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Received: June 19, 2021
Accepted: Nov 1, 2021

Citation:
Heywood S, Shrestha SD. Diagnosis and management of ectopic ureter in a low resource setting – A case series. Nep J Obstet Gynecol. 2021;16(33):105-112.
DOI: https://doi.org/10.3126/njog.v16i2.42111

Diagnosis and management of ectopic ureter in a low resource setting – A case series

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ABSTRACT

Aims: To review the experience of women and girls treated for ectopic ureter in Surkhet through the fistula camps and Fistula Treatment Centre and analyse the diagnostic method, management and outcome.

Methods: This was a retrospective study of the patients with ectopic ureter who received treatment in Surkhet since 2009. Data were analysed from patient records and interviews.

Results: Seven patients were diagnosed with ectopic ureter over a period of 12 years. Five patients were prepubertal with age range of 7 - 15 years. History of continuous incontinence with normal voiding was suggestive of ureteric fistula in six out of seven cases. The seventh case had massive ureterocele in the distal end of the duplex right ureter. Ultrasound suggested a diagnosis of duplex collecting system in five cases. IVU or CT IVU suggested duplex ureter in only three cases. Diagnosis was confirmed by examination under anaesthesia after i.v. frusemide in six cases. All cases were managed by implantation of the ectopic ureter into the bladder. In the case with ureterocele heminephrectomy was performed. All seven patients are well at follow-up between 6 months and five years after presentation.

Conclusion: Ectopic ureter is a treatable cause of urinary incontinence and the diagnosis may be overlooked. A lifelong history of continuous urinary leakage with normal voiding, in absence of trauma or surgical procedure should raise the suspicion. Examination under anaesthetic and ultrasound proved most effective in confirming the diagnosis.

Keywords: diagnosis; ectopic ureter; urinary incontinence

INTRODUCTION

Urinary incontinence causes considerable distress and psychological disturbance to children and their parents. Ectopic ureter is an uncommon and often overlooked cause of urine incontinence. Delay in diagnosis and treatment results in many years of suffering and disturbance of social

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development and schooling.[1] There has been a service for repair of obstetric fistula in Surkhet since 2009. Initially this was delivered through annual fistula repair camps, based at the Surkhet government hospital until a designated Fistula treatment centre was opened at Provincial Hospital, Karnali Province in 2018. The fistula repair services in Surkhet have been publicised by radio and outreach programmes to health workers and communities since 2011. Among the patients responding there were several young girls with a life long history of continuous urine incontinence where investigation and management at other centres had not led to a diagnosis or relieved the symptoms. This study is a review of seven cases of duplex ectopic ureter who have been successfully treated in Surkhet. The aim of this study is to identify the investigations that proved helpful in confirming the diagnosis.

METHODS

This was a retrospective case series of seven cases of ectopic ureter diagnosed and treated in Surkhet between 2012 and 2021. From the case notes and patient interviews the presenting history, examination findings and investigations were reviewed to identify the factors that led to the diagnosis of ectopic ureter.

RESULTS

There were seven cases diagnosed and treated for ectopic ureter (one bilateral) among 337 confirmed cases of fistula presenting to the fistula surgeon in Surkhet [Table-1]. Five of the seven patients were pre-pubertal girls with age range 7 to 15 years. Three of these children had been investigated in centres in India and Nepal but investigations, including intravenous urography were inconclusive and had not led to a diagnosis of ectopic ureter. One child had been prescribed anticholinergic medication. These children were all attending school and reported isolation and distress due to adverse comments from school friends and one child had been beaten by her teacher because the classroom bench was wet.

The five children all gave a classical history of continuous urine incontinence with normal voiding. On examination in each case the vulva and clothing were found to be wet. One child did appear to have a small orifice just below the urethral meatus. To avoid causing further distress a more detailed examination was deferred until the patients were under anaesthesia.

Ultra sound examination was performed on all children and the request form advised that ectopic ureter was suspected. In four out of five cases the ultrasound report suggested at least duplex collecting system. In all cases the radiologist advised intravenous urogram (IVU) or computerised tomography (CT) but though performed in four of the five cases this investigation did not add to the evidence for ectopic ureter.

Examination under anaesthesia was performed on all five children either as a separate procedure or immediately preceding laparotomy. In four cases the opening of the ectopic ureter could be seen at the vestibulum with urine spurting after giving a small dose of intravenous frusemide. The fifth case was the child whose ectopic ureter had been reported on ultrasound and suspected on IVU and the

opening of the ectopic ureter was thought to be in the distal urethra.

The two adult women, aged 21 and 30 did not present with the classical history. Patient B aged 21 reported a four-year history of slight urine incontinence which had become much worse following childbirth one year earlier. She had normal vaginal delivery of a live baby after three days in labour. For the past year she had suffered continuous incontinence but was also voiding normally. On examination vulva and vagina were normal with no fistula evident but the vagina was wet and there was smell of urine. Dye test was negative, excluding a vesico-vaginal fistula but raising suspicion of ureteric fistula. Ultrasound scan reported a duplicated collecting system on the left and possible ureteric fistula. IVU reported a duplex left kidney with minimal hydronephrotic changes in the upper moiety. Review of the images by the surgeons raised suspicion of bilateral ectopic ureters. Therefore, at operation bilateral exploration was undertaken and the diagnosis confirmed. At preliminary cystoscopy single left and right ureteric orifices were identified in the bladder. Both ectopic ureters were seen opening into the urethra.

These six patients were all cured immediately after reimplantation of the ectopic ureters into the bladder.

The seventh patient (G) had a very different history with intermittent incontinence, mainly in the day time and sometimes initiated by cough. The symptoms had improved since pregnancy. She had three children aged 8-13 all born by normal vaginal delivery. She described the incontinence as starting in childhood when "something burst'. On examination there was a small cystocele but otherwise vulva and vagina were normal with no sign of fistula, no wetness, and stress incontinence was not evident. Dye test was negative. An ultrasound examination in the clinic revealed a cystic swelling beside the bladder on the right side. Para- vesical cyst was suspected. At operation there was found to be a cystic structure in the midline of the vagina but extending into the right side of the pelvis with a pin hole opening in mid vagina. The cystic structure was marsupialised and there was drainage of clear fluid. Ectopic ureter was considered. Ultrasound examination revealed situs inversus totalis with features suggestive of right ectopic ureter with insertion into the vagina. IVU reported segmental dilatation of the right distal ureter.

Vaginal examination prior to laparotomy revealed a little clear fluid draining from the small opening in the vagina but there was no response to frusemide challenge suggesting that there was little function in the upper pole of the right kidney drained by the ectopic ureter. Laparotomy, as expected, revealed a dilated right ectopic ureter ending in a large ureterocele. To reimplant the short ectopic ureter proximal to the ureterocele into the bladder a Boari flap was raised. On day five post - operation a second laparotomy was performed because of heavy urine leakage from the drain site. There was leakage from the anastomosis of the ureter to the Boari flap. A length of fallopian tube with mesent-

ery was sutured between the lower end of the ureter and the opening in the Boari flap. Initially the patient made good progress but on day 19 she experienced fever with rigors and on day 23 there was paralytic ileus and urine leakage from the wound indicating breakdown of the anastomosis. She was referred to Kathmandu where, after further investigation, a right heminephrectomy was performed with closure of the bladder defect. From this point she made an excellent recovery and at follow up was found to be completely well.

The patients home districts were spread across the three western provinces of Nepal and follow-up was conducted by telephone interview if the patient did not wish to return to Surkhet. The length of follow - up in this series was between six months and five years. All patients reported complete resolution of the urinary symptoms and were very happy with the outcome of treatment.

DISCUSSION

The human kidney and urogenital tract develop from three principal embryonic structures: the metanephric mesenchyme, the mesonephric (Wolffian) duct and the cloaca. At four to five weeks of gestation the ureteric bud originates as an outgrowth of the mesonephric duct. It branches to give rise to the collecting ducts, the renal pelvis, the ureter, and the bladder trigone. Ureteral duplication can develop if there are duplicate ureteral buds or early division of these buds. Duplication may be complete, with two separate ureters or partial, resulting in bifid renal pelvis with distal confluence into a single ureter; and 50% of cases are asymptomatic.

Ectopic ureter is defined as a ureter opening into an area outside the posterolateral extremity of bladder trigone and more than 80% of the cases are associated with the complete duplex collecting system.[2] Ectopic ureters are generally from the upper pole of a duplex kidney.[3] In females an ectopic ureter may open into the bladder neck, the urethra, the uterus, the proximal part of the vagina or the vestibule. It is most likely to be found distal to the urethral sphincter, resulting in continuous incontinence. Single system ectopic ureters are less common than duplex system and may be difficult to diagnose as the ectopic ureter usually drains a small dysplastic kidney which may be missed on imaging.[4]

Ureterocele, a cystic dilation of the terminal ureter is rarely reported in adults and is often associated with some urological anomaly such as a duplicated system or stenosis of the ureteric orifice. It may present with a variety of symptoms which may not include incontinence.[5]

A number of young girls have presented to the Surkhet fistula programme complaining of urinary incontinence due to a variety of causes including vesico-vaginal fistula, overactive bladder and neuropathic bladder due to spina bifida. A detailed history of the urinary incontinence will help to differentiate the cause.

In a patient with ectopic ureter there will be a normal voiding pattern. The amount of incontinence will depend on the function of the renal moiety drained by the ectopic ureter but the incontinence will be continuous by day and night. Examination of the child in the outpatient clinic may be limited to avoid adding to her distress but it is possible to confirm that there is wetness of the clothing and vulva.

Table-1: Summary of patient details (Case A through to G)

Variable	**A**	**B**	**C**	**D**	**E**	**F**	**G**
Age	10	21	15	7	11	10	30
Past tests	yes	no	yes	no	no	yes	no
Year	2017	2015	2012	2014	2021	2020	2013
History *	Life long typical	4 years typical	Lifelong typical	Lifelong typical	Lifelong typical	Lifelong typical	Minimal urine leak
Examination	wet	wet	wet	wet Orifice in vestibule	wet	wet	dry
Suspected diagnosis	Ectopic ureter	Ectopic ureter	Ectopic ureter	Ectopic ureter	Ectopic ureter	Ectopic ureter	Paraurethral cyst
Pre-op US report	R duplex ureter	Duplex left kidney	normal	Duplex L ureter	Normal	L duplex kidney and ureter	Cystic swelling to right side of bladder
Pre-op IVP report	Lucknow – normal report	L duplex collecting system and ureter	normal	no	CT IVU duplex L kidney single ureter	L duplex collecting system	no
EUA ±frusemide change	Ureteric orifice in vestibule	Cystoscopy ectopic ureteric orifice in urethra	Ureteric orifice in vestibule	Ureteric orifice in vestibule	Ureteric orifice in vestibule	Ureteric orifice in urethra	Massive cystic structure extends into R pelvis, pin hole mid vaginal punctum draining clear fluid - ?ureterocele
OT findings	R duplex ureter tortuous and dilated	Bilateral duplex, little dilatation	L duplex, normal calibre ureter	L duplex normal calibre ureter	L duplex dilated ureter	L duplex not dilated	Massive ureterocele terminal R duplex ureter
Surgery	Reimplant ectopic ureter	Reimplant bilateral ectopic ureter	Reimplant ectopic ureter	Reimplant ectopic ureter	Reimplant ectopic ureter	Reimplant ectopic ureter	Reimplant short ectopic ureter through Boari flap. Post op leak- referred for hemi-nephrectomy
Follow up	1 year cured	5 years cured	18 months cured	6 months cured	Cured at discharge	6 months cured	8 months cured

*NOTE: *Typical history of ectopic ureter - Continuous urine incontinence with normal voiding and no history of trauma or surgery*

In the earlier years of the fistula programme in Surkhet there was no radiology service in Surkhet district. Ultrasound service was provided by the general practitioners, surgeons and gynaecologists at the Surkhet Regional Hospital. For examination by radiologist or for IVU the patients were referred to Nepalgunj or Kohalpur. In all cases the suspected diagnosis was indicated when the patient was referred for imaging investigation. Ultrasound examination was able to identify duplex renal collecting system and less frequently a duplex ureter. A duplex ureter was identified in some cases where the ureter at operation proved of normal caliber and missed in other cases where the ureter was dilated. IVU had been performed for two patients before presentation in Surkhet and ectopic ureter had not been suspected.

Where there was still doubt about diagnosis the children were examined under ketamine anaesthesia. The ectopic ureteric orifice was identified in the vestibule in four cases.

IVU is an important investigation in the assessment of urinary incontinence in childhood but in our series did not add to the confidence of the diagnosis of ectopic ureter. The duplex collecting system was reported in only three cases and one ectopic ureter was seen in the patient where the condition was bilateral. IVU is now being replaced by CT Urography and Magnetic Resonance Urography(MRU) as the examination of choice since these demonstrate function of the kidneys as well as visualization of the collecting system but a wide range of investigations have been employed.[6-9] In our study IVU and even one CT IVU did not add to the evidence for ectopic ureter from ultrasound examination. In case D the diagnosis of duplex ectopic ureter was suspected on history and examination and confirmed by ultrasound examination and IVP was not performed.

Where there is high index of suspicion from the patient's history the films should be reviewed by the surgeons also. If there is no evidence of duplex kidney the possibility of single system ectopic ureter should be considered especially if single kidney is reported. A second dysplastic kidney should be sought. Cystoscopy would reveal a single ureteric orifice on the side of the normal kidney.

In a low resource setting access to such investigations is limited but the experience in Surkhet would suggest where detailed history leads to suspicion of ectopic ureter the ultrasound examination and examination under anaesthetic will provide sufficient confirmation to proceed with curative surgery.

Ectopic ureter rarely presents in the adult.[10] In this series the two adult patients had unusual presentation. Patient B aged 21 only noted urine incontinence in the last four years and particularly since childbirth. The reason for this is not clear. The ectopic ureters opened into the urethra. It is possible that the process of childbirth resulted in some prolapse of the bladder neck but this would only produce the late onset of incontinence if the ectopic ureters opened at the bladder neck. There is little in the literature about this situation. In

one series of late presentation of ectopic ureter two women developed uretero-vaginal fistula after gynaecological surgery and one, similar to our patient, developed incontinence after prolonged labour.[11] The patient with ureterocele had minor complaint of incontinence. Ureteroceles may present in a number of ways, through pressure effects, development of stones or recurrent urinary tract infection.[5] In our case the patient reported leak after "something burst" in childhood. This resulted in a pinhole fistula draining into the mid vagina. We have seen rare patients with pin-hole vesico-vaginal fistula who have also given a history suggestive of stress incontinence rather than fistula, since they only experienced leakage of urine on coughing or other such stimulation.

Examination under anaesthesia was found to be the most effective investigation to confirm the diagnosis before laparotomy. The results of this series suggest that, with high index of suspicion, a detailed history of incontinence and the availability of ultrasound examination, supported by examination under anaesthesia with frusemide challenge, a confident diagnosis should be possible in most cases. Once the diagnosis is made surgery is curative.

The management principles for ectopic ureter are to preserve renal function, prevent infection and to restore continence.[2] The surgical options include hemi-nephrectomy, uretero-ureterostomy and ureteric reimplantation or uretero-neocystostomy whether through open surgery or when available by laparoscopic surgery.[9] Reimplantation of the ureter is ideally perfo-rmed with extraperitoneal approach. Where both ureters were of normal caliber insertion of ureteric catheter into the normally inserted ureter allowed confident identification of the ectopic ureter and in one case where the ureters were too small to allow passage of the available 5Fr ureteric catheter both duplex ureters were re-implanted.

This surgery is not within the normal practice of the general gynaecologist but a fistula treatment centre can expect to see such cases due to the common history of continuous urine incontinence. The Surkhet fistula project was fortunate to have the services of an expatriate visiting consultant uro-surgeon for many years.

CONCLUSIONS

This study showed that ectopic ureter though an uncommon cause of urinary incontinence, can be diagnosed with confidence and managed successfully in a low resource setting. The diagnosis should be considered where there is a history of life long urinary incontinence with a normal voiding pattern. Where access to CT Urography or MR Urography is not available the diagnosis can still be confirmed by ultrasound examination and examination under anaesthesia with frusemide challenge. This distressing condition can be cured immediately by appropriate surgical treatment.

REFERENCES

1. Wang MH. Persistent Urinary Incontinence: A Case Series of Missed Ectopic Ureters. Urol Case Rep. 2015;3(6).
2. Demirtas T, Tombul ST, Golbasi A, Sonmez G, Demirtas A. The ectopic ureter opening into the vulva, which is a rare cause of lifelong urinary incontinence: Treatment

with ureteroureterostomy. Urol Case Rep. 2021;36.

3. Mishra K, Elliott CS. A violation of the Weigert-Meyer law – an ectopic ureter arising from the lower renal pole. J Clin Urol. 2017;10(3).
4. Grover JK, Soni DK, Khan S. Successful management of single system ectopic ureter with preserved renal function in a female child: a case report. Int Surg J. 2021;8(3).
5. Yuri P, Utama ETP. A complete duplicated collecting system with giant ureterocele in adult: Case report. Int J Surg Case Rep. 2021;79:49–52.
6. Abrams,P, Cardozo, L, Wagg, A, Wein, A. (Eds) Incontinence 6th Edition (2017). ICI-ICS. International Continence Society, Bristol UK, ISBN: 978-0956960733. Available from: https://www.ics.org/publications/ici_6/Incontinence_6th_Edition_2017_eBook_v2.pdf
7. Demir M, Çiftçi H, Kılıçarslan N, Gümüş K, Oğur M, Gülüm M, et al. A case of an ectopic ureter with vaginal insertion diagnosed in adulthood. Turk Urol Derg. 2015;41(1).
8. Figueroa VH, Chavhan GB, Oudjhane K, Farhat W. Utility of MR urography in children suspected of having ectopic ureter. Pediatr Radiol. 2014;44(8).
9. Roy Choudhury S, Chadha R, Bagga D, Puri A, Debnath PR. Spectrum of ectopic ureters in children. Pediatr Surg Int. 2008;24(7).
10. Toia B, Pakzad M, Hamid R, Wood D, Greenwell T, Ockrim J. Diagnosis and surgical outcomes of ectopic ureters in adults: A case series and literature review. Neurourol Urodyn. 2019;38(6).
11. Blacklock A, Shaw R, Geddes J. Late Presentation of Ectopic Ureter. Br J Urol. 1982;54(2):106–10.