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Case Report

Unilateral multicystic dysplastic kidney disease associated with ipsilateral ureteric bud remnant and contralateral duplex collecting system ☆

*Harry Galuh Nugraha, MD, SpRad(K), Audy Sarah Putrini Adibrata, MD**

Department of Radiology, Padjadjaran University, Bandung, Indonesia

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ABSTRACT

Congenital anomalies of the kidney and urinary tract are among the most common developmental malformations. The heterogeneity of these anomalies is very high, some of them are rarely discussed in the literature. Herein, we present a case of a 5-year-old male who was found to have a combination of unilateral multicystic dysplastic kidney associated with ipsilateral ureteric bud remnant and contralateral duplex collecting system.

Introduction

Multiystic dysplastic kidney (MCDK) is commonly associated with an atretic or absent ureter. In a rare occurrence, MCDK may associated with persistent ureteric bud structures [1]. MCDK It is usually asymptomatic due to the compensatory hypertrophy of the contralateral kidney [2]. However, it can cause symptoms if it is associated with other congenital anomalies of the kidney and urinary tract (CAKUT) [3]. The common CAKUT associated with MCDK is vesicoureteric reflux (VUR), pelviureteric junction obstruction (PUJO) and posterior urethral valves (PUV). However, the occurrence of MDCK associated with contralateral duplex collecting system is extremely rare [4–6]. This case report aims to raise awareness of the possibility this unique phenomenon among radiologists and clinicians.

Case report

A 5-year-old male child with recurrent urinary tract infection was referred to our hospital for a urethrocystography and magnetic resonance imaging (MRI) of the abdominopelvic area. The child had a history of rectourethral fistula that had been repaired when he was 2 years old. There was no complaint of faeces-stained urine nor urinary drainage through the rectum following the repair surgery. An ultrasound per-

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* Corresponding author.
E-mail address: audy_sarah@yahoo.com (A.S.P. Adibrata).

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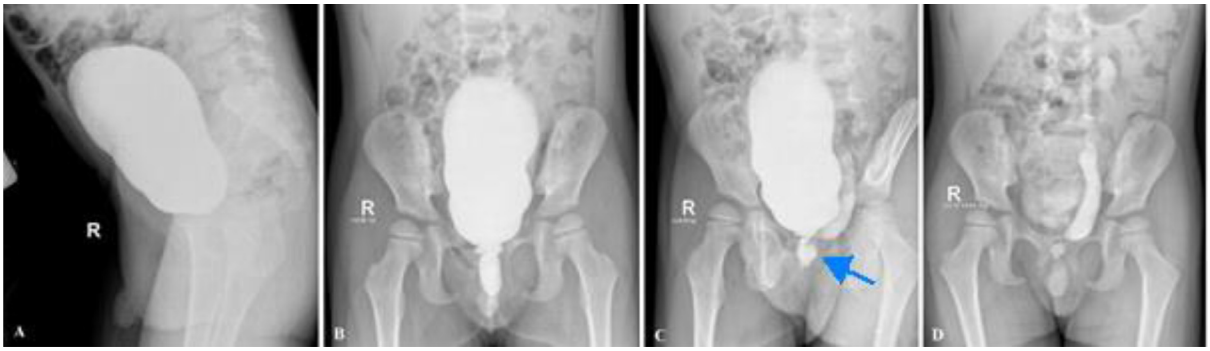

Fig 1 – Voiding urethrocystography. (A) The urinary bladder is large. Some trabeculation can be seen. (B) The prostatic urethra was dilated during voiding. (C) Oblique view showed a urethral diverticulum (blue arrow) at the posterior wall of prostatic urethra. (D) Left vesicoureteric reflux (VUR) was evident.

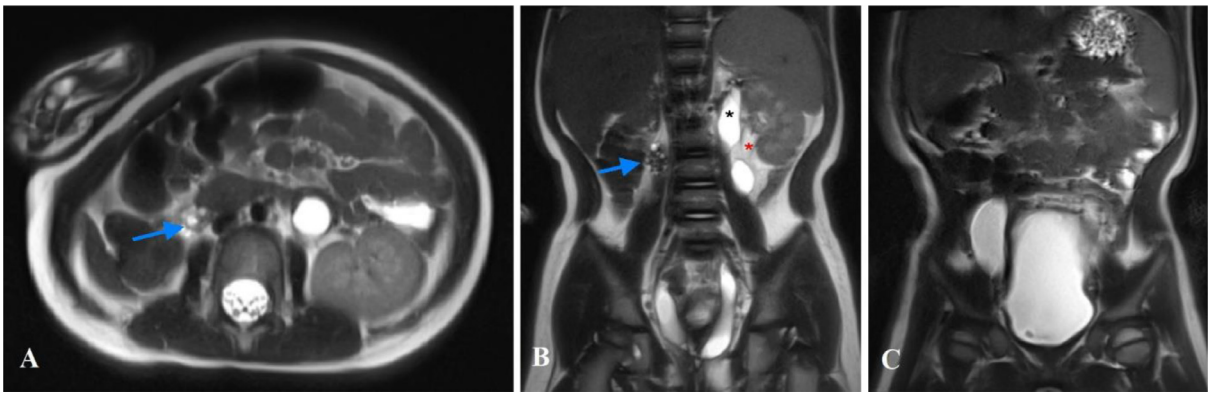

Fig. 2 – T2-weighted image. (A) and (B) Axial and coronal image showed a multicystic structure in the right flank area (blue arrows). The left kidney has 2 ureters, one at the upper pole (black asterisk) and the other one is at the middle pole (red asterisk). The ureter at the upper pole connected to the urethra. (C) Coronal image showed the blind ending ureter emerging from the urethra.

formed in other hospital disclosed suspicion of right renal agenesis and duplex collecting system of the left kidney.

Urethrocystography revealed a large and slightly trabeculated urinary bladder. During voiding, the posterior wall of the prostatic urethra was dilated, suggestive of PUV. There was also a urethral diverticulum on the posterior wall of the prostatic urethra. There was no connection between the structure to the colon. Fig. 1.

Magnetic resonance imaging revealed a retroperitoneal cluster of cystic structure in the right flank area. That structure was 0.91×1.21×1.91 cm in size. It showed no function on isotope renography, confirming diagnosis of multicystic dysplastic kidney (MCDK). There was a dilated, orthoptic ureter emerging from the urethra with a blind ending. There was no communicating structure between the ureter and the dysplastic kidney Fig. 2.

The left kidney had 2 ureters on the upper pole and the middle pole. The ureter on the upper pole was dilated and it entered the bladder at the urethra. The upper part of the middle pole ureter was also dilated, due to the mass effect of the upper pole ureter. The bladder insertion was at the bladder trigone. Delayed phase postcontrast scanning showed functional middle pole ureter. The bladder wall was thickened with hypertrophic bladder neck, resulting from recurrent cystitis Figs. 3 and 4.

The patient underwent a partial nephrectomy of the left kidney to remove the upper pole ureter. No further follow up on imaging at our hospital after the procedure. On the by phone follow up, the patient's parents stated no complaints after the surgery.

Discussion

CAKUT represents a various group of abnormalities involving the kidney, collecting system, urinary bladder and urethra, with the incidence of 1-3 in a general population of 500 [7]. Unilateral MCDK occurs in approximately 1 per 4300 live births [8]. There are 2 predominant theories about the MCDK.

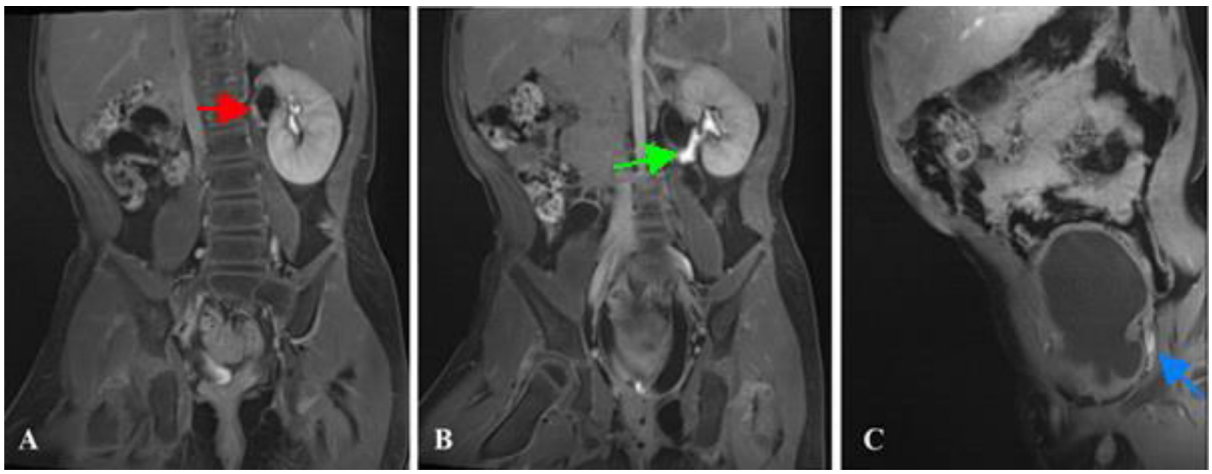

Fig. 3 – Delayed phase of contrast-enhanced T1-weighted image with fat saturation. (A) and (B) Coronal image showed the nonfunctional upper pole ureter (red arrow) and functional middle pole ureter (green arrow). (C) The functional left ureter (blue arrow) entered the bladder at the bladder trigone.

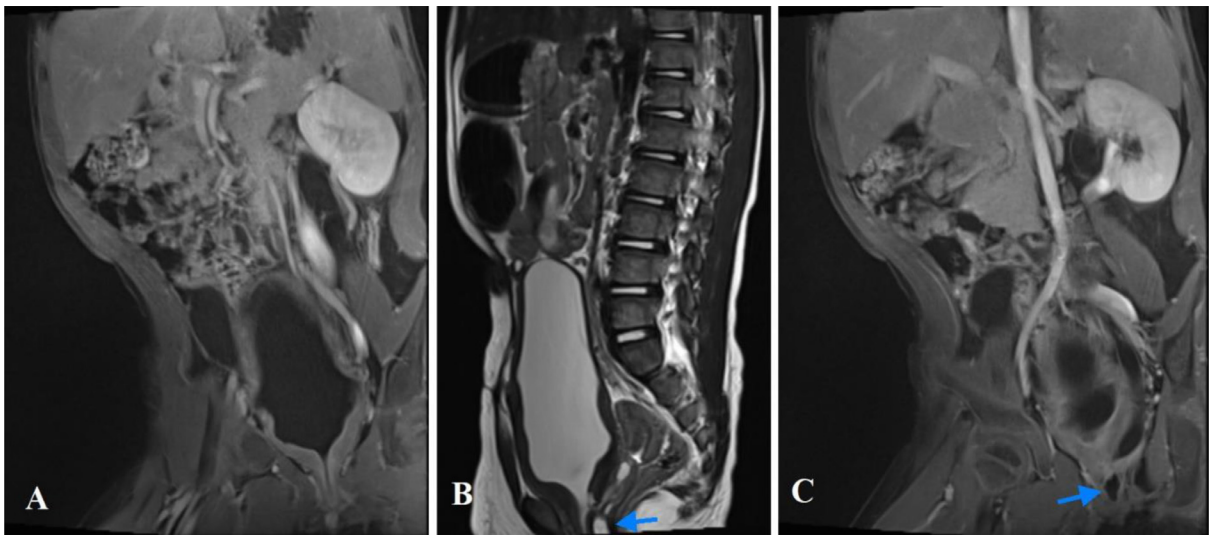

Fig. 4 – (A) Contrast-enhanced T1-weighted image with fat saturation showed the hypertrophic bladder neck. (B) Sagittal plane of T2-WI showed the urethral diverticulum on the posterior urethral wall (blue arrow). (C) The diverticulum (blue arrow) demonstrated no enhancement.

The first theory suggests that ureteral atresia leads to severe obstructive hydronephrosis and eventually MCDK. The other theory proposes that an abnormal interaction between the metanephric blastema and ureteric bud leads to a failure of normal differentiation of these structures [1,2].

The affected kidney tends to involute and some disappears before birth, especially if the original size of the kidney was already tiny. Hence it is possible to misdiagnosed regressed MCDK as renal agenesis [2,8]. The ureteric bud usually regresses in MDCK. When it rarely persists, it gives rise to tubular or cystic structure related to the urinary tract. The tubulocystic structure varies from a simple urethrocele to a complex abnormalities involving the mesonephric duct structures [1].

Concomitant PUV and VUR in MCDK are not necessarily unusual. It is the coexists duplex collecting system of the contralateral kidney that makes our case unique [4,5]. It has been reported only twice in the literature. One of the cases reported in a study in Turkey [6]. The other case was diagnosed antenatally in The United Kingdom [5].

Conclusion

The coalescence of unilateral multicystic dysplastic kidney associated with ipsilateral ureteric bud remnant and contralateral duplex collecting system is an exceedingly uncommon finding. It emphasizes the width of the spectrum, of which radiologists and clinicians must be aware of.

Patient consent

Complete written informed consent was obtained from the patient for the publication of this study and accompanying images.

REFERENCES

[1] Coleman R, Sanchez O, Ghattaura H, Green K, Chandran H, McCarthy L, et al. Tubulocystic anomalies of the mesonephric duct associated with ipsilateral renal dysgenesis. J Pediatr Urol 2019;15(1):46.e1–46.e6.

[2] Ferro F, Vezzali N, Comploj E, Pedron E, Di Serafino M, Esposito F, et al. Pediatric cystic diseases of the kidney. J Ultrasound 2019;22(3):381–93.

[3] Kohl S, Avni FE, Boor P, Capone V, Clapp WL, De Palma D, et al. Definition, diagnosis and clinical management of non-obstructive kidney dysplasia: a consensus statement by the ERKNet Working Group on Kidney Malformations. Nephrol Dial Transplant 2022;37(12):2351–62.

[4] Schreuder MF, Westland R, van Wijk JAE. Unilateral multicystic dysplastic kidney: a meta-analysis of observational studies on the incidence, associated urinary tract malformations and the contralateral kidney. Nephrol Dial Transplant 2009;24(6):1810–18.

[5] Damen-Elias HAM, Stoutenbeek PH, Visser GHA, Nikkels PGJ, de Jong TPVM. Concomitant anomalies in 100 children with unilateral multicystic kidney. Ultrasound Obstet Gynecol 2005;25(4):384–8. doi:10.1002/uog.1851.

[6] Aytaç B, Sehıtoğlu I, Vuruskan H. Multicystic dysplastic kidney: four-year evaluation. Turk patoloji dergisi 2011;27(3):210–14.

[7] Surabhi VR, Menias CO, George V, Matta E, Kaza RK, Hasapes J. MDCT and MR urogram spectrum of congenital anomalies of the kidney and urinary tract diagnosed in adulthood. Am J Roentgenol 2015;205(3):W294–304.

[8] Hiraoka M, Tsukahara H, Ohshima Y, Kasuga K, Ishihara Y, Mayumi M. Renal aplasia is the predominant cause of congenital solitary kidneys. Kidney Int 2002;61(5):1840–4.