Original Article

CROSSED FUSED RENAL ECTOPIA MULTIDETECTOR COMPUTED TOMOGRAPHY STUDY

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ABSTRACT

Crossed renal ectopia is one of the rarest congenital malformations where a kidney is located on the side opposite to the side of its ureteral insertion into the urinary bladder and is generally fused with the normally located ipsilateral mate. Generally this anomaly remains as a silent clinical entity and is incidentally detected during evaluation for other conditions. We report here three such cases of crossed fused renal ectopia detected by multidetector row contrast enhanced computed tomography. Crossed fused renal ectopia of inferior type was observed in a male on the right side with the ureter of the ectopic left kidney crossing the midline. In two female patients, L-shaped or tandem kidney was noted, one on the right and another on the left side. Over all in two cases the left kidney was ectopic and in one the right kidney. No renal pathologies like urinary tract infection, nephrolithiasis or hematuria were found in our patients.

KEYWORDS: Crossed fused renal ectopia, Tandem kidney, L-shaped kidney, Renal ectopia, Multidetector computed tomography, Renal fusion anomaly.

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INTRODUCTION

Congenital malformations of the urinary system are not uncommon and crossed renal ectopia (CRE) is one of the rare positional and fusion anomalies of the kidney. Crossed renal ectopia occurs when a kidney is located on the side opposite from which its ureter enter into the urinary bladder [1]. In about 90 % of cases, the crossed ectopic kidney fuses with its ipsilateral mate. Crossed fused renal ectopia is the second most common renal fusion anomaly after the horse-shoe kidney with an estimated incidence of 1: 2000 to 1: 7500 autopsies.[1,2]. The prevalence of the crossed renal ectopia with fusion was estimated to be 1 in 1000 live births [3]. In a review of 400 children evaluated by DMSA renal scan, crossed fused renal ectopia was found in 7 cases (1.75 %) [4]. In an another retrospective review, the incidence of CRE was reported as 1 out of 3078 CT scans and horse-shoe kidney in 1 out of 474 scans [5]. The true incidence of this anomaly is not known because a large majority of the patients having this anomaly remain asymptomatic and undetected.

Though CRE remains as a silent entity, in some cases it may be associated with recurrent urinary tract infections, nephrolithiasis, vesicoureteral reflux, uretero-pelvic junction obstruction, hydronephrosis and multicystic renal dysplasia and hence its importance to nephrologists, surgeons and radiologists. Moreover, the condition may also be associated with congenital malformations affecting skeletal, cardiovascular,

genitourinary and gastrointestinal systems [6,7,8,9]. In this case series, we review the radiological features of crossed fused renal ectopia detected by multidetector row computed tomographic (MDCT) examination in three patients. It is recently suggested that MDCT urography is the modality of choice comprehensively evaluating anatomical features of this renal fusion anomaly in a single examination [2].

MATERIALS AND METHODS

Three cases of crossed fused renal ectopia presented here were evaluated in a single diagnostic center during the period from October, 2012 to February, 2014 and the anomaly was detected incidentally when the patients were examined for other suspected conditions. The diagnostic center routinely obtains written informed consent from the patients before contrast injection. All patients underwent contrast enhanced computed tomography (CECT) by a 64 channel scanner (GE Optima-60) and received 85 – 100 ml of non-ionic contrast (Omnipaque, 300 mg I/ml) at the rate of 5 ml/s intravenously. Scans were obtained from diaphragm to upper part of thigh and delayed phase scans were also obtained. The scans were analyzed in a separate work station (AW volume share 4.5) with multiplanar reformatting and maximum intensity projection (MIP) and volume rendered (VR) images obtained.

OBSERVATIONS

Case-1: 58 year old man with empty left renal fossa and presence of two renal masses on the right side. Upper pole of the ectopic left kidney has fused with the lower pole of the orthotopic right kidney- crossed fused renal ectopia inferior type.(Fig.1) Left ureter crosses the midline and inserts into the urinary bladder on the left side. Hilum of the ectopic left kidney is directed anteriorly. The ectopic left kidney is supplied by an artery arising from aorta just above the level of its bifurcation and pass anterior to IVC (Precaval course) to reach the hilum.

Case-2: 28 year old woman with empty right renal fossa. The right kidney is placed transversely anterior to L-4 vertebra and fuses with the lower pole of the orthotopic left kidney resulting in a L-shaped crossed fused renal ectopia (Fig.2). The right ureter crosses the midline at L-5 vertebra. Hilum of the right kidney is directed anteriorly whereas that of left kidney anterolaterally. Slight dilatation of left pelvicalyceal system is seen. The ectopic right kidney receives its arterial supply from a recurrent branch having a curved course arising from the left common iliac artery and crossing anterior to aortic bifurcation. A short right renal vein and a longer left renal vein drain into left common iliac vein.

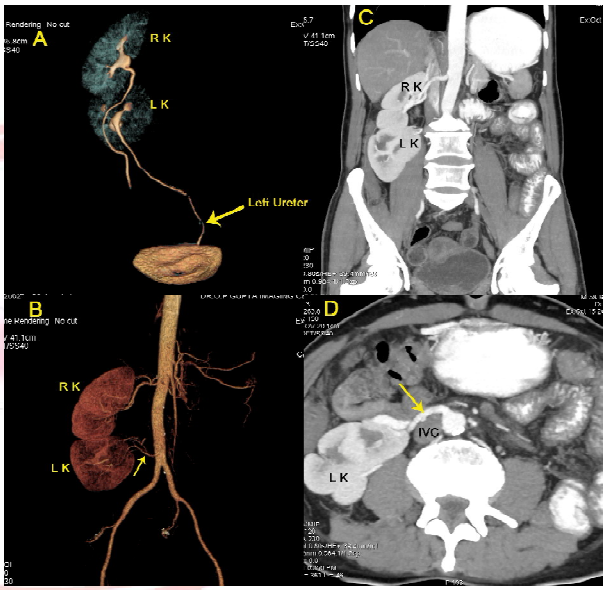

Fig.1: 58 year old male with crossed fused renal ectopia of the left kidney. 1-A:VR urographic image showing the fusion of upper pole of crossed ectopic left kidney (LK) with the lower pole of normally positioned right kidney (RK)- inferior ectopia type. The left ureter is crossing the midline and open into urinary bladder on the left side. 1-B: VR image showing the presence of two right renal arteries from the aorta entering the medially facing hilum of right kidney (1-C- coronal image). A single left renal artery (arrow in 1-B and 1-D) arising from anterior aspect of aorta just proximal to its bifurcation passes to the right and has a precaval course (passing anterior to inferior vena cava (IVC) to reach the hilum (1-D). Hilum of the left kidney faces anteriorly.

Case-3: 70 year old woman with L-shaped crossed fused renal ectopia. The left kidney is ectopic lying transversely anterior to L-4 and L-5 vertebrae and fusing with the lower pole of right kidney (Fig.3). The left ureter crosses the midline to insert into bladder on the left side. The hila of both the kidneys face anteriorly. The ectopic left kidney is supplied by 4 arteries, 2 from the aorta, one from left common iliac and one from the right common iliac artery.

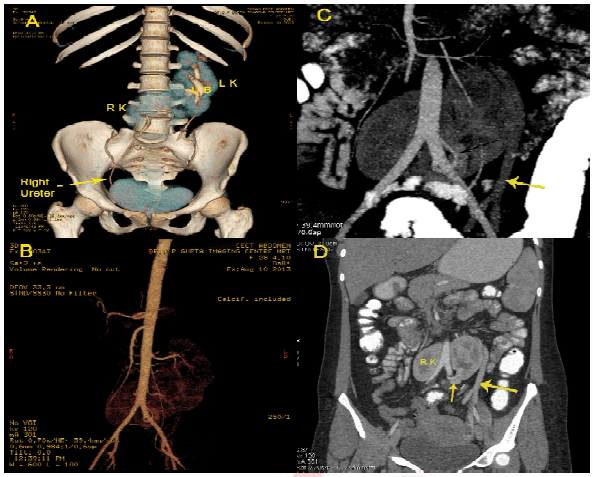

Fig.2: 28 year old female having crossed fused renal ectopia with L-shaped or Tandem kidney. 2-A:VR urographic image show ectopic right kidney, placed horizontally in front of L-4 vertebra, is fusing with the lower pole of the left kidney. The right ureter emerging from the anteriorly placed hilum is crossing the midline in front of L-5 vertebra and opens into urinary bladder on the right side. Mildly dilated left pelvicalyceal system, suggestive of hydronephrosis, is emerging from the anterolaterally directed hilum. 2-B: VR image shows a single left renal artery arise from aorta opposite to inferior mesenteric artery. 2-C:Coronal image. The right renal artery supplying the ectopic right kidney arises from the left common iliac artery and has a recurrent course to reach right kidney Arrow- left renal vein. 2-D: coronal image showing right renal vein (short arrow) and left renal vein (long arrow); both drain into left common iliac vein.

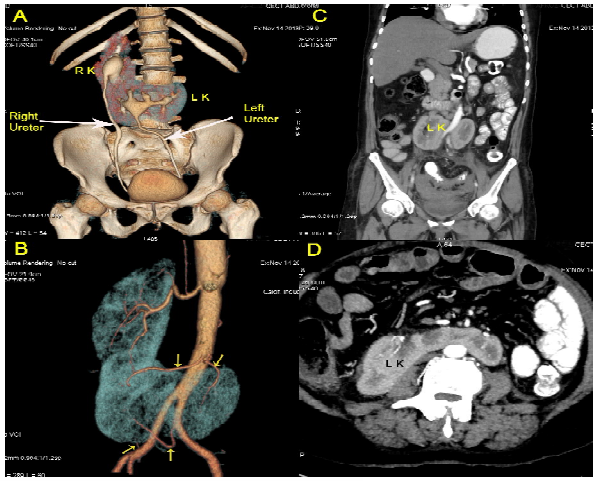

Fig.3: 70 years old female having crossed fused renal ectopia with L-shaped or Tandem kidney. 3-A: VR urographic image show ectopic left kidney (LK) placed horizontally anterior to L4 and L5 vertebrae is fusing with the lower pole of the normally placed right kidney (RK). The left ureter emerges from the anteriorly placed hilum and crosses the midline at sacral promontory to insert into urinary bladder in the left side. The hilum of the right kidney also faces anteriorly. 3-B: VR image show a single right renal artery, arising opposite to the level of superior mesenteric artery, supply the right kidney. The ectopic left kidney is supplied by 4 arteries (arrows); two of them arise from the anterior aspect of aorta just above its bifurcation. The third artery arises from the medial aspect of left common iliac and crosses the right common iliac artery to reach the left kidney. A fourth artery which is very short arise from the right common iliac artery to supply the left kidney. 3-C: Coronal image; 3-D: Axial image.

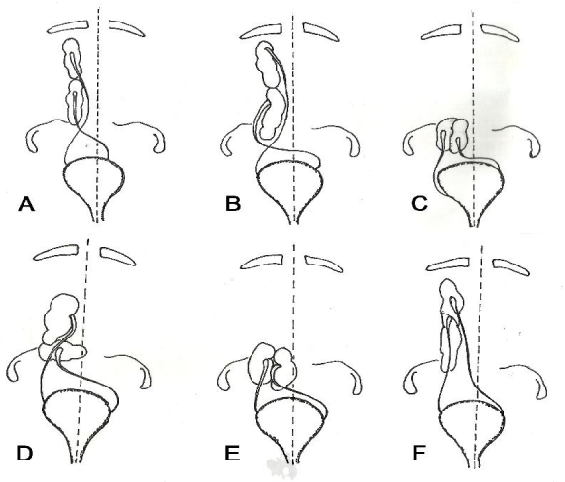

Fig.4: Six types of crossed fused renal ectopia. A- Unilateral fused kidney- inferior ectopia type; B- Sigmoid or S-shaped kidney; C- Lump kidney; D- L-shaped kidney; E- Disc kidney; F- Unilateral fused kidney- superior ectopia type.

DISCUSSION

Crossed renal ectopia (CRE) is a rare type of renal fusion anomaly in which both the kidneys are situated on one side and in about 90 % of such cases the crossed ectopic kidney is fused with the orthotopically located kidney. In this condition the ectopic kidney is located contralateral to the side of its ureteric orifice and the ureter of the ectopic kidney cross the midline which distinguishes this condition from horseshoe kidney. McDonald and McClellan [10] classified the crossed renal ectopia into (i) crossed ectopia with fusion (90% cases); (ii) Crossed ectopia without fusion' (iii) unilateral crossed ectopia (associated with unilateral renal agenesis) and (iv) bilateral crossed ectopia without fusion(both ureters cross the midline). In CRE, the left to right ectopia is more common (the left kidney crossing to the right side) and males are more commonly affected.

Crossed fused renal ectopia is further classified into 6 types (Fig.4). In decreasing order of frequency they are: (A) Unilateral fused kidney inferior ectopia with the upper pole of the crossed ectopic kidney fusing with the lower pole of the orthotopic ipsilateral mate. Both renal pelvis may be anterior. (B) Sigmoid or S-shaped

kidney in which the crossed kidney lies inferiorly with the renal pelvis directed laterally and the normally positioned kidney lies superiorly with the pelvis directed medially. (C) Unilateral Lump kidney with fusion occurring over a wide margin and both renal pelvis directed anteriorly; located more inferiorly. (D)L-Shaped or Tandem kidney in which the crossed kidney lies inferiorly and transversely fusing with the lower pole of the normal kidney. (E) Unilateral disc kidney in which the fusion occurs along the medial borders and (F) Unilateral fused kidney superior ectopia type is the least common type; the ectopic kidney is placed superiorly with its lower pole fusing with the upper pole of the normal kidney. Both renal pelvis are anterior.

The precise mechanism of occurrence of crossed renal ectopia is not fully understood and several theories have been put forward to explain this anomaly. Among them are the mechanical theory (abnormally placed umbilical arteries mechanically obstructing cephalad migration), the ureteral theory (wandering of the ureteral bud to the opposite side), the teratogenic theory, the genetic theory (observation of the anomaly in families) and theory of abnormal rotation of the caudal end of the fetus (increased prevalence of this anomaly with scoliosis).

CRE is sporadically reported in the literature because this anomaly may remain as a silent clinical entity without producing any signs and symptoms and this is supported by several case reports in cadavers. [11–14]. In these four case reports, male to female ratio and left to right ratio is 3:1. Case studies of patients investigated for nephrolithiasis and pyelonephritis with inferior type of crossed fused renal ectopia of the left kidney, have been reported [15-17]. Inferior ectopia type of CRE is the most common type. Only in one case inferior type of crossed fused renal ectopia of the right kidney was detected [18]. Sigmoid type of kidney, which is second common type of CRE with fusion, associated with staghorn calculus was reported by Amin et al. [19]. Superior ectopia, the rarest type of CRE with fusion, was reported in a female patient by Patel and Singh [20]. In our study L-shaped kidneys were found in two female patients and inferior type of CRE in one male patient.

A number of case series of CRE with fusion have been published. [21-24]. Analysis of these studies indicate that the CRE with fusion occurs more commonly in males and the left kidney is affected more than the right kidney. Many congenital anomalies are associated with CRE with fusion such as vaginal agenesis [6], VACTERL association [8], TAR syndrome [9], renal dyaplasia [25] and a single ureter [26]. Kulkarni etal detected intestinal malrotation associated with a lump kidney in a male cadaver. [7]. Crossed fused left renal ectopia with left sided polydactyly was found in a 24 week aborted male fetus [27]. We did not find any congenital anomaly in our cases.

CONCLUSION

Crossed fused renal ectopia is an uncommon congenital anomaly which can remain asymptomatic throughout life and hence undetected. It is generally found incidentally when patients are investigated for other abdominal pathologies. In some cases it may be associated with nephrolithiasis, recurrent infections , hydronephrosis and congenital malformations affecting skeletal, gastrointestinal and urogenital systems. We have reported two cases of a rare type of L-shaped or tandem kidneys, both found in females, though CRE is more common in males. Multidetector computed tomographic (MDCT) evaluation provides excellent anatomical details of this anomaly in a single examination important for surgeons, nephrologists and radiologists for proper management.

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