Persistent Incontinence Following Surgery for Ureteric Triplication With Contralateral Duplication—A Management Dilemma

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Ureteral triplication is a rare congenital anomaly of urinary tract with wide spectrum of presentation. The presentation with incontinence has usually been taken as a sign of ureteral ectopia in these cases. However, ectopia cannot explain all cases with incontinence. We report a case of a 14-year-old girl with ureteral triplication with contralateral duplication and vesicoureteral reflux of the upper moiety on the triplicate side and dysplastic upper moiety on duplicated side, who had persistent incontinence following a left ureteroureterostomy and right heminephrectomy. The clinical features of this rare case and the management of this peculiar postoperative situation is discussed herewith. UROLOGY 134: 221–224, 2019. © 2019 Elsevier Inc.

Ureteral triplication is a rare congenital anomaly with only 100 cases have been reported in the literature since the initial description by Wrany in 1870.[1] Pediatric cases are rare and it occurs in females more often.[2] Paucity of symptoms often lead to delayed or missed diagnosis.[3] The following case report discusses the management of a 14-year-old female with incontinence since birth who on evaluation was found to have ureteral triplication and contralateral duplication.

CASE REPORT

A 14-year-old female presented to our outpatient department with a history of continuous urinary incontinence since birth and left flank pain. She had undergone an intravenous pyelography which showed duplicated collecting system on the left side (Fig. 1A). On physical examination, the urethral meatus was found to be patulous with stress leak on coughing. We could not make out any vaginal pooling of urine. The patient was sent for urinalysis and a complete blood count which were essentially normal.

An ultrasound was performed in which a dilated upper moiety and gross hydroureter was detected on the left side with a dilated ureter on the right side. Computerized tomography urography and magnetic resonance urography showed a bilateral duplication of ureter. This was accompanied by a gross hydroureter of both the duplicated ureters leading to the upper pole moieties with the right-side upper moiety showing significant dysplasia and negligible excretion of contrast (Fig. 1B and C). While the left upper moiety ureter was draining into the bladder neck, the distal opening of the right upper moiety ureter could not be made out clearly. A radionucleotide isotope scan showed normal excreting kidneys with normal GFR and no obstruction on either side. Micturition cystourethrogram showed a left grade V vesicoureteral reflux in the ureter draining the left upper moiety which appeared to be opening separately in the bladder neck on delayed films (Fig. 1D).

A cystoscopy was performed which showed single ureteric orifice on right side and 2 ureteric orifices on the left side with one orthotopically placed and the other at the bladder neck. To our surprise, we also found another ureteric opening on the left side of the vestibule. On left side, all 3 ureteric orifices were catheterized (Fig. 2A) and retrograde pyelography (RGP) showed ureter opening at the bladder neck to be draining the dilated upper moiety and the orthotopically placed ureter to be draining the lower moiety. We could not identify the course of the ureter opening into the vestibule on RGP. So, we did a ureteroscopy through the bladder neck orifice and we found the ureteric catheter from the vestibule orifice entering the ureter from below and laterally. It was concluded that this case was likely to be a Smith Type-4 Triplication (Fig. 3). We were unable to identify the ectopic opening of the right upper moiety separately and in view of the negligible function of the right upper moiety, we decided to do a right heminephrectomy.

Guide wires were kept in all 3 orifices, and exploration through a left Gibson's incision revealed 2 separate ureters leading up to the bladder (Fig. 2B). We did not dissect

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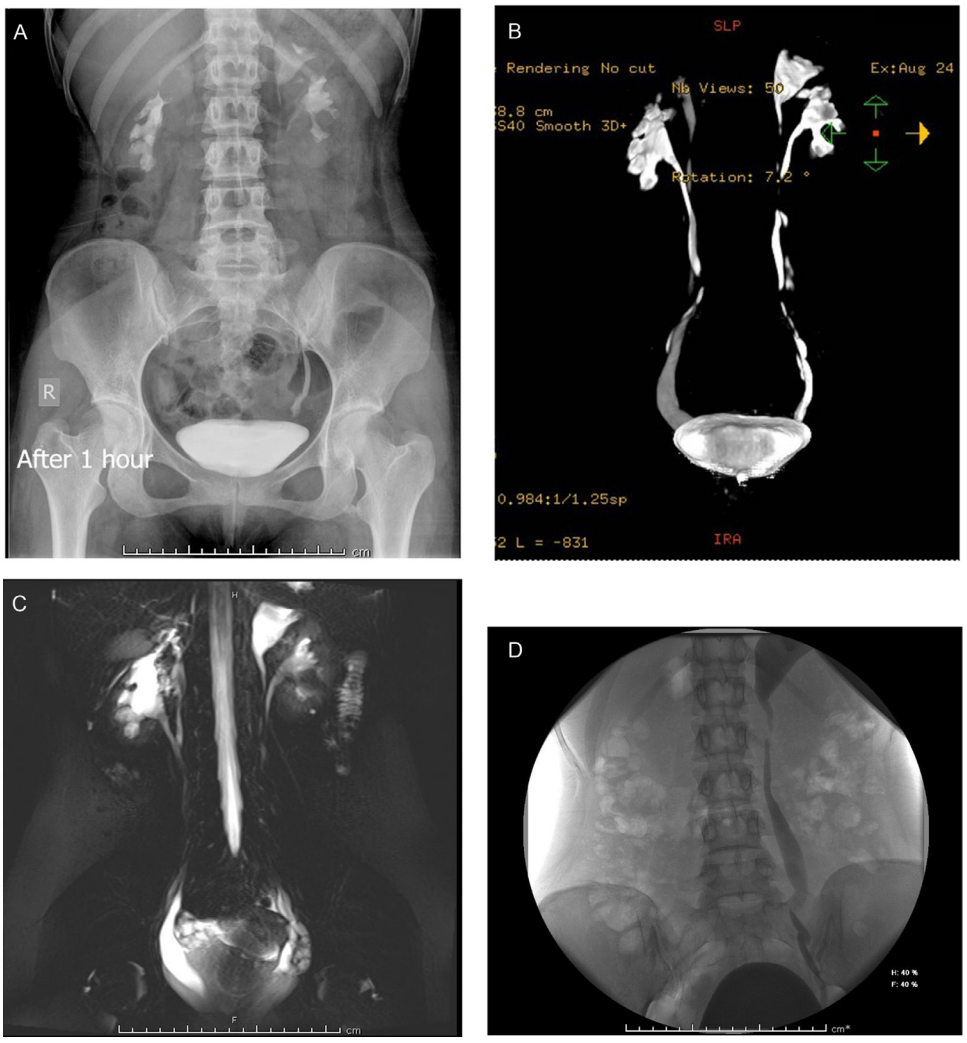

Figure 1. Pre-operative images. (**A**) Intravenous pyelogram (IVP) showing duplex system on the left side. (**B**) Excretory phase of CT urogram showing bilateral duplex system with poorly excreting dysplastic right upper moiety. (**C**) MR Urogram showing bilateral duplex system with dilated lower end of upper moiety ureters of both sides. (**D**) MCUG image showing grade V vesicoureteral reflux into the right upper moiety ureter. (Color version available online.)

further below to see for the bifurcation of the upper moiety ureter to avoid denervation of the bladder neck. A lower left ureteroureterostomy with lay open and ligation of distal ureter was performed (Fig. 2C) with right heminephrectomy. The patient had a complete recovery. Postoperatively, the patient continued to complain of incontinence. Our initial suspicion was whether following ligation of the common stem, the leak was due to the Y communication from bladder neck to the external meatus at the vestibule. However, when we examined there was no leakage from the vestibular opening and the incontinence was more like a severe stress leak due to a lax bladder neck. We started her empirically on anticholinergics (as she had urgency also probably due to detrusor instability) and taught her pelvic floor exercises. Fortunately, the patient responded and the incontinence gradually stopped. The anticholinergics were tapered and stopped over 1 month and at 3 months postsurgery she was doing well with no incontinence episodes with a postoperative ultrasound showed the disappearance of the left upper moiety hydroureteronephrosis.

DISCUSSION

Complete triplication of the ureter is a rare congenital anomaly of the urinary tract which was first described by Wrany in 1870.[1] In 1946, Smith presented a simple classification of Triplicate ureter, which is still in use today.[4] Smith classified it into 4 types: type I (35%), 3 separate ureters from the kidney with 3 separate draining orifices to the bladder; type II (21%), 3 ureters arising from the kidney with 2 of these joining and draining into 2 orifices; type III (31%), all 3 ureters joining together before reaching the bladder and draining through a single orifice; and type IV (9%), 2 ureters arising from the kidney with 1 becoming an inverse Y bifurcation and draining into 3 orifices. Our patient was the rarer type IV variant with

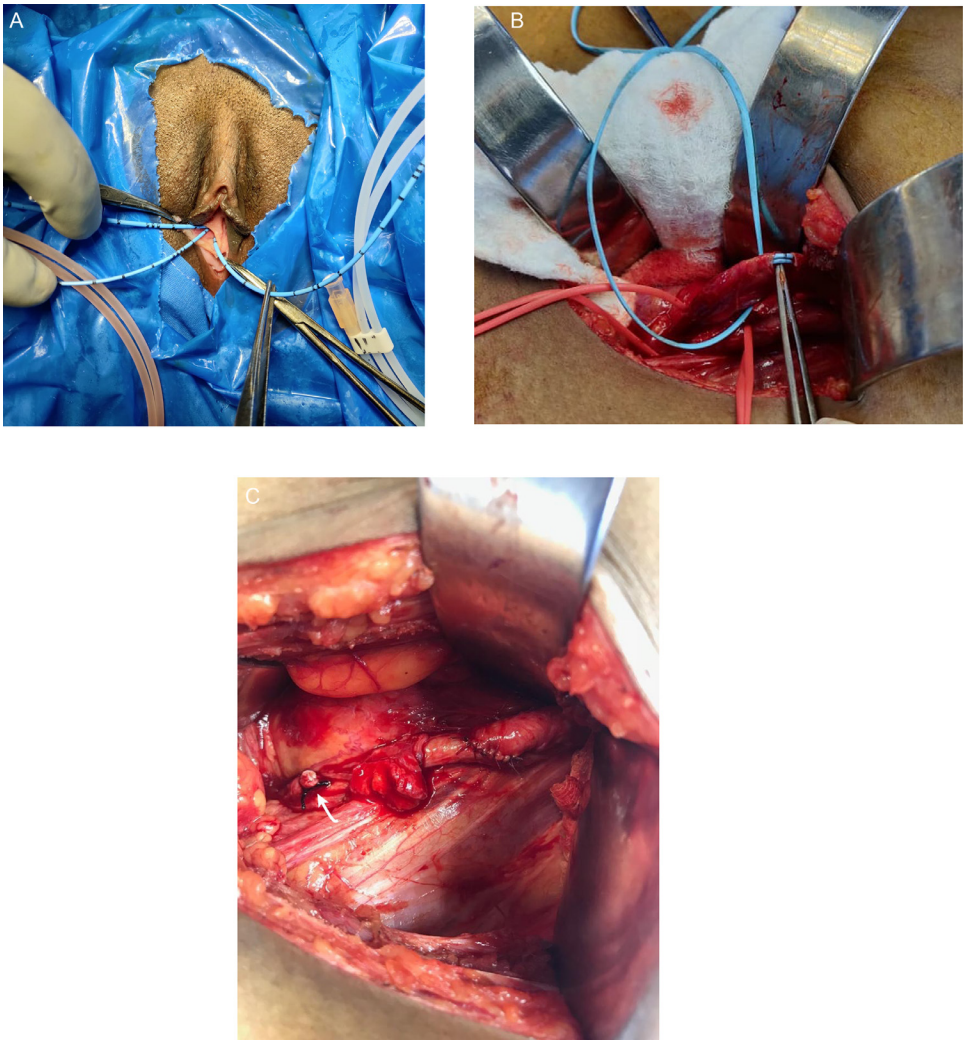

Figure 2. Intraoperative pictures. (**A**) Ureteric catheters placed through all three ureteric openings on the left side (orthotopic intravesical meatus, bladder neck, vestibule). (**B**) Upper moiety ureter containing two ureteric catheters and the lower moiety ureter looped and separately shown. (**C**) After completion of lower ureteroureterostomy. Distal ligated stump of the upper moiety ureter can be seen below (arrow). (Color version available online.)

1 draining the upper moiety bifurcating and opening into the bladder neck and the vestibule.

The basic problem of triplication is its own correct diagnosis, because it occurs in different forms, and it may be misdiagnosed as duplication.[5] Usually it is detected incidentally during surgery and autopsy. Noninvasive diagnostic tools include ultrasonography, intravenous pyelography, computerized tomography urography, and magnetic resonance urography.[2,6,7] However, in spite of all these investigations it can be misdiagnosed as a duplication as in our case, where only on examination under anesthesia and cystoscopy with RGP, the other orifice was cannulated and was confirmed as a type IV triplication.

Presentation of ureteral triplication is often delayed due to paucity of symptoms and signs. Clinical signs of triplication are nonspecific and the common presenting symptoms are the same as in duplication; urinary tract infection, urinary incontinence, and flank pain. Incontinence is usually taken as evidence of ureteric ectopia.[8] In our patient the primary complaint was incontinence. However not all incontinence in triplication is due to ureteral ectopia as we discovered in our patient as it was more a stress leak due to a lax bladder neck. We believe that in such triplication there is an element of bladder neck incompetence due to abnormal insertion of the ureters close to the bladder neck and this can manifest as stress leak. Children may not be able to give a proper history regarding incontinence and we should not assume all incontinence to be due to ectopic opening alone as in our case where the child had more of a stress leak from the beginning. This is very important in preoperative counselling and decision making for the patient as unless this stress component is addressed together with the ectopia these children may continue to leak causing considerable distress to the child and their caretakers. Fortunately, our patient responded

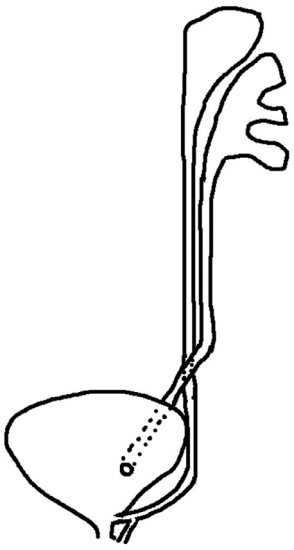

Figure 3. Schematic representation of Smith type IV triplication on left side.

to pelvic floor exercises and anticholinergics and was completely dry in a month. Further, we believe the parents have to be counselled about the possibility of return of incontinence following vaginal delivery when the pelvic floor muscles can be further weakened. This case report highlights the fact we have to consider all variable conditions once ureteral triplication has been diagnosed and every patient with ureteral triplication requires an individual strategy for management and treatment.

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