

Crossed Non-Fused Renal Ectopia with Bilateral Hydroureteronephrosis and Ureterocele in Ectopic Kidney- A Case Report

¹Dr. Leeladhar Agrawal, Professor, Department of Paediatric surgery, Jaipur National University and Medical College, Jaipur, Rajasthan,

²Dr. Anshu Kotia, Associate Professor, Department of Anaesthesia, Jaipur National University and Medical College, Jaipur, Rajasthan,

³Dr. Dinesh Kumar Barolia, Senior Resident, Department of Paediatric Surgery, S. M. S. Medical College Jaipur, Rajasthan

⁴Dr. Pradeep Gupta, Assistant Professor, Department of Paediatric Surgery, S. M. S. Medical College, Jaipur, Rajasthan.

Corresponding Author: Dr. Pradeep Gupta, Assistant Professor, Department of Paediatric Surgery, S. M. S. Medical College, Jaipur, Rajasthan.

Citation this Article: Dr. Leeladhar Agrawal, Dr. Anshu Kotia, Dr. Dinesh Kumar Barolia, Dr. Pradeep Gupta, “Crossed Non-Fused Renal Ectopia with Bilateral Hydroureteronephrosis and Ureterocele in Ectopic Kidney- A Case Report”, IJMSIR- July - 2020, Vol – 5, Issue - 4, P. No. 85 – 88.

Type of Publication: Case Report

Conflicts of Interest: Nil

Abstract

Crossed renal ectopia (CRE) with Ureterocele in ectopic kidney is extremely rare congenital anomaly. We are reporting here a neonate with urinary sepsis having non-fused Crossed renal ectopia with ureterocele and bilateral hydroureteronephrosis.

Keywords: Crossed renal ectopia, hydroureteronephrosis, ectopic kidney, kidney, and ureterocele.

Introduction

Crossed fused renal ectopia is an uncommon congenital anomaly wherein both kidneys whether fused or non-fused are located on one side, but the ureter of ectopic kidney crosses midline following normal course to open in its normal position in the bladder. CRE is the second most common fusion anomaly of the kidney after horseshoe kidney [1]. Blood supply of these

ectopic kidneys may be from lower aorta, or from common or external iliac arteries [2]. Here, we present a case of CRE without fusion, ureterocele and hydroureteronephrosis in ectopic kidney.

Case Report

A one month old male infant presented with complaints of purulent urine and mild grade fever since birth. Antenatal ultrasound suggests moderate degree of right hydroureteronephrosis with absent left kidney with a cystic area in bladder lumen.

Preoperative renal functions were normal. TLC was mildly raised. Urine was grossly purulent, with numerous pus cells on microscopy. Significant bacterial colony counts of E. Coli were found on urine culture. Postnatal USG showed right severe hydroureteronephrosis.

Voiding cystourethrogram suggestive of bladder outlet obstruction but with no VUR on either side.

In CT urography right sided grade III hydroureteronephrosis was noted (fig.1). Left kidney was small and ectopically located over right kidney (fig.2), anteromedial to right one, malrotated with hilum facing right laterally and showing gross left hydroureteronephrosis with mega ureter. Left renal pelvis and left ureter were not opacified in study.

DTPA scan had shown non visualization of left kidney with preserved perfusion and cortical functions on right side.

Cystourethroscopy and surgical removal of nonfunctioning ectopic pyonephrotic left kidney was planned.

Cystourethroscopy revealed a big ureterocele on left side that was incised. The right ureteral opening was isotopic and normal.

Right flank muscle splitting extraperitoneal incision given. The right kidney was dilated with dilated tortuous ureter. About 5cm×2.5cm cystic mass containing purulent fluid, was found anteromedial to right kidney, continuing in a 2.5 cm wide tortuous tubular structure to the left side of pelvis up to urinary bladder(fig.3), where it was ligated. The cystic mass was separated from right kidney and removed with its entire ureter. Right tube nephrostomy was performed for decompressing the dilated right upper tracts.

Post-operative period was uneventful. The nephrostomy tube was removed on 21st day of surgery. Patient was kept on prophylactic antibiotics.

Histopathology showed disorganized renal parenchyma that was distorted by variable sized cyst lined by flattened epithelium. Ureter was unremarkable.

During follow up for the last 21 months, patient doing well except occasional low grade UTI in initial period.

Serial USG monitoring showed gradual regression of right hydronephrosis. The renal biochemistry has been always found within normal limits.

CT Urography



Fig.1: Showed right gross hydroureteronephrosis with mega ureter.



Fig.2: Showed VUJO with ureterocele (arrow) obstructed ectopic opening.

Intraoperative view



Fig.3: Showed ectopic left kidney and dilated, tortuous ureter.

Discussion

CRE occurs in approximately 1 out of 7000 live births [3]. Pathogenesis is unknown. The most probable cause is abnormal development of ureteric bud during 4th to 8th weeks of gestation [4]. During their ascent if developing kidneys fuse in midline it results in horseshoe kidney. If one kidney advances slightly ahead of the other, inferior pole of advancing kidney fuses with the superior pole of lower kidney and results in fused CRE. In 90% of cases crossed ectopic kidney fuses to normally placed kidney. This anomaly is two times common in male and the cross from left to right [5].

As compare to available literature, our patient was male with left to right cross over and all the classically described findings were present, except that the crossed kidney was not fused with the normal kidney. However the final diagnosis was made after CT Urography. Other investigations like antenatal USG did not

conclude an accurate existing anomaly. Consistent with these finding, the DTPA studies demonstrated nonfunctioning of ectopic kidney.

The condition is generally asymptomatic and diagnosed as incidental finding or during autopsy [6]. VUR, urinary infections, ureterocele, megaureter, renal calculi, and renovascular hypertension can co-exist with ectopic kidneys [7]. Our patient was symptomatic in the form of urinary sepsis, so he was evaluated without delay. Bilateral hydroureteronephrosis in this patient might be caused by associated large left ureterocele.

Surgical plan was made according to radiological findings. On cystoscopy the incision of utererocele was made to avoid cystotomy and to decrease its size. The advantage of this procedure was achieved in the form of improvement in right sided hydroureteronephrosis in follow-up periods. Nephroureterectomy of ectopic nonfunctioning pyonephrotic left kidney was performed to get relief of urinary sepsis and to remove the diseased organ.

In one of the reported cases, the non-ectopic kidney was normal and no surgical intervention was performed for the crossed ectopic kidney [7]. However, we did tube nephrostomy to decompress the hydronephrotic but well-functioning solitary right kidney and achieved excellent result.

Conclusion

In the current English literature, there are only two cases of CRE and ureterocele. In best of our knowledge this patient is the third reported case with this association. In the first reported case, the crossed ectopic kidney was fused. In second reported case, there was crossed non-fused ectopic kidney with ureterocele but the affected kidney was dysplastic. So this is the first case ever, in available literature having

crossed non-fused renal ectopia but with bilateral hydroureteronephrosis and an ureterocele in nonfunctioning ectopic kidney.

References

1. Nussbaum AR, Hartman DS, Whitley N, et al. Multicystic dysplasia and crossed renal ectopia. *AJR Am J Roentgenol* .1987; 149: 407–410.
2. Caldamone A A, and Rabinowitz R. Crossed fused renal ectopia, orthotopic multicystic dysplasia and vaginal agenesis. *J Urol*.1981; 126: 105–107.
3. Robson WL, Leung AKC, Thomason MA. Multicystic dysplasia of the kidney. *Clin Pediatr*.1995; 34:32-40
4. Boyan N, Kubat H, Uzum A .Crossed renal ectopia with fusion: report of two patients. *Clin Anat*. 2007; 20:699-702.
5. Kwon TW, Sung KB, Kim GE. Experience of an abdominal aortic aneurysm in a patient having crossed ectopia with fusion anomaly of the kidney. *J Korean Med Soc*.2004; 19:309–310.
6. Sood R, Truong MX, Rossleigh MA, et al. Renal scintigraphy unraveled the diagnostic dilemma of antenatal hydronephrotic solitary kidney-crossed renal ectopia. *Clin Nucl Med*.2005; 30:621-2.
7. Siegel RL, Rosenfeld DL, and Leiman S .Complete regression of a multicystic dysplastic kidney in the setting of renal crossed fused ectopia. *J Clin Ultrasound* .1992; 20:466-469.