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Case Report

Isolated congenital urethrocutaneous fistula: Two case reports

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Abstract

A common complication after hypospadias surgery is a urethrocuteneous fistula. But when it is congenital and present in isolated form, it becomes extremely uncommon and usually present in the paediatric age group and associated with hypospadias, chordee, or anorectal malformation. In this report, we are presenting 2 cases of uncircumcised boys with this rare entity of Isolated congenital anterior uretrocuteneous fistula, highlighting etiology, embryology, and surgical reconstructions for management.

Keywords: Congenital, isolated urethrocutaneous fistula.

Introduction

An isolated congenital urethrocuteneous fistula is a very rare developmental anomaly where the prepuce, urethra, and meatus are normal, and the fistula is present on the ventral surface of the distal penile area, usually coronal or subcoronal without any association like chordee, hypospadias, anorectal malformation or any history of penile trauma or surgery. It is a rare anomaly of unknown etiology but reflects a focal defect in urethral plate which prevents fusion of urethral folds[1-4]. It was reported by Campbell in 1951, and the largest series was conducted by Caldamone et al. on 14 cases. Repair involves the principles of hypospadias surgery and may involve simple multilayer fistula closure to more complex reconstruction of the distal urethra and glans. It is a different entity from the posterior urethrocutaneous fistula, which usually represents Y Y-type duplication of the urethra with a proximal ventral limb and associated anorectal malformation. Here, we report 2 cases of isolated congenital anterior urethrocuteneous fistula. Meticulous clinical examination is stressed, and strategies are discussed with a literature review.

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Case Report

Case 1

A 6-year-old boy presented with a congenital abnormal opening on the ventral surface of the penis in addition to the normal external meatus. There was no previous history of circumcision, trauma, or surgical intervention. On examination, there was normal prepuce, glans, external urinary meatus, and penile shaft without chordee. Anorectum was normal, and no other local systemic abnormality was present. The distal urethra was present and surrounded by spongiosum. There was an oval opening about 1x0.5 cm on the ventral aspect of the distal penile shaft. A catheter was passed through the external urethral meatus, which came out through the fistula, suggesting a patent distal urethra. A congenital urethrocuteneous fistula was confirmed. Investigations like routine blood tests, urine examinations, micturating cystourethrogram, and abdominal ultrasonography were normal. The patient underwent surgical correction under general anesthesia. After placing the stay suture on the glans, the proposed circumferential incision incorporating the fistulous opening was tabularized in a multilayer on an 8 fr feeding tube and the neourethral suture line was

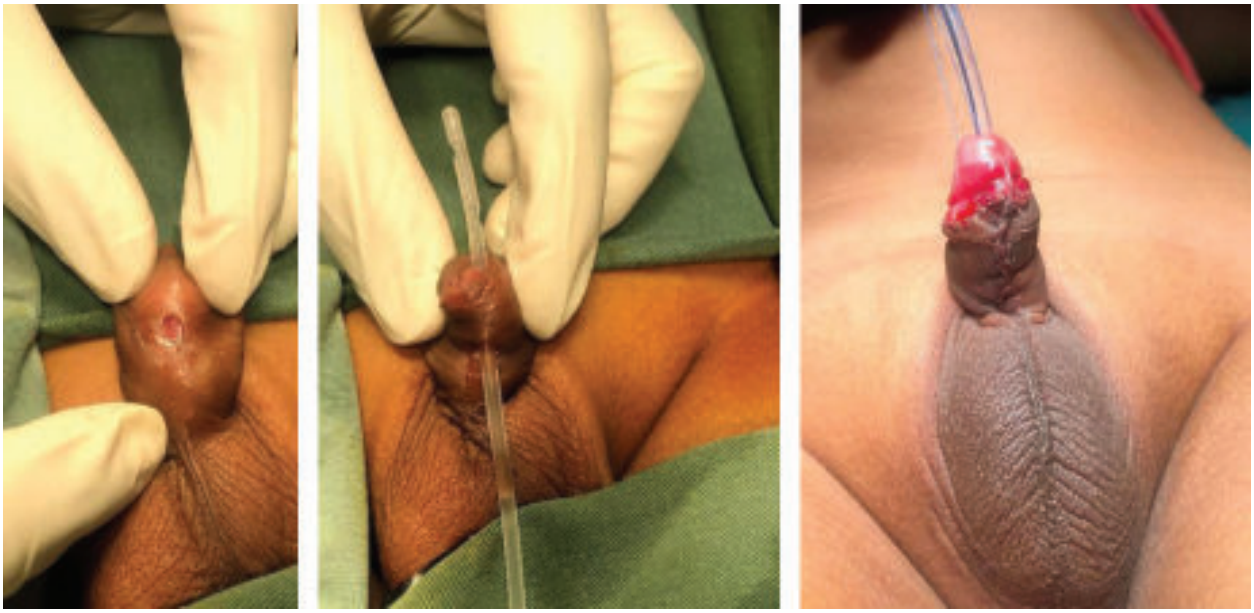

Fig: *Operative procedure of congenital urethrocutaneous fistula*

reinforced with a dartos flap after harvesting an additional subcutaneous dartos flap. Penile skin coverage was given. A feeding tube was kept in situ for seven days. The child voided normally after the removal of the feeding tube; the patient has normal voiding without recurrence or any other complications after two years of follow-up.

Case 2

A 5-year-old uncircumcised boy attended our outpatient clinic complaining of passing urine through an opening located on the ventral side of his penis since birth, according to his parents. He had neither a history of trauma nor surgical intervention. On inquiry about family history, there was no history of such condition or any other genitourinary tract malformations. He was voiding urine through both openings. On local examination, a clearly apparent fistula was seen on the ventral side of the shaft of the penis, just proximal to the coronal sulcus, measuring about 1x0.5 cm distal to that fistula; the prepuce and glans are intact with normal caliber urethral meatus at its tip. The rest of the local examination revealed the absence of chordee, normal scrotum with bilateral palpable testis, and normal anus. A catheter was passed through the external urethral meatus, which came out through the fistula, suggesting a patent distal urethra. The diagnosis was made for a congenital urethrocuteneuos fistula.

Abdominal ultrasound and micturating cystourethrogram revealed no other genito-urinary anomalies. The boy was prepared for elective surgery, and the Thiersch-Duplay urethroplasty method was planned for the surgical process to reconstruct a near-normal caliber urethra. At operation, Closure of the fistula was done in multiple layers on 8 fr feeding tube with additional subcutaneous dartos flap, and the neourethral suture line was reinforced with the flap. The penile skin coverage was given like a circumcised child. The feeding tube was kept in place for ten days, and after removal, the child voided normally. During the post-operative period, there was an infection in the penoscrotal region, which was treated with proper antibiotics. The patient is having regular voiding without any complications after 2 years following removing the feeding tube.

Discussion

An isolated congenital anterior urethrocutaneous fistula without anorectal malformations, hypospadias, or chordee is an extremely rare anomaly reported in few cases in the literature[5]. Till 2019, only 50 cases had been reported in the literature, where half of which were in the distal shaft, 15 at the mid-shaft, three at the penoscrotal junction, and at unspecified locations in 7 cases. It occurs more commonly in an isolated form (29/50; 58%), with a normal urethra, meatus, and foreskin, or associated with anorectal malformations, hypospadias, and other genitourinary anomalies[6,7]. Isolated congenital anterior

urethrocutaneous fistulas are usually posteriorly positioned, associated with anorectal malformation and often represented by the ventral functional limb of Y-type urethral duplication[1,4,8,9]. In this condition, history is crucial as previous trauma, circumcision, for example, or surgical procedures can give a clue about the cause of the fistula[6].

In the male fetus, urethral development and sexual differentiation begin approximately in the uterus at about eight weeks and are completed in 15 weeks. At approximately the 11th week of development, under the influence of testosterone, the urethral folds begin to fuse ventrally in the midline to form the urethra.[1] The male urethra is formed by a fold fusion of the urethra along the ventral surface of the penis, which is extended to the coronal sulcus. The embryonic urethra has three (3) different parts: Wolffian duct opening (proximal portion), including verumontanum. The second segment extends from the verumontanum to the base of the glans. Failure of fusion of urethral folds leads to various levels of hypospadias and, in extremely rare occasions, this may lead to a congenital anterior urethrocutaneous fistula. According to the level at which the lack of fusion occurs, this kind of anomaly can be isolated from the perineum to the glans[1].

The etiopathogenesis of congenital anterior urethrocutaneous fistula is not yet well established., but several etiologic hypotheses have been proposed. The association of imperforate anus with urethral abnormalities is known as the urogenital membrane forming the urethra is part of the cloacal membrane, which in turn forms the anorectal region[2,10]. Campbell stated that an isolated congenital anterior urethrocutaneous fistula represents a urethral blowout behind a congenital obstruction. Olbourne theorized that the urethral plate results in arrested distal migration of the urethral plate on the localized deficiency of the plate[2,4]. Cook and Stephen suggested an alternative mechanism, namely pressure atrophy of penile urethra by fetal parts leading to pressure necrosis by the heel of the baby's foot. These fistulae may also result from cyst development along the mediogenital raphe at the frenulum during the parental period (sir-12). K A Rashid et al. reported that one of their patients had been reported to have a small ventral penile cyst at birth, resembling an epithelial inclusion cyst, which ruptured at 1-2 weeks of age and became a small fistula[1,11]. Finally, in circumcised patients, these lesions might be the result of possible urethral injuries[5].

The management of isolated congenital anterior urethrocutaneous fistula is primarily surgical, aimed at reconstructing near normal caliber neo urethra with no recurrence rate. The choice of the method of repair should be individualized according to many variables, eg. the size and the location of the fistula as well as existing other abnormalities, such as hypospadias or chordee and the condition of the distal urethra[6,12]. These include local skin flap, preputial skin flap, Thiersch-Duplay urethroplasty, Denis Brown urethroplasty, buccal mucosal graft, tabularized incised plate urethroplasty with dartos flap, and primary closure. Small fistulas which are <0. 5 cm can be easily closed primarily after refreshing the edges and covering the skin.[13,14] Fistulas of size >0.5 cm but <1 cm can be closed with a turnover flap[1,5,6]. Larger fistulas that are >1 cm can be closed by tabularized incised plate urethroplasty using the Thiersch Duplay method[1,8,9,15,16]. Fistulae larger than 2 cm are associated with significant defects in the urethra and skin cover, which can be dealt with bilamellar preputial island flap[5,17]. If the fistula is an isolated abnormality with intact urethra beyond it, primary closure is indicated with success. However, if defective, then this method is abandoned, and the defect should be opened to be converted and managed as a hypospadias[6,17].

Conclusion

Although the isolated congenital anterior urethrocutaneous fistula is a very rare developmental anomaly it can be managed easily and effectively. Increased awareness, well-prepared history, and a thorough clinical examination are the most useful ways to diagnose (this rare anomaly) an isolated congenital anterior urethrocutaneous fistula appropriately and to dismiss concomitant irregularities/anomalies.

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