Case Report

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Successful management of single system ectopic ureter with preserved renal function in a female child: a case report

Jitendra Kumar Grover, Deepak Kumar Soni*, Shehtaj Khan

Department of General Surgery, L. N. Medical College, Bhopal, Madhya Pradesh, India

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*Correspondence:
Dr. Deepak Kumar Soni,

E-mail: dil.deepaksoni@gmail.com

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ABSTRACT

Ectopic ureter is a rare congenital anomaly of the genitourinary system, in which ureter does not insert into its normal anatomical position in bladder. Incidence is 1 in 4000 and has female predominance. Ectopic ureter is generally (80-85%) associated with a duplication of renal system, while in single system is found in 15-20%. If ectopic ureter drains a single kidney it is known as single system ectopic ureter (SSEU) in a female is extremely rarely found. Here, we report a case of single system ectopic ureter without renal dysplasia and other congenital abnormalities, which was diagnosed and managed successfully with uretero-neocystostomy. In conclusion, once a female is presented with urinary incontinence after toilet training then evaluation should be done for ectopic ureter along with other possible condition and management should be as early as possible & proper surgical intervention done.

Keywords: Continuous wetting, Single system ectopic ureter, Uretero neocystostomy, Congenital anomalies

INTRODUCTION

Ectopic ureter is a rare congenital anomaly of the genitourinary system, defined as a ureter that does not insert into its normal anatomical position. The occurrence of ectopic ureter is 1 in 4000. Gender ratio is 2 to 6:1 with female predominance & it shows racial differences.¹ In most of the cases (80-85%) ectopic ureter is associated with a duplicate renal collecting system, while in 15-20%, single system is found.² If ectopic ureter drains a single kidney it is known as single system ectopic ureter (SSEU). Ectopic ureter draining a single system in a case of a female is extremely rare.² The single system ectopic ureter is a rare condition which is usually associated with non-functioning dysplastic condition and other congenital anomalies like congenital heart disease, spinal cord malformation and ano-rectal malformation. In males, most common site of lower end of ectopic ureter is posterior urethra (47%) while in females the most common sites are bladder neck and upper urethra (33%), vaginal vestibule (33%), vagina (25%) and less commonly the cervix and uterus (<5%).^{1,2} Here, we report a case of single system ectopic ureter without renal dysplasia and other congenital abnormalities, which was diagnosed and managed successfully with ureteroneocystostomy.

CASE REPORT

A 3 year old female child presented to pediatric surgery department with history of continuous wetting inspite of normal micturation pattern since birth. Parents consulted a doctor when she complained of abdominal pain 2 months back, pain during micturitio for 15 days and fever for 5 days. On examination general condition of the child was satisfactory, vitals stable and no neurological deficits were seen. The external genitalia were female phenotype and urine dribbling was seen. On investigation Renal function test were normal. Computed tomography (CT)

urography showed grossly dilated right pelvicalyceal system and ureter, with thinning of right renal cortex. The distal portion of ureter showed extension into the expected location of upper vagina (Figure 1). Findings were suggestive of chronic ureteric obstruction and back pressure changes leading to gross hydronephrosis and hydroureter (Figure 1). Excretion of contrast was seen from the right kidney suggesting functioning of right kidney. Urinary bladder, left kidney and ureter were normal, so we planned for surgical intervention. On exploration, right ureter was dilated and terminated into right lateral wall of vagina. Right ureteric re-implantation (extravesical ureteroneocystostomy) was performed (Figure 2). Follow-up till 3 months was uneventful.



Figure 1: CT urography showing grossly dilated right renal pelvicalyceal system and ureter.



Figure 2: Right ureteric re-implantation.

DISCUSSION

Association of single system ectopic ureter with functioning bilateral kidney and absence of congenital anomalies is rarely reported in the literature. The term "ectopic ureter" has universally been used to describe the ureter that terminates at the bladder neck distally in one of the mesonephric duct structures or that is incorporated into any of the nearby mullerian structures. This anomaly comprises a variety of ureteral insertions shifting from nearly normal to extravesical location. In men, the extravesical ectopic orifice is always suprasphincteric and enters prostatic urethra, seminal vesicle, ejaculatory duct or vas deferens.³ In women, the ectopic orifice is either suprasphincteric or infrasphincteric, with most common insertion into urethra or vestibule and rarely in fallopian tube, cervix or vagina.3 In males single system ectopic ureter doesn't present with incontinence but as recurrent epididymitis, vesiculitis, prostatitis, orchitis, bloody and painful ejaculation. In females, suprasphincteric ureteral opening is diagnosed during investigation for etiology of recurrent urinary tract infection. An ectopic ureter that drain either into the urethra distal to the sphincter (infrasphincteric) or in to the vagina typically present with classical symptoms of urinary incontinence like continuous wetting, despite of normal micturition pattern.4 Other conditions associated with ectopic ureter are recurrent UTI and obstructive uropathy. These two risk factors finally leads to chronic renal failure.In females, differential diagnosis of this condition are voiding dysfunction, overactive bladder and ureterocele.⁵ Imaging studies are compulsory to confirm the diagnosis. USG KUB is the initial imaging modality. Contrast enhanced computed tomography (CECT) or magnetic resonance imaging (MRI) urography is investigation of choice.6 Dimercaptosuccinic acid (DMSA) scan is an accurate diagnostic method for associated dysplastic kidney.7

Symptoms, renal functions (most important), patient age and life quality decide the management of ectopic ureter.² Dysplastic kidney and those with function of less than 10% are normally removed.⁴ Surgical treatment consists of partial nephrectomy and laparoscopic ureteral ligation (clipping) in duplex system and ureteral reimplantation in case of preserved renal function. In case of single system ectopic ureter with dysplastic kidney, laparoscopic simple nephrectomy is recommended.⁷ In patient with significant renal function, either open or laparoscopic ureteral reimplantation is treatment of choice for single system.

When there is a preserved kidney function or functioning upper pole duplex system kidney, procedure consists of ureteroneocystostomy or distal and proximal ureteroureterostomy respectively. No clear co relation of renal function with the site of the ectopic ureteric orifice has been found, as most of patients with vaginal ectopic ureter have sufficient renal function to justify renal preservation. Ureteric re-implantations preserve renal function although improvement after surgery is

determined by the degree of renal dysplasia. In our case there was preserved renal function and because there was no dysplasia, we did ureteroneocystostomy. To our knowledge, Kumar et al, has reported a case of bilateral SSEU which was managed by bilateral ureteric reimplantation. Prakash et al reported a case of SSEU drainage into gartner's cyst which was managed by laparoscopic ureteral reimplantation.

CONCLUSION

Although, case of ectopic ureter are reported in literature, it is rare to have a normal kidney function without renal dysplasia, which may be cured with proper surgical management. Urinary incontinence in children after toilet training should be evaluated properly for ectopic ureter, especially in females so that management can be done early and patient may not suffer with psychological development.

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REFERENCES

- 1. Duicu C, Kiss E, Simu I, Aldea C. A rare case of double-system with ectopic ureteral openings into vagina. Front Pediatr. 2018;6:176.
- Kumar S, Bera MK, Bera KP, Vijay MK, Kundu AK. Laparoscopic ureteric reimplantation of a single-system ectopic ureter in a girl: A rarity. J Minim Access Surg. 2010;6(3):80-2.
- 3. Prakash J, Singh BP, Sankhwar S, Goel A. Normal functioning single system ectopic ureter draining

- into a Gartner's cyst: laparoscopic management. BMJ Case Rep. 2013;2013:bcr2013009563.
- Gangopadhyaya AN. Single system ectopic ureter in females: A single center study. J Indian Assoc Pediatr Surg. 2007;4:202-5.
- 5. Hanson GR, Gatti JM, Gittes GK, Murphy JP. Diagnosis of ectopic ureter as a cause of urinary incontinence. J Pediatr Urol. 2007;3(1):53-7.
- Figueroa VH, Chavhan GB, Oudjhane K, Farhat W. Utility of MR urography in children suspected of having ectopic ureter. Pediatr Radiol. 2014;44(8): 956-62.
- Lee YS, Im YJ, Kim SW, Kim MJ, Lee MJ, Lim NL, Han SW. The vagaries of proper imaging in diagnosing single-system ectopic ureter in children with continuous incontinence and outcomes of simple nephrectomy. J Pediatr Surg. 2016;51(3): 469-74.
- 8. Ghosh B, Shridhar K, Pal DK, Banerjee M. Ectopic ureter draining into the uterus. Urol Ann. 2016; 8(1):105-7.
- 9. Kumar A, Goyal NK, Trivedi S, Dwivedi US, Singh PB. Bilateral single system ectopic ureters: case report with literature review. Afr J Paediatr Surg. 2008;5(2):99-101.
- 10. Wakhlu A, Dalela D, Tandon RK, Chandra H, Wakhlu AK. The single ectopic ureter. Br J Urol. 1998;82(2):246-51.

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