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Original Article

Rare cases of ectopic ureter: Analysis from a single centre with review of the literature

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ABSTRACT

Objective: Purpose of our study was to aggregate and analyse rare cases of Ectopic Ureter (EU), their association with other anomalies, clinical features, diagnosis and management.

Material and methods: A total of nine patients with rare presentation of EU were evaluated. Combination of endoscopic and imaging modalities was used as required to define the anatomy and devise the best surgical approach in these cases.

Results: Among six females and three males with EU, four cases had bilateral EU, four unilateral EU and one case had EU of a solitary kidney. Urinary incontinence was encountered in five cases including one male patient whilst other cases presented with varied clinical features and associated anomalies. Two patients had anorectal malformations, and two had uterine anomalies in the form of bicornuate uterus. Other patients had multiple rare associations such as triplication of ureter, bilateral absence of seminal vesicles with infertility, multicystic dysplastic kidney, ureter draining in uterus, renal failure, absence of bladder trigone, and hypospadias etc. Ureteric reimplantation was performed in four cases, two required ureteroureterostomy, another two had undergone upper pole nephrectomy and in one case renal transplant had been carried out owing to chronic renal failure.

Conclusion: EU is among group of those congenital entities which remain shrouded until adulthood, when symptoms become distressing. Fortunately, prognosis is favourable after surgical correction, in spite of its rarity. Complexity arises when other associated anomalies are identified.

Keywords: Congenital abnormalities; ureteral diseases; urinary incontinence.

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Introduction

Ectopic ureter (EU) with an incidence of 1:2000 and 1:4000 in autopsies is defined as any ureter, single or duplex, that opens at places other than the bladder trigone.[1] More than 80% of females with EU have duplex systems and majority (75%) of males with EU had single collecting system.[2,3] Bilateral duplex system is found in approximately 17-33% the cases with female predominance.[4,5] An EU can drain into the urethra, Wolffian, or Müllerian structures. Apart from being a part of renal–urinary tract malformations which are classified under the term "congenital anomalies of the kidney and urinary tract", EU can present with various other anomalies. Urinary tract abnormalities in conjunction with anorectal malformations occur in approximately 60% of the cases, that are attributed to abnormal development of cloacal membrane, however, presence of EU is extremely rare.[6] Location of the ureteric buds, their interplay with the developing kidney and associated malformation is the basis of clinical significance of these anomalies. Presentation varies according to the sex, position of the ureteral opening and associated anomalies. Management in such cases should be clearly defined and factored into the clinical decisions. Our study is unique as it is a segregation and retrospection of some rare cases of EU, which are often found in the

literature as case reports only. This study will enquire into etiology, pathophysiology, diagnosis, and clinical management of such cases with some interesting images.

Material and methods

From January 2014 to 2018, nine patients with rare presentation of EU were treated in the Department of Urology at tertiary care hospital in Eastern India. The approval from institutional ethical committee (IPGME&R/IEC/215/27.03.2017) and informed consent from patients (parents in case of children) were received. Only those cases of EU which were rare and/or associated with some rare anomalies were included. Cases of EU which were common and not associated with any other anomalies and required conventional treatment were not included in the study. We scrutinized the available medical records of patients and analysed parameters including etiopathogenesis, clinical presentation, associated anomalies, diagnosis, treatment undertaken and outcome.

Task of seeking EU opening was accomplished by various radiological imaging like ultrasonography (USG), intravenous urogram (IVU), voiding cystourethrogram (VCUG), computed tomography (CT) urogram, and magnetic resonance (MR) urogram. Whenever required, to evaluate renal function radionuclide renal imaging was performed. Endoscopy and specific manoeuvres were used to further delineate anatomy.

After thorough evaluation and confirmation of adequacy of renal function, appropriate surgical procedure was planned. Accordingly laparoscopic and open surgeries were performed and followed up. Clavien-Dindo grading system was used to classify surgical complications.

Results

Enumeration of cases with demographic data, clinical presentation, associated anomalies and management in various cases of EU is depicted in Table 1. The study population consisted of 6 female and 3 male patients. Variegated clinical presentations were seen among patients aged from 6 to 45 years. All three male patients had single system EU while female patients had either unilateral or bilateral complete duplicated system. One of the female patients had Smith type-3 triplication of ureter, in which triple ureter joined into a single ureter which opened ectopically into vaginal vault.[7] All except one female patient had chief complaint of urinary incontinence with or without associated symptoms of recurrent urinary tract infection (UTI) and flank pain. A 22-year-old female had only complaint of left flank pain without urinary incontinence. All three male patients had complaints of flank pain and recurrent UTI; curiously, one of the male patients also had history of occasional urinary incontinence (Case-1).

Interestingly, a 15-year-old male patient (Case-1) had constellation of anomalies including anorectal malformations, hypospadias, bilateral single system EUs opening into prostatic urethra and absent trigone (Figure 1a). Anorectal malformation was repaired in infancy. For urological complaints he underwent bilateral ureteric reimplantation by Politano-Leadbetter technique and hypospadias repair.

Rare association of multicystic dysplastic kidney (MCDK) with pelvic kidney and EU was identified in a 16-year-old male (Case-2). Patients presented with growth retardation, renal failure, abdominal lump and recurrent pain abdomen. His left kidney was non-functioning MCDK and the right one was a small hydronephrotic pelvic kidney with ureter opening into prostatic urethra (Figure 1b-d). Since both kidneys were non-functioning, renal transplantation was contemplated and surgical correction was not attempted.

Another 45-year-old male (Case-3) presented with single hydronephrotic right kidney with EU opening into prostatic urethra and deranged renal function (Figure 2a, b). Associated anomalies were left renal agenesis, bilateral seminal vesicle agenesis and primary infertility. An open ureteric reimplantation was performed.

An atypical scenario of bilateral EU with right ureteric triplication in a 21-year-old female was successfully managed by laparoscopic right ureteric reimplantation (case-4/Figure 2c, d). This patient had complaints of urinary incontinence due to right triplicate ureter opening into a vaginal vault.

Bilateral complete duplex system was seen in two female patients (Case-5, 7). Case-5 had right upper moiety ectopic refluxing ureterocele and left upper moiety EU opening at urethrovaginal septum (Figure 3a, b). In this case, right common sheath reimplantation was done followed by left upper pole nephrectomy as it was non-functioning.

A 13-year-old female (Case-6) had an anarchic presentation with right upper moiety EU opening in uterus. The computed tomography urogram exhibited a right sided duplex moiety with an EU draining into the uterus (Figure 3c). Ureteric opening was not seen on vaginoscopy however there was continuous dribbling of urine from the cervix. A possible uterine opening of the EU was demonstrated as ureter was delineated on injecting contrast material into uterus. Laparoscopic end-to-side ureteroureterostomy into the normal ureter of the lower moiety was performed.

Among the two cases of unilateral complete duplication one had (Case-8) anorectal malformation with upper moiety EU opening into the vagina, so ureteroureterostomy was executed. A patient (Case-9) with bicornuate uterus had an upper moiety ureter

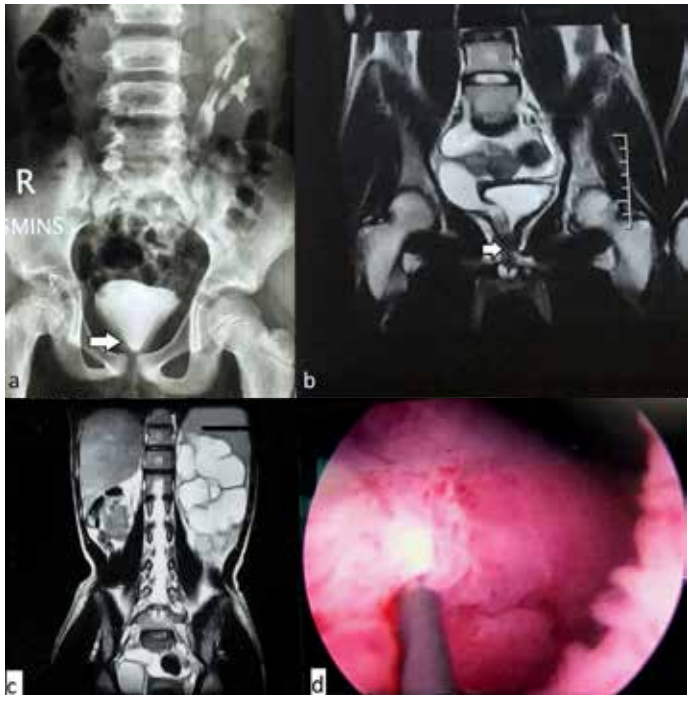

Figure 1. a-d. IVU in a patient with absent trigone (a); MR urogram depicting ectopic ureter of the right pelvic kidney draining into prostatic urethra and left kidney of same patient with multiple cysts (b, c); endoscopic view of the same patient showing right ureter opening into prostatic urethra (d)

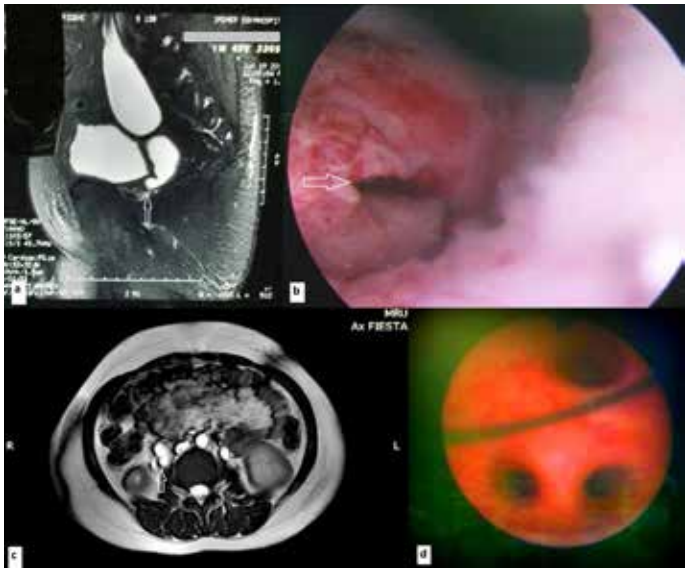

Figure 2. a-d. MR urogram showing ectopic ureter of a single kidney opening into prostatic urethra and endoscopic view of the same patient (a, b); MR urogram showing triplication of the right ureter and ureteroscopic view of the same patient (c, d)

opening at vaginal vault. She had sole complaint of flank pain. Renal angiogram was suggestive of non-functioning upper pole (Figure 3d) hence laparoscopic upper pole nephrectomy was

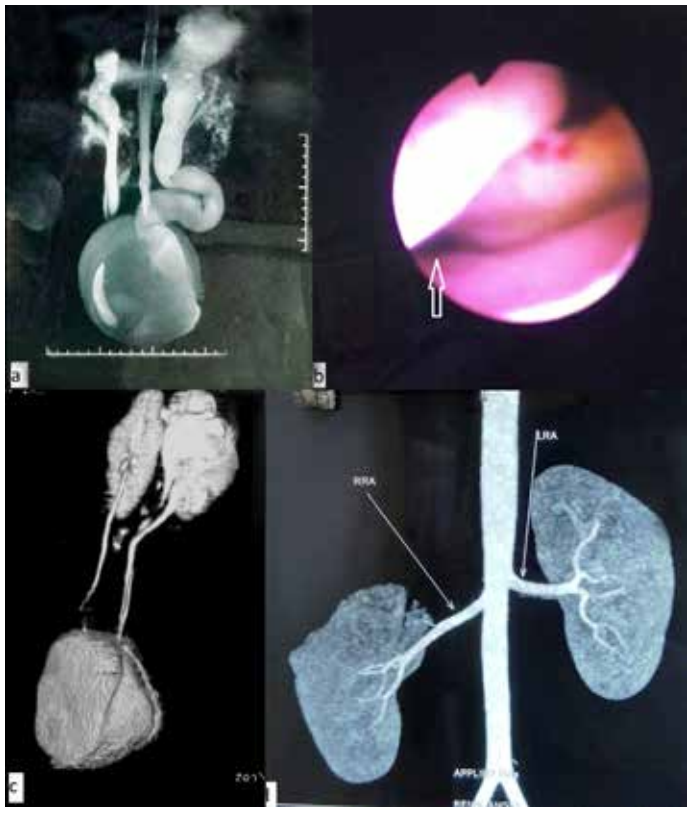

Figure 3. a-d. Right upper moiety ectopic ureterocele and left upper moiety ectopic ureter on MR urogram (a); Right ectopic refluxing ureterocele on cystoscopy of the same patient (b); CT urogram depicting ectopic ureter opening into uterus (a); Right nonfunctioning upper moiety on CT angiogram (d)

performed. At a minimum of 6-month-follow-up, a significant improvement in all surgically managed patients was imminent both anatomically and physiologically.

Discussion

If a ureter migrates with the Wolffian duct structures, it is embarked elsewhere other than the trigone, thus termed as an EU. Its true incidence remains obscured, owing to its asymptomatic course in the majority of cases. Most common congenital anomaly in the urinary tract is duplication with an incidence of 0.7% found in one series of more than 50,000 autopsies.[8] Apparently, EU are commonly encountered in duplicated system as compared to single system. Females are affected two to four times more commonly than males whereas right and left collecting systems are affected equally. Bilateral duplications occur in 17-33% of cases.[5] Triplication of ureter is extremely rare. Smith classified ureteral triplications in four types as follows: type-1 is complete triplication (35%), type-2 where three ureters join to form two ureters (21%); type-3 is trifid ureter (31%) and type-4 is double ureter, and one bifurcated one (9%).[7]

Table 1. Profile of the patients with ectopic ureter including presentation, associated anomalies, treatment and outcome

Case, Age (Years)/Sex	Ectopic ureter opening	Rare presentation and/or association	Treatment	Outcome (minimum follow-up 6 months)
Case-1, 15/Male	Bilateral ectopic ureter opening into prostatic urethra	Anorectal malformation, absent trigone, hypospadias, urinary incontinence	Politano-Leadbetter bilateral ureteric reimplantation and hypospadias repair	No complaints of incontinence, voiding well at 1-year follow-up
Case-2, 18/Male	Ectopic ureter of the right pelvic kidney opening in prostatic urethra	Left multicystic dysplastic kidney, renal failure	Renal transplant	On regular follow-up after renal transplant
Case-3, 45/Male	Ectopic ureter of the single system opening into prostatic urethra	Renal agenesis, absent seminal vesicles, primary infertility	Open ureteric reimplantation	Anatomic and functional improvement noted after 6-months
Case-4, 13/Female	Right upper moiety ureter draining in uterus	Ureter draining into uterus	Laparoscopic end-to-side ureteroureterostomy	Continent after surgery
Case-5, 20/Female	Triplication of the right ectopic ureter opening into vagina. Left upper moiety ureter opening in vaginal vault	Smith type-3 triplication of ectopic ureter opening into vagina. Incomplete trigone development	Right laparoscopic ureteric reimplantation	Continent with anatomic and functional improvement
Case-6, 21/Female	Right upper moiety refluxing ectopic ureterocele, left upper moiety ureter in urethrovaginal septum	Bicornuate uterus, right refluxing ectopic ureterocele	Left upper pole nephrectomy, Right common sheath reimplantation	No reflux, no incontinence
Case-7, 9/Female	Bilateral upper moiety ureter opening into vagina	Bilateral ectopic ureter draining into vagina	Bilateral common sheath reimplantation	Continent with anatomic and functional improvement
Case-8, 11/Female	Right upper moiety ureter opening into vagina	Anorectal malformation	Ureteroureterostomy	Continent with anatomic and functional improvement
Case-9, 22/Female	Left upper moiety ectopic ureter draining into vagina	Bicornuate uterus, ectopic ureter without incontinence	Laparoscopic left upper pole nephrectomy	Satisfactory improvement in pain at one year follow-up

A close embryologic relation exists between the development of the genitourinary organs and lower gastrointestinal tract.[9] EU is frequently found in association with vertebral, anorectal, cardiovascular, tracheoesophageal, renal and limb anomalies (VACTERL) and other associated syndrome is anorectal malformation (ARM).[10] However, majority of EU cases are not part of a syndrome and their etiology is assumed to be multi-factorial with a causal interplay of genetic and environmental factors. Clinical manifestations of syndromic forms may vary and genetics and underlying mechanisms remain elusive. Wolffian duct serves as a template for formation of paramesonephric duct hence justifying the association between renal tract and uterine malformations.[11,12] Congenital uterine malformations result from abnormal formation, fusion or re-absorption of the Müllerian ducts during fetal life. The process may be partial or total and affect one or multiple parts of the female urogenital tract.[13,14] Hence renal tract defects are likely to be found in women with Müllerian duct malformations. In our study, two out of the nine patients had ARM and two had concomitant uterine malformation. Congenital anomalies of the kidney and urinary tract are the most common cause of all birth defects, constituting 23% of all such defects and EU is part of it.[15] One of our patients presented with bilateral renal anomalies in the form of MCDK and nonfunctioning hydronephrotic kidney due to EU and presented with renal failure. In patients with duplicated collecting system and EU, it's not unusual to come across dysplastic upper pole of the kidney. A patient, who had presented to us with complaint of pain, was later found to have complete dysplasia of left upper moiety of kidney.[16]

Symptoms of EU in every patient are linked directly to the position of its outlet. Male patients with EU enjoy normal continence if the ureteric opening remains proximal to the sphincter and connected to the structures that continue to drain into the bladder. The most common outlet of EU in males is the prostatic urethra (57%), followed by seminal vesicle (33%), ejaculatory duct and vas (10%). While in females, it frequently opens into urethra, vagina and perineum. Incontinence of urine is the fundamental presentation in female population since the sphincter is bypassed.[17] Although, infection may be present, continuous dribbling of urine despite normal voiding is prototypical. Curiously, one of our female patients denied any symptoms of incontinence. This might be attributed to non-functioning of upper moiety. Bilateral single EU, associated with bladder neck malformation and lack of sphincter control, is one exception.[18] Usual complaints in males are flank pain, fever, urinary tract infection, abdominal mass or epididymitis.[19]

Rare presentations of EU present diagnostic dilemmas and difficulty in decision making. USG and VCUG are indicated in the initial evaluation of patient suspected of having a ureteral anomaly followed by IVU, CT urogram or MR urogram whenever required. Endoscopy with retrograde pyelogram is valuable to further delineate anatomy. Once detected, considering the different options in the management, a radionuclide renal scan becomes necessary in the decision-making.[20]

The surgical principle in the management of EU is to restore it anatomically while preserving the renal function. The prognosis in most cases is excellent. Ureteric reimplantation is performed in most cases if the part of kidney is functioning. Ureteroureterostomy is required, if one ureter is relatively short. Laparoscopic and open surgeries are both feasible with good long-term results. Collocation of such cases demonstrates melange of presentations, challenges in diagnosis and individualizing appropriate management.[21]

In conclusion, EU is a well-recognized entity, however close embryological relation between various urogenital and lower gastrointestinal tract can lead to multiple anomalies in a syndromic or non syndromic form. Presence of renal failure, MCDK, though rarely, all sites of opening of EU associated with anorectal and urogenital malformations can be seen with EU. Variety of surgical techniques are available for reconstruction of EU so urologist must remain familiar with the etiopathogenesis and well versed in the evolving recommendations for further management.

Ethics Committee Approval: Ethics committee approval was received for this study from the ethics committee of IPGME&R KolkataIEC (215/27.03.2017).

Informed Consent: Written informed consent was obtained from patients (parents in case of children) who participated in this study.

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