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Bilateral single system ectopic ureters – A rare variant

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ABSTRACT

Bilateral single system ectopic ureters are a rare entity in paediatric urology. We report a girl child with bilateral single system ectopic ureters with right system opening into the vagina, who presented at 3.5 years with continuous dribbling of urine & a small capacity bladder. Renal scans and MRI were done which indicated bilateral single system ectopic ureters with hydroureteronephrosis. We managed her surgically by a right nephro-ureterectomy, bladder augmentation, left ureteric reimplantation and Mitrofanoff. Post op patient had acute on chronic renal failure, stabalised by haemodialysis. It is a rare presentation if managed promptly can prevent renal replacement therapy.

1. Introduction

Ectopic ureters are associated with a duplicated system almost 80 % of times. Less common are single system ectopias accounting for 10–20 %[1] Rarest are bilateral single system ectopic ureters[2] These children are usually incontinent owing to non-functional bladder neck sphincter and small volume bladder[1]

Here we present a case of a girl child with bilateral single system ectopic ureters with right system opening into the vagina.

2. Case report

A 3.5 years girl child, known case of chronic kidney disease (CKD) diagnosed elsewhere, presented to us with dribbling of urine, continence with incontinence since neonatal life. She was antenatally diagnosed case of right hydronephrosis. Child did not pass urine till day of life 4 followed by spontaneous continuous dribbling of urine thereafter. She had multiple episodes of febrile UTI thereafter which were managed conservatively.

Ethylene Cysteine renogram was suggestive of hydronephrotic left kidney with grossly dilated, tortuous ureter with an obstructive curve. Right kidney was not functional (Fig. 1).

Her outside MCU showed mild trabeculated bladder with wide bladder neck and no evidence of vesico-ureteric reflux although the bladder capacity was not mentioned. (Fig. 2). We did a betadine MCU at our center which showed the bladder capacity to be 50 ml which was small for her age. Her serum creatinine at presentation was 3mg/dl thus magnetic resonance urography (MRU) was done.

MRU detected a small left kidney with hydroureteronephrosis, and left ureter revealed ectopic opening in the proximal urethra. Right kidney had cortical thinning with hydroureteronephrosis with distal ureter narrowing having an ectopic opening in the vagina suggestive of bilateral single system ectopic ureters with hydronephrosis and hydro-ureter (Fig. 3).

A Diagnostic cystoscopy was done to plan the further management for this child which revealed right ureteric orifice opening in the vagina and the left ureteric orifice was noted on the anterior bladder wall just above the bladder neck. Mild bladder trabeculations were noted and the capacity was small.

3. Surgical management

A Right nephro-ureterectomy with bladder augmentation (Fig. 4) with left ureteric reimplantation and Mitrofanoff (Fig. 5) was done.

Post-operatively child suffered acute on chronic renal failure, serum creatinine was increased to 5mg/dl with metabolic disturbances leading to seizures, hence underwent 2 cycles of haemodialysis after which she stabilised. On follow up after 1 month post op, her sr. creatinine was 2mg/dl. She is on chemoprophylaxis, tablet sodamint and continent on 2 hourly clean intermittent catheterisation, with overnight drainage.

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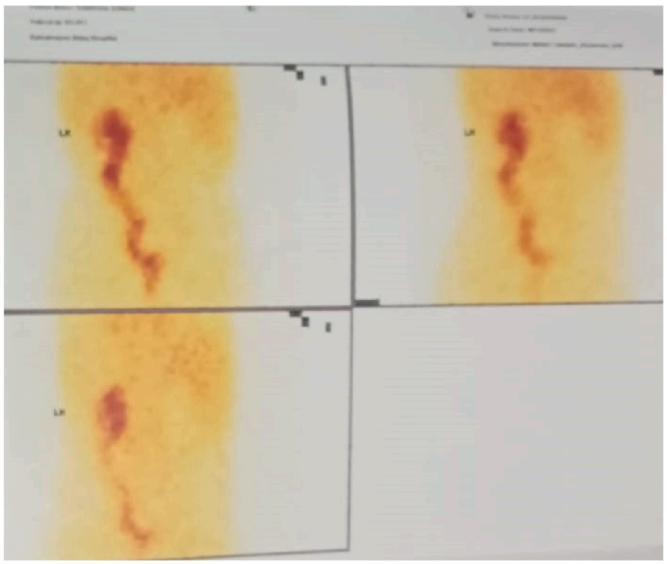

Fig. 1. EC-renogram.

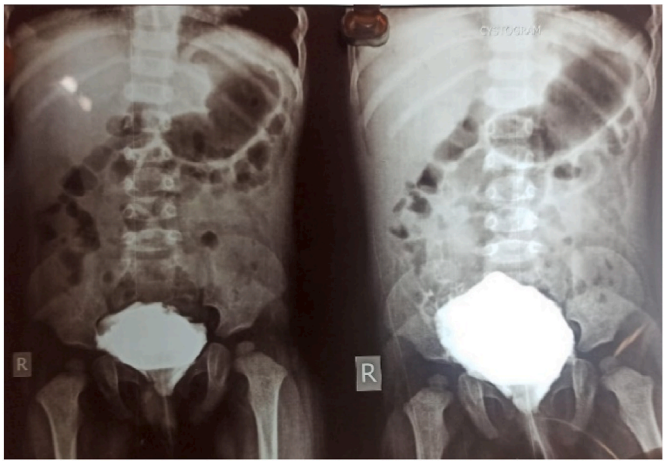

Fig. 2. Micturating cysto-urethrogram. (MCU).

4. Discussion

One of the rarest entities in urology with less than 80 cases reported is bilateral single system ectopic ureters.[3] Whether to do a ureteral re-implantation or bladder augmentation along with it, is a common controversy. Kesavan et al. reported that the bladder neck and trigone were maldeveloped in 75 % of bilateral ectopic ureters and in unilateral ectopic ureter it was 54 %[4] Only ureteral re-implant may not cure incontinence due to insufficient growth of bladder neck and trigone as per the study by Heuser et al.[5]

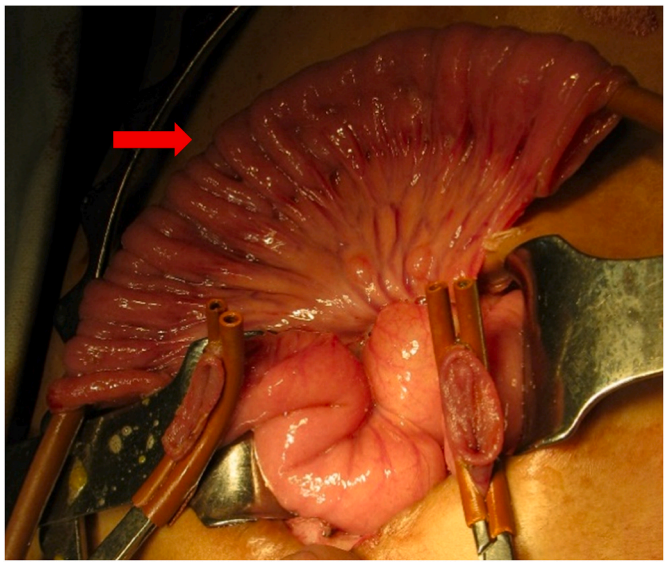

Fig. 4. Bladder augmentation.

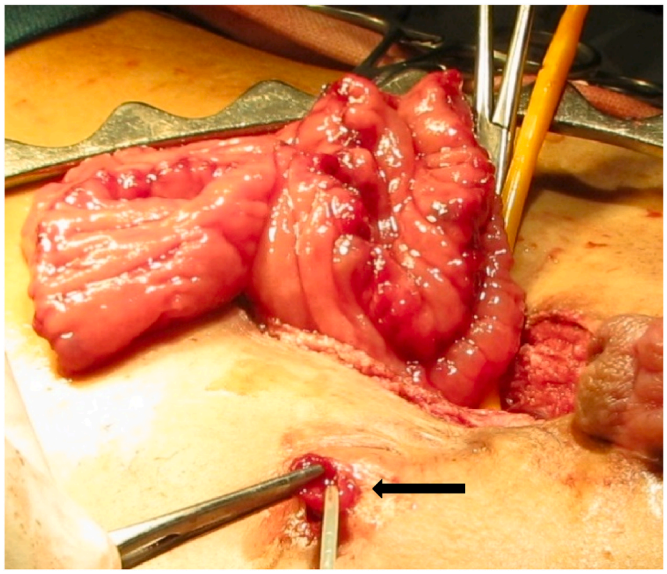

Fig. 5. Mitrofanoff procedure.

Various procedures have been described in literature like Kropp's procedure, artificial sphincter and pubo-vaginal sling to increase the bladder neck resistance, Young–Dees-Leadbetter bladder neck tightening. However, Jayanthi et al. proposed the concept of bladder neck

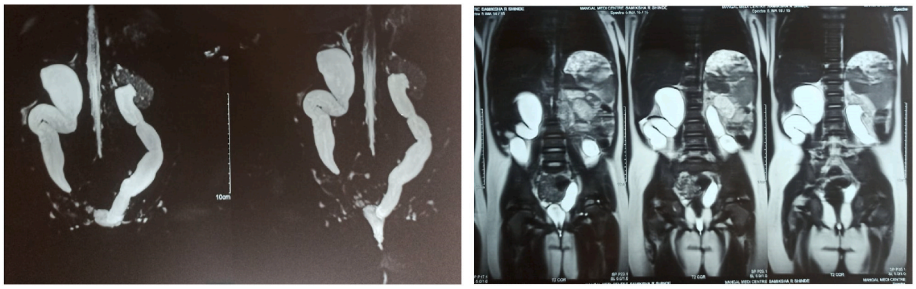

Fig. 3. MR- Urography.

closure, augmentation and appendico-vesicostomy to achieve total day and night time continence[6] There is a spontaneous increase in the bladder capacity with time and intestinal augmentation may not be necessary in all patient was proposed by William et al.[7]

We managed by doing a nephro-ureterectomy, bladder augmentation along with Mitrofanoff for our patient who is continent and on regular follow up. Such patients are usually in chronic renal failure and may require dialysis and renal replacement therapy in the long run.

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