KJU
Korean Journal of Urology

www.kjurology.org
DOI:10.4111/kju.2011.52.5.368

Case Report

Isolated Congenital Urethrocutaneous Fistula of the Anterior Urethra

Tarun Jindal, Mir Reza Kamal, Soumendra Nath Mandal, Dilip Karmakar

Department of Urology, Calcutta National Medical College, Kolkata, India

Isolated congenital urethrocutaneous fistula of the anterior urethra is an extremely rare anomaly in which, along with a normal urethra and meatus, a fistula is present. These cases usually present in the pediatric age group. To date, only one such case has been described in the adult population. We describe one such case in an adult and discuss the management of this uncommon anomaly.

Key Words: *Cutaneous fistula; Urethra; Urinary fistula*

Article History:
received 10 February, 2011
accepted 7 April, 2011

Corresponding Author:
Tarun Jindal
92-B, First floor, Jhowtala road, Above Apex Diagnostics, Kolkata-700017, India
TEL: +91-9674444929
FAX: +91-33-40081710
E-mail: drtarunjindal@gmail.com

Congenital, urethrocutaneous fistula of the anterior urethra is an uncommon anomaly. It is associated with chordee, hypospadias, and at times, anorectal malformations. An isolated congenital urethrocutaneous fistula of the anterior urethra is extremely rare and is usually present in the pediatric age group. Only one adult case has been described to date. There is no information to date about the effect of this deformity on the potency or the fertility of the patients. We present a case of an isolated adult congenital urethrocutaneous fistula of the anterior urethra and discuss the management of this uncommon deformity.

CASE REPORT

A 19-year-old male presented with an additional opening on the ventral surface of the penile shaft along with a normally placed urethral meatus at the tip of the glans. The additional opening had been present since birth. The patient had dribbling of urine from this opening at the time of micturition. There was no history of trauma during or after birth. There was no history of swelling or stone impaction at the site of the opening. The only surgical history was of a ceremonial circumcision at the age of 5 years that was done by a local priest. The patient was married and had normal erectile function and a normal sexual life. He had fathered a child. On examination, the glans was normal in size and shape with a normally placed external urinary meatus. There was a small opening near the corona on the ventral aspect of the penile shaft, just lateral to the midline. The patient had a normal erection. There was no visible chordee and the length of the penis was normal. At the time of micturition, the patient voided mainly through the normally placed meatus along with dribbling of urine from the ventral opening (Fig. 1). The results of investigations, such as hemogram, biochemical profile, urine analysis, abdominal ultrasound, intravenous pyelography, and other radiological investigations to rule out coexistent anomalies,

FIG. 1. The patient voiding mainly through the normally placed meatus with dribbling of urine from the ventral fistulous opening.

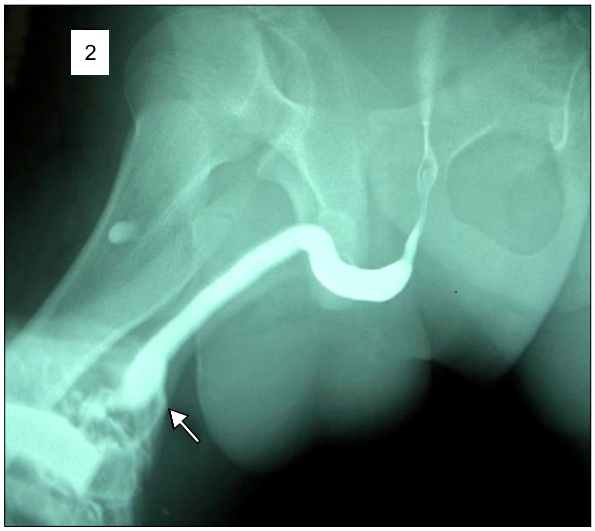

FIG. 2. Retrograde urethrogram showing the fistulous tract (arrow) communicating with the ventral part of the anterior urethra.

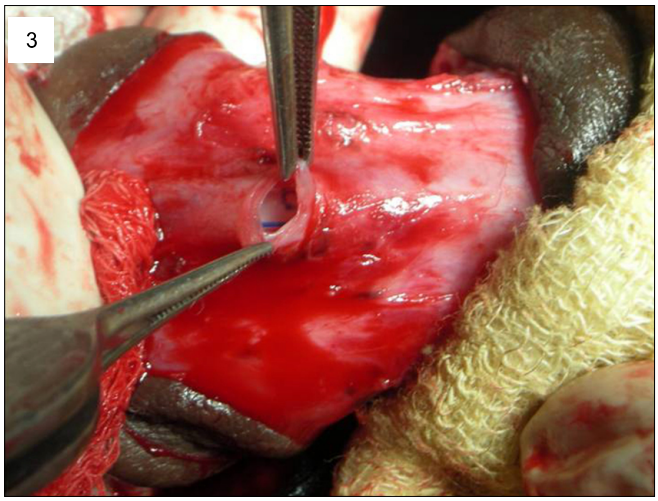

FIG. 3. The defect in the normally placed urethra that was closed primarily.

were normal. There was no anorectal malformation.

A diagnosis of isolated congenital urethrocutaneous fistula was considered. A retrograde urethrogram revealed the fistulous tract communicating with the anterior urethra. The normally placed urethra did not reveal any abnormality (Fig. 2).

At surgery, an infant feeding tube could be easily passed into the fistulous opening via the external urinary meatus. A degloving incision was made, and the fistulous tract was identified. There was a lack of spongiosus tissue around the fistulous tract, whereas the distal urethra had normal spongiosus covering. The tract was excised, and double layered repair of the defect was done (Fig. 3). A 14 Fr catheter was inserted from the external urinary meatus. The patient was able to pass urine normally after removal of the catheter and the fistula had completely healed. The patient is doing well after 9 months of follow-up.

DISCUSSION

Congenital anterior urethrocutaneous fistula is a rare condition. A review of the literature indicates that there are two variants of congenital anterior urethrocutaneous fistula. The more common type is associated with chordee, a dorsal hood, distal hypospadias with or without a distal urethral or spongiosal defect, and anorectal anomalies. The rarer variety, as in the case reported here, develops in an isolated fashion with no chordee or hypospadias and an intact distal urethra and spongiosum. Only 24 cases of this variety have been reported in the English literature to date. All cases apart from one were described in the pediatric age group [1-8]. Chen et al reported this deformity in a 26-year-old male [8]. The fistula was at the proximal part of the anterior urethra. There was no mention of the effect of this deformity on the patient's virility or his ability to procreate [8]. Our patient had normal erectile function and was a father, thus proving that this deformity has no effect on the reproductive function of the patient. The patient did not seek medical advice for his condition until late because of the minimal symptoms associated with his condition.

Considerable confusion about this entity exists, and there have been claims that it is in fact a result of trauma during circumcision. However, due to the lack of urothelium in the pathological examination of the circumscribed skin and also because of the reports of its existence in uncircumscribed patients, it is now being considered as a separate entity. The exact mechanism by which this anomaly occurs is not clear. The arrest of the growth of the proximal urethra and compensatory overgrowth of the glandular urethra has been proposed to be the mechanism for the formation of the isolated variety of fistula [2,6]. A deficiency of spongiosum in the fistula, as occurred in our case, can also be explained by this, because only the proximal urethra is associated with the formation of corpus spongiosum [2,4-6].

Before deciding on surgical correction, it is important to assess the patency of the urethra beyond the fistula. Probing, dye studies, and cystourethroscopic examination may be helpful in substantiating the diagnosis. Several methods of surgical repair have been described in the literature, including local skin flap, preputial skin flap, Denis Brown urethroplasty, Thiersch-Duplay urethroplasty, primary closure, buccal mucosal graft, and tubularized incised plate urethroplasty with dartos flap [1-8]. The method of repair must be individualized depending on the location and the size of the fistula. In the present case, the fistula was small, and thus primary closure was used for repair with success. In larger fistulas, a flap may be better to prevent recurrence. The incidence of recurrence following repair is about 17% as per the reported literature (4 recurrences in 24 cases) [1-8].

To conclude, isolated congenital anterior urethrocutaneous fistula is a condition that can be easily and effectively managed. Increased awareness, good history, meticulous

examination, and imaging studies can help in the correct diagnosis and treatment of this rare clinical entity.

Conflicts of Interest
The authors have nothing to disclose.

REFERENCES

1. Betalli P, Carretto E, Midrio P, Zanon GF, Gamba PG. A new indication for buccal mucosal graft: isolated congenital fistula of the penile urethra. Pediatr Surg Int 2003;19:586-7.
2. Olbourne NA. Congenital urethral fistula. Case reports. Plast Reconstr Surg 1976;57:237-9.
3. Akman RY, Cam K, Akyuz O, Erol A. Isolated congenital urethrocutaneous fistula. Int J Urol 2005;12:417-8.
4. Karnak I, Tanyel FC, Hiçsönmez A. Congenital urethrocutaneous fistula: a case report and literature review, with a nomenclature proposal. J Pediatr Surg 1995;30:1504-5.
5. Gupta V, Sharma SB. A new indication for tubularized incised plate urethroplasty: isolated congenital penile urethrocutaneous fistula. J Indian Assoc Pediatr Surg 2006;11:148-50.
6. Caldamone AA, Chen SC, Elder JS, Ritchey ML, Diamond DA, Koyle MA. Congenital anterior urethrocutaneous fistula. J Urol 1999;162:1430-2.
7. Arena F, Scalfari G, Impellizzeri P, Antonuccio P, Romeo C. Case report of congenital fistula of the penile urethra. Minerva Pediatr 2008;60:255-7.
8. Chen Q, Xiao WL, Jiang YM, Sun LJ. Adult isolated congenital anterior urethrocutaneous fistula. Urol Int 2010;Epub ahead of print