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SUPERIOR CROSSED RENAL ECTOPIA- THE RAREST OF THE RARE

Radiology

Dr. Vaidehi A Patel Associate Prof. in Department of Radio Diagnosis and Imaging, G. R. Doshi and K.M. Mehta Institute of Kidney Diseases & Research Centre (IKDRC)- Dr. H. L. Trivedi Institute Of Transplantation Sciences (ITS), Civil Hospital Campus, Asarwa, Ahmedabad - 380016, Gujarat, India.

Harsh C Sutariya* Associate Professor. Department of Radio Diagnosis and Imaging. G.R. Doshi and K.M. Mehta Institute of Kidney Diseases & Research Centre (IKDRC) & Dr.H.L. Trivedi Institute of Transplantation Sciences (ITS). *Corresponding Author

ABSTRACT

Various renal anomalies pertaining to number, volume, structure, form, rotation, vascular attachments and fusion are seen. Fusion anomalies are uncommonly seen. Fusion anomalies show two types; partial fusion and complete fusion. Crossed renal ectopia is second most common fusion anomaly and is further divided in six sub types out of which superior crossed renal ectopia is the rarest variant. Only few cases have been reported. Here we report a case of 8 years old male with right superior crossed renal ectopia with gross hydrouretero-nephrosis.

KEYWORDS

Fusion anomaly, Crossed renal ectopia, superior

INTRODUCTION

About 30-40% developmental anomalies are seen in genito-urinary system [1]. Wide range of anomalies varying from complete absence to ectopic location, mal-rotation, altered shape, fusion and altered vascular attachments are included in the wide range of anomalies. Common fusion anomalies are Horse-shoe kidney, crossed renal ectopia and pan cake kidney. Crossed renal ectopia is further sub divided in six groups; they are unilateral fused kidney with inferior ectopia, sigmoid or S- shaped kidney, lump kidney, L shaped kidney, disc or doughnut shaped kidney and Unilateral fused kidney with superior ectopia in order according to their incidence. Crossed renal ectopia (CRE) is the second common fusion anomaly with prevalence estimated to be 1 in 1000 live births [2].

Case Report

About 8 years old male patient presented with right flank pain and difficulty in micturation. Clinical examination was un-remarkable except right flank pain. Lab investigations showed increased total leukocyte count and serum creatinine. On primary ultrasound examination the left kidney was not seen in left renal fossa and a large kidney mass was seen in right renal fossa which showed duplex PC system and double ureters. The upper moiety ureter crossed the mid line to drain in to bladder via left vesico-ureteric junction while the lower moiety ureter drains in to bladder via right vesico-ureteric junction. Both the moiety showed presence of gross hydronephrosis and hydroureter up to lower end. The patient was further investigated by CT IVP [Figure 1]. CT IVP showed inferior pole of left kidney fused with superior pole of the normally positioned right kidney. Both the PC systems from duplex PC system showed gross hydronephrosis and hydroureter up to lower end. The upper moiety ureter crossed mid line and drained in to bladder via left Vesico-ureteric junction. So the diagnosis of left superior crossed renal ectopia was made.

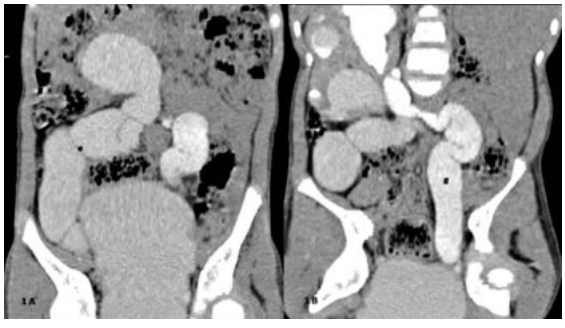

Figure 1: (A) Delayed phase of CT IVP showing lower moiety ureter(*) opening in to Bladder via right VUJ (B) delayed phase of CT IVP showing gross hydronephrosis in both moiety and upper moiety ureter crossing mid line and opening in to bladder via left VUJ

DISCUSSION

The estimated incidence of crossed fused renal ectopia is approximately 1:13001:7500[3]. It is fusion as well as ectopic anomaly occurring in about 0.08% - 0.01% cases. The crossed ectopic kidney crosses the mid line and fuses with its ipsilateral mate in about 90 % cases. Its prevalence is estimated to be 1 in 1000 live births [2]. Halaseh M reported only 7 cases (1.75%) of CRE in a review of 400 children evaluated by DMSA renal scan [4]. In a retrospective study done by Glodny B et al, they reported the incidence of CRE as 1 out of 3078 CT scans [5]. Although the true incidence of this anomaly is not known as a large number of asymptomatic patients with CRE go undetected.The male-to-female incidence ratio estimated is 2:1 and it is three times more common on the left side [6,7].

The precise mechanism of CRE is not fully understood; various theories are put forward. Mechanical theory, ureteral theory, theory of abnormal rotation are explained. Moreover, the teratogenic theory and the genetic theory are also explained.

In every case of CRE, the ureter from normal kidney follows normal course and enters bladder via normal opening, whereas that of ectopic kidney crosses the midline and enters the bladder via its normal opening on the contralateral side.

Various types of CRE are identified according to the type of fusion. Six variations of CRE have been described: inferior crossed fused ectopia; sigmoid or S-shaped kidney; unilateral lump kidney; L-shaped or tandem kidney; unilateral disc kidney; superior crossed fused ectopia [8]

Figure 2

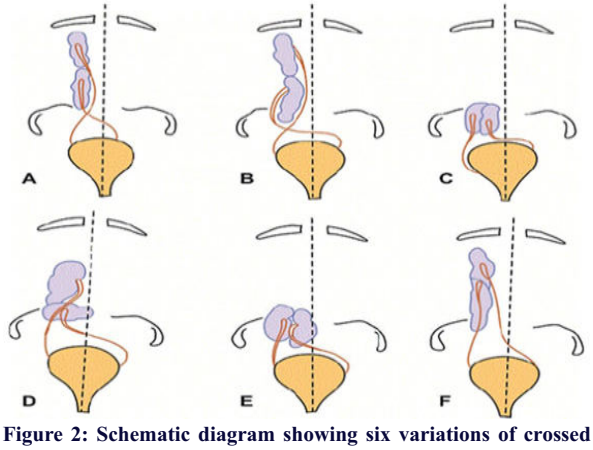

Figure 2: Schematic diagram showing six variations of crossed renal ectopia

***Inferior crossed fused ectopia*:**
The upper pole of the crossed kidney is attached to the inferior pole of the normal kidney. Both the renal pelvis may face anteriorly.

Sigmoid or S-shaped kidney:
The crossed kidney lies inferiorly with its pelvis facing laterally and the normal kidney lies superiorly with its pelvis facing medially thus each pelvis is oriented correctly.

Unilateral Lump kidney:
Here, the fusion occurs in a wide renal region hence the total kidney mass is irregular and lobulated and the ascent is prevented beyond the sacral promontory. Both renal pelvis draining separate areas of renal parenchyma face anteriorly. The ureters cross the midline.

***L-shaped or tandem kidney*:**
Crossed kidney lies in a transverse and inferior plane fusing with the lower pole of normal kidney and assuming the shape of "L" in front of L4 vertebra to lie in the midline or contra lateral para median space. The ureters enter the bladder via their normal openings.

Unilateral disc kidney:
In this type, kidneys are fused at medial aspect along their entire length. The pelvis faces anteriorly and ureters coarse along their normal pathway to drain in to bladder via their normal openings. Each half of the kidney has its own collecting system drains and does not communicate with the opposite half.

Superior crossed fused kidney:
it is least common variant. Here the lower pole of ectopic kidney fuses with the upper pole of normally positioned kidney and the ureter crosses the midline to enter in bladder via its normal opening.

The blood supply of normal as well as ectopic kidney is frequently anomalous. Blood supply to the ectopic kidneyarises from the ipsilateral vessels but occasionally arise from contralateral side. The renal artery arises from upper abdominal aorta in 25% cases and from lower part of aorta or iliac arteries in 75 % cases. Total 1 to 6 renal arteries may be seen in two kidneys while 2 to 4 major arteries are present at various levels [8, 9].

Frequently associated anomalies are: Imperforate anus (4%), orthopedic anomalies- mostly involving bony pelvis and vertebrae (4%), septal cardiovascular anomalies, gastrointestinal and other genitourinary abnormality. Abnormality is commonly seen in the ectopic kidney which includes PUJ Obstruction, cystic dysplasia and carcinoma [2,10]. Vaginal agenesis, VACTERL association, TAR syndrome, renaldysplasia and single ureter are also proposed to be associated to crossed renal ectopia.

Usually the patients are asymptomatic, but when present, the symptoms are abdominal or flank pain, a palpable abdominal mass, hematuria, dysuria and other symptoms due to urinary tract infection. The abnormal position of kidney and aberrant vascular structures lead to poor drainage and stasis which can predispose to hydronephrosis, calculus formation and infection. Other urological complications seen with CRE are Pelvi ureteric junction obstruction, reflux, and ectopic ureteroceles [8]. Sporadic cases of mass lesions in CRE have been reported [11].

CONCLUSION
The diagnosis of fused kidney requires long-term follow-up, early diagnosis of complications and to look for associated congenital anomalies. MDCT urography provides better evaluation of such patients in a single examination. Three-dimensional reformatted images can provide particularly good delineation of congenital fusion anomalies of the kidney along with its vascular supply information which is very important to the surgeon.

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Conflicting Interests
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