CASE REPORT

Rare association of congenital penile urethrocutaneous fistula with Y-type urethral duplication

Ashok Kumar Gupta, Manoj Kumar, Kawaljit Singh, Ashok Kumar Sokhal

Department of Urology, King George's Medical University, Lucknow, Uttar Pradesh, India

Correspondence to

Dr Ashok Kumar Gupta,
dr.akgupta555@gmail.com

Accepted 14 November 2016

SUMMARY

A 24-year-old man presented with symptoms of difficulty in passage of urine, mixed with gas and stool from penoscrotal junction since birth. He had a history of surgery (posterior sagittal anorectoplasty) for imperforate anus in first week of his life. On physical examination, external meatus was adequate with small 5×5 mm fistulous opening noted at penoscrotal junction with normal anal tone and opening. On evaluation with retrograde urethrography, voiding cystourethrogram, cystoscopy and barium enema, he was found to have penile urethrocutaneous fistula of 0.5×0.5 cm with Y-type incomplete urethral duplication. Urethroanal fistula (Y-type urethral duplication) was of small size (<5 mm), so closed primarily through perineal route with interposition of ischioanal fat. Penile urethrocutaneous fistula (4 mm) too was closed primarily and covered with tunica vaginalis flap in single sitting. It has been 6 months since surgery and the patient is fully continent and voiding well with no urinary or faecal leak.

BACKGROUND

Coexistence of congenital penile urethrocutaneous fistula and Y-type urethral duplication (YUD) has not been reported in the literature previously. Both conditions were reconstructed in single sitting with minimum morbidity and successful outcome, which is also unique about the management of this case.

CASE PRESENTATION

A 24-year-old man presented with difficulty in passage of urine, mixed with gas and stool from the undersurface of penis since fourth day of his life. He had a history of surgery (posterior sagittal anorectoplasty) for imperforate anus in first week of his life. There is no history of trauma, strangulation or stone impaction. On physical examination, external meatus was normal in position, adequate in size, prepuce was circumcised with no chordee or hypospadias. A small 5×5 mm urethrocutaneous fistula noted at penoscrotal junction (figure 1) with normally developed phallus and scrotum. Examination of back revealed well-developed gluteal folds, midline groove with normal anal tone on digital rectal examination. No fistulous opening was noted in perineum.

INVESTIGATIONS

On retrograde urethrogram through orthotropic meatus, an urethrocutaneous fistula was found at the penoscrotal junction and another urethrorectal fistula

between the proximal bulbar urethra and anus just 2 cm above the anal verge was found (figures 2 and 3). Duplication length appeared small (<1 cm) in retrograde urethrography. voiding cystourethrogram showed normal contour of the bladder wall with no evidence of diverticula or fistula or vesicoureteric reflux, with a normal bladder neck opening and single non-dilated posterior urethra (figure 4). It also did not reveal any duplication of bladder or septated bladder. On cystoscopy through orthotropic meatus, penile urethra was normal with mucosal blanching starting from penoscrotal junction upto proximal bulbar urethra, where a ventral mucosal fold with channel was seen. It was traversing ventrally separate from the normal dorsal posterior urethra for about 4 cm before opening into anus, just 2 cm above anal verge. Rest of the cystoscopy was unremarkable. On barium enema thin fistulous tract seen between the lower rectum and posterior urethra was suggestive of low-type urethroanal fistula. Ultrasonography of the abdomen showed normal bilateral kidneys, normal thickness urinary bladder wall with no vesical or renal calculi and was largely unremarkable. Haemogram (haemoglobin 13.4 g/dL, total leukocyte count 5400 mm³), serum biochemistry (urea 37 mg/dL, creatinine 0.9 mg/dL, Na 136, K 4.5 mEq/L) and liver function were within normal limits.

Based on these findings, diagnosis of congenital penile urethrocutaneous fistula and YUD, an incomplete type of duplication were made. According to Effmann's classification, YUD is categorised under type IIA2 anomaly.

DIFFERENTIAL DIAGNOSIS

1. Congenital vesicorectal fistula
2. Distal urethral stricture with proximal urethrocutaneous fistula

TREATMENT

Surgery was carried out under spinal anaesthesia in lithotomy position. The fistulous connection between posterior urethra and anus was excised through a midline perineal incision after placing a 10 French infant feeding tube from normally placed urethral meatus to anus through the fistulous tract. The fistula tract was separated from the adjacent subcutaneous tissue, all around, from its urethral end to distal end. Then rectal and urethral defects were closed in two layers with a 3-0 vicryl (figure 5). The penile urethra cutaneous fistula was excised after making a circumferential incision around the external fistulous opening at the

CrossMark

To cite: Gupta AK, Kumar M, Singh K, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2016-217331

Figure 1 Normal glanular external meatus with small 5×5 mm fistulous opening noted at penoscrotal junction.

penoscrotal junction and mobilising this fistulous tract upto penoscrotal urethra (figure 6). The resultant urethral defect was small hence closed in layers and reinforced with tunica vaginalis flap.

OUTCOME AND FOLLOW-UP

The patient was allowed to take feeds orally from postoperative day 1 and discharged after 3 days of uneventful postoperative hospital stay. Pericatheter retrograde urethrogram performed after 4 weeks; no contrast leak was found and the per urethral catheter was removed. It has been 6 months since surgery and patient is fully continent and voiding well with no urinary or faecal leak.

DISCUSSION

Until now only 52 cases of anterior congenital penile urethral fistula are mentioned in the literature¹⁻³. Congenital penile urethrocutaneous fistula is an unusual developmental anomaly in children who present with an abnormal opening on the ventral aspect of penis with a normal foreskin and an absence of chordee and hypospadias.

On the basis of development, *congenital urethral fistula* can be categorised into three groups. The commonest type is rectourethral fistula and it is associated with anorectal

Figure 2 RGU through orthotropic meatus, showing urethrocutaneous fistula at penoscrotal junction and urethrorectal fistula between proximal bulbar urethra and anal canal. RGU, retrograde urethrogram.

Figure 3 RGU through penoscrotal fistula, showing narrow lumen penile urethrocutaneous fistula and urethrorectal fistula between proximal bulbar urethra and anal canal. RGU, retrograde urethrogram.

malformation. The second common type is posterior urethro-perineal fistula or YUD. The third type is distal urethrocutaneous type, the rarest one.⁴ Embryologically urogenital membrane is the anterior portion of the cloacal membrane, from which the urethra develops after it has been divided by the urorectal septum. So, a primary defect in the urogenital membrane and cloacal membrane may explain the associated imperforate anus and congenital urethrocutaneous fistula. This abnormality usually is an isolated deformity, but may be associated with imperforate anus, hypospadias or ventral chordee. The cause is unclear, but probably reflects a focal defect in the urethral plate that prevents fusion of the urethral folds.^{5 6} The

Figure 4 VCUG showing normal bladder neck, single proximal urethral channel with incomplete type urethral duplication. VCUG, voiding cystourethrogram.

Figure 5 Fistulous tract excision through a midline perineal incision after putting a 10 French infant feeding tube from urethra to anus through the fistulous tract.

management of these anterior urethrocutaneous fistulae depends on size and location. Small fistula which are <0.5 cm can be easily closed primarily after refreshing the edges and covered by skin. Fistulae of size >0.5 cm but <1 cm can be closed with turnover flap. Larger fistulae which were more than 1 cm can be closed by tubularised incised plate urethroplasty using the Thiersch-Duplay technique.⁷ Fistulae larger than 2 cm are associated with significant defect in urethra and skin cover which can be dealt with bilamellar preputial island flap.

Based on location, distal urethra fistula with intact urethra and spongiosum are primarily repaired and reinforced with Bayer's flap. In the mid-penile region primary repair is reinforced with tunica vaginalis flap.

The YUD is an uncommon congenital anomaly comprising 6–30% of all urethral duplication.^{8,9} In YUD, ventral urethra (VU) originates from the bladder neck/or posterior urethra and opens either within the rectum/anus or onto perineum. Urethral duplication has been divided by Williams and Kenawi into epispadiac, hypospadiac, spindle and YUD type. According to Effmann's classification, YUD is categorised under type IIA2 anomaly.¹⁰

In YUD, VU originates from bladder neck/or posterior urethra and opens either into the perineum or within the rectum/anus. Urethral duplication has been divided by Williams and Kenawi into epispadiac, hypospadiac, spindle and YUD type. This probably would have caused a backward effect on the development of urorectal septum causing misalignment of its components that results in the persistence of a communicating cloacal lumen between the urethra and the anal canal. Williams and Kenawi

Figure 6 Circumferential incision around external fistulous opening at penoscrotal junction and excision of fistulous tract.

considered this communicating urethroanal tract to be primarily a fistula that developed early enough to acquire some of the properties of the urethra. According to Effmann's classification, YUD is categorised under type IIA2 anomaly.^{8,9,11}

The different choices of management for YUD are: (1) mobilisation of the fistulous tract anteriorly then its anastomosis to midline scrotal skin tube. The orthotopic urethra is then laid open followed by urethroplasty in next stage. (2) The orthotopic urethra can be laid open along with perineal urethrostomy of the rectal urethra and then staged reconstruction of the orthotopic urethra. (3) For the creation of diversion colostomy, the ectopic urethra is lengthened with strip of rectal mucosa, reconstruction of the orthotopic urethra in first stage, followed by scrotal skin tubularisation for mid-urethra, reconstruction of orthotopic urethra and maintaining the urethral continuity, and colostomy closure in second stage; (4) progressive augmentation by dilation of the urethra anterior; (5) single-stage reconstruction by VU up to penoscrotal junction then mid-scrotal skin flap for bulbar and membranous urethra and preputial flap for reconstruction of penile urethra.

Association of anterior urethrocutaneous fistula with epispadiac-type urethral duplication has been reported by Merrot *et al.*¹² In this case, they excised the urethrocuteaneous fistula with successful outcome. Our case is unique in the sense that urethral duplication was of Y type, communicating with the anal canal. Both the penile urethrocutaneous fistula and the ventral limb of YUD were excised and repaired successfully in the same sitting.

The urethroanal fistula (YUD) was of small size <5 mm so it was closed primarily with the interposition of ischioirectal fat. The penile urethrocutaneous fistula was also small (5 mm) so it was closed primarily and then covered with tunica vaginalis flap in single sitting.

Patient's perspective

Getting cured from long-standing problem restored my faith in medical system. I express my sincerest thanks to the urology department, KGMU.

Learning points

- ▶ Incomplete urethral duplication of Y type can rarely be associated with penile urethrocutaneous fistula. The cause is not defined but local defect of urethral development may be the reason.
- ▶ Developmental anomalies like congenital anterior urethrocutaneous fistula in association with Y-type urethral duplication should be thoroughly investigated with voiding cystourethrogram, retrograde urethrography, barium enema and cystoscopy before proceeding with surgery.
- ▶ Even if these conditions coexist, single-stage reconstruction can be performed with successful results.

Twitter Follow Ashok Gupta at @ashok gupta

Acknowledgements The authors acknowledge the contribution of Dr JD Rawat MCh (paediatric surgery) for his help in managing this case.

Contributors AKG and MK were involved in planning, investigation, interpretation of data and reporting of this case. KS and AKS contributed in conception and design of the study and worked on drafting the article or revising it critically for important intellectual content.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Elder JS. Abnormalities of the genitalia in boys and their surgical management. In: Wein AJ, ed. *Campbell-Walsh urology*. 9th edn. Philadelphia: Saunders, Elsevier, 2007:3745–6.
- 2 Harjai MM. Congenital urethrocutaneous fistula. *Pediatr Surg Int* 2000;16:386–7.
- 3 Biswas S, Ghosh D, Das S. Congenital urethrocutaneous fistula-our experience with nine cases. *Indian J Surg* 2014;76:156–8.
- 4 Caldamone AA, Chen SC, Elder JS, et al. Congenital anterior urethrocutaneous fistula. *J Urol* 1999;162:1430–2.
- 5 Ibrahim K, Tanyel FC, Hiçsönmez A. Congenital urethrocutaneous fistula: a case report and literature review with a nomenclature proposal. *J Paed Surg* 1995;30:1504–5. [http://dx.doi.org/10.1016/0022-3468\(95\)90422-0](http://dx.doi.org/10.1016/0022-3468(95)90422-0)
- 6 Ritchey ML, Sinha A, Argueso L. Congenital fistula of the penile urethra. *J Urol* 1994;151:1061–2.
- 7 Rashid KA, Kureel SN, Tandon RK. Congenital anterior isolated urethrocutaneous fistula: a case report. *Afr J Paediatr Surg* 2008;5:52–3.
- 8 Bogaert GA. Urethral duplication and other urethral anomalies. In: Gearheart JP, Rink RC, Mouriquand PD, eds. *Pediatric urology*. 2nd edn. Philadelphia: Saunders, 1976:446–58.5.
- 9 Olbourne NA. Congenital urethral fistula: case reports. *Plast Reconstr Surg* 1976;57:237–9.
- 10 Effmann EL, Lebowitz RL, Collodny AH. Duplication of the urethra. *Radiology* 1976;119:179–185.
- 11 Sánchez MM, Vellibre RM, Castelo JL, et al. A new case of male Y-type urethral duplication and review of literature. *J Pediatr Surg* 2006;41:e69–71.
- 12 Merrot T, Pankevych T, Chaumoitre K, et al. Congenital anterior urethrocutaneous fistula associated with urethral duplication. *Pediatr Surg Int* 2003;19:744–5.

Copyright 2016 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- Submit as many cases as you like
- Enjoy fast sympathetic peer review and rapid publication of accepted articles
- Access all the published articles
- Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow