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CASE REPORT

UNDIAGNOSED URETEROVAGINAL FISTULA IN CONGENITAL DUPLEX KIDNEY WITH ECTOPIC URETERIC INSERTION TO URETHRA; SOMETHING TO LOOK FOR?

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ABSTRACT

Continuous urinary incontinence in young female with normal voiding pattern should prompt proper assessment for ectopic ureter.

We report a young female who presents with continuous urinary incontinence and malodourous urine. She was frequently treated as recurrent urinary tract infection and subsequently referred to our urology unit. Initial imaging revealed right duplex kidney with ectopic ureter, hence she had undergone right heminephrectomy. However her symptoms does not improve significantly. Further evaluation revealed uretero–urethral fistula and uretero–vaginal fistula. She then underwent urethroureteral and ureterovaginal fistula repair, which resolved her distressing urinary incontinence.

Keywords: *Ectopic ureter; urinary incontinence; female; urinary tract fistula*

INTRODUCTION

The term "ectopic ureter" denotes a ureter that inserts at or distal to the bladder neck. Ectopic ureters most commonly occur in females and often drain into upper moiety of a duplex kidney (1). In most females with an ectopic ureter, the ureter has its insertion either into genital tract or urethra (2). This leads to typical symptom of continuous urinary incontinence with an otherwise normal voiding pattern. Screening with ultrasonography and/or intravenous urography (IVU) could assist in confirming the affected system which likely to be upper pole moiety of a duplex collecting system (3,4). We report a case of young lady who presents with urinary incontinence since childhood and frequent urinary tract infection, then found out to have ectopic ureter with uretero–urethral fistula and uretero –vaginal fistula.

Case Presentation

We report a 31-year-old lady presented to us with a complaint of urinary incontinence since the age of 8, without any prior history of trauma or fall. She denied having bowel or neurological symptoms. She had frequent visits to the hospital and was treated as a recurrent urinary tract infection. Micturition cystourethrogram (MCUG) revealed right distal ureteric bulbous distension and right duplex kidney, with no demonstrable fistulous connection. Contrasted enhanced computed tomography (CT) of the abdomen/pelvis revealed complex right duplex kidney. Right upper moiety revealed hydronephrosis and hydroureter with possible ectopic ureteric insertion. Subsequently intravenous urography (IVU) revealed the poorly function of the right upper moiety with normal function of right lower moiety.

She had undergone right retrograde pyelogram with ureterorenoscope and right ureteric stenting, followed by right heminephrectomy and ureterectomy at the age of 24. Histopathological examination report revealed ureteritis in right upper moiety ureter and chronic pyelonephritis in right upper moiety nephrectomy.

Postoperatively she continued to complain of foul–smelling urine dribbling from the vagina that persisted for many years. Repeated MCUG revealed residual right ureteric stump with evidence of reflux from the urethra during micturition phase and likely urethrovaginal fistula (Figure 1).

Rigid cystoscopy and contrast study revealed urethrorectal fistula at 7 o'clock just distal to the bladder neck. There were 2 openings seen at 4 o'clock and 10 o'clock location in the ectopic ureter, contrast injection showed communication with the vagina (Figure 2A and 2B). On table retrograde urethrography done through the urethroureteral fistula revealed remaining right ectopic ureter up to mid part of ureter.

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At the age of 30-years, she was subjected to urethroureteral and ureterovaginal fistula repair by transvaginal approach.

Intraoperatively the opening of urethroureteral fistula was identified just below the bladder neck and separated from the normal urethra opening.

The fistula was ligated and closed with four layers followed by Martius Flap to prevent recurrence (Figure 3).

Postoperatively, she did not complain of urinary incontinence anymore during clinic follow up.

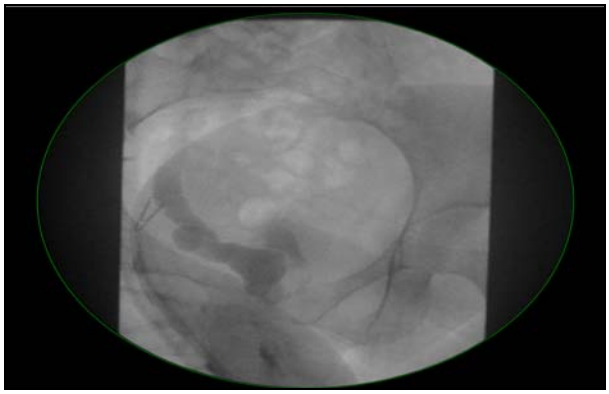

Figure 1: Repeated MCUG after initial surgery revealed reflux to the remnant of stump on the right side. The stump is seen arising from the urethra and extending superiorly into the lower border of sacral ala. Posteriorly, there is fistulous communication between the urethra and vagina with opacification of the vagina and uterine cavity.

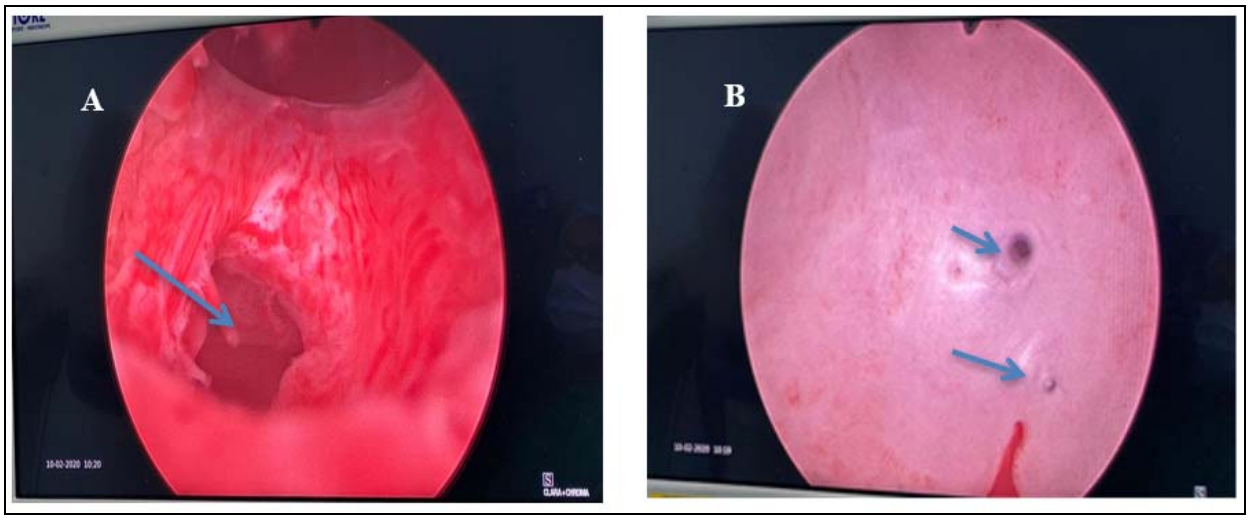

Figure 2 (A): Cystoscopy view showing (arrow) opening of right ectopic ureter below the bladder neck.

Figure 2 (B): Cystoscopy view showing image inside the right ectopic ureter, there is ureterovaginal fistula with 2 opening (arrow).

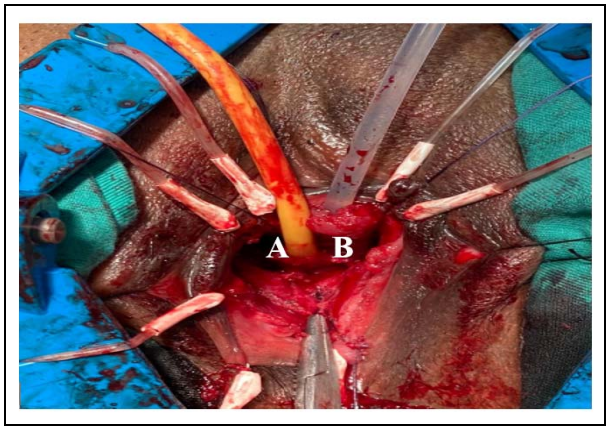

Figure 3: Intraoperative view: right ectopic ureter was separated from urethra and ligated, primary repair of urethra with Martius fat Flap done. (A)Ectopic ureter with urethroureteral fistula, (B)Normal urethra opening

DISCUSSION

An ectopic ureter is known as a ureter that abnormally place at or distal to the bladder neck, or outside of the urinary tract entirely. This is commonly due to embryological malformation, where the ureteric bud arises more cephalad than usual on the mesonephric duct, leading to distal ectopic ureteric insertion. The ureter remains attached to the mesonephric duct for longer, migrates more caudally than usual and inserts into either the urinary tract distal to the bladder or the genital tract (5).

The mesonephric duct remnant will turn into the epoophoron, oophoron and Gartner's duct in the female. When the ectopic ureteric bud included into the adjacent structures of paramesonephric duct origin, this will lead to urinary drainage into the female genital tract (1,5). Usual sites of ectopic ureter in the female include urethra (35%), vestibule (34%) and vagina (25%) (2), which is demonstrated in our case who was found to have urethra – ureter fistula and uretero vaginal fistula. She complained of continuous urinary incontinence due to the insertion of ectopic ureter distal to the urethral sphincter as well as into vagina.

The female with an ectopic ureter has continuous urinary incontinence with otherwise normal urinary pattern, often associated with abnormal urine odour which warrants a thorough assessment. They often suffer from wet and erythematous perineal rash due to persistent incontinence from an ectopic ureter. Detailed inspection may demonstrate urine leak from the vestibule and / or vaginal orifice (1).

The imaging studies should aim to recognize any additional poorly functioning renal parenchyma, particularly when initial imaging reported a "solitary" kidney. IVU is useful to demonstrate if duplex system is present, though occasionally required a delayed phase (6).

Studies had showed that laparoscopic nephroureterectomy is comparable to its open counterpart (6 – 8). Irrespective of the technique used, it is imperative to identify the ectopic / dysplastic kidney and / or ureter involved. Urinary incontinence can be cured with removal of poorly functioning ectopic kidney and its ectopic ureter.

Conclusions

An ectopic ureter assessment should be considered in females with continuous urinary incontinence with normal voiding patterns, irrespective of age. Imaging studies can be helpful, from initial ultrasonography and/or IVU, followed by detailed imaging such as CT or fluoroscopy.

Once the ectopic ureter and affected renal unit are identified, we can offer a cure for this disturbing symptom and provide a better quality of life in these affected patients.

Competing interests

There was no funding for the study and no conflicts of interest to disclose.

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