

Solitary Crossed Renal Ectopia with Vesicoureteric Reflux Presenting with Impaired Renal Function in a Neonate

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ABSTRACT

Solitary crossed renal ectopia (SCRE) is an exceedingly rare anomaly of the urinary tract. So far, only 34 cases have been reported in the literature. It usually presents after infancy. Most of these cases are diagnosed incidentally while patients are undergoing evaluation for associated genitourinary, cardiovascular, hematological or vertebral abnormalities. We report the first case of SCRE presenting in neonatal age with impaired renal function and vesico-ureteric reflux.

Key words:

Impaired renal function, neonate, solitary crossed renal ectopia, vesico-ureteric reflux

INTRODUCTION

Solitary crossed renal ectopia (SCRE) is a congenital abnormality that occurs due to combination of unilateral renal agenesis and renal ectopia.^[1] Most of the patients are asymptomatic and usually present with complaints related to genitourinary, cardiovascular, hematological or vertebral abnormalities.^[1-4] SCRE is usually incidentally diagnosed on routine ultrasound or autopsies. We report a case of SCRE with an unusual presentation in neonatal age with vesico-ureteric reflux (VUR) and impaired renal function.

CASE REPORT

A 15-day-old male neonate was brought to us by parents with an antenatal ultrasound scan suggestive of right sided hydronephrosis and hydroureter. At the time of presentation, the general and systemic examination of the neonate was normal. His renal functions were deranged (blood urea nitrogen (BUN): 16.2 mmol/L [8.0-16.4 mmol/L] and serum creat: 331 µmol/L [50-110 µmol/L]). Patient was having grade V VUR. There were no associated, cardiovascular and vertebral abnormalities.

Routine urine examination revealed plenty of pus cells and urine culture showed growth of *Escherichia coli*. Ultrasound of abdomen showed right sided hydronephrosis with right upper hydroureter with non-visualization of the left kidney. There were mobile internal echoes in urinary bladder. Patient was catheterized perurethrally and urinary tract infection (UTI) was treated with intravenous antibiotics.

Renal radionuclide scintigraphy, technetium-99m-L, L-ethylenedicycysteine (EC scan) was done which was suggestive of absent left kidney and incomplete pelviureteric

junction obstruction on the right side with adequate cortical function [Figure 1].

After obtaining a sterile urine culture report, a voiding cystourethrogram (VCUG) was done. It revealed grade IV VUR into the left ureter. The VCUG showed the ureter crossing the midline and entering the kidney on the right side [Figure 2a and b]. Magnetic resonance urography, done to confirm the anatomy, revealed a solitary right kidney with the ureter crossing midline and opening on the left side of urinary bladder along with presence of hydroureteronephrosis [Figure 3]. Diagnostic cystoscopy revealed absent right ureteric orifice and a wide left ureteric orifice with left hemitrigone confirming the diagnosis of SCRE.

Patient was managed initially with conservative treatment (antibiotics and catheterization). Once renal functions improved (BUN: 8 mg/dL and serum creatine: 1.2 mg/dL), the patient was discharged on chemoprophylaxis.

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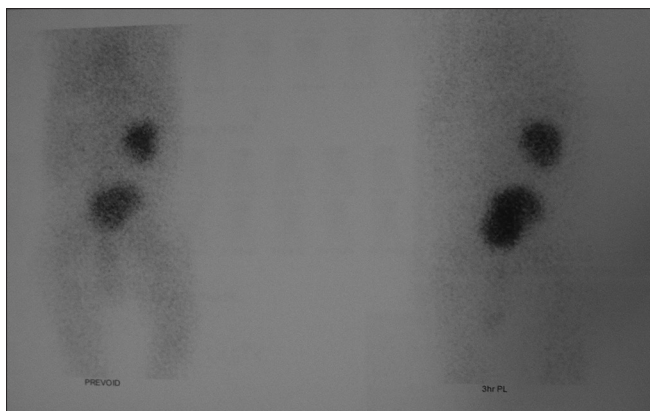


Figure 1: EC scan showing absent left kidney and right PUJ obstruction

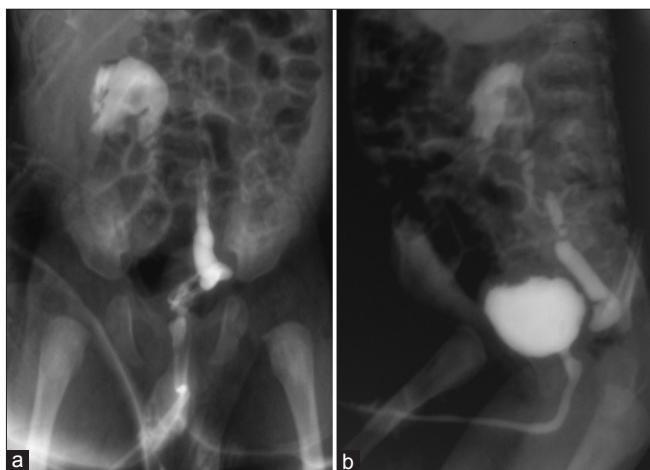


Figure 2: (a) Voiding cystourethrogram anteroposterior view showing left to right renal ectopia, (b) Voiding cystourethrogram lateral view showing left to right renal ectopia

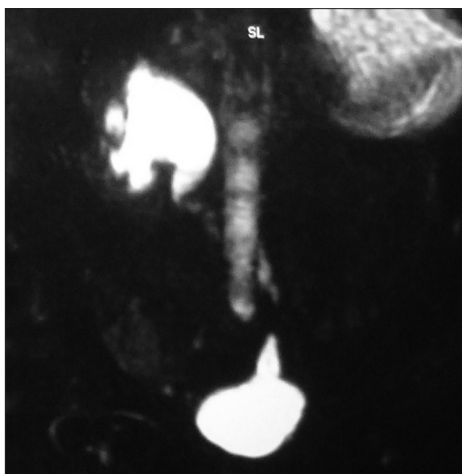


Figure 3: Magnetic resonance urography showing solitary crossed renal ectopia

However, in view of breakthrough UTIs with VUR in a solitary functioning renal unit, ureteric reimplantation was done at 6 months of age. Patient is asymptomatic and renal functions are normal at follow of 16 months.

DISCUSSION

Crossed renal ectopia is a condition in which kidney is located on side opposite from which its ureter inserts into bladder. McDonald and McClellan classified renal ectopia into 4 types (i) crossed renal ectopia with fusion, (ii) crossed renal ectopia without fusion; (iii) SCORE and (iv) bilaterally crossed renal ectopia. Renal agenesis of one side and contra lateral displacement of the ureter draining the only kidney is termed as SCORE. Only 34 patients of SCORE has been reported in the literature until date.^[1-6] Males predominate with a ratio of 2:1. Left to right ectopia is 2 times more common than right to left ectopia. Our patient was having left to right renal ectopia with single right kidney.

The various embryological theories that have been proposed for renal ectopia are (i) the metanephros and ureter are displaced by pressure of the umbilical arteries, (ii) the ureter wanders to induce the contra lateral metanephros, (iii) strong forces attract a ureter to the opposite metanephros and (iv) deviation of the median axis of renal vasculature.^[7-9]

SCORE is frequently incidentally diagnosed. Most of the cases reported in literature are associated with orthopedic deformities, genital abnormalities, hematological disorders and anorectal malformation.^[4-6,10-12] The most common genitl abnormality in the male is cryptorchidism or absence of the vas deferens; in the female, it is vaginal atresia or a unilateral uterine abnormality. Most individuals with crossed ectopic anomalies are asymptomatic and are often discovered incidentally at autopsy, during routine perinatal ultrasound screening, or after bone scanning. Our case presented with impaired renal function in neonatal age with VUR, which is an unusual presentation of SCORE.

SCORE can be diagnosed by ultrasonography and radionuclide scintigraphy using ^{99m}Tc- dimercaptosuccinic acid (Volkan *et al.*, 2003) in most of the asymptomatic cases. Excretory urography and multi-detector three-dimensional (3D) computed tomography (CT) urography is excellent for delineating renal ectopia. But in our patient, excretory urography and CT urography could not be done due to deranged renal functions. VCUG, magnetic resonance imaging urthogram and diagnostic cystoscopy confirmed the diagnosis of SCORE (left to right ectopia) in our patient.

Most of the individuals with SCORE do not require any treatment for this condition. Our patient required treatment for UTI, deranged renal function and then ureteric reimplantation for grade IV VUR.

Patients with SCORE have a good prognosis. Malignancy in SCORE is unusual and the incidence of renal cell carcinoma

in the CT era in a solitary crossed ectopic kidney is approximately 1 in 22 million.^[13]

CONCLUSION

SCRE is very rare disorder and our patient is probably the 35th case to be reported in literature. SCRE is often an incidental diagnosis but we report an unusual presentation of SCRE in neonatal age with impaired renal function, UTI, and VUR without any associated abnormality. CT urography or magnetic resonance urthogram is gold standard investigations for diagnosis of SCRE. SCRE usually requires treatment for associated abnormalities rather than for renal problems. Our patient required treatment initially for deranged renal function, UTI and then ureteric reimplantation for persistent grade IV VUR with breakthrough UTIs.

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