CROSS-FUSED RENAL ECTOPIA- A CASE REPORT

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PRESENTATION OF CASE

A case of cross-fused renal ectopia was found during a routine cadaveric dissection at Department of Anatomy, Government T.D. Medical College, Vandanam, Alappuzha. The right kidney crossed the midline and fused with the lower pole of left kidney. Cross-fused renal ectopia seems to be one of the rarest of the fusion anomalies of kidney. Though the symptoms particular to cross-fused ectopia are rare, patients generally present in adulthood with symptoms of various urinary tract infection or calculi, which is believed to be secondary to stasis of urine. The condition is readily detected on conventional urography, CT and US are very useful.

DIFFERENTIAL DIAGNOSIS

Congenital anomalies of the kidneys include a group of so-called fusion anomalies in which both kidneys are fused together in early embryonic life either partially or completely. Partial fusion anomalies of the kidney can generally be placed into two categories- 1) Horse-shoe kidney and its variants and 2) Cross-renal ectopia. Complete fusion represented by 'cake' kidney or fused pelvic kidney. These renal fusion anomalies exhibit abnormalities of position (ectopia), migration, rotation and vascular supply.

Cross-Fused Renal Ectopia (CFRE) is the second most common renal fusion anomaly next to horseshoe kidney where the ectopic kidney crosses the midline and fused with the orthotropically located kidney. It is both a fusion and ectopic anomaly and its prevalence was estimated to be 1 in 1000 livebirths.[1] Left to right ectopy is more common and the condition is more in males. The present report is a case of right to left ectopy, which is rare.

CLINICAL DIAGNOSIS

During the routine cadaveric dissection of abdomen for the medical students in the Department of Anatomy, Government TD Medical College, Alappuzha, we observed that the right kidney was absent in the right lumbar region of an old male cadaver. On further dissection, on the left side of the posterior abdominal wall, a large mass was observed. Careful observation of the specimen revealed that it was the right kidney crossing the midline and fusing with the lower pole of left kidney with aberrant renal vessels. It was a rare case of cross-fused renal ectopia. The fused renal mass has the following dimensions.

Length (from upper pole of normal left kidney to lower pole of fused right kidney - 15 cms.

Breadth (in the upper part 1.5 cm below the upper pole of left kidney) - 7 cms.

Breadth (in the middle between the two hila) - 4.5 cms.

Breadth (in the lower part below the hilum of fused kidney) - 7 cms.

Thickness (of left kidney) - 4 cm.

Thickness ((of fused right kidney) - 3 cm.

The upper border of the fused mass was at the level of T12 and lower border was at the level of L5. The fused mass was lying in front of left psoas major, left quadratus lumborum and bifurcation of aorta. The left kidney was in normal vertical position with all its normal relations showing normal ascent. It was the right kidney, which crossed the midline and fused with the lower pole of left kidney. The case represented inferior type of cross-fused renal ectopia. The posterior surface of the fused mass was smooth and continuous. The anterior surface was rough and presented a groove separating the left and right kidney through, which passed the inferior mesenteric artery, which prevented the further ascent. When compared with the left, right kidney was small and it was the medial aspect of upper pole, which fused with the lower pole of left kidney.

The fused renal mass presented two hilum on the anterior surface, upper one representing the hilum of left kidney and lower one representing that of right. In both hilar, the major calyces were the anterior most structure. The calyces unite to form renal pelvis. The right ureter emerged through the medial border of the fused right kidney, passed below its lower pole, crossed the midline and entered the lesser pelvis by passing medial to right external iliac vessels. It opened on the posterior surface of urinary bladder on its normal site. The left ureter passed through the anterior surface of left kidney over the left psoas major, the lateral pelvic wall and opened on the urinary bladder at its normal site.

Vasculature of fused renal mass also showed marked variations. The left renal artery arose from the abdominal aorta and reached the hilum of the left kidney behind the left renal vein. Below the origin of inferior mesenteric artery, another artery was seen originating from the left side of aorta and opened into the right kidney on its posterior surface. In addition to this, a very small aberrant renal artery arose from the abdominal aorta and opened into the upper part of fused right kidney. The inferior mesenteric artery was seen originating from the abdominal aorta close to its bifurcation and passes anterior to the fused kidney grooving

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its anterior surface and then continue into the pelvis. The origin of inferior mesenteric artery prevented the further ascent of fused kidney. The left renal vein was seen passing between the major calyces and opened into IVC. The right renal vein emerged through the hilum between major calyces joined the left renal vein by a recurrent course deep to inferior mesenteric artery. Before joining IVC, the common renal vein received the left suprarenal vein.

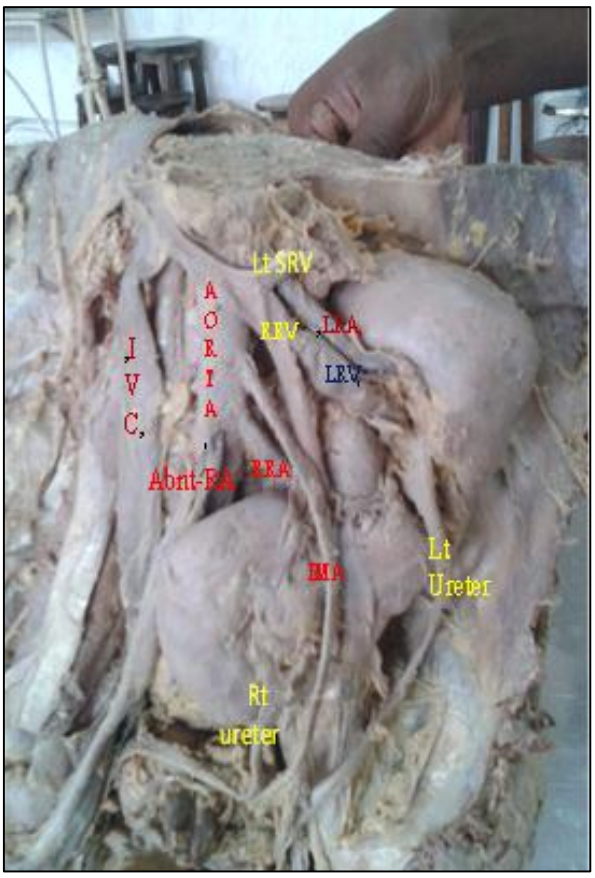

Figure 1. IVC - Inferior Vena Cava, IMA - Inferior Mesenteric Artery, Lt. SRV - Left Suprarenal Vein, LRA - Left Renal Artery, LRV - Left Renal Vein, RRV - Right Renal Vein, RRA - Right Renal Artery, Abnt. RA - Aberrant Renal Artery

PATHOLOGICAL DISCUSSION

Crossed fused ectopia is the second commonest congenital fusion anomaly of kidney, the first being horseshoe kidney.[2] In this abnormality, both kidneys are located on the same side with two separate ureters arising from the respective kidneys. The ureters arising from the crossed over kidneys travels back to opposite side and inserts in the normal site of urinary bladder. The blood supply maybe atypical.

Crossed renal ectopia was first described by Pamarolus in 1654 as quoted by[3] refers to a condition where one kidney has crossed to the opposite side of the body. Renal fusion anomalies were first studied and classified by Wilmer (1938). Later, it was revised by McDonald and McClellan into 4 groups.[4] 1) CRE with fusion (CFRE 90%), 2) CRE without fusion (uncommon), 3) Solitary CRE (very rare) and 4) Unfused bilateral CRE (extremely rare).

Subsequent studies led to the establishment of a CFRE classification into 6 types.[1,5] They are-

1. Inferior ectopia- commonest type. The crossing kidney lies inferiorly. The upper pole of the crossed kidney is fused to the lower pole of the resident kidney. Both renal pelvis are directed anteriorly.
2. Sigmoid or S-shaped kidney- The crossed kidney lies inferiorly with its pelvis directed laterally. The pelvis of the superiorly located resident kidney is directed medially.
3. Unilateral lump kidney- The two kidneys are completely fused to form an irregular lump. Both renal pelvis are directed anteriorly.
4. Unilateral disc kidney- The kidneys are fused along their medial border. The renal pelvis of the normal kidney is directed anteromedially. The crossing renal pelvis is directed laterally.
5. Unilateral L-shaped kidney- The crossing kidney lies inferiorly and transversely.
6. Superior ectopia- The kidney crosses the midline and lies superior to the resident kidney. Both renal pelvis are anteriorly rotated.

Based on the above classification, the present case is an example for CFRE of inferior type. According to Asghar and Wazir, 2004, there is a male predominance and the left to right crossover occurs more frequently with the inferior ectopia as a common variety.[6] But, here it is right to left crossover, which is rare.[7] In general population, CFRE occurs more frequent in men than women from 3:2[6] to 2.3:1.[8]

EMBRYOLOGICAL BASIS

The embryological basis of CFRE is still a controversy. CFRE may be due to aberrant metanephros development in which the normal displacement of foetal kidneys from the pelvis to the lumbar fossae is altered or inhibited.

The formation of kidneys depend on the presence of both the ureteric bud and the metanephric blastema. The ureteric bud arises from the lower portion of the Wolffian duct and the metanephric blastema is a mesoderm tissue. Development of kidney begins from 4th week of gestation by the interaction between the ureteric bud and nephrogenic cord at the level of S2 vertebra in the pelvis.[9] The ureteric bud enters the metanephric blastema to induce the changes in the developing kidney from 4th week to 8th week of IU life. Further during 6th to 9th week[8] of development, the foetal kidney ascends along the posterior abdominal wall from pelvis to its normal position in lumbar region close to each other with hila directed anteriorly[10] to reach the definitive position in the lumbar region involves a well-orchestrated series of movements including ascent, lateral migration, axial deflection and medial rotation (almost 90 degrees).[5,8,11] According to Khan et al,[11] the ascend of the kidney is caused by the diminution of body curvature and by growth of the body in lumbar and sacral regions in the 9th week of development. Kidney attains adult position adjacent to adrenal gland.[12]

The upward migration of ascend of kidney could be arrested by several causes. It is postulated that if compression factor of the umbilical arteries persist at the beginning of the cranial migration in the presence of two unequal metanephric masses the result will be crossed ectopia.[13] The ascend of the fused kidney are limited to the origin of the inferior mesenteric artery from the abdominal aorta. Ashley and Mostofi[9] suggested that undue persistence of caecum in right lumbar region before reaching its final position in right iliac fossa may favour the migration of the right renal unit to the left. Above is considered as the mechanical theory behind CFRE. In present case, the inferior mesenteric artery is seen grooving the anterior surface of the fused mass forming an anterior fork with abdominal aorta preventing further ascend.

According to theory of abnormal caudal rotation, the over bending and rotation of the caudal end of the embryo prevents the ureteric bud from merging with the ipsilateral metanephric blastema and it turns towards the more closer contralateral side. The migrated and the normally placed ureteric bud induces the metanephric blastema twice to form two kidneys on the same side. This theory is supported by the fact that there is increased prevalence of this anomaly with scoliosis. But, in the present case, this theory cannot be followed as it was a normal cadaver without any abnormality.

According to ureteral theory, the crossover is strictly a ureteral phenomenon with the developing ureteral bud wandering to the opposite side and inducing the differentiation of the contralateral metanephric blastema. According to Rinat et al, this process may result in the inability of the ureteric bud to communicate with more distant ipsilateral metanephric blastema. The nephrogenic tissue from the side that does not receive a ureteric bud completely regresses.[14]

In addition to this, teratogenic theory and genetic theory (observation of anomalies in families) is also postulated to explain the CFRE phenomenon. It is suggested that the sonic hedgehog gene signal is critical for kidney positioning along the media lateral axis and its disruption will result in renal fusion. Intense research is going on to unravel the genetic mechanisms underlying the development of congenital anomalies of kidney and urinary tract. In this case report, the ectopic right kidney crosses the midline and fuses inferiorly with the orthotropic left kidney with aberrant renal vessels. The lower part of ureters and trig one of the bladder all are normal, which indicates the normal caudal part of mesonephric ducts.

During its normal migration, the metanephros acquires its vascular supply form pelvic branches of the iliac arteries and from branches of the dorsal aorta.[15] In the developing kidney, the vascular supply is re-established progressively as it migrates superiorly. If migration is arrested, the temporary blood supply will become permanent.[5] In the CFRE, the superior kidney is usually supplied by one renal artery, branch of abdominal aorta while the remaining kidney gain its blood supply from the middle group of the mesonephric arteries. Turkvatan et al[16] demonstrated that a total number of 1-6 arteries supplies the two kidneys that form CFRE in which 2-4 are major arteries. In the present case, the superior kidney, which is found in normal position is supplied by a single renal artery originating from abdominal aorta at the level of L1 vertebra. The inferior kidney is supplied by two renal arteries, one main renal arose from the left side of aorta below the origin of inferior mesenteric artery. The additional artery entering the upper posterior part of fused inferior kidney arising from the abdominal aorta maybe persisting mesonephric artery. The development of renal veins is a part of complex developmental process of IVC. In most cases, the venous drainage of crossed ectopic kidney is into distal part of IVC or into common iliac veins, but in present case, the common renal vein opened to the proximal part.

CONCLUSION

Crossed-fused renal ectopia is a rare congenital anomaly, which can remain asymptomatic throughout life and hence undetected. Most of the time, crossed renal ectopias are diagnosed at autopsy or incidentally during radiological investigation or during cadaveric dissection. They are of immense clinical importance because of their concomitant congenital anomalies involving other organ systems. The most frequent anomalies with crossed ectopia are imperforate anus, skeletal abnormalities and septal cardiovascular defects. However, the present case has no such anomalies. Though asymptomatic, these patients are prone to chronic and recurrent urinary tract infection, urolithiasis, hydronephrosis, hypertension and present asymptomatic abdominal mass. The anomalous vascular pattern could provide a formidable challenge during aortic reconstructive surgery for post-renal aortic and aortofemoral aneurysms. Various imaging modalities used to investigate renal fusion anomalies include sonography, intravenous pyelography, computed tomography, renal scintigraphy, MDCT angiography and MRI.

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