

Obstructing Calculus in a Single Limb of a Duplicated Ureter: A Recall of the Surgical Anatomy and the Variable Ureter Anomalies

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Abstract

The ureter is a common site of congenital anomalies which may be associated with a considerable morbidity particularly among young patient. The congenital anomalies of the ureter coexist with multitude of other urinary tract anomalies, but it may occur independently. It is more common in females. The complete duplication of the ureter may not produce symptoms which would suggest the presence of malformation. Therefore, such anomaly may not become apparent until later in life. Further, this anomaly might not be recognized prior to the surgery and hence, missing of the stone is highly possible. Herein we present a case of complete ureter duplicate with an obstructive stone located close to VUJ of one limb of the duplicate. A sound knowledge of the surgical anatomy and of the congenital ureter anomalies is essential for correct diagnosis and appropriate management.

Keywords: Duplicated ureter; Unenhanced CT KUB; Vesico-ureteric junction; Ultrasonography; Ureteroscopy; Calculus

Introduction

The ureter is a derivative of the wolffian (mesonephric) duct. Ureteral bud branches off from the caudal portion of Wolffian duct at the fourth and fifth week of gestation. Alteration in the bud number, position or time of development results in ureteral anomalies. A combination of genetic mutation and environmental factors contribute to the formation of urinary tract anomalies [1,2]. Ureter duplication is inherited by an autosomal dominant gene of variable penetration [3]. The highest prevalence has been reported in Caucasian females [4,5]. Duplication of the ureter may be incomplete (bifid ureter) or complete in which each limb of the duplicate open separately in the bladder and this is a rare anomaly [6,7]. The VUJ orifice of the limb which drain the upper renal moiety (the first limb of the duplicate) has ectopic insertion usually medial and inferior to the lower pole moiety ureter (the second limb of the duplicate) which has orthotopic insertion in the bladder (Weigert-Mayer law) [1]. The complete duplication is present in approximately in 1 from 125 individual [6]. It is more common

in females than males with a ratio of 2:15. In the English literature few cases of calculi found in one limb or in the two limbs of the duplicated ureter [8,9]. More few cases reported with bilateral ureteric duplication in which stones were found in all of the limbs of the duplicate [10]. Some cases of ureteral duplication with ureterocele stone have been reported [5,7]. A similar case of duplex was also detected in a research study and another seen during a routine dissection of a cadaver [6,11]. Generally speaking, most of the aberrations detected incidentally during URS surgery for obstructing stone or during another pelvic organ surgery [5]. Herewith we present an additional case of complete duplicated left ureter with an impacted calculus found in a single limb of the duplicate. Meanwhile some related elegant images selected from many others are also presented.

Case Presentation

A 22-year-old female patient newly married repeatedly she attended the emergency room (ER) with a complaint of left sided abdominal pain, dysuria and difficulty in urine void. Her abdomen was mildly tender on the left iliac fossa. She had microscopic hematuria with few RBC's seen in her sterile urine. Urine pregnancy test was negative. Blood work including kidney profile were all normal. Ultrasound scan revealed obstructive 7.8 mm calculus close to the anatomical site

of left VUJ (Figure 1). Subsequent plain CT KUB scan emphasize the presence of the calculus impacted at the same described location. In addition, a bilateral non obstructive tiny renal gravels (3-4 mm each) were also seen (Figure 2). Subsequently left ureteroscopy was suggested, therefore informed written consent was taken. Her bowel evacuated with low rectal enema. Eventually smooth rigid left ureteroscopy was done through the normally appearing left VUJ orifice. Surprisingly the previously described stone was not seen in the scoped limb of the duplicated ureter and considered as missing. This raised a high index of suspicion for ureteric anomaly. Therefore, a thorough cystoscopy guided us to a pinpoint, with small caliper, congested and edematous another orifice 1 cm inferior and medial to the normally located orifice (weigert-Mayer law applies) (Figure 3). This second left VUJ orifice was poorly compliant and negotiated with difficulty. Ultimately it led us to the second limb of the duplicated left ureter with an impacted stone encountered inside and just close to the ureteric orifice of this limb (Figure 4). The calculus broken with holmium laser and the fragments were removed with complete clearance. The two limbs stented separately with a ureteric catheter F5 for a day (Figure 5). The recovery was uneventful. The catheters were removed next day, and she discharged after spontaneous void.

Discussion

Ureteral duplication usually is asymptomatic, and its diagnosis is incidental. It can be associated with urinary tract infection, vesicoureteric reflux, ureterocele, stone formation, obstructive uropathy, ectopic ureter and other congenital complications [3-7]. In such cases renal morphology, ureteral condition and renal function warranted to be evaluated. The initial evaluation of a patient with a suspected obstructive stone usually occur in the emergency room (ER). As a gold standard in

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Received August 17, 2019; **Accepted** August 27, 2019; **Published** September 03, 2019

Citation: Shawish FA, Shawish WA (2019) Obstructing Calculus in a Single Limb of a Duplicated Ureter: A Recall of the Surgical Anatomy and the Variable Ureter Anomalies. J Clin Case Rep 9: 1271.

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Figure 1: 7.8 mm left VUJ stone.

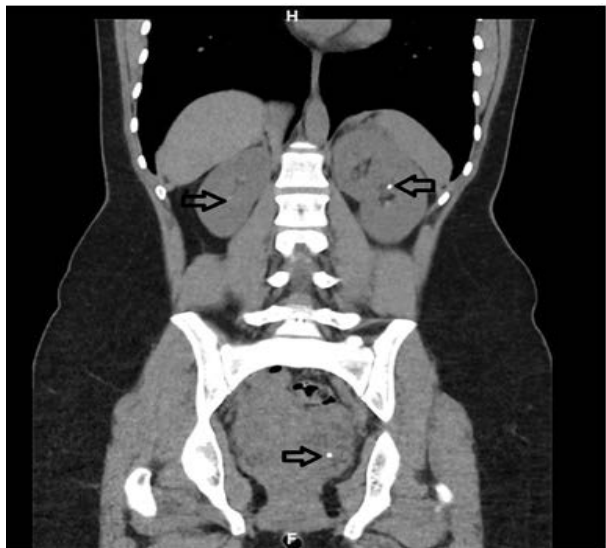


Figure 2: Left VUJ 7.8 mm stone.

the clinical practice currently this is limited to unenhanced CT KUB scan [12]. However, CT scan is associated with hazards of radiation and high cost [12]. Nevertheless, American Urological Association (AUA) and American College of Radiology (ACR), recommends the non-contrast CT KUB scan as a first line investigation for all of the adult patients presenting with symptoms suggesting of obstructive stones [13]. It has the highest sensitivity (94-97%) and the highest specificity (96-100%) and consistently provide the most accurate diagnosis [12,13]. In addition Hounsfield unit can be measured if ESWL is considered [7,14]. On the other side European Association of Urology

(EAU) differ regarding the optimal initial imaging modal, considering the ultrasound KUB scan as the first line of investigation for similar conditions [13]. Ultrasound scan has 40% sensitivity and 84% specificity [12]. However, all the main guideline providers (AUA, ACR and EAU) agreed on the ultrasound KUB scan to be the first line of investigation in pregnant women and paediatric patients [13]. Currently the sensitivity and specificity of standard KUB radiography estimated to be 57% and 76% respectively [13]. In a combined study for ultrasound scan and KUB radiography a wide variation exists in the sensitivity and specificity. It estimates the combined sensitivity and specificity with 58% to 100% and 37% to 100% respectively. However, none of the main bodies providing guidance recommend the contrast material to be used as a first line investigation in a patient with obstructive ureteric stones. Therefore the surgical anatomy of the aberrant ureter without contrast may not be visualized and hence any duplication of the ureter mostly will not be illustrated prior to the surgical procedure. So, in most of the patients with impacted ureteric stone of the duplicated ureter, similar to our case, most of the anomalies can be detected only in the theatre

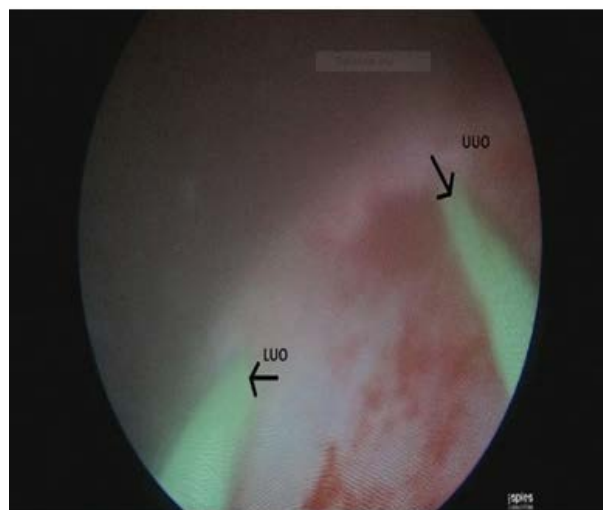


Figure 3: LUO: Left lower ureteric orifice; UJO: Left upper ureteric orifice with separate guide wire in.



Figure 4: 7.8 mm stone at VUJ of one limb of duplicated left ureter.



Figure 5: Two ureteric catheters F5 in each limb of duplicated left ureter.

during the procedure of ureteroscopic stone extraction. In such cases there is a high possibility for the ureteric anomaly to be missed and hence the calculus will not be accessed [8]. Indeed, in our search in the English medical literature many of the treated patients of the impacted calculus in a duplicated ureter in a single limb or in both limbs patient required a subsequent definitive further treatment [7-15]. Usually this may prolong the hospital stay and consequently increase the cost of treatment.

Conclusion

Further it may subject the patient to a possible but avoidable complication. Therefore, a solid knowledge of the surgical anatomy and the awareness of the congenital ureteric anomaly particularly with complete duplicated ureter might change the pitfall to a successful

surgery with stone removal and in a single session. Recall of the congenital anomalies of a duplicated ureter prior to the surgery of an obstructing calculus is crucial to reduce both morbidity rate and the cost of the treatment.

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