Annals of Medicine and Surgery 84 (2022) 104824

Contents lists available at ScienceDirect

Annals of Medicine and Surgery

journal homepage: www.elsevier.com/locate/amsu

Case Report

Bilateral complete duplication of ureter with ectopic ureter presenting as persistent urinary dribbling with normal voiding pattern in 17-year-old female: Case report

Sampurna Singh [a,*], Narayan Prasad Bhusal [a], Sarmendra Mishra [a], Santosh Singh [b], Kabindra Rai [a], Shiwani Bista [a], Suprabha Amatya [a], Shreeja Kisee [a]

[a] Department of Surgery, Kist Medical College, Kathmandu, Nepal
[b] Department of Surgery, Gandaki Medical College, Pokhara, Nepal

ARTICLE INFO

Keywords:
Recurrent urinary tract infections
Urinary incontinence
Upper pole heminephrectomy
Ureterectomy

ABSTRACT

Introduction: and importance: Bilateral complete ureteral duplication is a rare urinary tract abnormality and presence of unilateral ectopic ureter makes it a rarest entity. Continuous urinary dribbling with normal voiding pattern and recurrent UTI are frequent presenting complains in case of ectopic ureter. Long term childhood problem of urinary incontinence with delayed diagnosis in adult in this case makes it a perfect case to report.
Presentation of case: We present a case of 17-year-old girl with continuous urinary dribbling and constant wetting with normal voiding pattern since childhood. She also had recurrent history of urinary tract infections.
Clinical findings and investigations: Local external examination of genitourinary system was found to be normal. There were skin lesions on buttocks and thighs suggestive of urine dermatitis. There were no examination findings of other congenital anomalies. Complete blood count, Renal function test, Serology, Urine R/ME, Urine culture and sensitivity, Ultrasound abdomen and pelvis, Computed Tomography scan, intraoperative cystoscopy were done.
Intervention and outcome: Right upper pole hemi nephroureterectomy was performed. She was post operatively managed with IV Fluids, antibiotics, analgesics, antipyretics and antiemetics. She is asymptomatic now and regularly followed up on OPD basis.
Relevance and impact: Congenital abnormalities of the genitourinary system like ectopic ureter should be clinically suspected in case of persistent urinary problems like urinary dribbling with normal voiding pattern and recurrent urinary tract infections. Upper pole heminephrectomy is an ideal choice of treatment in case of nonfunctional upper moiety. This case emphasizes the early detection of genitourinary abnormalities and provide the perspective on late diagnosis and management in such cases.

1. Introduction

1.1. Background and rationale

Ureteric duplication is urinary tract anomalies with the incidence in between 0.5% and 3%. Union of two proximal branches of ureter occurs before inserting into the bladder in partial duplication while two ureters insert separately into the bladder in complete one with later being rarer type [1]. Duplication of ureter is more common in female than male with higher prevalence in Caucasian females [2]. In a study of 5196 children and adults, the duplex systems were present in 1.8% (95) of the patients; complete duplication was present in < one third of the duplicated systems and bilateral duplex systems were present in 0.3% (16) patients [3]. Duplex collecting system is autosomal dominant in inheritance with incomplete penetrance [4]. Single mesonephric duct developing two separate ureteral buds best explains the development of complete duplex system with duplicated ureters inserting separately into the urinary bladder [5]. An ectopic ureter is one that inserts outside of bladder trigone. Incidence of ectopic ureter is 1 in 2000–4000 with female to male ratio of 2–6: 1 [6]. Lower pole ureter is more prone to reflux as it makes a shorter tunnel into the bladder while the anomalous insertion of the upper pole ureter makes it more prone to obstruction [7]. In our

* Corresponding author.
E-mail address: sampsin687@gmail.com (S. Singh).

https://doi.org/10.1016/j.amsu.2022.104824
Received 26 July 2022; Received in revised form 29 September 2022; Accepted 30 October 2022
Available online 5 November 2022

case, there was obstruction in the upper pole ureter presenting as hydroureteronephrosis in right side only. Here, we present a case of a 17-year-old girl who had presented with a history of continuous urinary dribbling and recurrent urinary tract infections. This case emphasizes the early detection of genitourinary abnormalities and provide the perspective on late diagnosis and management in such cases (see Figs. 1–9).

1.2. Guidelines: SCARE 2020 paper

This case report has been reported in line with the SCARE Criteria [8].

1.3. Patient information: demographics and presentation

A 17-year-old female, unmarried student, hindu by religion presented with the chief complain of continuous dribbling of urine since childhood and had to change 6 to 7 pads per day. However, she had normal voiding pattern and had no history of fever, burning micturition, dysuria, blood in urine, per vaginal bleeding and discharge at the time of presentation. She was deprived of necessitated health checkup and services due to financial restraints.

1.4. Past medical and surgical history

She had history of recurrent UTI which was treated in local medical centers. She had no known history of TB, Hypertension, Diabetes Mellitus. She had no surgical interventions done in the past.

1.5. Menstrual history

Menarche (k) = 15 years Menstrual cycle = 3–4 days/28 ± 2 days Regular, 4–5 pads partially soaked.

Dysmenorrhea absentIntermenstrual bleeding absent.

1.6. Drug and allergy history

No history of long-term drug intake and no known allergies till date.

1.7. Family history

She had no history of similar illness in the family.

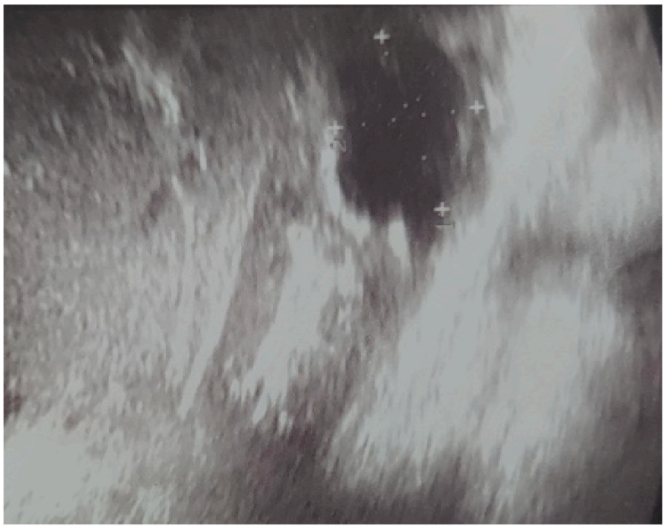

Fig. 1. USG image of right kidney showing dilated upper moiety and normal lower moiety.

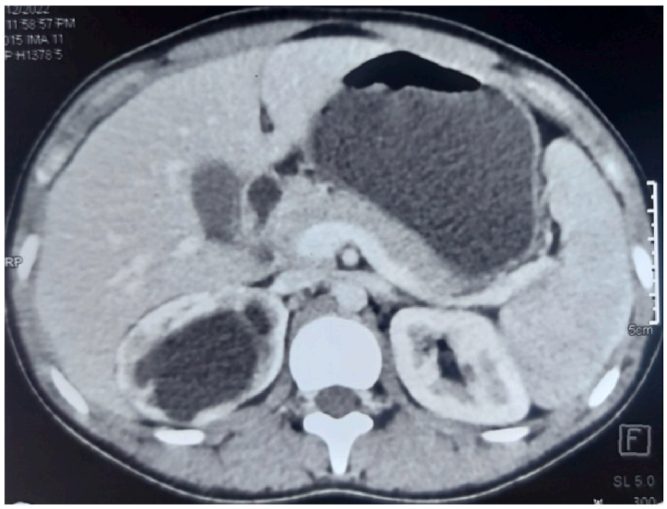

Fig. 2. CT IVU showing dilated upper moiety pelvicalyces in right kidney.

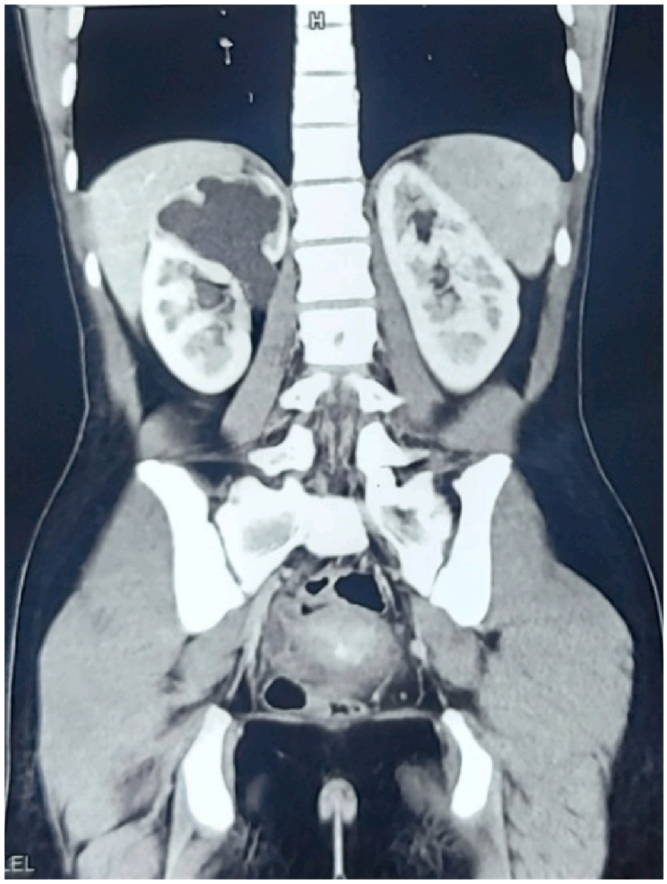

Fig. 3. CT IVU showing dilated upper moiety pelvicalyces in right side.

1.8. Social history

Does not consume alcohol, smokes 4 to 5 cigarettes per day.

1.9. Clinical findings

Patient was thin built with weight of 35 kg. Her vitals at the time of presentation were normal with pulse rate of 92 beats per minute, respiratory rate of 16 breaths per minute, body temperature of 97.8 F,

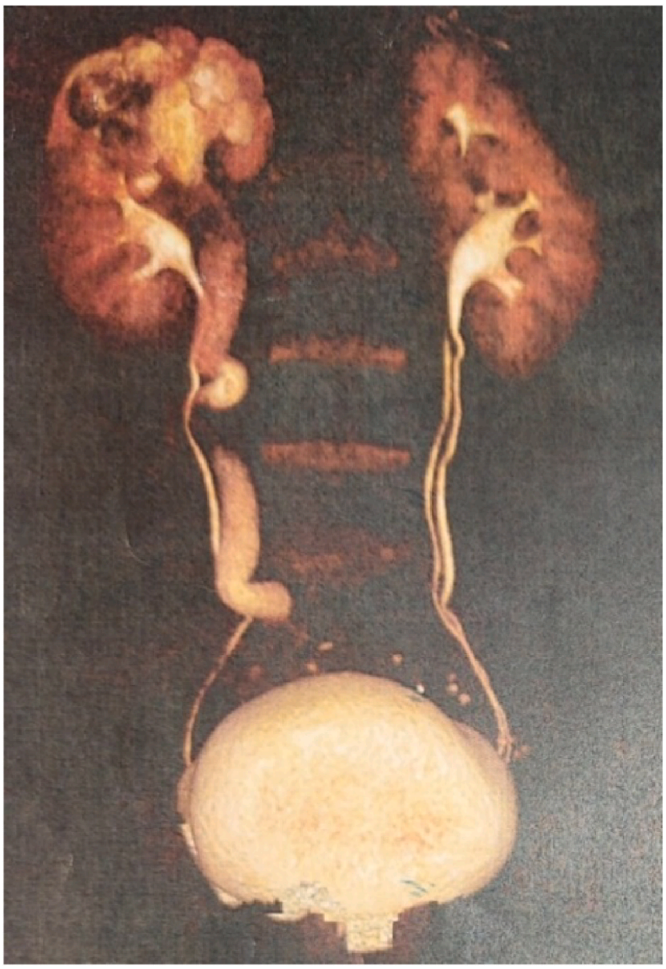

Fig. 4. 3D reconstruction of CT IVU showing bilaterally duplicated ureters and dilated upper moiety pelvicalyces with tortuous and dilated upper moiety ureter in right side.

blood pressure of 100/70 mm Hg and SpO2 of 96% in room air. Pallor, icterus, cyanosis, clubbing, lymphadenopathy, edema and dehydration were absent. Per abdominal examination was found to be normal.

1.10. Diagnostic assessment and interpretation

Relevant preoperative investigations were sent (Table 1).

Ultrasound of the abdomen and pelvis shows dilated upper moiety with normal lower moiety in right kidney however the left kidney shows no abnormal findings. In CT IVU, right and left kidneys were normal in size. On bilateral sides, duplex collecting systems with double ureters were present. On right side, there was dilated and tortuous upper moiety pelvicalyces with dilated upper moiety ureter draining below bladder neck into upper urethra below external urethral sphincter. However lower moiety ureter was normal in course and caliber and draining into normal site of urinary bladder. While on left side upper moiety ureter was seen draining separately into urinary bladder 5 mm inferomedially to the site of the opening of lower moiety ureter.

1.11. Intervention

After thorough history and relevant investigations, provisional diagnosis of bilateral complete duplication of ureter with right upper moiety gross hydroureteronephrosis was made. The patient was planned for right hemi nephroureterectomy as upper moiety in right kidney was nonfunctional. Intra operative cystoscopy was performed which showed

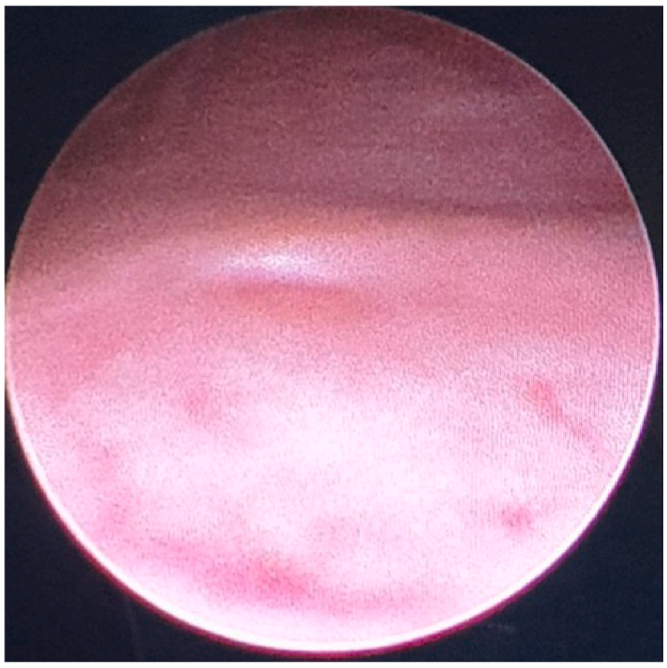

Fig. 5. Cystoscopy image showing one lower moiety ureteral orifice into the urinary bladder on right side.

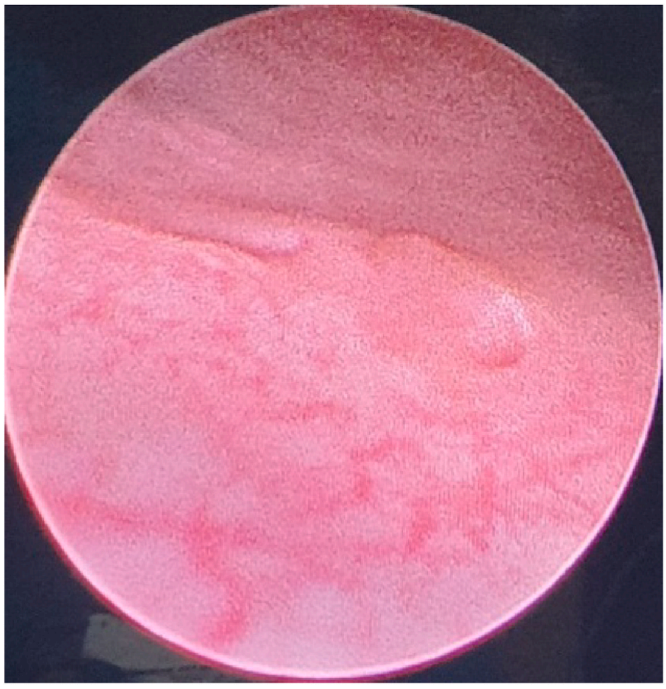

Fig. 6. Cystoscopy image showing two orifices into the urinary bladder on left side.

normal double ureteral orifices openings into the bladder in the left side while single lower moiety ureteral orifice opening in right side. Intra operatively, there was thinned out renal parenchyma with dilated upper moiety and ureter. Right hemi nephroureterectomy was done. Upper moiety ureter was excised as distal as possible and remaining small lower portion of ureter was left open to avoid the risk of cyst formation. The operation was uneventful and blood loss was minimal. As the left kidney was functioning properly and there were no complications, it was left untouched. The problem of urinary dribbling and incontinence was

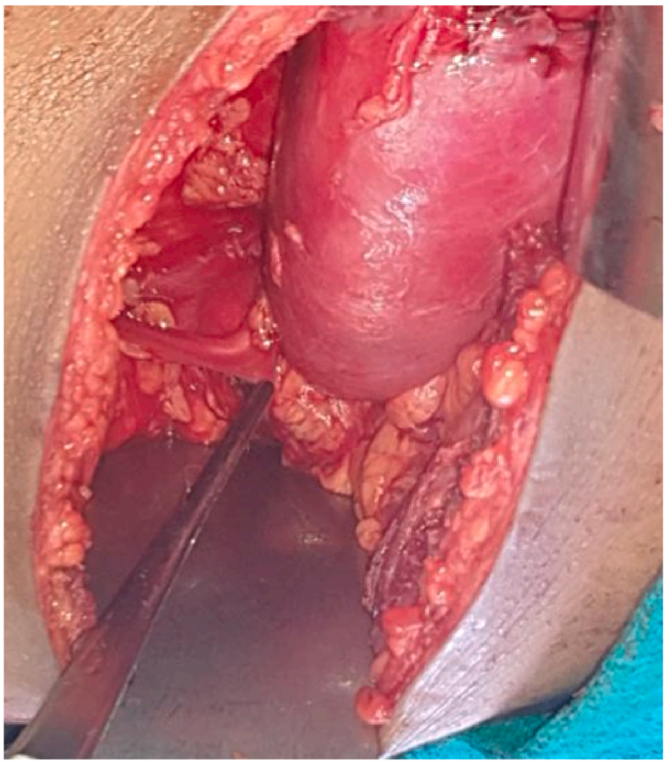

Fig. 7. Image showing right kidney with dilated upper moiety ureter.

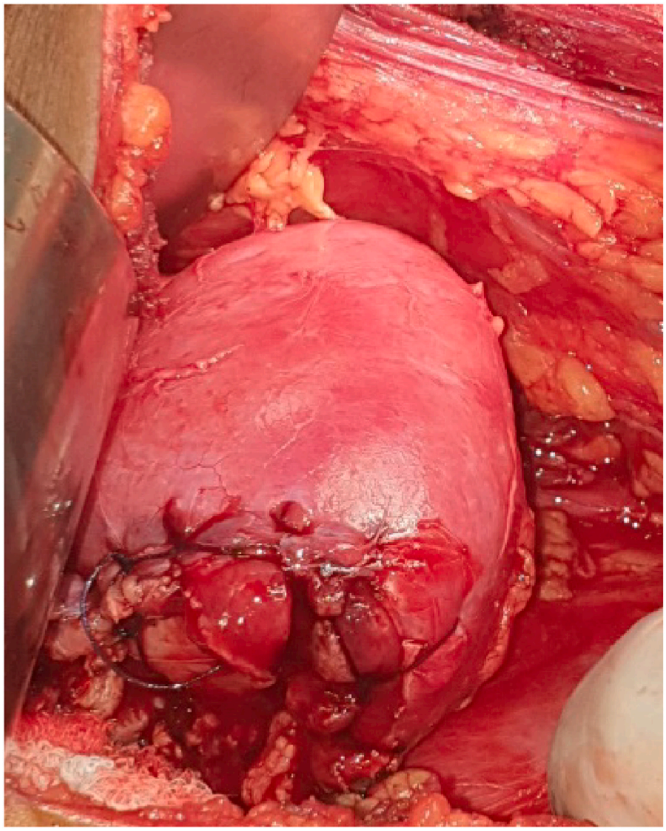

Fig. 8. Image showing right kidney after upper pole hemi nephroureterectomy.

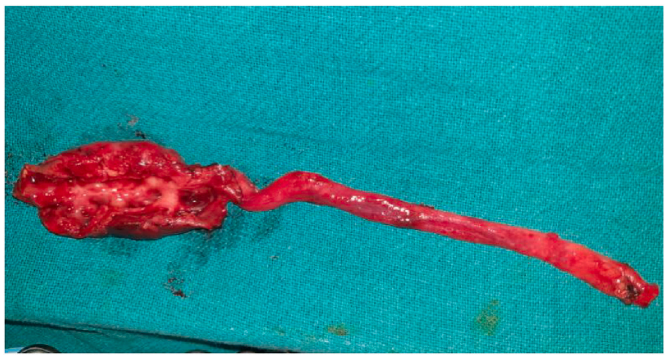

Fig. 9. Excised upper pole and upper moiety ureter of right kidney.

Table 1
Summary of laboratory test.

Laboratory test	Patient level	Unit	Reference range
Urea	13	Mg/dl	10–40
Creatinine	0.5	Mg/dl	0.3–1.2
Sodium	138	Mmol/L	135–146
Potassium	4.3	Mmol/L	3.5–5.2
Total leucocyte count	6900	/cumm	4000–11,000
Hemoglobin	11.5	Gm/dl	12–15
Blood sugar random	77	Mg/dl	70–140
Prothrombin time test	14.0		
International normalized ratio	1.0		
Blood grouping and Rh typing	AB positive		
Serology	Non-reactive		
Clotting time	6	Minutes	5–12
Bleeding time	2	Minutes	2–6
Urine culture and sensitivity	No growth after 24 hours of incubation at 37C		
Urine routine	WBC: 2–4/HPF RBC: Nil/HPF		

relieved post operatively and the patient was managed with IV fluids, antibiotics, analgesics, antiemetic and antipyretic. Post operative renal function tests were normal. Post operative period was uneventful and patient had recovered well.

1.12. Outcome

The patient has been followed up regularly in OPD basis and she had no complains thereafter.

1.13. Intervention settings and name of clinician

Dr. Narayan Prasad Bhusal, Department of Surgery, Kist Medical College.

2. Discussion

Ureteric duplication can be asymptomatic. However, the patients can present with abdominal or flank pain, hematuria and complications such as urolithiasis, urinary tract infections, vesicoureteral reflux, urinary incontinence, ureteroceles and associated other congenital anomalies like ectopic ureter. Recurrent UTI in pediatric patients may suggest duplication with the risk of UTI increased 20 times in pediatric age group. Iatrogenic ureteral injuries can be a serious complication in abdominal or pelvic operation; more commonly during gynecological and general surgical procedures due to duplication of the ureter [1,7,8].

In complete duplication, ureter usually from the upper moiety insert into the vagina, urethra, epididymal region or vestibule and presents with the problems of persistent urinary incontinence, continuous urinary dribbling and wetting and can cause ureteroceles [9]. However, ureteroureteral reflux, vesicoureteral reflux and ureteropelvic junction obstruction are most commonly associated with incomplete duplication [10].

An ectopic ureter may insert into bladder neck, urethra, ejaculatory track, vas deferens, seminal vesicles, vagina or uterus other than the bladder [11]. An ectopic ureter with single collecting system is more often found in male while ectopic ureter with duplex collecting system is more common in female [12]. Ectopic ureter usually originates from the upper renal pole of the duplex system (80%) while ectopic ureter in single system is exceptional (20%) [13]. Insertion of the upper pole moiety's ureter follows the Weigert-Meyer rule and is inferior and medial as compared to the lower pole moiety's ureter [10]. The dilated upper pole infundibulum tends to push down on the functioning lower pole collecting system creating the classically described "drooping lily" appearance [7]. Urinary incontinence is the commonest presenting complain of an ectopic ureter, usually in female as insertion of an ectopic ureter bypass the external urethral sphincter; however urinary incontinence may not be present in male as ectopic ureter usually inserts above the external urethral sphincter in them. Both sexes may present with antenatal hydronephrosis and UTIs. Only girls with a ureteral insertion at or above the bladder neck and upper urethra will be continent [6]. One should suspect an ectopic ureter in female with persistent urinary incontinence and normal voiding pattern following toilet training [13]. Most cases are diagnosed in early childhood [1]. Imaging diagnostic modalities are must to confirm the diagnosis in all cases [14]. Ultrasound shows dual pelvises or hydronephrosis particularly in lower pole during prenatal scan when ureteropelvic junction obstruction is present [10]. Ultrasound is an excellent diagnostic modality after birth; asymmetry between two sides indicates unilateral duplication while non uniform hydronephrosis between upper and lower poles indicates complete duplication and uniformity is non conclusive [15]. Complete duplication without obstruction may evade ultrasound detection and ultrasound almost never detect an ectopic insertion of the ureter as it does not clearly delineate the anatomical relationship between ureter, bladder, urethra and vagina [14,16]. In such cases, contrast-enhanced computed tomography (CT) or magnetic resonance imaging urography should be used to detect or rule out an ectopic ureter [16].

Renal function and damage on kidney by duplication can be assessed by intravenous pyelogram (IVP) which appears as delayed excretion or even no excretion [17]. Intravenous pyelogram could also be used to clearly determine the level of convergence between ureters when diagnosis is in doubt between complete versus partial duplication [10]. Renal Scintigraphy may be done to assess scar tissue, upper and lower pole function in complete duplication to help in choice of therapy [17]. Micturating cystourethrogram (MCUG) could detect vesicoureteral reflux [10]. Cystoscopy could help in detecting anomalies like ectopic ureter [14]. Surgery is the best available treatment as it resolves the incontinence, prevent complications and preserve renal functions. Surgical options include upper pole partial nephrectomy or heminephrectomy in nonfunctional duplex system, laparoscopic ureteral ligation (clipping), ureteral reimplantation in preserved renal function [18]. When there is preserved kidney function or a functioning upper pole in duplex system kidney, the surgery technique consists of ureteroneocystostomy or distal and proximal ureteroureterostomy respectively [7]. Ureteral clipping is faster, safer and more efficient option compared to heminephrectomy or reconstructive procedures in children [19]. Loss of function in the remaining ipsilateral moiety occurs in 5% of patients that underwent heminephrectomy due to vascular injury or torsion of the renal pedicle from extensive renal mobilization [20].

3. Take away lessons

Bilateral duplex collecting system with unilateral ectopic ureter is rare congenital abnormality and it is predominant in female. One should suspect an ectopic ureter in female patients who present with persistent urinary incontinence with normal voiding pattern despite toilet training. Ultrasound is the best initial diagnostic workup to detect duplex collecting system while CT or MR urography should be used to delineate the anatomical relationship of genitourinary organs and establish the ectopic presence. Upper pole heminephrectomy is an ideal choice of treatment in case of nonfunctional duplex system. However, laparoscopic ureteral ligation or ureteral reimplantation should be done if renal function is preserved.

Provenance and peer review

Not commissioned, externally peer reviewed.

Ethical approval

Research studies involving patients require ethical approval. Please state whether approval has been given, name the relevant ethics committee and the state the reference number for their judgement.

Please state any sources of funding for your research

This case report hasn't been funded by any person or any institutions.

Author contribution

1. Sampurna Singh, Internship at Department of Surgery, Kist Medical College, Kathmandu, First Author: he has conceptualized, collected and analyse data and wrote the case, done all the discussion part.
2. Narayan Prasad Bhusal, Department of Surgery, Kist Medical College, Kathmandu, Second author (Guarantor): he has presented the case.
3. Sarmendra Mishra, Department of Surgery, Kist Medical College, Kathmandu, Third Author: he has collected the imaging data.
4. Santosh Singh, Department of surgery, Gandaki Medical College, Pokhara, Fourth Author: he has helped in literature review and discussion portion.
5. Kabindra Rai, Internship at Department of Surgery, Kist Medical College, Kathmandu, Fifth Author: he has worked in history and examination portion.
6. Shiwani Bista, Internship at Department of Surgery, Kist Medical College, Kathmandu, Sixth Author: Editor of the article.
7. Suprabha Amatya, Internship at Department of Surgery, Kist Medical College, Kathmandu, Seventh Author: Editor of the article.
8. Shreeja Kisee, Internship at Department of surgery, Kist Medical College, Kathmandu, Eight Author: Editor of the article.

Please state any conflicts of interest

There is no any conflicts of interest with this article.

Registration of research studies

1. Name of the registry:
2. Unique Identifying number or registration ID:
3. Hyperlink to your specific registration (must be publicly accessible and will be checked):

Guarantor

Narayan Prasad Bhusal, Department of Surgery, Kist Medical College, Kathmandu, Nepal.

Consent

Written informed consent was obtained from the patient's mother for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

References

[1] J. Raja, A.M. Mohareb, B. Bilori, Recurrent urinary tract infections in an adult with a duplicated renal collecting system, Radiol. Case Rep. 11 (2016) 328–331, https://doi.org/10.1016/j.radcr.2016.08.015.

[2] E. Siomou, F. Papadopoulou, K.D. Kollios, A. Photopoulos, E. Evagelidou, P. Androulakakis, et al., Duplex collecting system diagnosed during the first 6 years of life after a first urinary tract infection: a study of 63 children, J. Urol. 175 (2006) 678–682, https://doi.org/10.1016/S0022-5347(05)00184-9.

[3] Privett JTJ, W.D. Jeans, J. Roylance, THE INCIDENCE AND IMPORTANCE OF RENAL DUPLICATION, vol. 27, 1976.

[4] N. Cohen, M. Berant, Duplications of the renal collecting system in the hereditary osteo-onycho-dysplasia syndrome, J. Pediatr. 89 (1976) 261–263, https://doi.org/10.1016/s0022-3476(76)80462-3.

[5] A. Ghobashy, M. El-Shazly, A. Lari, O. Al-Hunaidy, A. Allam, N. Alenezy, et al., A case of complete renal duplex with H-shaped ureter, Case Rep. Urol. 2012 (2012) 1–2, https://doi.org/10.1155/2012/643207.

[6] S. Baskin Laurence, Ectopic Ureter. Up to Date, 2016.

[7] C. Ak, A. Traoré, M. Jalloh, M. Ndoye, L. Niang, G. Sm, Bilateral Ureteral Duplication and Right Ectopic Ureter Presenting with Incontinence: A Case Report, 2019, https://doi.org/10.35248/2168-9857.19.8.216.

[8] R.A. Agha, T. Franchi, C. Sohrabi, G. Mathew, A. Kerwan, A. Thoma, et al., The SCARE 2020 guideline: updating consensus surgical CAse REport (SCARE) guidelines, Int. J. Surg. 84 (2020) 226–230, https://doi.org/10.1016/j.ijsu.2020.10.034.

[9] Fernbach SK, Feinstein KA, Spencer K, Lindstrom CA. Ureteral duplication and its complications. Radiographics n.d.;17:109–127. https://doi.org/10.1148/radiographics.17.1.9017803.

[10] D. Scantling, C. Ross, H. Altman, A 52-year-old male with bilaterally duplicated collecting systems with obstructing ureteral stones: a case report, Curr. Urol. 7 (2013) 104–106, https://doi.org/10.1159/000356257.

[11] I. Kuliniec, P. Mitura, P. Płaza, D. Widz, D. Sudoł, M. Godzisz, et al., Urinary incontinence in adulthood in a course of ectopic ureter—description of two clinical cases with review of literature, Int. J. Environ. Res. Publ. Health 18 (2021), https://doi.org/10.3390/ijerph18137084.

[12] E.F. Avni, C. Matos, F. Rypens, C.C. Schulman, Ectopic vaginal insertion of an upper pole ureter: demonstration by special sequences of magnetic resonance imaging, J. Urol. 158 (1997) 1931–1932, https://doi.org/10.1016/s0022-5347(01)64183-1.

[13] C. Duicu, E. Kiss, I. Simu, C. Aldea, A rare case of double-system with ectopic ureteral openings into vagina, Front. Pediatr. 6 (2018), https://doi.org/10.3389/fped.2018.00176.

[14] M.S. Muhamad, M.A. Mousa, M. Oukan, A. Razzok, Giant hydronephrosis secondary to an ectopic ureter associated with bilateral duplex collecting system: a case report, Oxf. Med. Case Rep. 2022 (2022) 139–141, https://doi.org/10.1093/omcr/omac034.

[15] B. Adiego, P. Martinez-Ten, J. Perez-Pedregosa, T. Illescas, E. Barron, A.E. Wong, et al., Antenatally diagnosed renal duplex anomalies: sonographic features and long-term postnatal outcome, J. Ultrasound Med. 30 (2011) 809–815, https://doi.org/10.7863/jum.2011.30.6.809.

[16] V.H. Figueroa, G.B. Chavhan, K. Oudjhane, W. Farhat, Utility of MR urography in children suspected of having ectopic ureter, Pediatr. Radiol. 44 (2014) 956–962, https://doi.org/10.1007/s00247-014-2905-4.

[17] P. Yuri, E.T.P. Utama, A complete duplicated collecting system with giant ureterocele in adult: case report, Int. J. Surg. Case Rep. 79 (2021) 49–52, https://doi.org/10.1016/j.ijscr.2020.12.083.

[18] R.L.P. Romao, V. Figueroa, J.L.P. Salle, M.A. Koyle, D.J. Bägli, A.J. Lorenzo, Laparoscopic ureteral ligation (clipping): a novel, simple procedure for pediatric urinary incontinence due to ectopic ureters associated with non-functioning upper pole renal moieties, J. Pediatr. Urol. 10 (2014) 1089–1094, https://doi.org/10.1016/j.jpurol.2014.04.008.

[19] R.I. Lopes, N. Fernandez, M.A. Koyle, J. dos Santos, J. Perez, R. Romao, et al., Clinical outcomes of the upper urinary tract after ureteral clipping for treatment of low functioning or nonfunctioning renal moieties, J. Urol. 199 (2018) 558–564, https://doi.org/10.1016/j.juro.2017.09.080.

[20] M.J. Biles, J.B. Finkelstein, M.v. Silva, S.M. Lambert, P. Casale, Innovation in robotics and pediatric urology: robotic ureteroureterostomy for duplex systems with ureteral ectopia, J. Endourol. 30 (2016) 1041–1048, https://doi.org/10.1089/end.2015.0645. Mary Ann Liebert Inc.