopia and if it is fused with the normally located kidney then it is defined as crossed fused renal ectopia. More than 90% of crossed ectopic kidneys are fused to one another. The estimated incidence is around 1 per 1000 births.^[1] There is a male predilection with a 2:1 male to female ratio. We present an unusual case

of 58-year-old woman who not only had crossed fused renal ectopia but also fused ureters.

CASE REPORT

A 58-year-old female presented to us with requisition for contrast-enhanced computed tomography (CECT) of abdomen. The patient had upper abdominal pain and jaundice. She had undergone ultrasound examination by sonologist in her village, who reported contracted gallbladder

and prominent common bile duct (CBD). Additionally, left renal fossa was mentioned to be empty.

CECT abdomen revealed two calculi in CBD with contracted gallbladder explaining the patient's symptoms. Incidentally, right-sided crossed fused renal ectopia was also detected [Figure 1]. The kidneys were malrotated with their pelvis lying anterolaterally. The inferolateral pole of the superior kidney (measuring 7.2 cm in its greatest dimension) was fused with the superomedial pole of the inferior one (7.9 cm in its greatest dimension) in shape of "figure of three". Together, the kidneys were lying along L2 to L5 vertebrae. The proximal ureters of both the kidneys were also fused and single ureter

continued below to open normally into ipsilateral side of urinary bladder. Mild hydronephrosis involving the lower kidney was present. No renal/uretric calculus or perinephric fat stranding was present. The parenchyma of both kidneys showed normal enhancement. Superior kidney was supplied from normally originating ipsilateral renal artery. The arterial supply of the inferior one could not be properly delineated. However, both the kidneys drained through separate renal veins into the ipsilateral side of inferior vena cava.

The empty renal fossa showed no defined presence of renal fascias. Instead, the jejunal loop was seen extending into the region. There was no associated skeletal abnormality.

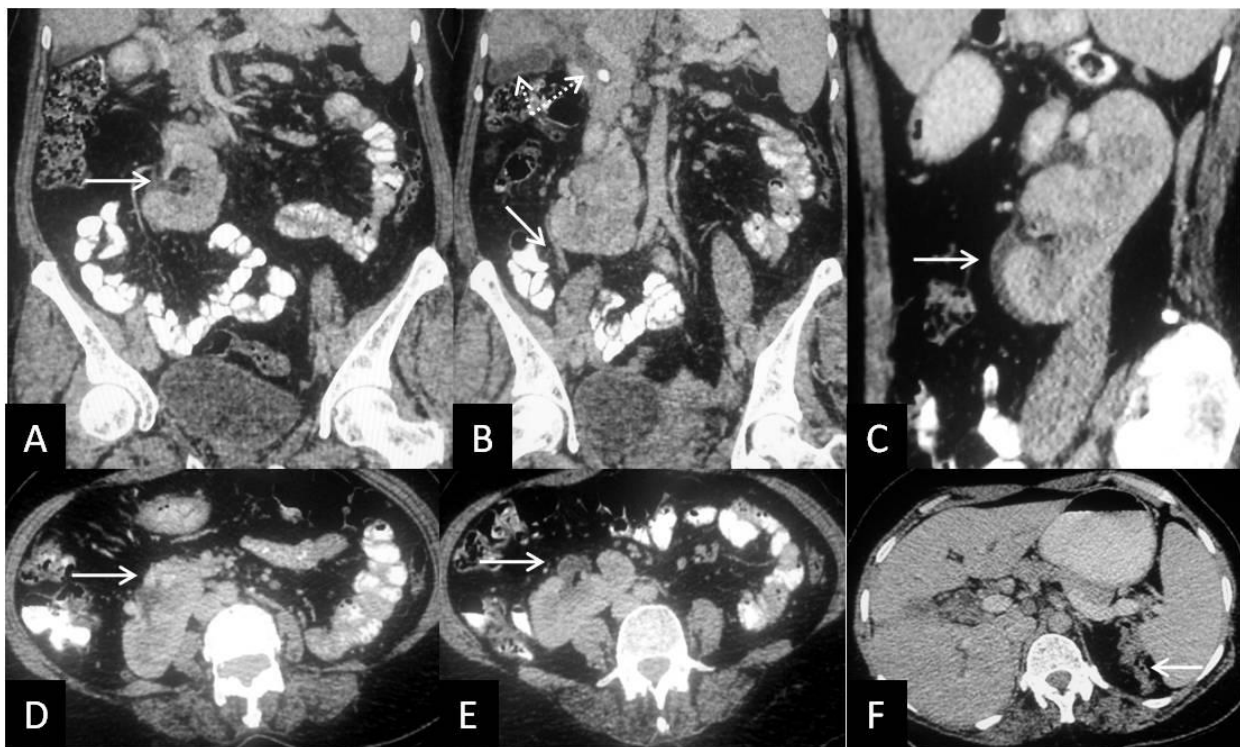


Figure 1 (A-F): Coronal (A-B), sagittal (C) and axial (D-F) sections of CECT abdomen show presence of right crossed renal ectopia with fusion of supero-inferior poles. Their proximal ureters are fused (arrows in A-C) leading to single ureter draining into bladder. Superior (arrow in D) & inferior (arrow in E) kidneys are malrotated & show normal parenchymal enhancement with mild hydronephrosis of the inferior one. Contralateral renal fossa shows jejunal loops & loss of defined renal fascias (arrow in F). Cholelithiasis is also seen with dilated CBD & contracted gallbladder (dashed arrows in B).

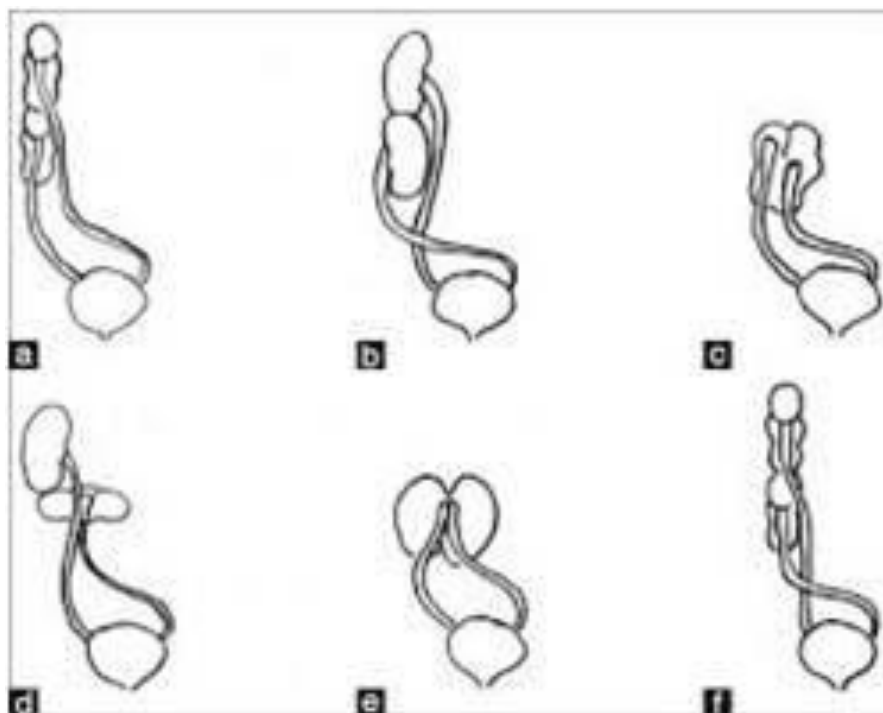


Figure 2: Current classification of crossed renal ectopia. a: inferior ectopia, b: sigmoid kidney, c: lump kidney, d: L-shaped kidney, e: disc kidney, f: superior ectopia

DISCUSSION

Crossed fused renal ectopia is the second most frequent fusion abnormality of the urinary tract, horse-shoe kidney being the most common. Wilmer, in 1938, first categorized the fusion anomalies of the kidney while McDonald and McClellan, in 1957, included crossed ectopia with fusion, crossed ectopia without fusion, solitary crossed ectopia and bilateral crossed ectopia in a modified classification. The current classification includes six types of crossed renal ectopia : a- inferior ectopia, b- sigmoid kidney, c- lump kidney, d- L-shaped kidney, e- disc kidney, f- superior ectopia [Figure 2].^[2-5] Left to right ectopy is about three times more common than vice-versa. In most cases, it is the upper pole of the crossed ectopic kidney that is fused to the lower pole of the normally located kidney.

In this classification, the important feature is that the ureters are present on either side of the urinary bladder. The crossed fused renal ectopia in our case was unusual as there was a single ureter, and this type, to our knowledge, has been documented earlier in only one case report so far by Kaur N et al, 2013.^[6] Their case had symptomatic renal

anomaly which led to its detection. We not only reaffirm such variation but also bring to light that this anomaly with single ureter can remain asymptomatic even till sixth decade of life.

The embryological basis of crossed renal ectopia has not been clearly established. The formation of kidneys depends on the presence of both the ureteric buds and the metanephric blastema. The ureteric bud arises from the lower portion of the Wolffian duct and the metanephric blastema is a mesodermal tissue. Both these tissues migrate towards each other and merge to form the kidney and the urinary tract.

Crossed renal ectopia results as a consequence of abnormal renal ascent in embryogenesis with fusion of the kidneys within the pelvis. It is thought to occur in the first trimester, at around 4th-8th week of fetal life (normally, the kidney reaches L2 level by 2nd month). One school of thoughts opine that an abnormally situated umbilical artery prevents normal cephalic migration. Normal location of the kidneys is required for formation of the extraperitoneal perirenal fascial planes and therefore ectopia (or renal agenesis) results in failure of development of

fascial layers in the flanks on the side not occupied by renal tissue. The lack of restraining fascia causes relaxation of mesenteric supports for bowel loops in this region and thus may lead to malposition of bowel into the extra-peritoneal fat of the empty renal fossa.

Other theory suggests that overbending and rotation of the caudal end of the embryo prevents one of the ureteric buds from merging with the ipsilateral metanephric blastema and it turns toward the more-closer contralateral side. Both the migrated and the normally placed ureteric buds induce the metanephric blastema twice to form two kidneys on one side.^[7,8] However, this fails to explain our case of crossed fused renal ectopia that also had fusion of ureters leading to a single ureter opening into urinary bladder. This case challenges the embryological basis of crossed renal ectopia and gives a reason to reconsider it.

The anomaly is readily detected on conventional urography though fusion of kidneys cannot be commented upon. On ultrasound there may be a characteristic anterior or posterior "notch" between the two fused kidneys. The parenchymal band joining the two kidneys can be better visualized on CT scan. Also, anatomical relationship with adjacent structures and positions of the ureter can be better assessed.

Most cases of renal ectopia remain asymptomatic during life and are diagnosed incidentally.^[9] When symptoms do occur, the most common symptoms reported are abdominal or flank pain, a palpable mass, hematuria and dysuria. In a crossed fused renal ectopic kidney, complications such as nephrolithiasis, infection, and hydronephrosis approaches ~50%. Signs or symptoms of urinary tract infection occur in 30% of the patients. In children, urinary tract infection is the most common presentation. Ureteropelvic junction obstruction causing significant hydronephrosis occurs in one-third of the individuals. Stones are thought to be due to associated hydronephrosis or ureteropelvic junction obstruction, causing stasis of urine. The most frequent anomalies associated with crossed ectopia are imperforate anus (4%),

skeletal abnormalities (4%) and septal cardiovascular defects.^[2]

Crossed fused ectopia usually doesn't require any primary treatment. The fused renal units need not be separated. Complications such as pelviureteric junction obstruction or vesicoureteral reflux may require treatment.

Renal cell carcinoma, transitional cell carcinoma and Wilm's tumor have been reported in crossed fused renal ectopia, which were managed by resection of the involved renal unit.^[10-12] Renal angiography or CT angiography is essential in these cases to provide key information about the renal vasculature as the blood supply to cross-fused kidney is usually anomalous. Therefore, it is essential to be aware of this anomaly before planning any surgical intervention in the renal region.

CONCLUSION

Careful assessment of an incidental finding on imaging also becomes sometimes very important for overall prognosis of the patient.

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