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Bilateral ureteral ectopia with duplex collecting system and hypoplastic bladder: a rare variant

Swapnil Singh Kushwaha, Sidhartha Kalra*, Dorairajan L. N., Manikandan R., Sreerag K. S. and Mujahid Ali

Abstract

Background: Bilateral single system ectopic ureter is a rare congenital anomaly, but bilateral ureteral ectopia with duplex collecting system with bladder hypoplasia is even rarer. Urinary incontinence in such cases is dealt with various reconstructive procedures.

Case presentation: A 20-year-old female presented with continuous urinary incontinence without normal voiding since birth. Imaging revealed bilateral duplex collecting system with ectopic ureter draining into the vestibule, and hypoplastic bladder. The patient was managed with Mainz-II pouch creation with serous lined extramural ureteral reimplantation technique with complete resolution of incontinence.

Conclusion: Our article highlights the challenges in surgical planning, treatment and the patient perspectives for this rare congenital malformation presenting with incontinence.

Keywords: Ectopic ureter, Hypoplastic bladder, Mainz-II pouch, Duplex collecting system

1 Background

Duplex collecting systems can be described as the kidneys with two pelvicalyceal systems associated either with a single ureter (bifid renal pelvis or partial ureteric duplication) or double ureters (complete ureteric duplication). Ectopic ureter in females classically presents with continuous urinary incontinence. There can be two distinct forms; one can be associated with a duplex collecting system in 80–85% of the cases, while the other draining a single system is found in around 20%. Bilateral ectopic ureter draining duplex collecting system is an uncommon congenital anomaly. Here, we describe our experience of managing bilateral duplex collecting system with ureteral ectopia and hypoplastic bladder in an adult female.

*Correspondence: sid6121984@gmail.com

Department of Urology, Jawaharlal Institute of Postgraduate Medical Education and Research, Puducherry 605006, India

2 Case presentation

A 20-year-old female presented to our institution with continuous urinary incontinence since birth. There is no history of normal voiding or recurrent UTIs. On examination, low-volume continuous leak of urine was present, but the urethral and vaginal orifices appeared normal. The renal function test was normal. CT urogram revealed a bilateral duplex collecting system with dilated bifid ectopic ureter draining into the vestibule and hypoplastic urinary bladder. Examination under anesthesia revealed a short urethra and small capacity bladder (< 50 mL) with an underdeveloped bladder neck but without trigonal anatomy or ureteric openings. Bilateral pinpoint ureteric orifices were visualized in the vestibule at 5°clock and 7°clock positions of the urethra and confirmed by cannulation (Fig. 1).

A urinary diversion procedure was considered for achieving urinary continence. After explaining the pros and cons of urinary diversion options, the patient being, a 20-year-old girl with her religious beliefs and cosmesis, was neither willing for stoma nor clean intermittent

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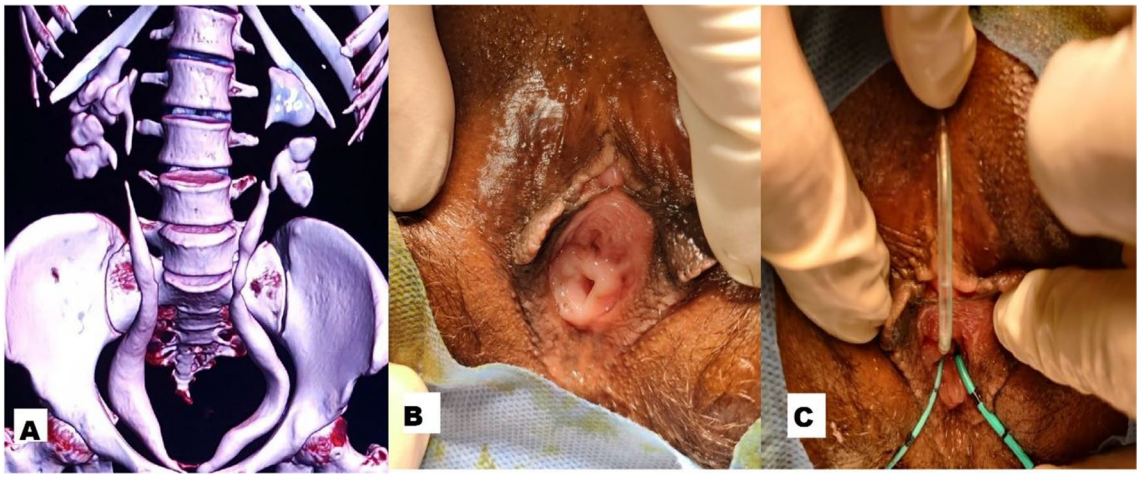

Fig. 1 **A** 3D reconstructed CT showing bilateral duplex collecting system with ectopic ureter. 3D, three-dimensional, **B** Urethral and vaginal orifices, **C** Ectopic ureteral orifices cannulated with ureteric catheters

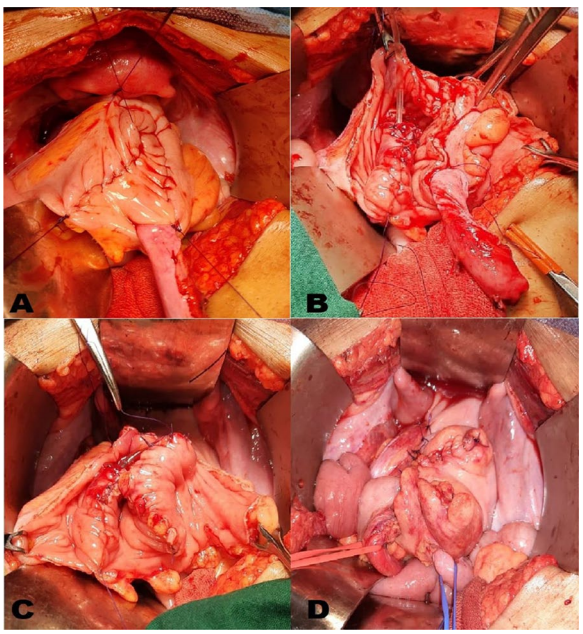

Fig. 2 **A** Bed for ureteral implantation, **B** Serosa lined extra-mucosal tunnelling, **C** Implanted ureters, **D** Rectosigmoid pouch closure

catheterization and opted for the Mainz-II procedure. Before carrying out the proposed diversion, the colonoscopy and warm saline enema test were done to exclude any large bowel pathology and to evaluate the anal sphincter competence, respectively. Both ureters were dilated and tortuous on laparotomy, and on tracing distally, they opened in the vestibule. Mobilized ureters were then implanted by serous lined extramural technique, and a rectosigmoid pouch was created (Fig. 2). Ureteral catheters were used as splints, and a rectal tube and an abdominal drain were kept. Postoperative recovery was uneventful. Per rectal voiding with continence for 3 to 4 h of the dry interval was achieved in the post-operative period at six months. Postoperatively she was started on alkalinizing drugs and antibiotic prophylaxis.

3 Discussion

Any ureter whose orifice does not insert into the normal trigonal position of the urinary bladder is considered ectopic. The incidence of ureteral ectopia is 1/2,000–4000 [1]. The most common sites of ectopic ureteral insertion in females are the upper urethra (33%) and vestibule (33%), followed by the vagina (25%), and less commonly in the cervix, uterus or rectum (5%). Ectopic ureters may coexist with many other abnormalities like bladder agenesis, renal dysplasia and non-genitourinary anomalies (cardiac, spinal cord or anorectal malformations, etc.) [2]. None of the other anomalies was detected in our patient.

The ureteral ectopia arises from the cranial origin of the ureteric buds from the mesonephric ducts resulting in a delay in their incorporation into the urogenital sinus. In bilateral ectopic ureter, there is a lack of bladder distension with urine, so trigone and bladder neck development do not occur. Furthermore, the bladder growth is prevented by an improperly functioning short urethra and poorly developed bladder neck leading to the hypoplastic bladder [3]. The hypoplastic bladder poses an obstacle in performing successful ureteral reimplantation.

After an extensive literature review regarding the bilateral duplex collecting system with ectopic ureter, only a handful of bilateral single-system ectopic ureter reports were found. This is the first reported case

representing a rare congenital anomaly of the genitourinary system to the best of the author's knowledge.

We emphasized the importance of preoperative imaging in the diagnosis, surgical planning, treatment challenges and the patient perspectives for this rare congenital malformation presenting with incontinence.

Imaging studies are mandatory in all cases to confirm the diagnosis. Ultrasound, an initial diagnostic test, has a limited role as it can rarely detect an ectopic insertion of the ureter. The method of choice for diagnosing an ectopic ureter is a CT or MR urography [4].

The best treatment for such a patient is surgery, as it resolves incontinence, preserves renal function and eliminates recurrent UTIs. Treatment of such cases is challenging owing to their rarity and has many considerations. The primary aim should be renal function preservation. Secondly, adequate bladder capacity is necessary to reimplant the ectopic ureters. The surgical management of ectopic ureter depends on surgeon experience, preference and patient perspective. The primary goal in managing bilateral ureteric ectopia is the attainment of continence [1, 3]. Hypoplastic bladder and underdeveloped bladder neck are the major difficulties in management. Whether to perform only ureteral reimplantation, ureteral reimplantation with augmentation or continent and non-continent urinary diversion remains a controversy [3]. The most accepted and popular procedure is augmentation cystoplasty with ureteral reimplantation. When the bladder is hypoplastic and neck is underdeveloped, the last resort is to abandon the native bladder and select urinary diversion. Continent urinary diversion procedures are associated with adequate self-perception of cosmesis and global satisfaction, but with the added risk of continence mechanism failure.

Mainz-II procedure creates a high-capacity and low-pressure urinary reservoir without any cutaneous stoma requiring catheterization and has a limited risk of continence failure. In this case, the application of serous-lined extramural tunnel technique provides an additional advantage in creating an anti-refluxing anastomosis for dilated ureters [5, 6]. The metabolic complications and the potential risk of developing malignancy must be addressed. In long-term follow-up of ≥ 10 years, the risk of malignancy at the ureteric implantation site described in the literature is 5–13%. Thus, there is an obvious need for the detection and prevention of metabolic complications, and colon cancer screening. Screening by annual colonoscopy is usually started ten years after the surgery [6–8].

4 Conclusion

Ectopic ureter should be kept in mind for patients with continuous urinary incontinence and recurrent UTIs. Anal diversion surgeries such as the Mainz-II can be a viable alternative for continent urinary diversion in patients not willing for cutaneous continent stoma or clean intermittent catheterization.

Abbreviations

CT: Computed tomography; MR: Magnetic resonance; UTIs: Urinary tract infections.

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Author contributions

The authors alone are responsible for the content and writing of this article. SK, LND and RM performed the surgery. SSK, SK, SKS and MA were the major contributors in preparing the manuscript.

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Data Availability

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Declarations

Competing interests

The authors declare that they have no competing interests.

Consent for publication

Written informed consent for publication of details and images was obtained from the patient.

Ethics approval and consent to participate

Ethics approval was obtained from the Institutional Ethical Committee, JIPMER, on 24/01/2022 and consent to participate was obtained from the patient.

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