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Case Report

Bladder and Urethral Agenesis: A Report of 2 Cases

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Keywords
Bladder agenesis
Ureterosigmoidostomy
Urethral agenesis
Urogenital anomalies

Abbreviations
CT - Computed tomography
DMSA - Dimercapto succinic acid

Abstract

Bladder and urethral agenesis are among the rarest congenital anomalies, and to our knowledge, only 27 cases have been reported in English literature until now. The authors treated two such cases in the last one year. The first case was diagnosed at 2 months of age, and the second case at 10 months of age. Both were females. In The first case ureterosigmoidostomy was done at 1 year of age. The second case had features of chronic kidney disease due to a stenosed ectopic ureter. Therefore, diverting cutaneous ureterostomy was done. Bladder and urethral agenesis are associated with other congenital anomalies, mostly with upper urinary tract anomalies. To prevent diagnostic delay and to reduce mortality cum mortality by early management, awareness of this rare condition is essential.

BACKGROUND

Bladder and urethral agenesis, one of the rarest urogenital anomalies, has an incidence of 1 in 60,000,000.[1] There are many theories regarding the etiology of bladder agenesis. One of them suggests that it may result from the failure of mesonephric duct and ureteric bud integration in the trigone resulting in lack of bladder filling.[2] Another theory suggested that this may be due to complete atresia of urogenital sinus.[3] In females with absence of bladder, ureter may insert into the Mullerian derivatives such as the uterus, anterior vaginal walls, or vestibule as the Mullerian ducts also open into urogenital sinus.[2] On the contrary, males with the described disease are mostly stillborn, as they can only survive if the ectopic ureters drain into either rectum or patent urachus, which rarely occurs.[4] Surgical intervention to establish adequate urinary drainage is required to protect kidneys from obstructive uropathy and urinary tract infection. Herein, we report two cases of bladder and urethral agenesis in female infants, where both of them had ectopic insertion of the ureter into the vagina.

CASE REPORT - 1

A 2-month-old girl presented with continuous dribbling of urine since birth. A history of periodic normal voiding is absent. Her perinatal period was uneventful. Antenatal ultrasound did not detect any urological anomaly. The anus was in normal position. The external genitalia was that of a female, with a single opening in the vestibule.

Retrograde contrast radiography showed bilateral hydronephrotic kidneys with ureters opening into

a common channel indicating the absence of bladder and urethra.(Fig.1) CT urogram revealed bilateral incomplete duplex pelvi-calyceal system. (Fig.2) DMSA scan showed bilateral renal scarring. Magnetic resonance imaging (MRI) of the lumbar spine did not show any abnormality. The patient was discharged with antibiotic prophylaxis. Subsequently, we did bilateral ureterosigmoidostomy with anti-reflux sub-serosal tunneling. (Fig.3) Double-J stents were kept in both ureters; but both of them got spontaneously expelled within the second post-operative day. The patient was discharged with prophylactic antibiotics and oral sodium bicarbonate as metabolic acidosis was expected. On follow-up after a month, mother complained of incontinent passage of urine per anus, and there were clinical features of metabolic acidosis. However, in the subsequent visits the blood gas analysis showed normal finding.

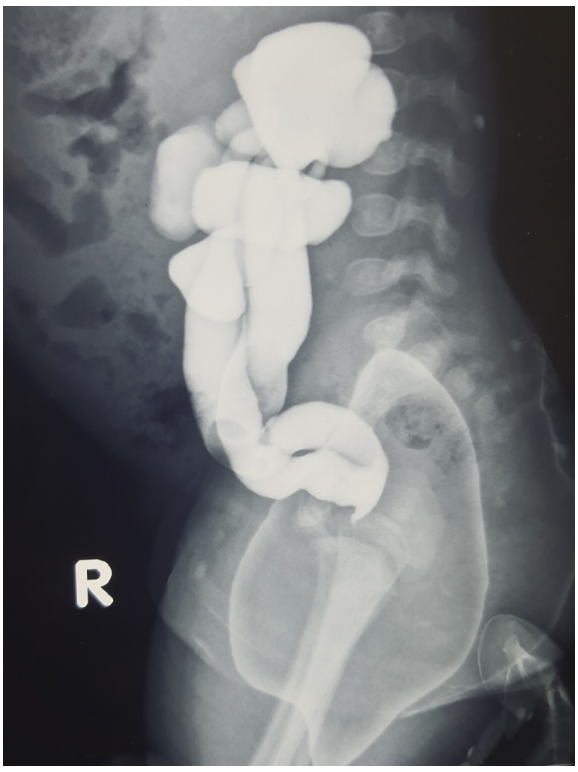

Fig 1. *Retrograde contrast film showing the ureters entering into a common channel in the absence of bladder (Case 1)*

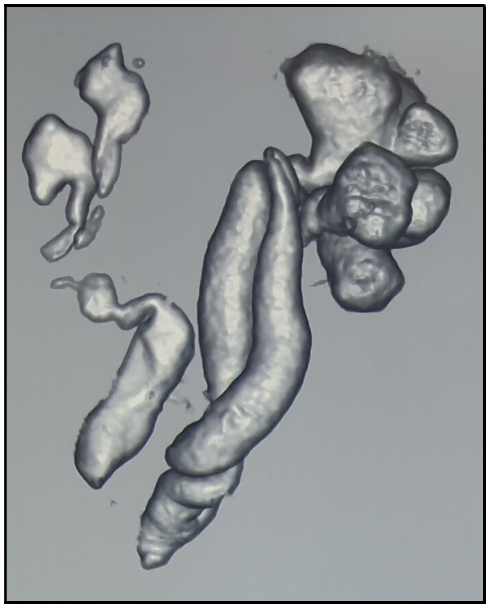

Fig 2. *CT urogram showing incomplete duplex ureter (Case 1)*

CASE REPORT - 2

A 10-month-old girl presented with continuous dribbling of urine since birth without any normal voiding pattern. She had been admitted to hospitals several times for episodes of dehydration, electrolyte imbalance and acute kidney injury which subsequently turned into chronic kidney disease. Prior to the current admission under our care, she had been managed by nephrologists for renal impairment. She had no symptoms pertinent to bowel movements.

During admission, we found her dehydrated and lethargic. Her weight was 5.5 kg which fell below the third percentile. Perineal examination showed a single opening in the vestibule from which urine was coming out. (Fig.4) Spine was normal. Ultrasonography couldn't trace the right kidney and found duplex left kidney with hydroureteronephrosis. In intravenous pyelography, delayed excretion of the left kidney with hydroureteronephrosis and non-visualization of the right kidney were noted. Contrast instillation through the single opening in the vestibule revealed incomplete duplex ureter and pelvis.

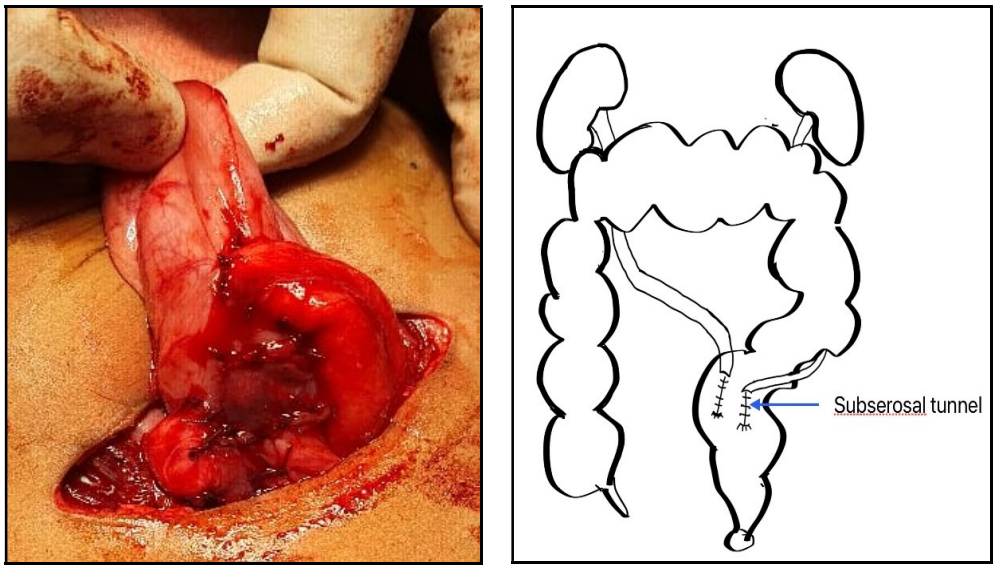

Fig 3. *(A) Intra-operative photograph showing dilated ureters implanted into the sigmoid colon. (B) Schematic diagram showing ureters implanted in sigmoid after creating sub-serosal tunneling*

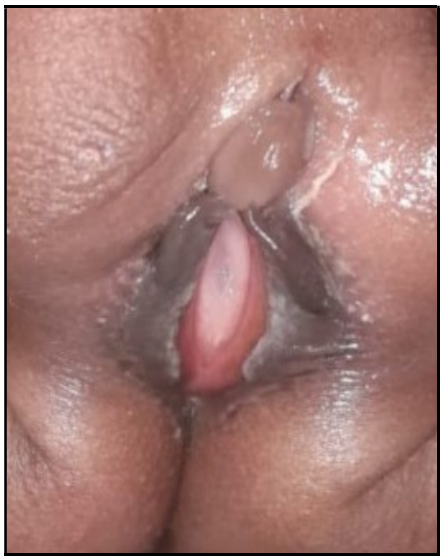

Fig 4. *Single opening in the vestibule (Case 2)*

on the left side. In magnetic resonance urogram, right kidney was not seen in normal position; rather it was located at lower abdominal cavity at level of L3 and L4. The right kidney was smaller and malrotated with anomalous insertion of the right ureter into the neck of small bladder. The left kidney showed multiple small cortical cysts. Left ureter was dilated, tortuous (maximum diameter 2cm) with smooth narrowing at its distal part. Laparotomy revealed absence of bladder and both the ureters opened into anterior vaginal wall. There was a pelvic right kidney with left sided duplex pelvi-calyceal system with grossly dilated ureter. We did urinary diversion by bilateral cutaneous ureterostomy as the patient's condition was not conducive for extensive surgery.

DISCUSSION

Bladder agenesis is thought to occur due to insult at 5-7 weeks of embryogenesis.[2] Most patients with bladder agenesis are females. In males, bladder agenesis is compatible with live birth only if the ureters open into rectum, patent urachus or seminal vesicle.[5] Both of our cases were female and in them the ureters opened into the anterior vaginal wall.

Usually, patients with bladder agenesis presents soon after birth; however, there are reports of delayed diagnosis as late as second and third decades of life.[6] Continuous dribbling of urine and repeated urinary tract infection are the

frequent symptoms. In our patients we initially suspected a commoner pathology, namely ectopic ureter. Nevertheless, absence of normal voiding did not support this initial suspicion. CT Urogram of case-1 was conclusive of bladder agenesis. But, all the investigations of case-2 pointed towards ectopic ureteric rather than bladder agenesis. Seemingly, the dilated vagina was mistaken for small capacity bladder by the radiologists. Examination under anesthesia revealed a single opening in the vestibule. According to Metoki, both of our cases were vaginal subtype of bladder agenesis.[7]

Prognosis of bladder agenesis is variable, and it depends mostly on associated anomalies. Upper tract anomalies such as the solitary kidney, ectopic kidney and unilateral duplex collecting system have been reported. Both of our cases had duplex collecting system. Only one of the previous author mentioned association of bilateral duplex kidney.[8] In addition to this, vascular anomalies and bony deformities has been reported. There are four published cases of bladder agenesis where continent urinary diversion was made, which were either Penn pouch or ileal reservoir along with a Mitrofanoff channel using the appendix. We did ureterosigmoidostomy in case-1 with the expectation that when she achieves bowel control it will also act as a continent diversion.

CONCLUSION

Preservation of renal function, adequate urinary drainage, and continence are aims of the management of bladder and urethral agenesis. Diagnostic dilemmas can delay the initiation of appropriate treatment in these patients. Collaboration of nephrologists, urologist, and radiologists are essential in preventing morbidity and mortality of this rare entity.

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