Case Report

Congenital Urethrocutaneous Fistula

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Abstract

Male congenital urethrocutaneous fistula is an extremely rare anomaly that is commonly associated with chordee or anorectal malformations. It is characterised by an abnormal urethral opening on the ventral aspect of the penis with well-formed distal urethra and meatus at the tip of the glans. The treatment is individualised according to the site of the fistula, associated anomalies and condition of the distal urethra. The principles of hypospadias surgery should be strictly followed.

Keywords: Abnormalities, congenital, paediatric, penis, urethra

Introduction

The congenital urethrocutaneous fistula is an extremely rare condition that may present in an isolated fashion or in association with hypospadias or chordee.[1-3] It is characterised by the fistulisation of the penile urethra to the skin with a urethral meatus at its normal position.[3] It may be confused with hypospadias.[4] We are presenting a case of 4-year-old boy with congenital urethrocutaneous fistula without associated penile anomalies. It has been described in the literature as an unusual type of hypospadias and congenital urethrocutaneous fistula.

Case Report

A 4-year-old boy presented to the outpatient department with the passage of urine from the undersurface of the penis as well as from normal urethral meatus since birth. He was passing majority of urine through this fistula and rest from the normal urethral meatus. There was no previous history of trauma, circumcision, surgical intervention or urinary retention. On examination, there was mid-penile ventral opening of size 0.4 cm × 0.5 cm with normal external urethral meatus. The prepuce and glans were normal without ventral chordee and deficit ventral prepucial skin. Urethra distal to ventral opening was patent and of normal calibre. Ventral skin proximal to the fistula was thinned out [Figure 1]. Infant feeding tube of 6 FR was passed from the normal urinary meatus up to the fistulous opening without resistance suggestive of patent distal urethra [Figure 2]. Anorectum was normal and the testis bilaterally descended in the scrotum. No other genital abnormality was seen. Routine blood investigations and abdominal ultrasound were normal. The patient underwent surgical correction. The surgical techniques included (a) Penile degloving upto root of the phallus, (b) Urethra from the fistula to the corona was laid open, (c) Urethroplasty was done over 6FR infant feeding tube passed, (d) Spongioplasty was done, (e) Vascularised inner preputial flap rotated ventrally over spongioplasty as a Byar's flap and (f) Perurethral catheter fixed with glans and connected



Figure 1: Clinical picture showing congenital urethrocutaneous fistula

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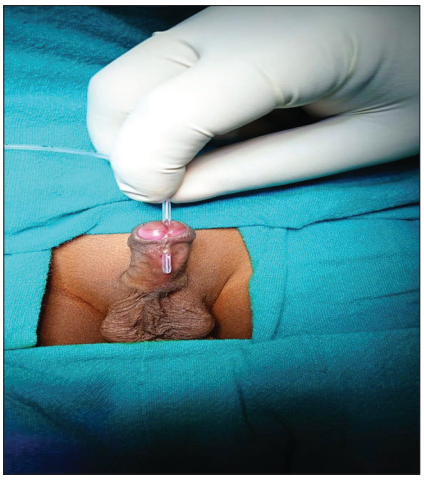


Figure 2: Patent distal urethra after calibrating the external urethral meatus



Figure 3: Post-operative clinical picture of congenital urethrocutaneous fistula

with urobag [Figure 3]. The glanular urethra and meatus were normal and hence left untouched. The post-operative period was uneventful. We removed perurethral catheter on post-operative day 8, the child passed urine from the tip of the glans in the single stream without any evidence of urethrocutaneous fistula. We were kept patient on regular follow-ups. First follow-up after 2 weeks for evaluation of oedema and scar. At 6-month follow-up, the child was able to passed in single stream with no evidence of urethrocutaneous fistula.

Discussion

Congenital urethrocutaneous fistula with normal urethral meatus is a rare condition. There is a paucity of literature about this entity.[3] The aetiology and pathogenesis are uncertain, and several theories have been proposed. The male genitalia develop in a proximal-to-distal manner. As the penis forms from the elongation and enlargement of the phallus, the lateral walls of the urethral groove form from the ventrally located genital folds, which then fuse at the midline.[5] Embryologically, congenital anterior urethrocutaneous fistula results from urethral plate blowout due to distal urethral obstruction.[6] The other theory suggested that a focal defect in the urethral plate may result in arrested distal migration of the urethral plate.[2] There are two varieties of congenital anterior urethrocutaneous fistula. First is an isolated fistula associated with normal foreskin, an intact distal urethra and spongiosum without chordee or hypospadias as in our case. The second type is congenital urethrocutaneous fistula associated with hypospadias-like characteristics, that is, chordee, a dorsal hood with or without distal urethra or spongiosal defect. Congenital anterior urethrocutaneous fistula is uncommon as compared with posterior urethrocutaneous fistula, which usually represents Y-type duplication of the urethra with anorectal atretic malformations.[7]

Surgical approach to repair congenital anterior urethrocutaneous fistula depends upon the type of fistula. A number of authors have described different methods of repairing of congenital urethral fistula. They include pedicle flap,[8] prepucial bound skin flap, modified Denis Brown urethroplasty or direct closure.[2] All of them have different merits and demerits. In our case, we performed a direct closure, as it was easy and more physiological. The extra layer of vascularised tissue from the inner surface of the prepuce gave extra support to heal without any recurrence.

Conclusion

Congenital urethrocutaneous fistula is a very rare clinical entity. Careful diagnosis and evaluation for associated anomalies are necessary for management. The surgical approach depends on the type and size of a fistula with good functional and aesthetical results.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the legal guardian has given his consent for images and other clinical information to be reported in the journal. The guardian understands that names and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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