Case Report

Bilateral ectopic ureters with bladder agenesis

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Abstract

Agenesis of Urinary bladder is an extremely rare congenital genitourinary anomaly and is associated with other severe malformations that are incompatible with life. About 60 cases and 22 live births only have been reported in English literature. We report an eight year old girl with triad of complete agenesis of bladder and urethra, solitary functioning left kidney and an ectopic ureter opening into the vagina. The management of the child along with embryo-pathogenesis and literature review is discussed.

Keywords: Bladder agenesis, Ectopic ureter, Continent urinary diversion.

Introduction

Bladder agenesis is an extremely rare congenital anomaly of urogenital system and is associated with other severe malformations that are incompatible with life. The vast majority (90%) of viable children with this anomaly is female.[1] About 60 cases and 22 live births only have been reported in English literature. We report an eight year old girl with triad of complete agenesis of bladder and urethra, solitary functioning left kidney and an ectopic ureter opening into the vagina.

Case Report

An eight year old girl presented with the history of persistent dribbling of urine and recurrent urinary tract infections. She had never voided with stream on her own. There was no history of any bowel irregularities. She was the only child of her parents and was born full term with simple vaginal delivery. There was no familial history of any congenital abnormalities. Her mother denied use of any drug

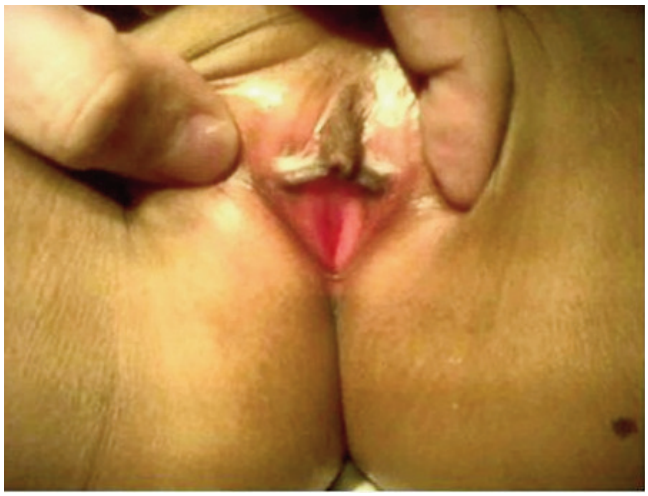

Figure-1: Single opening in the vulva.

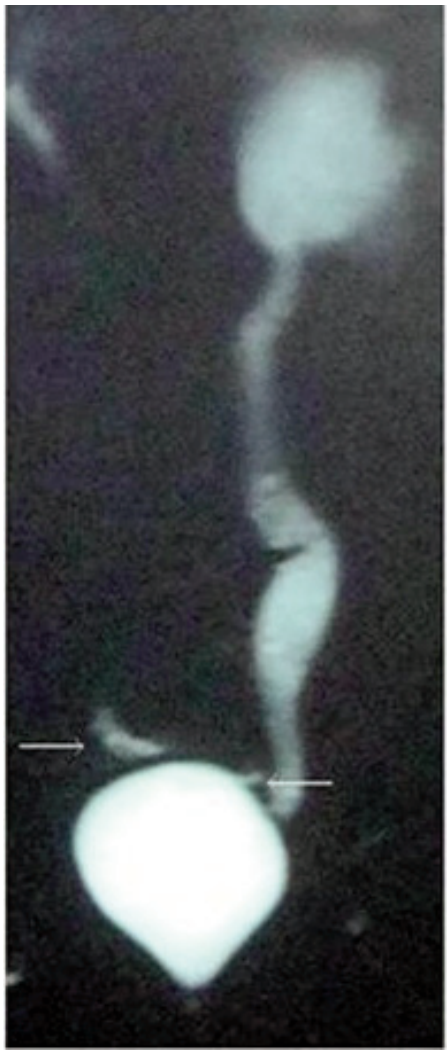

Figure-2: Sinogram showing contrast filled cavity with grade 4 VUR into left kidney. (Arrows show reflux into both fallopian tubes).

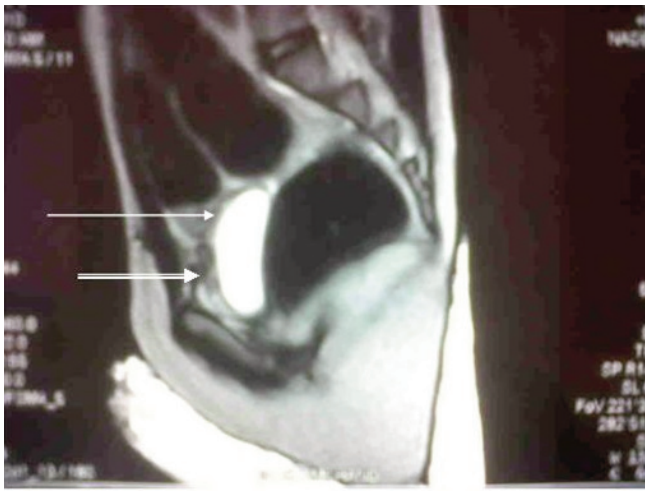

Figure-3: MRI scan: sagittal T2-W sequence shows dilated contrast containing vagina (single arrow) just behind the pubic bone (double lined arrow) resembling the bladder and absence of bladder.

or history of fever during pregnancy. The child's milestones were not delayed. She was investigated and found to have bilateral VUR and right nonfunctioning kidney and underwent right nephrectomy elsewhere but her incontinence was missed and was referred for further management.

On examination, she was haemo-dynamically stable, and fully mobile. The external genitalia were phenotypically female with excoriation of vulva and a single opening in the vestibule with continuous dribbling of urine (Figure-1). There was no obvious skeletal or spine deformity. The anal tone and neurological examination were normal.

Routine investigations i.e. complete blood counts and electrolytes were normal however serum creatinine was borderline high with a value of 1.4 mg/dl and Urine culture showed growth of E.Coli. Chromosomal analysis showed 46 XX karyotype. On abdominal ultrasound left kidney showed moderate hydronephrosis, there was absence of right kidney and a rounded full bladder was seen. Sinogram was done which showed a dilated cavity resembling urinary bladder with grade 4 vesico-ureteric reflux and reflux in both fallopian tubes (Figure-2). She had an MRI scan of pelvis which showed dilated vagina just behind the pubic bone resembling the bladder and absence of urinary bladder (Figure-3).

After treating her UTI an examination under anaesthesia and endoscopy was done which revealed single opening in perineum leading into a cavity with no obvious bladder neck or trigone. There was a definitive cervical os leading into the cavity and an opening was identified on the left side of the cavity that admitted 10 Fr cystoscope sheath. Retrograde pyelography confirmed it to be left ureter leading into the left kidney. No other openings were identified. A

diagnosis of bladder agenesis with ectopic ureter opening into vaginalized urogenital sinus was made.

Exploratory laparotomy was done by lower midline incision. Uterus was identified just behind the symphysis pubis along with fallopian tubes and ovaries; however no urinary bladder or any other remanant tissue was found. The left ureter was dilated and was opening into a dilated vagina. Ureter was dissected off the vagina and an orthotopic urinary reservoir was made from sigmoid colon and ileum after de-tubularization. Ureter was attached in an anti-reflux manner via a sub-mucosal tunnel and appendix dissected from the caecum and anastomosed to right leaf of colonic patch and brought out to right iliac fossa as a catheterizable Mitrofanoff stoma.

Postoperatively the child remained asymptomatic. Urinary reservoir was kept on drainage for 3 weeks and mucous aspirated. She was started on CIC with 10 Fr tube at 3 hourly intervals and she remained dry in between without any leakage of urine. Her renal functions remained stable with normal upper urinary tracts on imaging. She did not have any episode of pyelonephritis. She was continued on oral soda bicarb and did not show any metabolic derangements on follow up evaluation.

Discussion

Bladder agenesis is an extremely rare congenital anomaly. The exact determination of such cases is extremely difficult however Glenn reported occurrence of bladder agenesis in one out of 600,000 hospitalized patients.[1] This condition is usually not compatible with life and only 22 live cases have been reported so far. The vast majority (90%) of viable children with this anomaly are female.[2]

Almost all patients had other associated congenital anomalies especially orthopaedic, neurologic or other urological anomalies like renal agenesis, dysplastic or ectopic kidneys, ectopic ureters, bicornuate and absent uterus or vaginal atresia in females and absent prostate, seminal vesicles or penis in males.[3-5] The exact embryological mal-development leading to bladder agenesis is difficult to explain but it is proposed that these anomalies occur during weeks 5-7 of embryogenesis. Bladder starts to develop by 5th week of gestation when cloaca is divided by a uro-rectal septum into urogenital sinus and anorectal canal.[6]

In patients with bladder agenesis and without any hind gut abnormalities like in our case, it is proposed that the division of cloaca is normal but there is either a primary developmental failure or secondary atrophy of the urogenital sinus.[6] The latter phenomenon is due to failure to incorporate the mesonephric ducts and ureter into the trigone leading to lack of distension with urine and consequent atrophy. This is also associated with the most severe form of ureteric ectopia. However, ectopic ureter is essential for compatibility with life because it preserves the renal function.[7] Because of the associated abnormalities, the successful treatment and long term prognosis are very poor.

There is a dearth of paediatric urological facilities in Pakistan. This includes poorly developed health infrastructure, lack of support facilities like anaesthesiology and intensive care and extreme paucity of trained urologists and paramedical staff. Patients with complex urogenital anomalies requiring highly skilled reconstructive surgeries are managed by alternate therapies, by non-medical practitioners and surgeons with lack of training in paediatric urology.[8]

Our patient was an eight year old girl with history of Right nephrectomy due to a non-functioning kidney. Her diagnosis and management got delayed till eight years of age and she remained with incontinence of urine with significant physical and emotional stress.

She had a solitary functioning left kidney with ectopic ureter opening into the vagina with preservation of renal function. Fortunately, she did not have any other associated abnormalities making the management relatively easy.

The management of bladder agenesis depends upon the severity and extent of the anomaly, however the general principle includes relief of obstruction and preservation of renal function by making a continent or non-continent urinary diversion. This can be accomplished by uretro-sigmoidostomy i.e. internal stoma or an external stoma.[6,9]

It is also necessary to consider the physical status of the child, preferences of parents and their social situation in the management plan. We opted for a single stage definitive procedure, using both sigmoid colon and ileum to form an orthotopic catheterizable reservoir and appendix was used as a continence mechanism using the Mitrofanoff principle.

This case report adds to the pool of the available literature on bladder agenesis. During the literature review, we could only identify a single case of a 5 year old child who had similar triad of bladder agenesis with solitary kidney and ectopic ureter opening into the vestibule.[5] The author managed the patient using the continent ileo-caecal pouch (Penn pouch) with Mitrofanoff principle. No other publication described lower urinary tract reconstruction and attainment of continence in patients with bladder agenesis.

Singh BP et al[10] described the first report of a 5 year old girl with bilateral single system ectopic ureters that were opening into a vaginalized urogenital sinus with a capacity of 20-cc and without any definitive trigone or bladder neck.[10] The dilated vagina seen in our case could also represent the so called Vaginalized urogenital sinus.

Our patient managed well on CIC with well-preserved

renal functions and without any metabolic abnormality on subsequent follow up.

Conflict of Interest:

The authors declare that they have no conflict of interest.

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