

Case Report

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Duplex Moiety Kidney With Ureteral Ectopia; A Case Series

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Introduction

An ectopic ureter is characterized by an orifice that is outside the posterolateral extremity of the bladder trigone [1]. They are associated with either duplex kidney or less commonly a single system (20-25%) [2,3]. They are usually asymptomatic or detected incidentally in males. The diagnosis is complex due to variations in the anatomy of the ectopic ureter. Therefore, we would like to present a series of three cases of ectopic ureters and discussing their management.

Case Report

Case 1: A 6 year-old girl presented with incontinence. Examination had normal findings. CT scan-Intravenous pyelography (CT-IVP) showed left duplex kidney with upper moiety ureter opening ectopically in the anterior vaginal wall (Fig. 1-2). Voiding cystourethrography (VCUG) was unremarkable. There was adequate

Ureteral ectopia is a common condition presenting as incontinence in females and incidentally detected in males. The diagnosis requires thorough radiological investigations to delineate the anatomy and formulate the treatment. The prognosis is generally excellent. We presented a series of three cases of ureteral ectopia and discussion regarding their management.

Keywords: Female; Urinary Incontinence; Ectopic Ureter; Duplex Moiety Kidney.

Running Title: Duplex moiety kidney with ureteral ectopia

function in the both kidneys on Renal Scan (separate function of upper and lower moiety was not performed).

On examination under general anesthesia, a distended tubular structure was seen on the left anterior vaginal wall; however, the opening could not be visualized. A left end to side ureteroureterostomy was performed (Fig. 3), where the adequately functioning upper moiety ureter was anastomosed to the side of the lower moiety ureter.

Case 2: A 6 year-old female presented with continuous dampness of undergarments. Clinical examination had normal findings. Ultrasonography was suggestive of right complete duplex kidney with hydronephrosis of the upper moiety. VCUG had normal results. On CT-IVP, right complete duplex kidney with hydronephrosis of the upper moiety and hydroureter was seen (Fig. 4-5). The ectopic ureter was seen opening in the

upper part of the vagina. Renal scan showed 46.6% function in the left kidney with 53.4% function in the right kidney. Of this the upper moiety contributed to 12% function on the right, which was 6.4% of the total function. Cystogenitoscopy showed impression along the anterior wall of the vagina with flow of urine. Considering the above findings, right upper moiety heminephrectomy was performed (Fig. 6).

Case 3: A 5 year-old female presented with dribbling of urine since birth. General examination and local examination had normal findings. Ultrasonography (USG) showed Duplex moiety on the right side with moderate hydronephrosis in upper moiety and mild hydronephrosis in lower moiety. Left kidney was not visualized. VCUG had normal results. Renal scan showed 15.52% function in the upper moiety and 84.48% function in the lower moiety. Magnetic resonance urography (MRU) showed left crossed fused kidney with its ureter draining normally in the bladder on the left side (Fig. 7-8). However, the right ureter had an ectopic opening. On Cystogenitoscopy the left ectopic ureter was visualized at its normal site in the bladder. Opening of the right ureter was not seen in the bladder or vagina. At surgery the right ectopic ureter was anastomosed to the pelvis of normally draining left crossed fused kidney (Fig. 9). All three patients had an uneventful postoperative recovery.

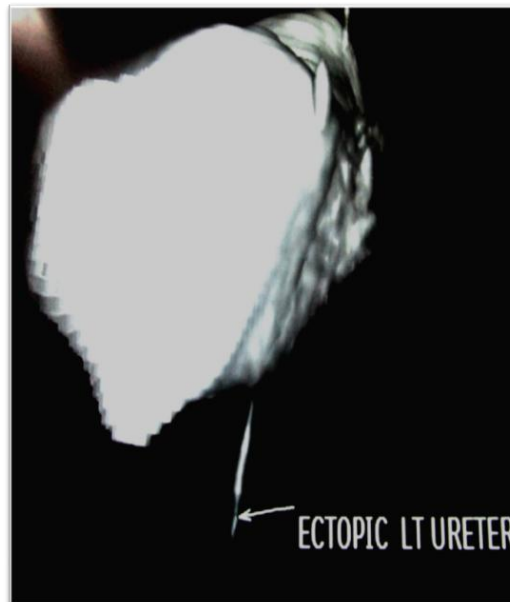


Figure 2. CT IVP showing the left ectopic ureter

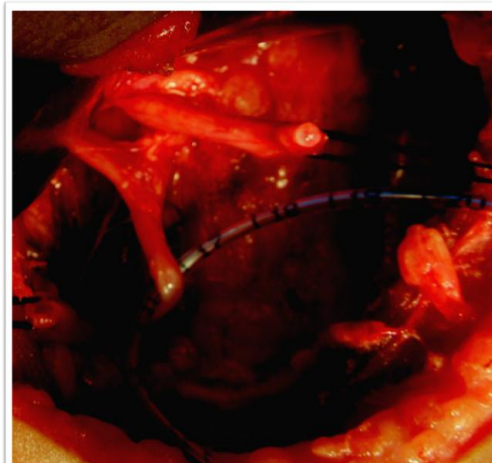


Figure 3. Intra-operative picture of transected upper moiety ureter



Figure 1. CT IVP showing left duplex moiety kidney

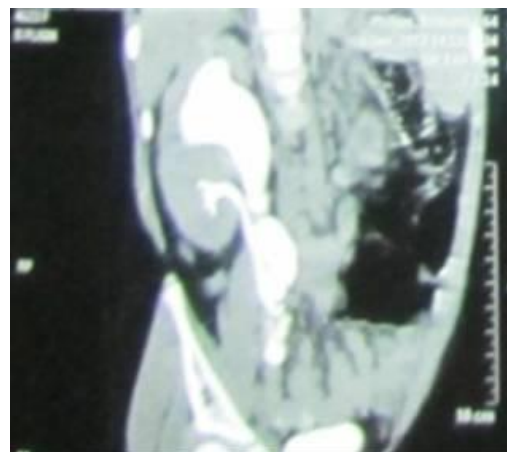


Figure 4. CT IVP showing right duplex moiety



Figure 5. CT IVP reconstruction showing ectopic ureter alongside normal ureter

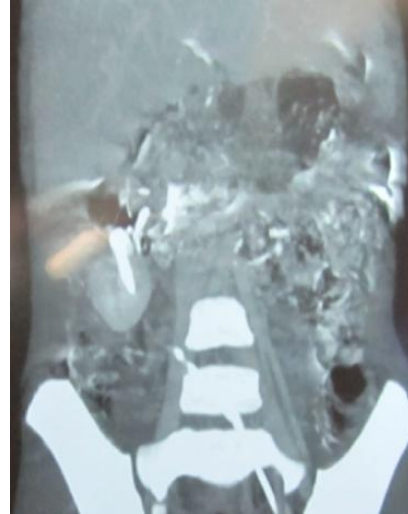


Figure 8. MRU showing left ureter crossing to drain in the bladder



Figure 6. Intra-operative image showing both Duplex moieties

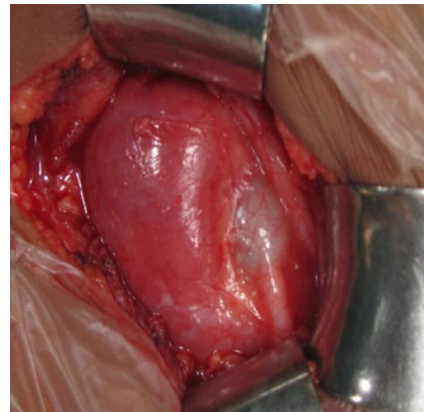


Figure 9. Intra-operative image showing dilated right renal pelvis with ureter along with left crossed fused ureter.



Figure 7. MRU showing both kidneys on right side

Discussion

Ureteral duplication is the most common urinary tract abnormality, occurring in approximately 1% of the population [4].

Ureteral ectopia in the bladder is usually detected incidentally and warrants no further treatment. However in toilet trained girls with normal voiding pattern constant dribbling of urine is indicative of ectopic ureteric opening [5]. In contrast, boys rarely present with incontinence. This is because the ureteral bud is a Wolffian structure and would not get inserted directly into the mullerian duct structures, rather it courses alongside these structures resulting in various ectopic sites for its opening namely urethra below the neck, vagina or perineum. Because the Wolffian duct is not associated with any structures

below the pelvic floor in males, they do not present with dribbling of urine [6]. Furthermore, the relationship between ureteric openings of the duplex moieties was put forth by Carl Weigert and Meyer, popularly known as the Weigert-Meyer rule, which says that the ectopic ureter or the ureterocele associated with the upper moiety is caudal to the ureteral opening of the lower moiety [7]. The ectopic ureter is associated with dysplasia of the draining renal unit [8]. According to the Stephens hypothesis, the further the ectopic orifice located from the trigone, more severe is the dysplasia [9].

As discussed earlier, asymptomatic duplication can be left alone. Complex duplications warrant a thorough history taking and evaluation. Although USG and CT-IVU suffice to delineate the anatomy in most cases, MRU is the investigation of choice, limited by its cost and availability [10]. Because ureteral ectopia is associated with VUR, VCUg should be performed in every case [11]. However in all cases discussed above, there was no VUR. Renal scans are imperative to know the function of individual moieties as well the kidneys to plan surgical management and assess the prognosis.

The management in these cases is complex and has to be tailored according to the individual case scenario. There are various surgical options that can be broadly divided as renal level surgeries, ureter level surgeries or bladder level surgeries. As previously discussed, ectopy with duplication is associated with dysplasia hence upper moiety nephrectomy is advocated [12], after confirming the function on renal scans. In case the function in the dysplastic moiety is considerable, then ureteropyelostomy should be preferred. The third case discussed above is interesting because ectopia was associated with cross fused kidney, which is rare [13].

Ureteral ectopy with duplication although common are complex entities, which require a detailed clinical evaluation. MRU is the preferred diagnostic modality. The surgical principles of management are to treat ectopic ureter while preserving the renal function. The prognosis in most cases is excellent.

Conflict of Interest

None declared

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