Pediatr Urol Case Rep 2020; 7(1):18-22 DOI: 10.14534/j-pucr.2020156927

PEDIATRIC UROLOGY CASE REPORTS
ISSN 2148-2969
http://www.pediatricurologycasereports.com

Bilateral single system vaginal ectopic ureters: A rare variant

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ABSTRACT

Bilateral single-system vaginal ectopic ureter (BSSVEU) is a very rare entity in pediatric urology. A 1-year-old girl was brought to outpatient clinic with continuous dribbling without normal voiding. The patient was diagnosed as BSSVEU by clinical findings, CT intravenous urography, genitoscopy and cystoscopy. Bilateral ureteric reimplantation was performed. Postoperative dry time was 2-3 hours after 3 months of follow-up and renal function was preserved at 3 months. Here, we report such a case and briefly discuss its diagnosis and management.

Key Words: Single-system ectopic ureter, vaginal ectopic ureter, ureteric re-implantation.

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Received: 2019-11-27 / Accepted: 2019-12-07
Publication Date: 2020-01-01

Introduction

An ectopic ureter opens other than the postero-lateral aspect of the trigone, sometimes the ectopic opening is outside of the urinary system. Ectopic ureters are usually associated with a duplex kidney and, in general, 80% of ectopic ureters arise from the upper pole of the kidney. If an ectopic ureter drains a single kidney, it is called a single system ectopic ureter, which occurs in only 20% of cases [1]. Bilateral single-system ectopic ureters (BSSEU) are even rare. This is the first case of opening into the vagina with normal urethral opening and normal bladder neck. In this case, both ureters were opening in the vagina leading to the very small unused urinary bladder with very small capacity (< 5 ml). The objective of reporting this case is to describe the challenge of management in a young girl whose small urinary bladder has never been exposed to urine.

Case report

A 1-year-old female presented with continuous dribbling of urine and never passed urine in the stream since birth. Parents were of poor socioeconomic status and an illiterate rural background. There was no history of high-grade fever, chills & rigor, swelling over back, altered bowel habit. Mother was unbooked and antenatal history was uneventful.

On general examination, child laying comfortably in mother lap, afebrile, heart rate 110/min, Blood pressure 90/60 mmHg,

respiratory rate 38/min, no pallor, icterus, cyanosis or lymphadenopathy present. There were no skeletal defects, neurological examination and other systems were normal. The abdomen was soft, nontender, no organomegaly, Bowel sounds were normal.

On per rectal examination anal tone normal, no mass felt, no sacral agenesis. Three orifices in the perineum (urethra, vagina, anus). On separating the labia majora, no urine coming per urethrae and urine pouring out through vaginal orifice (Fig. 1).

Fig. 1. Perineum of the child showing three openings (urethra, vagina, anus).

All routine investigations such as hemoglobin, total leucocyte count, renal function tests, and urine microscopic examination were within the normal range.

Abdominal sonography revealed bilateral moderate hydroureteronephrosis (Right >> Left), small contracted urinary bladder, uterus, and ovaries were normal, no other anomaly detected. Micturating cystourethrogram was not possible. Genitogram was done showing ectopic ureteric opening in the vagina with reflux on the left side, the right system not visualized (Fig. 2). A CT intravenous pyelography showed right hydro-uretero-nephrosis is more in comparison to the left kidney, bilateral ureters inserted to the lateral vaginal wall and contrast collection in the vaginal cavity and no calculus (Fig. 3).

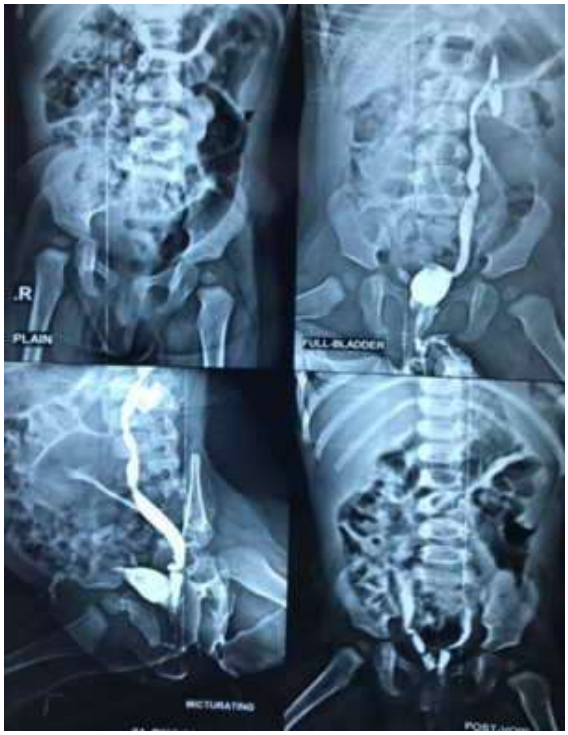

Fig. 2. Genitogram showing ectopic ureteric opening in the vagina with reflux on the left side.

On genitoscopy and cystoscopy, the bladder neck was developed with an Ill-defined trigone. Bladder capacity was approximately < 5 ml, normal vagina and ureteric orifice could not be visualized in the bladder or vagina. Bladder and vagina were catheterized for 24 hours, urine came out only from the vagina. The definitive diagnosis of Bilateral Single System Vaginal Ectopic Ureter (BSSVEU) was made and planned for single stage bilateral ureteric re-implantation.

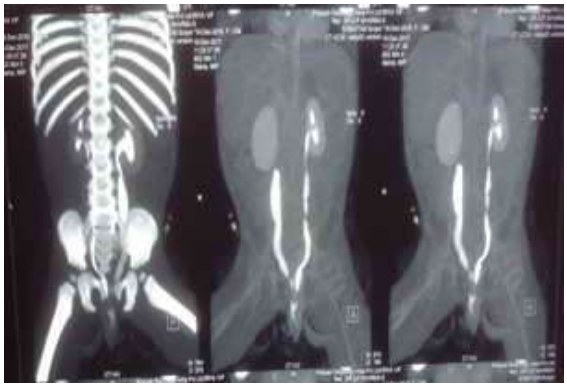

Fig. 3. CT intravenous pyelography showing right hydroureteronephrosis more in comparison to the left kidney, bilateral Ureters inserted to the lateral vaginal wall and contrast collection in the vaginal cavity.

At surgery, the ureters were identified, traced and found to be opening into the postero-lateral wall of the upper vagina [Fig. 4]. Ureters were dissected and isolated from vaginal insertion, bilateral reimplantation was done over DJ stents. The bladder was too small to accommodate both DJ stents and urethral catheter so a 6 Fr infant feeding tube inserted and fixed. Infant feeding tube comes out on day 2, both DJ stents come out per urethra first on day 5 and the second one on day 7 itself.

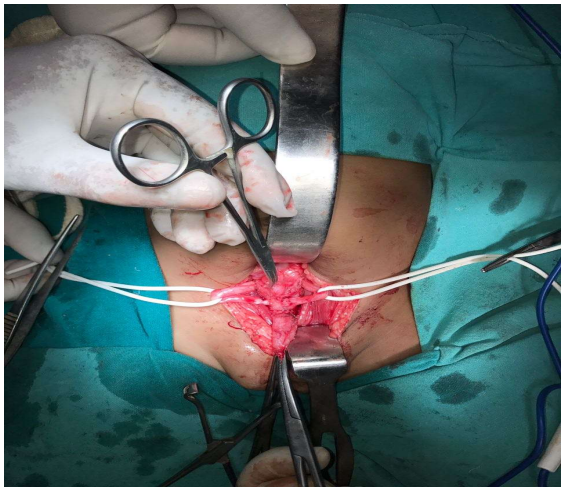

Fig. 4. Intraoperative picture showing ureters opening into the postero-lateral wall of the upper vagina.

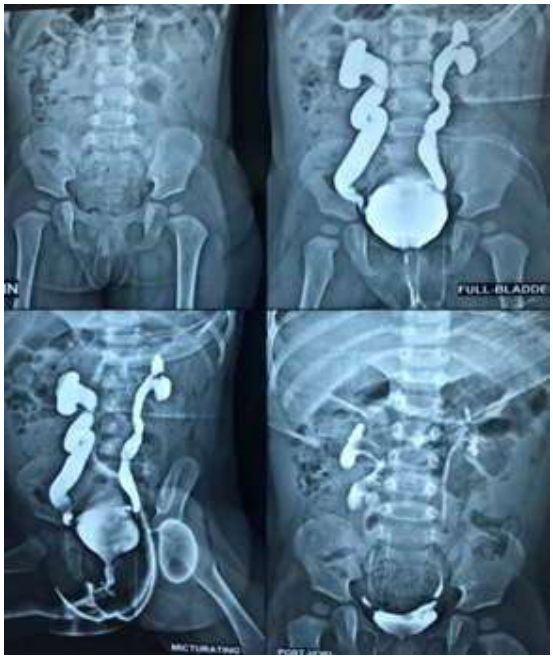

Fig. 5. Post-operative MCU at 3 months showing increased bladder capacity (30ml) with bilateral VUR, but no contrast retention after micturition.

On follow-up, after 1-month child passing urine in the stream and dry time for the child was 30 min. After 3 months the child remained dry for two hours. Renal function preserved. MCU show increased bladder capacity (30ml) with B/L VUR, but no contrast retention after micturition (Fig. 5). Antibiotic prophylaxis was given to the child.

The mother was very happy to report that the excoriation around the perineum had healed, and the child was passing urine in the stream.

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Discussion

We report a case of bilateral single-system ectopic ureters opening into the vagina associated with the hypoplastic bladder, well developed vesical neck and urethra, and bilateral ureteric reflux.

This is an unusual location for bilateral single-system ectopic ureters. There is 3 other reports of bilateral single-system ectopic ureters

opening into urogenital sinus but not in well-developed vagina. First by Sheldon et al. [2], in their case, urogenital anomalies were more severe i.e., the vagina was rudimentary and both kidneys were dysplastic, resulting in end-stage renal disease. Second by Bhupendra P. Singh et al. [3], in their case, ectopic ureters opening into urogenital sinus associated with the absence of the urethra, hypoplastic bladder, incontinent vesical neck, and bilateral ureteric reflux. Third by Farzeen Sharaf et al. [4], in their case anatomy was similar to Sheldon and Welch except for the renal abnormalities. All three case reports urethral openings were in urogenital sinus. However, in our case the vagina was normal, urethra and vesical neck were well developed and both kidneys were normal.

The primary management of BSSEU may vary from bilateral ureteric re-implantation only for the attainment of continence, to adding a bladder augmentation and bladder neck reconstruction, at the same sitting or subsequently. Kesavan, et al. [5] showed the vesical neck and trigone not well developed in 54% of unilateral and 75% of bilateral ectopic ureters.

Bhupendra P. Singh et al. [3] stated that ureteric re-implantation alone may not attain continence in patients with BSSEU. On contrary to them Kumar et al. [6] report that a bladder with BSSEU may not necessarily be useless and bladder capacity increases with the passage of time without the need for augmentation.

In this case, the increase in bladder capacity and improvement in continence was achieved only by ureteric re-implantation. The well-developed vesical neck and adequate urethra helped us to consider this option. Hence, in cases of bilateral single-system ectopic ureters, management modalities should be individualized from patient to patient depending upon the anomalies in the lower urinary tract as well as the reproductive tract.

A child presenting with dribbling of urine since birth, with or without normal micturition, needs to be investigated early and thoroughly.

In our opinion, primary ureteral re-implant is an option to avoid staged procedure and augmentation surgery complications. Adequate time should be given and after re-implantation, bladder response to urine challenge should be observed.

Compliance with ethical statements

Conflicts of Interest: None.

Financial disclosure: None.

Consent: All photos were taken with parental consent.

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