JOURNAL OF
Endoluminal Endourology

CASE REPORT
DOI:10.22374/jeleu.v6.i1.154

INVERTED Y URETERAL DUPLICATION WITH DISTAL OBSTRUCTING CALCULI IN THE ECTOPIC URETER IN THE PROSTATIC URETHRA AND DUPLEX KIDNEY

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Abstract

Duplication of the ureter and renal pelvis is a relatively common congenital anomaly with an incidence of approximately 1 in 150 births. Different anatomical variations of ureteric duplication exist. Ectopic ureters are usually associated with ureteral duplication and a duplex collecting system. Duplication of the ureter is more common unilaterally, has a female preponderance, and occurs more commonly on the left side.
We present the case of a 58-year-old male with inverted Y ureteral duplication who had distal obstructing ureteric calculi in the ectopic distal ureteral opening in the prostatic urethra.

Keywords: *duplex kidney, ureter duplication, inverted Y ureterm, ectopic ureter, urinary calculi, prostatic urethera*

INTRODUCTION

Embryologically, the development of the kidney occurs when the ureteric bud originates as an unbranched diverticulum from the distal part of the mesonephric duct. It grows cranially and invades the metanephros to form the primitive kidney and collecting system.

In the case of duplex ureter, incomplete ureteric duplication occurs when the ureteric bud bifurcates, and complete ureteric duplication occurs when two separate ureteric buds arise.

The ureteric bud that inserts more cranially into the metanephros will drain the upper moiety. This ureteric bud will have arisen from a more caudal position on the mesonephric duct, and as the bladder develops, it will maintain its lower position.[1]

This is the ethos of the Weigert-Meyer principle which dictates that the upper moiety will insert lower and more medially into the bladder. The upper pole ureter often obstructs, whereas the lower pole is more susceptible to reflux.[2]

Incomplete ureteric duplication forms a Y shape, which can have two conformations. The incomplete Y ureteral conformation describes two ureters with separate insertions in the kidney, and fuse distally to form a single ureter inserting into the bladder at the usual anatomical location in the trigone. Inverted Y ureteral conformation describes a single ureter originating at the renal pelvis that bifurcates distally, with one ureter inserting ectopically.

Sites of insertion of an ectopic ureter occur in the urethra or vagina in a female (resulting in incontinence and thus earlier presentation). In males, insertion sites include the posterior urethra, seminal vesicle, or ejaculatory duct.[3] Males do not present with incontinence as the ectopic ureter always inserts proximally to the external urethral sphincter.

CASE

The patient initially presented with recurrent visible haematuria in 2019. He was subsequently investigated with flexible cystoscopy and computed

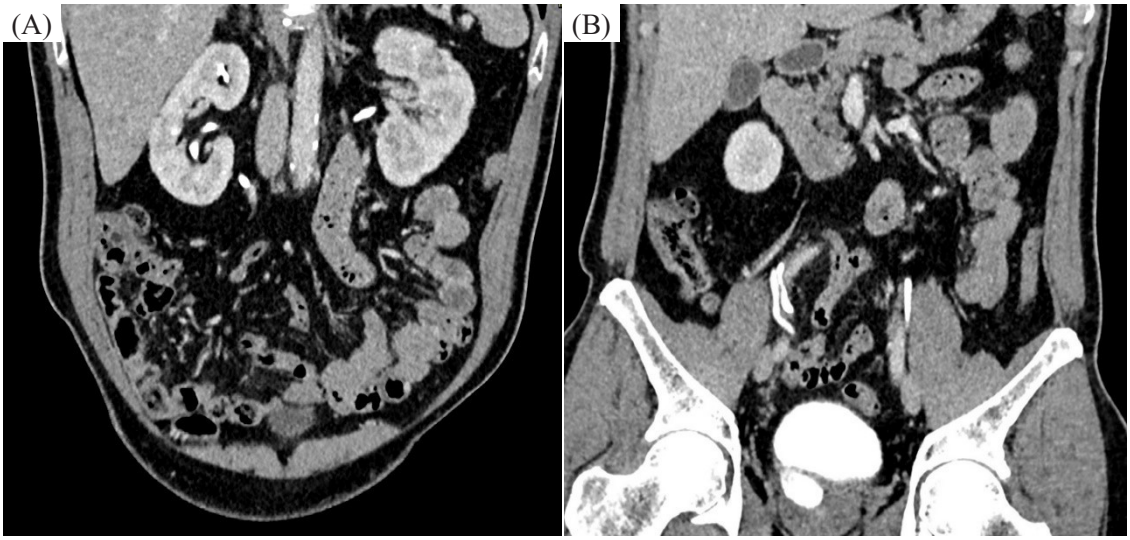

FIGURE 1 (A) Contrast CT imaging showing duplex kidney with contrast in upper and lower moieties draining via a single ureter and (B) inverted Y ureteral duplication with contrast drained into bladder and prostatic ureter via ectopic ureterocele.

tomography (CT) urogram. The flexible cystoscopy did not reveal any abnormalities, and both ureteric orifices were visualized. The CT urogram performed at the time was reported as a complete duplex system with the upper pole moiety draining ectopically into the prostatic urethra. The upper pole moiety was reported to be associated with a ureterocele, within which a 7 mm distal stone was visualized. The CT images are demonstrated in Figure 1.

The patient did not attend a further follow-up appointment and was not seen again until the following year when he presented acutely with left loin pain. This time he was investigated with *non-contrast* imaging.

The CT KUB reported a prominent right distal ureter, possibly representing a ureterocele. What was previously reported as a 7mm calculus in the ectopic ureter, was now reported as two large areas of calcification within the prostate. It also showed left renal calculi in the lower pole. The patient continued to miss follow-up appointments after this presentation though he still had persistent loin pain. Further repeat imaging studies performed were also non-contrast studies, and the scans were reported by different radiologists at each attendance. These non-contrast scans were reported as not showing a duplex system or duplication.

Eventually, the patient underwent right rigid ureteroscopy and retrograde studies in October 2020. On ureteroscopy, no stones were identified in the right ureter, and retrograde studies, as demonstrated in Figure 2 did not show any filling

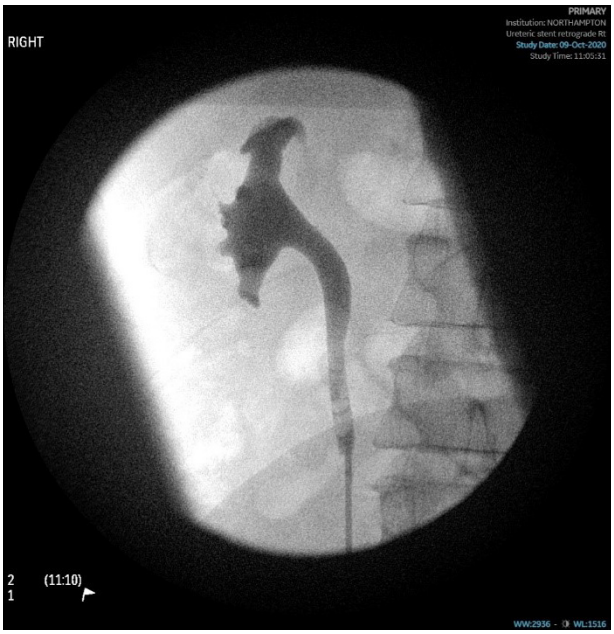

FIGURE 2 Right retrograde study performed in October 2020.

defect, hydronephrosis, or any evidence of duplex system. Therefore, it was assumed the patient had passed his right distal stones. This information was updated on the patient's letters for future reference and was in keeping with the non-contrast imaging studies performed prior to operative intervention.

Despite this, the patient continued to suffer from bilateral loin pain and was re-imaged in June 2021 with a CT KUB. This revealed the right distal ureteric stones were still present despite not being identified during the ureteroscopy. They had also increased in size and a right renal stone had also formed.

Taking into account the persistence and increase in size of the distal ureteric stones, it was suspected the patient had a duplex system, as diagnosed on his initial imaging in 2019, and the stones were present in the ectopic ureter, which opened into the prostatic urethra. This was associated with a ureterocele.

As a result, he was counseled about all potential approaches in planning the patient's definitive surgical procedure. The retrograde approach would involve identifying the ectopic ureteral opening in the urethra. He was informed as the ectopic ureteral opening could be covered by mucosa, resection of the bladder base, prostate or urethra may be necessitated. However, if this failed, we could also attempt an antegrade approach.

The procedure took place in March 2022 and on initial cystoscopy bilateral ureteric orifices were identified at the trigone, and a right retrograde study was performed which did not reveal any calculi. Furthermore, the prostate and prostatic urethra were carefully inspected for any ureteral openings, but none were identified. Therefore, with the Weigert-Meyer principle in mind, we decided to resect the prostatic urethra to identify the ectopic ureteral opening.

Using a standard 24French unipolar resectoscope, the prostatic urethra was resected, and almost immediately, the ectopic ureteral orifice was revealed, containing multiple large calculi in a patulous ureter. The calculi were extracted with a stone crusher, and a urethral catheter was left in place post-operatively.

The patient had a CT KUB 4 months later, which showed the three VUJ calculi were no longer present, and the right kidney was of normal size with no hydronephrosis. He subsequently attended for a flexible cystoscopy, and we could clearly see the ectopic ureteral opening in the resected prostatic urethra and the anatomical ureteric orifice in the

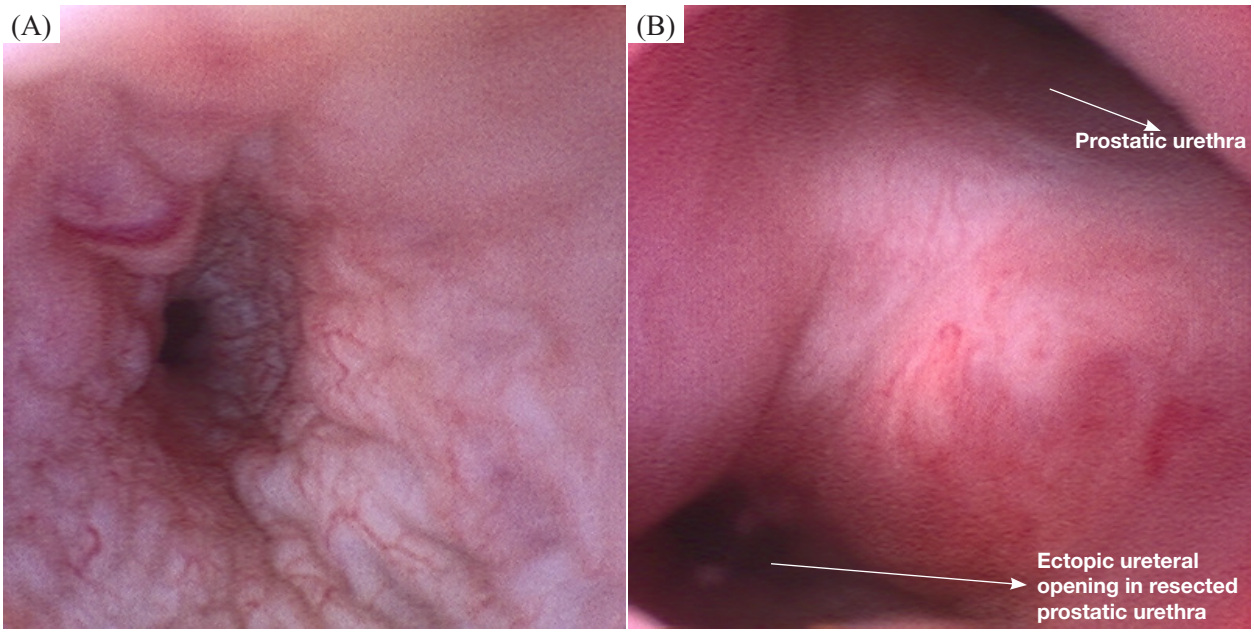

FIGURE 3 (A) Ectopic ureteral opening in resected prostatic urethra and (B) ectopic ureteral opening below and prostatic urethra above.

bladder at the trigone (Figure 3). At this time, he had no further pain or other urological symptoms.

DISCUSSION

Inverted Y ureteral duplication is the least frequently encountered duplication malformation in the urinary tract, and is much more common in females. There are few reported cases in the literature, and diagnosis is a challenge. The symptoms are variable and non-specific, and the key factors in making the correct diagnosis are a high suspicion index and an appropriate imaging modality.

Our patient presented on multiple occasions with urological symptoms of loin pain and visible haematuria and despite imaging on multiple separate attendances, an accurate diagnosis was not made for at least 18 months after the initial presentation. Even after undergoing ureteroscopy for the first time, the patient was misdiagnosed.

For these cases, imaging is paramount in making an accurate diagnosis. The patient's initial CT urogram in 2019 reported a complete duplex system, a *eutopic* ureter draining the lower pole moiety at the vesicoureteric junction and an *ectopic* ureterocele draining the upper pole moiety into the prostatic urethra. Whilst correctly identifying the duplex system, the inverted Y ureteral malformation was not picked up on this contrast study. Despite this, subsequent *non-contrast* CT images incorrectly report that the patient does not have duplex kidney/ ectopic ureter and even report the VUJ stones as prostatic calcification.

The patient's non-attendances to follow-up appointments were certainly a contributory factor to misdiagnosis. Though he presented multiple times with pain and imaging confirming the presence of stones, it was difficult to plan further investigation and treatment beyond these acute attendances. In addition, because the patient did not speak English and relied on his children to translate for him, it may have been difficult for him to receive or interpret correspondence to attend future appointments.

Indeed, a letter on our system shows that after multiple non-attendances, our consultant had appealed to the patient's GP to make contact with the patient to urge him to attend for treatment of his stones.

At each acute attendance the patient had to be investigated de novo, and with a likely differential diagnosis of stones, the patient had multiple non-contrast CTs. Though this is the gold standard of diagnosis for calculi, it is not the most appropriate form of imaging to identify duplex system and/or ectopic ureter. Furthermore, at each new presentation, imaging was reviewed by different radiologists.

In fact, radiology proved to be the main confounding factor in reaching the final diagnosis. The initial CT urogram performed in 2019 correctly identified the presence of a duplex kidney and ectopic ureter, with the opening into the prostatic urethra. However, all subsequent scans were non-contrast imaging, and therefore it is much more difficult to trace the ureter and delineate the point of bifurcation. The first non-contrast scan performed after the CT urogram contradicts this initial scan's findings, and reports no duplex system, no ectopic ureter, and reports the distal ureteric calculi as prostatic calcification.

It is crucial, therefore, that where duplex kidney and/ or ureteric duplication is suspected, contrast imaging of the urinary tract is obtained. Contrast imaging is likely to be the gold standard imaging modality for diagnosing a duplex system and ureteric malformations. The final diagnosis became evident after a retrospective review of the images post- operatively by a uro-radiologist.

Furthermore, operative studies alone may not be sufficient in making a diagnosis and also contributed to misdiagnosis in this case. The retrograde images obtained during the patient's first ureteroscopy suggest no ureteric malformation or duplex system. This was in line with the patient's non-contrast imaging prior to the procedure. Though rigid ureteroscopy was performed, direct visualization of the flexible ureteroscopy may have revealed the elusive diagnosis all along.

Furthermore, as the condition is rare, there is no standardised management. Ours is the first reported case of multiple urinary calculi in an ectopic ureter with inverted Y ureter malformation opening into the prostatic urethra.

Ye WX and Ren LG reported a case in February 2022 of multiple urinary calculi in an ectopic ureter with inverted Y ureter duplication. However, the ectopic ureter was *blind-ending,* and as such, the calculi were accessed via flexible ureteroscopy and were treated with transurethral ureteral holmium laser lithotripsy.[3]

In our case, due to the ectopic ureter opening into the prostatic urethra we were able to resect this part of the urethra which revealed a wide, patulous distal ureter with multiple calculi. Unfortunately, due to the calculi's size, retrieval proved difficult. We initially attempted to remove them with cold cup biopsy forceps, but these did not provide sufficient grip and could not close all the way around the calculi due to their large diameter. Eventually a Mauermayer Stone Punch was successfully utilized to remove the calculi. However, caution was used to avoid damaging the urethral tissue or sphincter.

We inserted a urethral catheter immediately post-operatively but again encountered difficulty due to the widely patent resected prostatic urethra. Eventually, the catheter was inserted over an optical urethrotome.

Post-operative CT imaging did not reveal any further calculi and flexible cystoscopy 4 months later showed the resected ureteral was still open and draining. The patient was asymptomatic following this procedure.

This method of managing ureteric calculi in an ectopic ureter is less complex and has fewer associated risks than that described by Ye and Ren. By resecting the prostatic urethra, the obstruction to drainage in the ectopic ureter was permanently cleared as seen in the flexible cystoscopy images. The resected ureteric orifice has a greater diameter than the anatomical ureteric orifice. Though long-term follow-up is required to know for sure, this potentially provides a pathway for stones to drain more easily, or may even reduce the precipitation and formation of stones in the urinary tract altogether.

CONCLUSION

Inverted Y ureteral malformation is a rare ureteric duplication abnormality for which the pathophysiology is unclear. It often occurs in isolation, rather than with an associated duplex kidney; our case is the first known case in the literature to show inverted Y ureteral malformation with ectopic prostatic ureteral opening and distal calculi.

Due to its rarity and few described cases in the literature, diagnosis is difficult and should be suspected when symptoms persist after intervention and discrepancies on serial imaging. Contrast imaging is the most useful imaging modality when duplex systems or duplications are suspected. For ectopic ureters that open into the urethra, resection may be the least invasive and most effective treatment of calculi.

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