

Research article

Duplex kidney with calculus obstruction of lower moiety lessons learnt

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ABSTRACT

Duplex kidneys are one of the commonest congenital malformations of the urogenital system that one comes across in urological practice. It can be complete or incomplete. Ureteral duplication is usually asymptomatic but can accompany by urinary infections, ureterocele, vesicoureteral reflux or other congenital complications. The presence of stone in one of the systems may sometimes pose a therapeutic challenge to the urologists, especially when they attempt to remove the ureteric stone endoscopically. We report a 33-year-female who presented with acute left ureteric colic. Axial imaging revealed a large distal ureteric calculus. During ureteroscopy, no calculi were seen in the entire course of ureter upto renal pelvis. Screening with an image intensifier revealed a stone adjacent to the line of ureteroscope, raising suspicion of a duplex system or radio-opaque shadow outside the lumen of ureter. On further evaluation, a mucosal elevation with oedema was seen above and lateral to the ureteric orifice, raising suspicion of complete duplication of the ureter with pseudo ureterocele. However, no other ureteric orifice was observed. Deroofing of the edematous mucosa revealed the underlying large ureteric calculus. Retrograde pyelogram confirmed a duplex system and two ureters with stone in the upper moiety. Our report highlights the unusual presentation and the hard lessons learnt. It is prudent to search for accessory ureteric orifice especially when stone is not seen during ureteroscopy but seen under image intensifier.

Keywords: Duplex Kidney, Urolithiasis, Hydronephrosis, Ureterocele.

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INTRODUCTION

Duplex kidneys are congenital malformations of the urogenital system. The incidence is reported to be approximately 1–3% [1]. Duplex kidneys can be classified as complete or incomplete according to ureteral morphology [2]. Meanwhile, such duplications also can be unilateral or bilateral. Bilateral duplex systems are reported in 0.3% by excretory urography [3].

The cause of duplication is an incomplete fusion of the two poles [4]. Females are more commonly affected [5]. Ureteral duplications are asymptomatic and identified accidentally but can accompany by urinary infections, stone, ureterocele, vesicoureteral reflux or other congenital complications. Partial urinary stasis causes occurrence of stones, but different irrelevant factors to ureteral duplication may also influence it.

Junction and an upper moiety that is dysplastic and pseudo ureterocele around both orifices that were treated by ureterocele excision/deroofing and ureteroreno scopy and laser lithotripsy of giant calculi.

CASE PRESENTATION

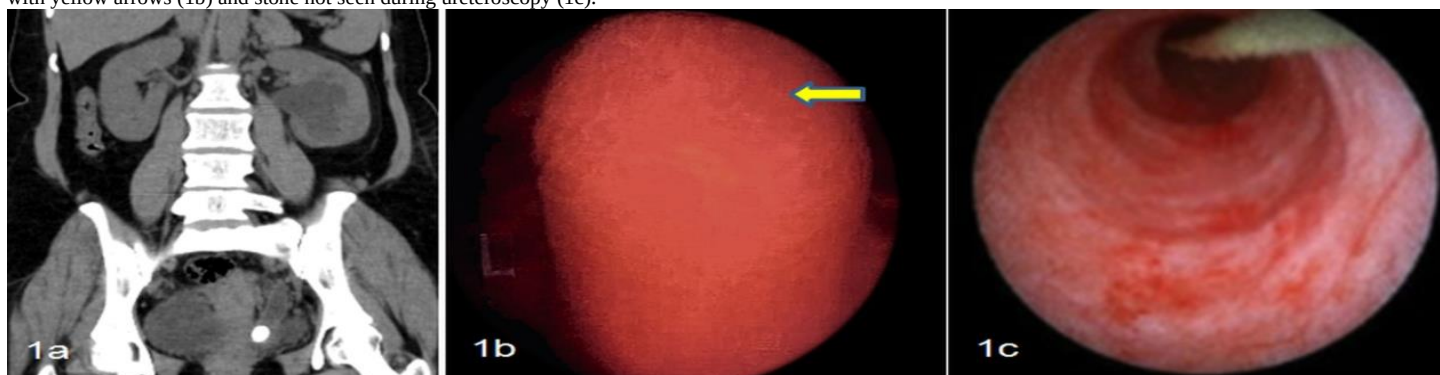
A 33-year-old woman was suffering from left loin pain for more than three weeks. The pain was colicky and aggravated for a

week. She also had nausea for 1 week but no febrile episodes. On examination, the left renal angle was tender. Renal parameters, absolute leukocyte counts were normal. Urine microscopy revealed micro-hematuria of 8 to 10 RBCs in each high power. Computed tomography (CT) scan demonstrated a 2 cm giant calculus in the left distal ureter (Figure 1a).

Patient was planned for ureteroscopy and stone fragmentation using Holmium Laser. Cystoscopy showed large mucosal protrusion in and around the left ureteric orifice (Figure 1b). However, during ureteroscopy, no stone was seen (Figure 1c).

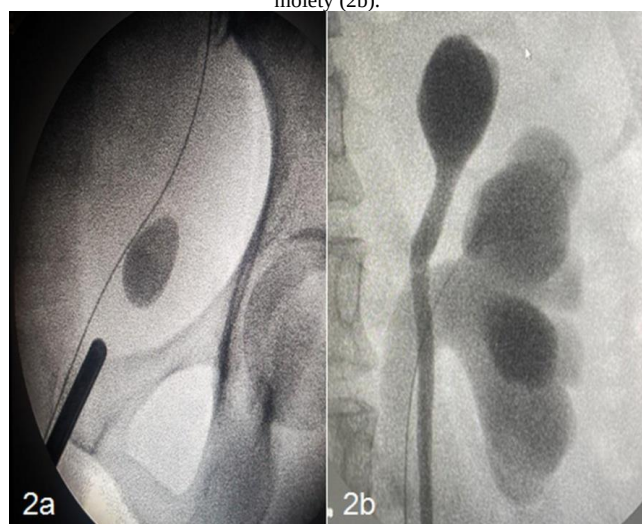
Imaging with an image intensifier revealed a big radio-opaque shadow adjacent to the ureteroscope (Figure 2a), suggesting a duplicated system with stone in the adjacent moiety. Repeat Cystoscopy did not reveal the second orifice, but demonstrated only a large mucosal protrusion, suggesting a pseudo-ureterocele. Deroofing of the pseudo-ureterocele demonstrated the large stone in left vesico ureteric junction, suggesting a complete duplication of left system. Retrograde ureterogram (RGU) confirmed the duplex system, with a dysplastic upper portion of kidney and a stone in lower moiety ureter (Figure 2b).

Figure 1: CT and endoscopic vision of stone in left ureter. CT scan (KUB region) showing the large left distal ureteric calculus (1a). The pseudo-ureterocele is marked with yellow arrows (1b) and stone not seen during ureteroscopy (1c).



We report a unilateral complete left ureteral duplication with obstructing giant calculi in lower moiety at the vesicoureteric

Figure 2: Intra-operative fluoro images. Fluoroscopic image showing the guidewire in upper moiety while the stone adjacent to it (2a). RGU image showing the attenuated upper moiety and the hydroureteronephrosis of lower moiety (2b).



The stone was levered out from the ureter into the bladder and removed in toto after laser lithotripsy. Double J stent was left in the lower moiety and removed after 14 days.

RESULTS AND DISCUSSION

Duplex kidneys are anatomical malformations where the renal units are made up of two distinct and discreet pelvicalyceal systems [6]. The renal vasculature, collecting and draining systems are separate. The overall occurrence of duplicate kidney and ureter is about 1~3% [7]. Duplications occurring on one side has been reported at 0.8% in American autopsies. Bilateral occurrences are rare and seen in 20–40% of all ureteral duplications [8].

Duplex kidneys are classified into complete and incomplete. Complete duplex systems involve two distinctive pelvicalyceal systems that arise from separate ureteric buds of the wolffian duct [9]. The resulting two separate ureters fuse to the adjoining mesenchyme and cause isolated and independent drainage of the ipsilateral kidney. These two ureters were identified as lower or upper moiety ureters according to their corresponding locations. The locations of ureteric orifices follow Weigert–Meyer law [10]. The

ureter of upper moiety opens below and medial to that from the one that drains the lower moiety [11]. This knowledge is of paramount importance especially when one performs ureteroscopy in duplex systems.

The clinical presentations of ureteral duplication are various and age-related. Most patients with duplex kidneys have no significant symptoms, but complications are relatively frequent [12]. The most common clinical presentation of ureteral duplication is recurrent UTI in children and VUR, ureterocele and obstruction in adults. Stone formation is a potential complication in such patients. Ultrasound can, many a time, miss a complete ureteral duplication without obstruction. Unequal hydronephrosis strongly raises suspicion of complete duplication. Excretory urogram (EU) is very helpful in differentiating the types of duplications. EU gives an indirect assessment of renal function too but spiral CT urogram and magnetic resonance imaging (MRI) much better can reveal the locations of ureteral orifices.

As patients with the stone disease are taken up for a definitive procedure after a non-enhanced CT scan, a pre-operative diagnosis of this congenital anomaly is missed out more often. In most instances, a duplex system is diagnosed on the table. During ureteroscopy, when the stone is not identifiable during routine ureteroscopy, it is prudent to look for another ureter harboring the stone.

Our report illustrates a rare unilateral left complete ureteral duplication with the large stone obstructing lower moiety drainage. The call for for a thorough knowledge of this congenital anatomical anomaly and a proper judgment of manifestations of a duplex system while performing ureteroscopy are the lessons to be learnt by every aspiring urologist and trainee while performing endo-urological procedures. Moreover, as most centers perform ureteroscopy after an unenhanced Computed Tomography scan, such congenital anomalies may be overlooked, posing a challenge intra-operatively. Surgeons and urologists should bear in mind such congenital anomalies, especially when they come across large volume stones. In such instances, it is sensible to do a contrast-enhanced CT scan to

circumvent such intra-operative surprises.

CONCLUSIONS

Duplex systems are common anomalies observed in urology practice. Stones obstructing the duplicated ureter are not so uncommon. One should show a constant vigil to make an intra-operative diagnosis of such congenital anomalies. It is practical to suspect a duplex kidney, especially when large distal ureteric calculi are identified under image intensifier but stones are not visualized during ureteroscopy.

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