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Crossed Fused Renal Ectopia: Varied Presentation, Diagnostic Challenges and Management

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Abstract: ***Aim: Crossed fused renal ectopia (CFRE) is a rare congenital anomaly, majority being asymptomatic but varied presentations have been reported. This study was conducted to evaluate the clinical profile and the challenges posed in the management of this entity. Materials and Methods: This was a descriptive study analysing the varied presentation, challenges in the diagnosis and management of ten patients diagnosed with crossed renal ectopia with fusion during the period from January 2019 to June 2021. Patients were diagnosed as a case of CFRE during investigations for abdominal pain, abdominal mass, genitourinary malformation and urinary tract infection. Results: The left moiety was crossed and fused with the right moiety in 5 cases and opposite in rest. Ultrasonography was found to be a good screening investigation with useful diagnostic contributions from CT scan, radionuclide scintigraphy and magnetic resonance urography. Out of ten, total six patients required intervention, rest were managed conservatively. In three patients, there was hydronephrosis secondary to ureteropelvic junction obstruction and underwent pyeloplasty. In one of the patients there was bilateral undescended testis and underwent bilateral orchidopexy. Two patients had calculus in ectopic kidney for which one had undergone lap pyelolithotomy and other patient underwent percutaneous nephrolithotomy (PCNL). All patients were asymptomatic at last follow-up with stable renal functions. Conclusions: Crossed fused renal ectopia was detected in most patients as an incidental finding. Associated urological problems were found and appropriate surgical management were done, in minimal invasive way.***

Keywords: Congenital anomalies, crossed fused renal ectopia, fusion anomalies

1. Introduction

Crossed fused renal ectopia (CFRE) is a rare congenital malformation, which is reported to be usually asymptomatic but may have varied presentations. When location of kidney is opposite to its ureteral insertion in to the bladder is defined as crossed renal ectopia and if it is fused with the kidney on the opposite side then it is defined as crossed fused renal ectopia. Crossed ectopic kidneys are fused to their contralateral kidney in ninety percent of the cases. It is the second most common renal fusion anomaly following horseshoe kidney (Kwon et al., 2004), and in different autopsy series it is detected in 1 in 1000 to 1 in 7000 (Hwang et al., 2002, Yano et al, 2003, Guarino et al., 2004). Most patients with real ectopia are asymptomatic during life and the number found clinically is estimated to be only 1 in 10000 patients (Guarino et al., 2004). The management of the condition includes diagnosing the type of fusion anomaly with associated urinary tract anomalies by choosing an appropriate imaging technique. CFRE develops during the fourth to eighth weeks of gestation. The condition results due to aberrant migration and crossing of the midline of the metanephric blastema and the ureteral bud. Most remain aymptomatic and detected as an incidental finding during imaging studies. Many anatomical variations of CFRE have been reported. (1, 2)

2. Materials and Methods

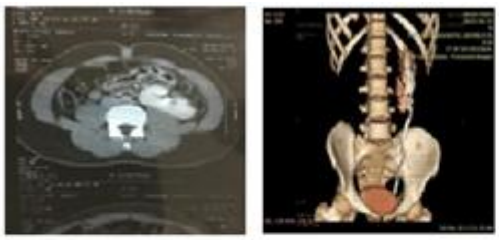
This is a descriptive study conducted at a tertiary care centre based on the retrospective analysis of ten patients diagnosed with crossed fused renal ectopia during the period January 2019to June2021. Of the total cases of CFRE (n= 10), six patients were symptomatic requiring intervention, and four had minor symptoms. Elaborate history was taken and thorough clinical examination was performed. All the patients were investigated for renal function test with electrolytes, complete blood picture, urine microscopy, urine culture and sensitivity, Ultrasound (USG) of the abdomen and pelvis was done for all the patients. Radiological investigations comprised of contrast enhanced Computed Tomography (CECT), Magnetic Resonance imaging (MRI), non contrast CT (NCCT), retrograde pyelogram (RGP), micturating cystourethrogram/MCU) and intravenous urogram (IVU). According to the type of the fusion anomaly and associated abnormality, further imaging techniques were done.

3. Results

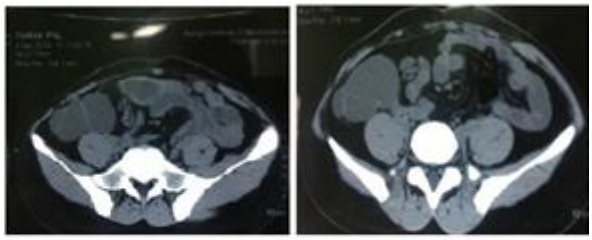
Enumeration of cases with demographic data, clinical presentation, associated anomalies and management in various cases of CFRE is depicted in Table 1. The study population consisted of seven males and three female patients. Varied clinical presentations were seen among patients aged from 10 to 45 years with mostly presenting in first and second decade of life. The study population had equal distribution of right and left CFRE. All were of inferior CFRE variety. Six patients required surgical intervention and rest were managed conservatively with symptomatic treatment and appropriate antibiotics. All patients were followed up in outpatient department. Those who underwent surgical intervention were followed closely with physical examinations and imaging at 3month interval for 1 year and 6 month interval thereafter. Patents that were conservatively managed were followed at 6 monthly interval or as and when patient presented with any complaint. All patients fared better in follow up with no complications.

Table 1: Summary of cases in the study

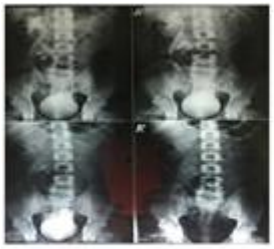
Case	Age/ Sex	Clinical profile	Lab Investigations	Imaging	Crossed renal ectopia	Associated anomaly	Management
1.	20/F	Left flank pain for 6 months, intermittent, mild to moderate, non radiating, not associated with any diurnal or postural variation. No significant past history	Creat-0.9 mg/dl Urine Culture-No growth Blood parameters normal	USG CECT	Right to left	Left moiety hydronephrosis with left PUJO	Left upper moeity laproscopic pyeloplasty
2.	13/M	Left flank pain and lower abdominal pain for 2 years, pain was intermittent, mild to moderate, non radiating. No h/o recurrent UTI. No significant past history	Creat-0.6 mg/dl Urine Culture-No growth Blood parameters normal	USG CECT	Right to left	Left hydro-nephrosis. Lower moiety malrotated with pelvis facing anteriorly, single ureter with normal insertion in urinary bladder with calculus **12x6** mm in lower moiety pelvis	Left laproscopic lower moiety pyelo-lithotomy
3.	45/M	Right flank pain for 6 months, intermittent, mild to moderate, non radiating, not associated with any diurnal or postural variation. No significant past history	Creat-2.02 mg/dl Urine Culture-No growth Blood parameters normal	USG NCCT KUB DTPA	Left to right	Right moiety hydronephrosis	Right moiety open pyeloplasty
4.	10/M	Bilateral undescended testis since birth. No history of recurrent urinary tract infections or haematuria. No history of any renal surgery or chronic renal disease in the past	Creat-0.5mg/dl Urine Culture-No growth Blood parameters normal	USG CECT RGP	Left to right	Bilateral undescended testes.	Bilateral orchidopexy
5.	25/M	Left flank pain for 6 months and gross intermittent painless haematuria for 4 months. No history of fever, dysuria or trauma. No history of previous renal surgery or chronic renal disease in the past	Creat-0.9 mg/dl Urine Culture-No growth Blood parameters normal	USG CECT	Left to right	Nephrolithiasis 15 x 1cm calculus in left moiety pelvis with hydronephrosis	Left Moiety PCNL
6.	26/M	Right flank pain for 6 months, intermittent, mild to moderate, non radiating, not associated with any diurnal or postural variation. No significant past history	Creat-1.2 mg/dl Urine Culture-No growth	USG CECT	Left to right	Right sided hydronephrosis	Right upper moeity laproscopic pyeloplasty
7.	34/F	Burning micturition with supra pubic pain off and on No history of fever	Creat-0.9 mg/dl Urine Culture-E. Coli+	USG CECT	Right to left	Empty right renal fossa on USG	Antibiotic course for 7 days
8.	12/M	Dysuria off and on. No history of fever	Creat-0.9 mg/dl Urine Culture-No growth	USG CECT	Right to left	Empty right renal fossa on USG	Conservative
9.	23/M	Recurrent urinary tract infections	Creat-1.0 mg/dl Urine Culture-No growth	USG CECT MCU RGP	Left to right	Empty left renal fossa on USG	Conservative
10.	21/F	Incidental finding	Creat-0.87 mg/dl Urine Culture-No growth	USG CECT	Right to left	Empty right renal fossa on USG	Conservative



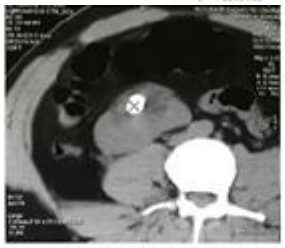
A. Crossed fused ectopic kidney with empty right renal fossa Lower moiety is malrotated with pelvis facing anteriorly. Single ureter is noted with normal insertion in urinary bladder



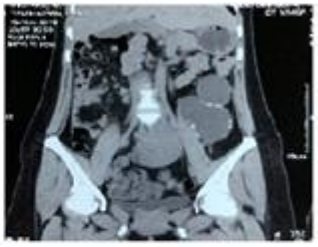
B At its renal pole right kidney is fused with upper pole of malrotated ectopic kidney. Left renal fossa is empty suggestive of crossed fused left ectopic kidney. Right ureter is grossly dilated and measures 1.9 cm near right pelvis. And ectopic ureter from malrotated kidney also dilated and measured 2 cm near pelvis. Both ureter are seen to fuse at the level of L2 vertebral body. the fused ureter is grossly dilated and traverses downwards in a tortuous course with abrupt narrowing noted about 4.5 cm proximal to right Vesicoureteric



C IVU suggestive of ectopic left kidney



D Crossed fused ectopia of left kidney with orthotopic insertion of left ureterwith left sided obstructive nephrolithiasis



E, Axial view of CT showing emptyright renal fossa with right crossed fused ectopia

4. Discussion

Crossed fused renal ectopia occurs during fourth to eight weeks of gestation. Itis thought to result from the abnormal development of the ureteric bud and metanephric blastema. After horseshoe kidney, crossed fused ectopia of the kidneys is the most frequent fusion abnormality of the urinary tract with male predominance of 3: 2. It occurs due to abnormal renal ascent during embryogenesis with the fusion of the kidneys within the pelvis.

One school of thought says that an abnormally situated umbilical artery prevents normal cephalic migration. Another studies believe that the ureteric bud crosses to the opposite side and induces nephron formation in the contralateral metanephric blastema which results in single renal mass with two collecting systems being located on one side of the abdomen. For the formation of the extraperitoneal perirenal fascial planes, the normal ascent of the kidneys is required. There is failure of the development of fascial layers in the flanks on the side not occupied by renal tissue due to renal agenesis. The malposition of bowel into the extraperitoneal fat of the empty renal fossa and relaxation of mesenteric supports for bowel loops in this region occurs due to lack of restraining fascia.

They are sub classified into six subtypes in decreasing order of frequency[7] type a: inferior crossed fusion; type b: sigmoid kidney; type c: lump kidney; type d: disc kidney; type e: L-shaped kidney; type f: superiorly crossed fused. While the inferior CFRE is the most frequent type observed, the superior CFRE is reported to be least common. Similarly in our study we had all inferior CFRE. The lower pole of the superiorly normally positioned kidney is fused with the upper pole of the inferiorly situated crossed ectopic kidney in inferior CRFE type. Another characteristic feature of CFRE is the three times more common occurrence of left to right ectopy but in our study we found equal distribution. In our study we found symptomatic cases that required minimal invasive interventions (6/10). CFRE is often found incidentally during routine ultrasound screening or at other imagingor during autopsy and are usually asymptomatic, but may present with vague lower abdominal pain, hematuria, fever, urinary tract infection, hypertension, renal failure, and palpable abdominal mass. [8] In our study most common presentation was left flank pain,

The characteristic ultrasonographic 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 [3, 9, 10]. In addition, ultrasonography can give vital information on the arterial supply and venous drainage, which can be grossly abnormal, CT is widely utilised in preoperative assessment as it may be utilized to precisely identify the anatomical characteristics of the anomaly and the anatomical relationship between the kidney and surrounding structures (4). Crossed renal ectopia with fusion are associated with abnormal vascularity (5). If surgery is contemplated, contrast based CT is needed to characterize vascular anatomy, as the vascular supply can be anomalous to both the ectopic & non ectopic kidneys. The fused renal units need not to be separated. The treatment guided towards the associated problems that lead to either symptoms or the deterioration of the upper tracts eg. pelvi-ureteric junction obstruction would require a pyeloplasty or vesicoureteral reflux would require either injection of a bulking agent or reimplantation of ureters. In our study, three patients required pyeloplasty, one required pyelolthotomy and one required left moiety PCNL. In all the types of fusion anomaly the ureteral orifice associated with each kidney is usually orthotopic. Renal ectopy has high incidence of association with other urological abnormalities. Vesicoureteral reflux (VUR) is the most common succeeded by other genitourinary tract aberrations such as cryptorchidism, hypospadias, renal multicystic dysplasia & hydronephrosis [6]. In our study though vesicoureteral reflux was not seen but cryptorchism and hydronephrosis were associated with renal ectopia.

Due to abnormal anatomical structure of the kidney and ureter, as well as the abnormal relationship with the surrounding structures (including the small bowel, vertebral column and blood vessels), the treatment of upper urinary tract calculi is technically challenging for urologists [11]. Few studies have reported the treatment of kidney stones in patients with CFRE and several treatment methods are available, including extracorporeal shock wave lithotripsy (ESWL) [12, 13], percutaneous nephrolithotomy (PCNL) [11, 14–18], laparoscopic nephrolithotomy [19] and retrograde intrarenal surgery (RIRS) [20, 21]. In our study, we managed two patients of renal stone disease, one with lap pyelolithotomy and one with PCNL.

Laproscopy surgery is feasible in these cases of CFRE and we have managed three cases with laproscopic intervention. Most appropriate surgical management is dependent on anatomical configuration of kidney in CFRE and hence a flexibility of operative planning is required as per situation.

5. Conclusion

In conclusion, crossed fused renal ectopia is mostly detected incidentally during investigation for other problem. The management of crossed renal ectopia with fusion is individualised as per case according to the underlying urological anomaly and its sequelae. Care is taken to preserve the renal function whenever possible. So it's important for surgeon to know about such renal anomalies.

Abbreviations

CFRE-Crossed Fused Renal Ectopia
CECT KUB-Contrast Enhanced Computed Tomogram scan of Kidney, Urere, Bladder
NCCT-Non contrast CT
USG-Ultrasonography
MRI-Magnetic Resonance Imaging
IVU-intravenous Urogram
RGP-Retrograde pyelogram
DTPA-Diethylene-triamino-penta-acetic acid
ESWL-Extracorporeal shock wave lithotripsy
PCNL-Percutaneous Nephrolithotomy
RIRS-Retrograde intrarenal surgery

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