Collaborative management of pediatric duplex collecting system with upper pole hydronephrosis, vesicoureteral reflux, and ureterocele: A case report

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ABSTRACT

Congenital anomalies of the kidney and urinary tract (CAKUT) is an uncommon finding with challenging diagnosis establishment and potentially devastating long-term renal outcome. We describe a case of unilateral duplex collecting system with upper pole vesicoureteral reflux and lower pole ureterocele. This 4-year-old female with prior antenatal assessment of kidney abnormality presented with recurrent urinary tract infection. A multidisciplinary approach was employed to encourage comprehensive diagnostic workup and management strategy. The complexity of such a rare case warrants higher index of suspicion and coordinated collaboration of a multispecialistic team.

Keywords: Congenital anomalies of the kidney and urinary tract, CAKUT; duplex collecting system; management; multidisciplinary; case report

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INTRODUCTION

Congenital anomalies of the kidney and urinary tract (CAKUT) is an umbrella term that comprises developmental defects of variable spectrum in the urinary system. The anomalies are mainly categorized by the organ involvement (i.e. kidney or its outflow tract) and disorder (i.e. macroscopic or microscopic changes). However, temporal perspective in developmental timeline better represents the severity of each defect. Heterogenous pattern of occurrence, whether it be in isolation, in combination, or as an element of other syndromes, further complicates its diagnostic aspect.1-2

The estimated prevalence of CAKUT ranged from 4 to 60 per 10,000 births.3 Despite its relatively rare occurrence, it is responsible for the poor long-term renal outcome in terms of chronic kidney disease and end-stage renal disease progression when left untreated.3-5 Duplex collecting system is amongst the most common congenital malformation (up to 0.9%) and generally accompanied with ureterocele and primary vesicoureteral reflux which altogether predispose to persistent or recurrent urinary tract infection (UTI).6 We introduce the diagnostic and management challenges of duplex collecting system along with its associated pathologies in which multidisciplinary paradigm was essential.

CASE PRESENTATION

A 4-year-old girl was admitted due to recurrent fever and cloudy urine since the age of 6 month. She also complained of dysuria with intermittent fever and cloudy urine since the age of 6 month. She was assessed with complicated UTI and infection. Urology ultrasound (Figure 1) showed duplex collecting system of the left kidney with upper moiety grade IV hydronephrosis, vesicoureteral reflux, and ureterocele. A multidisciplinary approach was employed to encourage comprehensive diagnostic workup and management strategy. The complexity of such a rare case warrants higher index of suspicion and coordinated collaboration of a multispecialistic team.

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Laboratory examination revealed increased white blood cell count (10.82 × 10³/L; neutrophil count 6.01 × 10³/L) and urinalysis suggestive of infection. Urology ultrasound (Figure 1) showed duplex collecting system of the left kidney with upper moiety hydronephrosis. The cystic lesion in the bladder was suspected to be ureterocele. Voiding cystoureterography (VCUG, Figure 2) showed left grade IV-V vesicoureteral reflux (VUR) with ureterocele.

She was assessed with complicated UTI and duplication of the collecting system of the left kidney with upper moiety grade IV hydronephrosis, grade IV-V left VUR, and left ureterocele. Empirical intravenous antimicrobial therapy consisting of ampicillin 100 mg/kg/day every 6 hour and gentamicin 7.5 mg/kg/day every 24 hour was administrated. Three months following initial treatment
she underwent dimercaptosuccinic acid (DMSA) renography which revealed cortical defect or renal scarring on the upper pole of the left kidney with the split function of 44.41% (1% upper, 16.39% middle, and 27.02% lower) for the left kidney and 55.59% for the right kidney (Figure 3). Endoscopic incision of the ureterocele and subsequent left open heminephrectomy were performed with one month interval in between. At present, she is in good condition without fever and urinary symptoms.

**DISCUSSION**

Diagnostic workup of CAKUT extends through the antenatal and postnatal timeframe. While majority of cases (60–85%) are detected by antenatal ultrasound imaging in the third trimester, the remainder will be left undiagnosed postnatally until significant symptom prompts further investigation.1 While UTI is often (30−76.2%) the first presentation indicative of underlying CAKUT in pediatric patients,7–9 imaging workup in UTI has limited role and is only reserved if the result would affect subsequent management. Imaging studies begin with ultrasound in all UTI cases and abnormal result will require VCUG or DMSA scans.10 In this case, kidney abnormality detected by antenatal ultrasound examination was not further investigated until current episode of care. The patient was repeatedly admitted with complicated UTI and finally underwent detailed workup for her condition.

Ureterocele is responsible for VUR and upper pole hydronephrosis as it may enlarge and disrupt other orifices, bladder muscular support, and ultimately leading to obstruction. Urine stasis and/or reflux leads to the development of infection.6 Recurrent UTI results in acute renal parenchymal damage which is best assessed with DMSA.10 Significant upper cortical segment functional discrepancy of the left kidney in this patient serves as a foundation to direct decision making in management strategy.

Duplex collecting system with high-grade reflux and ureterocele would unlikely respond to antibiotic treatment alone. Initial management should be directed to release the obstruction through endoscopic incision or puncture of the ureterocele. This patient underwent an endoscopic ureterocele incision to release the built-up pressure owing to hydronephrosis, prevent systemic infection, as well as to halt renal damage progression. Re-evaluation after decompression is necessary to determine the need for secondary surgery. Aforementioned upper pole function demonstrated by DMSA scan provided an
insight to dictate the extent of secondary surgery (i.e. partial nephrectomy or complete reconstruction). Upper pole heminephrectomy of the left kidney was chosen in current management because of existing reflux as well as severely declined function of the upper segment of the left kidney even after decompensation of the dilated system.

Notwithstanding numerous CAKUT case reports applying a multidisciplinary approach, it was seldom raised as the topic of interest. Considering the intricacy of this group of disease resulting from its diverse pathologies and unique combination of them, individually-tailored treatment strategies developed by a multidisciplinary team of pediatric nephrologists, pediatric surgeons or urologists, and specialized imaging expert, among others, offer a promising insight into thorough and timely management of CAKUT.

CONCLUSION

The complexity of multiple congenital anomalies of the kidney and urinary tract highlights the importance of comprehensive diagnostic workup and management to promote good clinical outcome. Future development of guidance for implementing a multidisciplinary approach is warranted.

AUTHOR CONTRIBUTIONS

All authors contributed to the concept, definition of intellectual content, clinical studies, data acquisition and analysis, along with manuscript editing; K.S., G.W.K.D., and P.P.Y.A. worked on design and manuscript review; K.D.L.B. worked on literature search and manuscript preparation; all authors served as guarantors for this study.

CONFLICT OF INTEREST

The authors have nothing to disclose.

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ETHICAL CONSIDERATIONS

Written informed consent was obtained from the patient and its copy was available to be reviewed by the Editor-in-Chief of this journal.

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