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Case Report

Duplication of ureter in a 7-year-old: a case report☆,☆☆,★,★★

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ABSTRACT

Duplication of ureter is one of the most common anomalies of the urinary tract. Early detection has dramatically increased due to advance in technology of imaging to detect the anomalies during antenatal period; however, numbers of undiagnosed adult still exist. It may remain asymptomatic, but may cause repeated urinary tract infections or calculi. This case report presents a case of a 7-year-old female who had duplex collecting systems. This patient has repeating symptoms throughout the years, referred to several medical specialties. Then ultrasonography and computed tomography showed that she had complete duplex collecting system on her left kidney. She was then undergone left partial nephrectomy surgery and removal of dilated left ureter. Congenital anomaly of the urogenital system should be considered in patients with chronic infection. Multimodal imaging technique such as ultrasonography, computed tomography, or magnetic resonance imaging should be done to confirm the diagnosis especially before surgical management.

Introduction

The ureters are bilateral fibromuscular tubes that transport urine from renal pelvis to the posterior surface to the urinary bladder. They are generally 22 to 30 cm in length and course through the retroperitoneum. Along the length of the ureter, there are 3 segments that in physiologically narrow: the ureteropelvic junction, the ureterovesical junction, and where the ureter cross the common iliac vessels [1,2]. Duplications of the ureter represent one of the most common anomalies of the urinary tract. The incidence of duplex renal collecting

Abbreviations. CT, computed tomography; MRI, magnetic resonance imaging; UTI, urinary tract infection.

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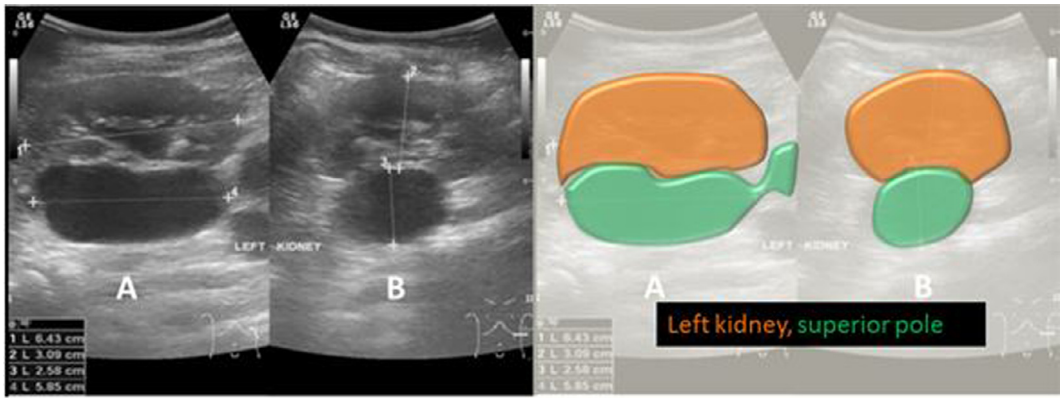

Fig. 1 – Ultrasonography longitudinal (A) and transverse (B) view with schematic drawing of the left kidney.

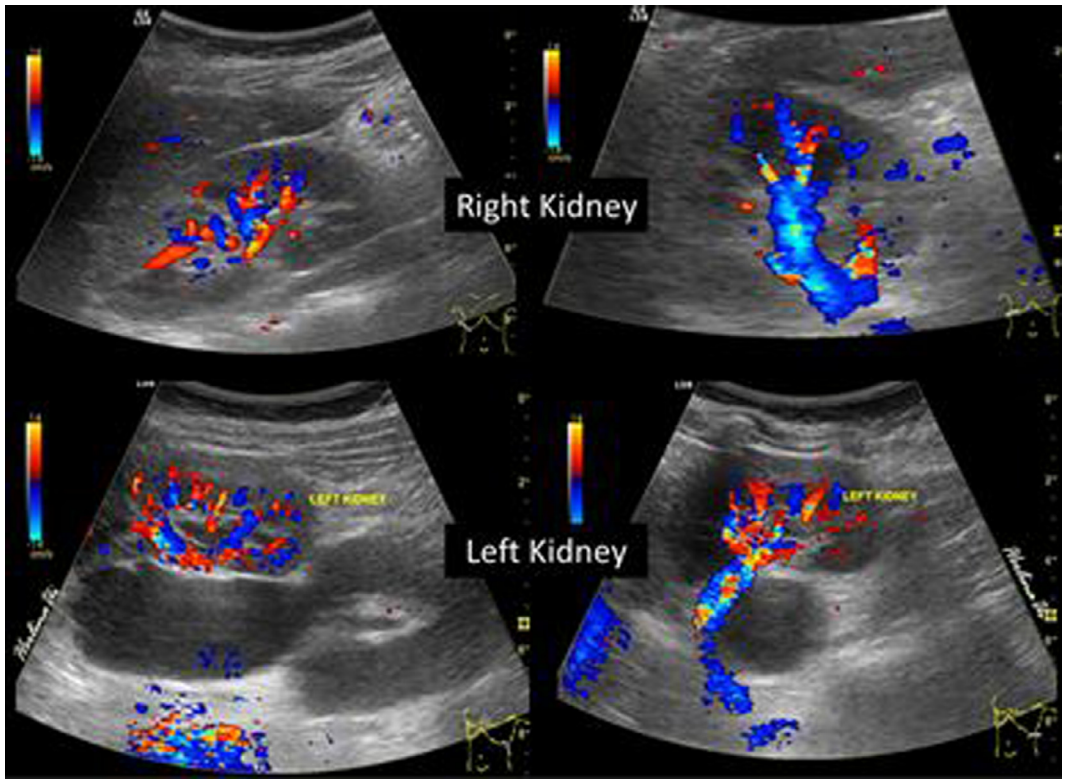

Fig. 2 – The color Doppler ultrasound image of longitudinal and transverse view of right and left kidney. Right kidney appeared normal, left kidney appeared abnormal.

system ad ureter ranges from 0.5% to 3%. Complete ureteral duplication is more rarely seen compared to single ureter or partial duplication. Complete duplicated systems are those where 2 ureters arise from the same kidney and drain separately into the bladder, while partial duplication is where 2 proximal branches drain the same pelvis but join together distally to form 1 common ureteric branch prior to emptying into the bladder [2,3]. One consequence of a nonfunctional renal collecting system is ureteric orifice malpositioning, such that the ureter of the inferior pole implants with a shorter tunnel into the bladder, thereby predisposing to vesicoureteral reflux. On the other hand, the positioning of the ureter of the superior pole of the kidney makes it more prone to ureteroceles and obstruction at the ureterovesicular junction [4]. The prevalence of duplication of ureter is higher in female than male [5].

Childhood detection of such renal anomalies has dramatically increased; however, a significant number of undiagnosed adult still exist [4]. Although duplication of ureter may remain asymptomatic, it may cause repeated urinary tract infections (UTIs) or calculi, also get injured during pelvic surgeries [6]. The presence recurrent UTI, most commonly due to vesicoureteral reflux and urinary incontinence in females occurs in cases of ectopic ureter entering the vagina, urethra,

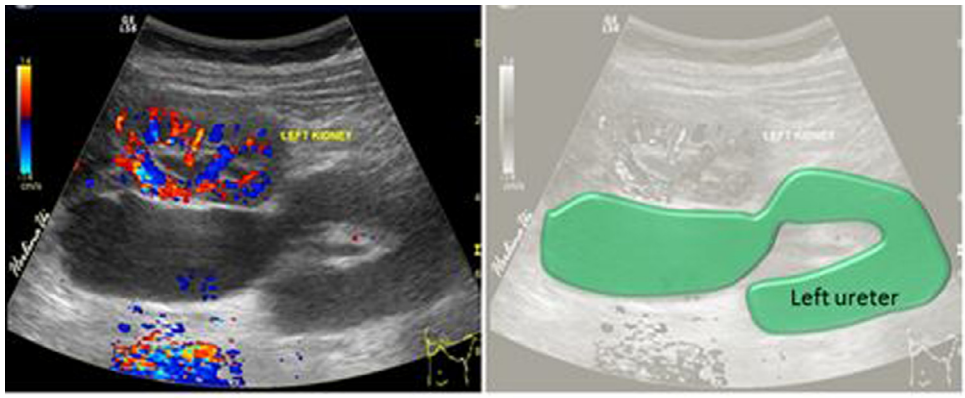

Fig. 3 – The color Doppler ultrasound image with schematic drawing of the longitudinal view of left kidney shows cyst-like structure surrounded by a rim of renal parenchyma in the upper pole of the kidney merging with the dilated and tortuous ureter.

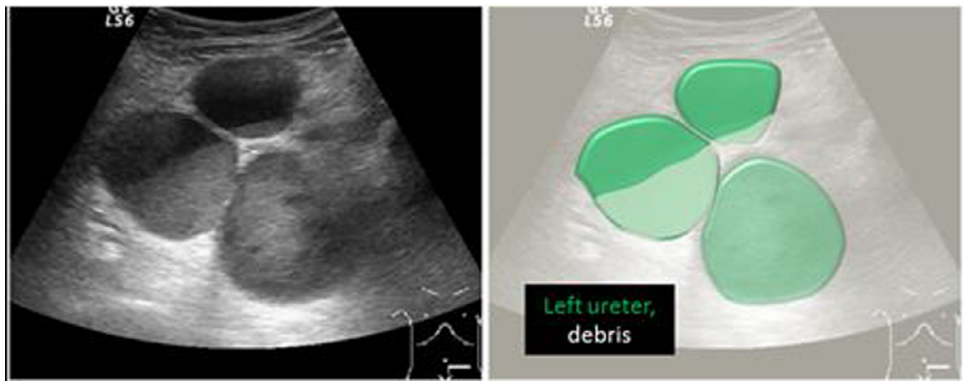

Fig. 4 – Transverse view of the ultrasound with schematic drawing, shows left dilated and tortuous ureter with prominent internal echo debris/"sludge" in the ureter.

or vestibule [5]. We introduce a case of unilateral complete ureteral duplication with recurrent UTI, which was treated by partial nephrectomy.

Case report

A 7-year-old female complained for worsening left-sided abdominal pain in the last 6 days before hospital admission. The pain intensified while urinating. She also experienced fever for 4 days. She also had occasional bed-wetting as a 7-year-old. On her medical history, she had frequently visited pediatrician for similar symptoms since she was 3 years old, and treated for UTI. She undergone an ultrasound examination in her previous hometown, then diagnosed with ovarian cyst. She was referred to a gynecologist. After visiting gynecologist, she was then referred to a pediatric radiology specialist for more detailed examination.

Physical examination revealed that she weighed 26.7 kg and was 105 cm tall. Vital signs were normal and on abdominal examination; she had left lower quadrant and left costovertebral angle tenderness. Laboratory values showed that she had normal blood urea nitrogen and serum creatinine, but had evidence of infection in the urinalysis result. Other laboratory results were unremarkable.

Abdominal ultrasound was done (Figs. 1-4). The ultrasound resulted duplication of left pyelocaliceal system, severe hydronephrosis on upper poles of left kidney and left tortuous ureteral dilatation with prominent internal echo debris/sludge in the ureter. The dilated and tortuous ureter simulated multiple cysts. The bladder was empty therefore not visualized. CT scan was then obtained (Figs. 5 and 6) an excretory phase image is shown. On the left kidney shows duplex ureter coming out from the superior and inferior renal pelvis. The ureter exiting the superior pole was dilated to the distal area, tortuous, reaching the urethral base while the ureter exiting the inferior pole shows normal excretion phase, with the distal-third part pushed by the ureter exiting the superior pole. The patient was then hospitalized and planned for surgical consultation.

This patient had left duplex collecting systems and undergone left partial nephrectomy surgery and removal of left dilated ureter that exited from the superior pole (Fig. 7). While hospitalized she was treated with antibiotic and pain medications.

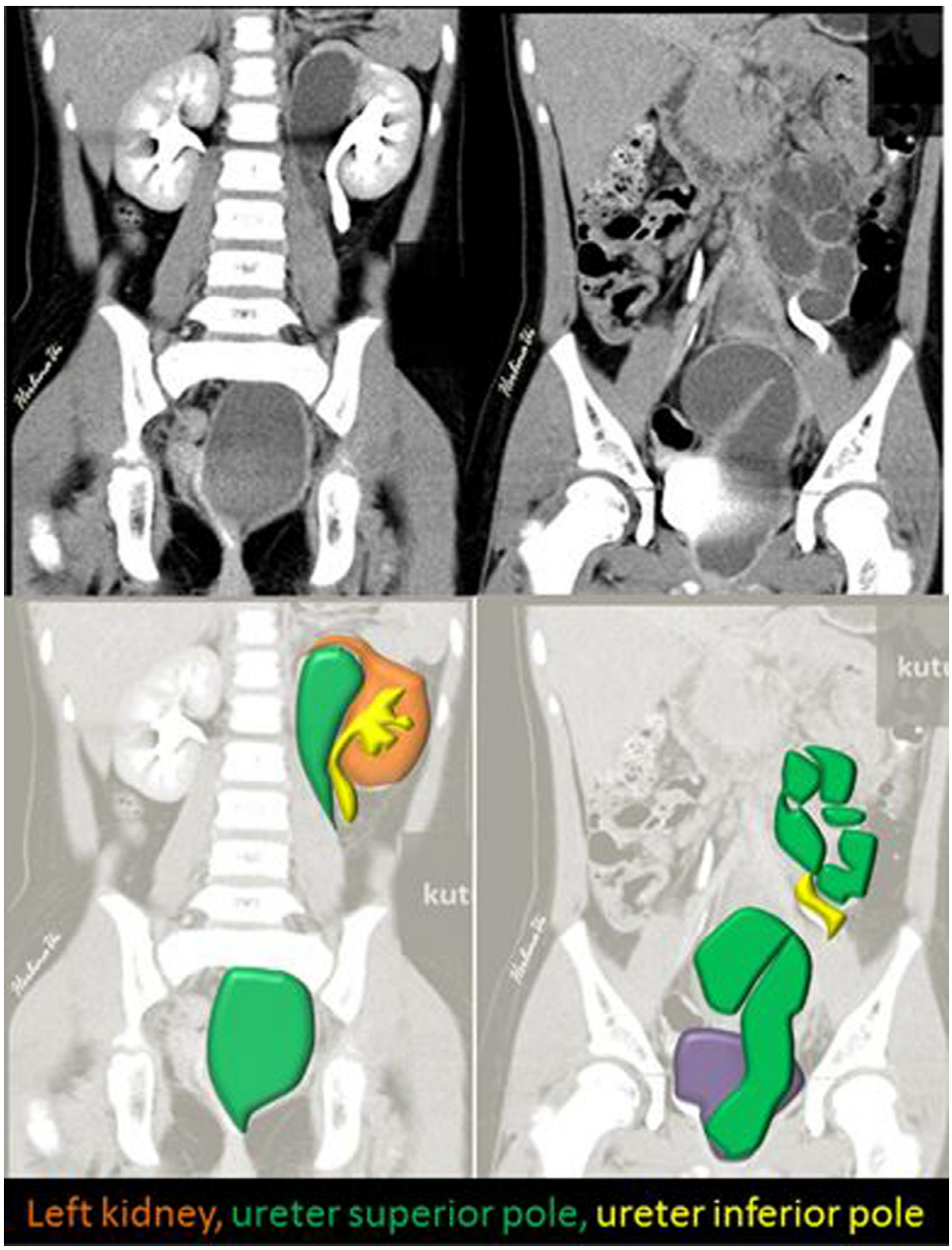

Fig. 5 – Multiplanar urogram-phase CT serial images coronal view and schematic drawing (below) shows dilatation of the proximal and distal parts of the superior pole of the left ureter. A dilated upper-pole ureter causes extrinsic compression and partial obstruction of the lower-pole ureter in their crossing part.

No serious complications were observed in both preoperative and postoperative period. She was discharged 5 days after the surgery.

Discussion

Duplication of ureter develops during the 4th to 5th weeks of gestation when there was failure to repress ectopic budding from the Wolffian Duct or multiple Ureteric Buds Induction during metanephros development. In complete ureter duplication, the ureteric buds rotate 108° when incorporating into the urogenital sinus, known as Weigert-Meyer rule. A duplex collecting system is one of the most common congenital genitourinary tract abnormalities, even though it has a rare occurrence [7–9]. It is 2 to 4 times more common in female than male, either be asymptomatic or causing recurrent UTI in children [6]. In this case, the patient is female and had been treated for UTI multiple times. Bed-wetting complaints may be caused by dribbling of urine due to infrasphincteric insertion of the upper-pole ureter. Duplex systems have potential for future complications, such as obstructive uropathy, stone formation, ureterocele, and vesicoureteral reflux. Early detec-

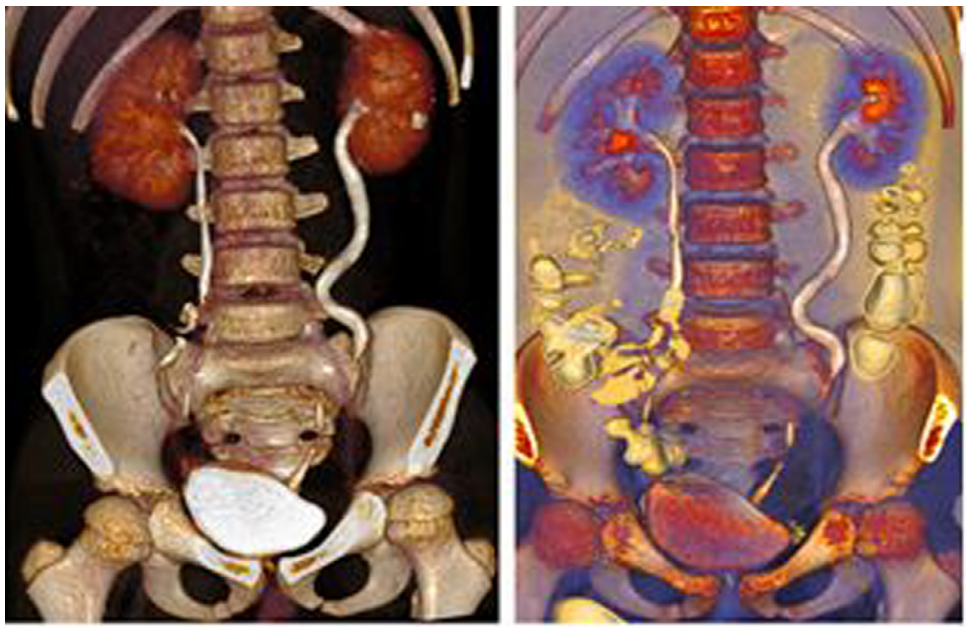

Fig. 6 – CT-scan urogram-phase, coronal volume rendering image.

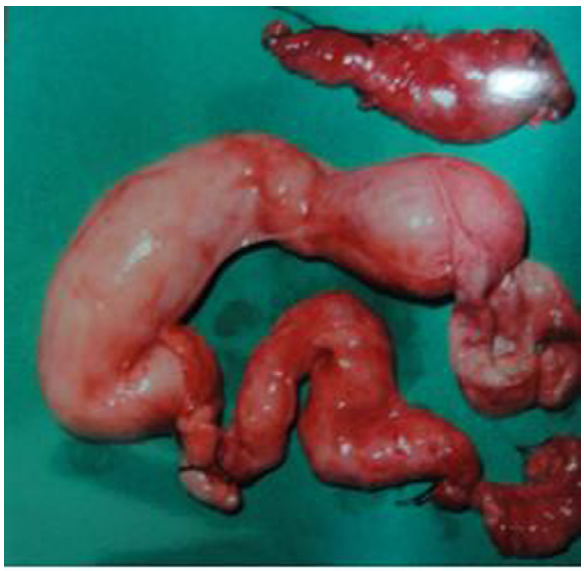

Fig. 7 – Surgical specimen of partial left nephrectomy and dilated left ureter exiting from the superior pole removal.

tion of this anomaly is helpful to prevent comorbidities and complications [10].

In all cases, imaging is mandatory to confirm the diagnosis. In children, kidney ultrasound represents the initial diagnostic test, but it has some limitations and may not be so helpful. Renal ultrasound and excretory urography can almost never detect an ectopic insertion of the ureter and they do not provide enough data regarding the precise anatomy as well as delineating the relationship between the ureter, bladder, and vagina [11]. Cross sectional imaging including computed tomography (CT) and magnetic resonance imaging (MRI) help in resolving the complex anatomy, including duplex collecting systems and to look for complication like pyelonephritis, renal stones, and malignancies. Noncontrast CT can detect renal stones and nephrocalcinosis. CT urography is useful in the case of duplex collecting system, visualization of their complex course, distal opening, and associated other genitourinary malformations such as ectopic kidney complicated by stones, infection, or hypertension arising from multiple anomalous arteries [12]. Other than ultrasound and CT, MRI has the advantages of lack of ionizing radiation, better soft tissue contrast, and detection of the collecting system abnormalities even without contrast. Disadvantage is requirement of sedation in small children, cost, and availability [12]. In this case, contrast CT urography was done to confirm the diagnosis. This case is the same as what has been shown by Weigert-Meyer's about the relationship between the upper and lower kidney parts in a duplex collecting system with their drainage. CT urography showed a dilated upper-pole ureter causing extrinsic compression and partial obstruction of the lower-pole ureter. CT could also be a better imaging technique to show the site of the ureter orifices.

The best treatment in children and symptomatic patients is surgery, and it tries to resolve the incontinence, prevent further complications, preserve renal function, and eliminate UTIs. Surgical treatment consists in upper pole heminephrectomy in the non-functional duplex system or recently laparoscopic ureteral ligation (clipping) or ureteral reimplantation in case of a preserved renal function [11]. The nonfunctioning or poorly functioning kidney is usually dilated and hydronephrotic [13]. Surgical management depends on surgeon experience and preference, laparoscopic experience, and pediatric material investments [11]. In patients with reflux to the upper pole, referred for upper pole

heminephrectomy, it is necessary to consider the removal of the ureter to the level of the vesicoureteral junction [14]. In this case, we found the patient's left kidney to be hydronephrotic or poorly functioning, therefore surgical upper pole heminephrectomy and removal of the dilated ureter exiting the upper pole were done to improve drainage and eliminate symptoms.

While sometimes came up as asymptomatic, congenital anomaly of the urogenital system should be considered in patients with chronic infection. Multimodal imaging technique such as ultrasonography, CT, or MRI should be done to confirm the diagnosis, to provide good anatomical image before the surgical management.

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