

Duplex Collecting System of Right Kidney in 3-Year-Old Child: A Case Report

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Abstract: *Background:* The duplex collecting system is one of the congenital anomalies of kidney and urinary tract. It occurs in 0.2-2% of the general population. It can remain asymptomatic or become symptomatic. Affected children may present with urinary tract infections. Treatment options include pharmacological and surgical approaches. *Case:* We reported a 3-year-old male child, who was referred to Sanglah Hospital with recurrent fever. From the physical examination, we found positive costovertebral angle tenderness. Urinalysis showed high leukocyte sediments (1655/field of view), positive urine culture (*Enterococcus gallinarum*), and decreased glomerular filtration rate (68.75 ml/minute/1.73 m²). Ultrasonography of the right kidney showed two pelvicalyceal systems with severe hydronephrosis of upper moiety. The intravenous pyelography showed the drooping lily sign in the right kidney. We diagnosed the patient with a complicated urinary tract infection caused by *Enterococcus gallinarum*, duplex collecting system of right kidney, chronic kidney disease stage II. Antibiotics were administered due to complicated UTI. Open heminephrectomy was done. The patient was discharged in stable condition. *Conclusion:* The duplex collecting system may cause recurrent urinary tract infection. It can be a risk factor for renal scarring and cause chronic kidney disease. Comprehensive diagnostic workup and multidisciplinary management are needed to promote a good clinical outcome.

Keywords: Duplex Collecting System, Urinary Tract Infection, Chronic Kidney Disease

1. Introduction

Congenital anomalies of the kidney and urinary tract (CAKUT) consist of a considerable number of diseases caused by defect in the morphogenesis or anatomy of the urinary system. The duplex collecting system falls under the broader classification of CAKUT in which the kidney has two pelvicalyceal systems with an upper and lower pole (moieties). The kidney may have either single or bifid ureter. The prevalence is estimated between 0.2 and 2% in the general population [1, 2].

The duplex collecting system has abnormal embryonic origins. The etiology of duplex kidneys can be traced back to

the initial induction steps of the ureter formation. The urogenital tract is derived from two independent germ layers. The kidneys and ureters develop from the intermediate mesoderm, and the bladder and urethra develop from the cloacal endoderm. In incomplete duplication, the two poles of a duplex kidney share the same ureteral orifice of the bladder. In most cases, the lower pole has normal function and the upper pole is non-functioning. In complete duplication, the upper pole drains into the bladder at a site distal to the normal orifice of the lower kidney pole. The kidneys may appear fused due to their development in the same metanephric mesenchyme [2, 3].

Most cases are diagnosed from antenatal ultrasound

imaging, which examines the kidneys, the outflows tracts, and amniotic fluid volume. Prenatal diagnose is based on ultrasonography which shows dilatation of the upper and lower pole. Some children with duplex collecting system acquire urinary tract infections (UTIs) secondary to reflux. Treatment strategies include administration of antibiotics to prevent recurrent UTIs, complete bladder reconstruction, or heminephrectomy of the non-functioning moiety [4, 5]. We present the rare case, diagnostic and management aspects challenges in a child with duplex collecting system of kidney.

2. Case

A 3-year-old male child was referred from a district hospital with diagnose of right kidney hydronephrosis. The patient had fever for two weeks, pain when urinating, and cloudy urine. From physical examination, we found positive costovertebral angle tenderness. There was no phimosis. Laboratory examinations revealed leukocyte $9.43/\text{mm}^3$, neutrophil 59.6%, lymphocyte 32.4%, decreased of hemoglobin level $9.43/\text{mm}^3$, mcv/mch 82.79/24.56, platelet $447.50/\text{mm}^3$. Renal function test showed urea 26 mg/dl, creatinine 0.8 and decreased of glomerular filtration rate (GFR) $68.75 \text{ ml/minute}/1.73 \text{ m}^2$. The procalcitonin level was 0.81, and urinalysis showed nitrite positive, elevated of leukocyte sediment level (1655/field of view), bacteria +++. Urine culture showed positive growth of *Enterococcus gallinarum* with 100.000 colony forming unit/mL.

Radiology examinations were performed, the first ultrasonography showed two pelvicalyceal systems in the right kidney (shown in Figure 1). In the upper moiety found severe hydronephrosis (Society of Fetal Urology (SFU) grade IV) with internal echo, while in the lower moiety we found mild hydronephrosis. Micturating cystourethrogram (MCUG) demonstrated no vesicoureteral reflux (shown in Figure 2). Intravenous pyelogram showed a drooping lily sign, which appears because the lower moiety is pushed by the enlarge non-functioning upper moiety (shown in Figure 3).

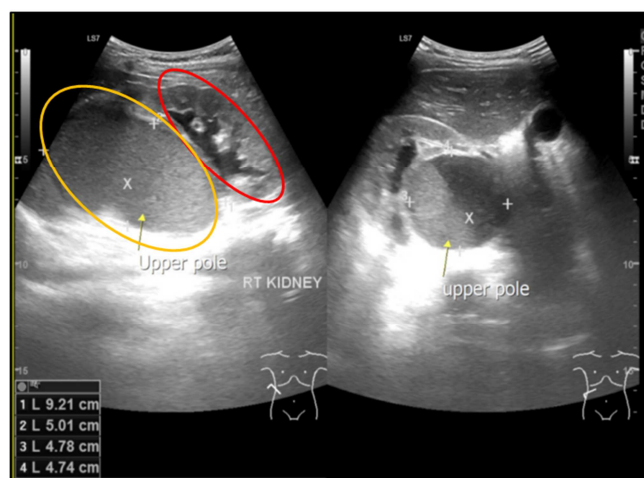


Figure 1. First USG of the right kidney. Yellow circle: severe hydronephrosis of upper moiety with internal echo (SFU grade IV). Red cycle: another pelvicalyceal system in the lower moiety.

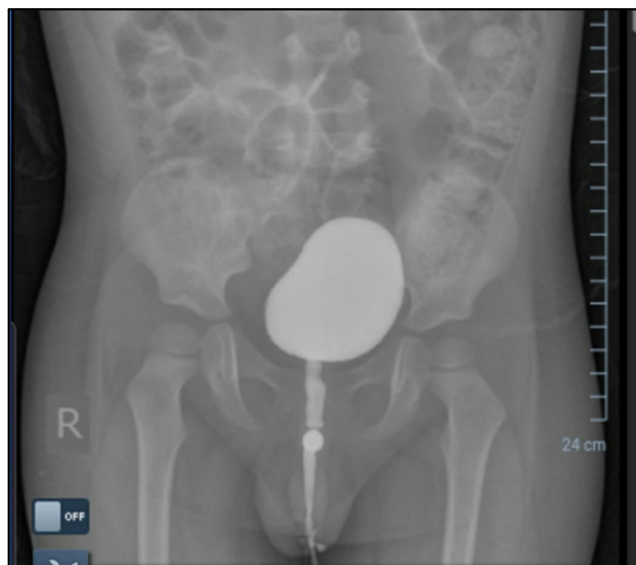


Figure 2. The MCUG examination showing: no vesicoureteral reflux.

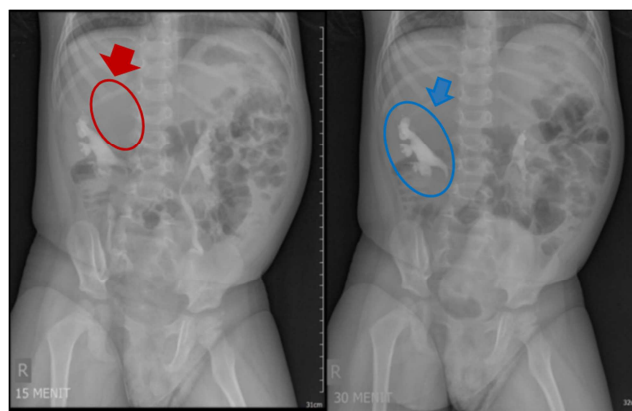


Figure 3. Intravenous pyelogram. Red cycle: non-functioning upper moiety right kidney. Blue cycle: drooping lily sign.

The patient diagnosed with complicated UTI caused by *Enterococcus gallinarum*, duplex collecting system of right kidney, CKD stage II. Patient was consulted to the Urology Department of Surgery. Heminephrectomy was done to remove the non-functioning upper moiety of the right kidney with its ureter. Empiric antibiotic, cefotaxime 900 mg four times a day was administered and changed to ampicillin 450 mg four times a day as a definitive antibiotic. Cotrimoxazole 40 mg daily at night was administered as a prophylactic antibiotic until the surgery was performed. Open heminephrectomy was performed successfully. During the surgery, it was revealed that the patient had complete duplication collecting system of right kidney. The patient was followed for 5 days after surgery, symptoms of UTI was resolved and GFR evaluation was improved to normal level.

3. Discussion

Duplex collecting system is a set of congenital anomalies with variety of phenotypes can be partial or complete duplication of the collecting system, pelvis, and renal

parenchyma. A duplex kidney has an upper and lower pole called moieties, which make the kidney look divided into two halves. Each moiety is drained by a ureter. The lower moiety is usually having normal function and drains most of the urine [6, 7]. The upper moiety usually non-functioning becomes obstructed and enlarged, resulting upper moiety mass effect (hydronephrosis). There is an increase incidence of infection because both VUR and obstruction are much more common in duplication systems [8, 9].

Symptoms associated with duplex kidney include urinary tract infection symptoms. In children ages 2 to 5 years old with UTI, fever and abdominal pain are the most common symptoms. After 5 years of the age, the most common symptoms are fever, dysuria, urgency, and costovertebral angle tenderness. In this case, the patient is 3-year-old male, complained recurrent fever, pain when urinating, and cloudy urine. The patient's USG showed a duplex collecting system of the right kidney with severe hydronephrosis (SFU grade IV) in the upper moiety and mild hydronephrosis in the lower moiety. Children with urinary tract malformations are prone to developing recurrent UTI with prevalence ranging from 25 to 59%. In duplex collecting system ureters typically follows the Wiegert-Meyer rule of the upper pole inserting caudomedial (may obstructed), lower pole inserting cranio-lateral. Ureter from the non-functioning upper moiety is often obstructed at the ureterovesical junction and develops ureterocele. Duplex collecting system result in natural free unidirectional flow of urine causing stasis of urine and causing the growth of pathogenic microorganisms [7, 10-12]. In this case, urine culture showed positive growth of *Enterococcus gallinarum*.

In a male infant, the recurrence of UTI needs some urogenital evaluation for detection congenital abnormalities. Analysis of such urinary infections includes urine culture sensitivity, ultrasonography, micturating cystourethrogram (MCUG), cystoscopy for a complete evaluation. Preventing recurrent UTIs in patients with duplex collecting system is one of the most effective ways to prevent renal scarring which progress to end stage renal disease [13]. According to the North American Pediatric Renal Trials and Collaborative Studies (NAPRTCS) and Europe, the most common cause of CKD in children are CAKUT (48-58%) [14, 15].

The treatment for renal duplication anomalies depends on the clinical problem. In children with complicated UTI prophylactic antibiotic is administered until surgical is performed. Therapeutic intervention, either surgical or conservative is needed to prevent further renal impairment. Treatment choice is influenced by several factors, such as age, presentation, ureterocele type, severity of hydronephrosis, grade of VUR (if present), UTIs, the remaining function of affected moiety, and surgeon's preference [16]. In this case, cotrimoxazole as a prophylactic antibiotic was administered until the heminephrectomy of the non-function moiety of right kidney is performed.

The goal of management is to preserve renal function, to eliminate infection and obstruction. The primary indication of heminephrectomy in children is to remove a non-

functioning upper or lower pole of kidney resulting from complicated duplex anomaly of kidney. There are several surgery methods that may be performed, open surgery, laparoscopic approach or robotic assisted approach [16].

4. Conclusion

Duplex collecting system of kidney is a rare congenital anomaly which can cause recurrent urinary tract infections in children. Furthermore, this condition associated with renal damage and can lead to chronic kidney disease. Early comprehensive diagnostic workup and multidisciplinary management are needed to promote a good clinical outcome.

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