Congenital anterior penile isolated urethrocutaneous fistula: A case report

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ABSTRACT

Urethrocutaneous fistula is a common complication after hypospadias repair. If congenital, it is usually associated with other genitourinary and gastrointestinal anomalies. Isolated congenital urethral fistula is a very rare anomaly. We present a 4-year old circumcised boy with this unusual anomaly. Etiology, embryology, and management are discussed. We emphasize meticulous clinical examination for the diagnosis and to rule out other associated anomalies.

Key words: Congenital, urethrocutaneous fistula, urethroplasty

INTRODUCTION

Congenital urethral fistula is a very rare anomaly where the urethra and meatus are normal and a urethrocutaneous fistula is present, typically coronal or subcoronal.[1] This abnormality usually is an isolated deformity, but may be associated with imperforate anus, hypospadias, or ventral chordee.[1-4] The cause is unclear, but probably reflects a focal defect in the urethral plate that prevents fusion of the urethral folds.[4] Repair is individualized that involves the techniques employed in hypospadias surgery and may involve simple multilayer fistula closure or more complex reconstruction of the distal urethra and glans.[1-5] Thirty-two cases of congenital urethral fistula were reported till 1999 that included cases with associated anomalies also.[5] Since then around half a dozen more such cases have been reported, most of them being isolated ones without any associated anomalies.[6-11] We report a case of isolated congenital anterior penile urethrocutaneous fistula; meticulous clinical examination is stressed and treatment strategies are discussed with literature review.

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CASE REPORT

A 4-year-old circumcised Muslim boy presented with an opening on the undersurface of the distal penile shaft in addition to an opening at the tip of glans. The opening, as reported by parents, was present since birth. The circumcision was done in his infancy by a barber convincing the parents that it would treat the defect also. There was no history of any swelling or injury at the site of the defect. On local examination, there was an oval opening measuring about 8×3 mm^2 on the ventral aspect of the distal penile shaft. There was a normal strip of skin intervening the opening and the frenulum. Penile length was normal with no visible chordee. Glans shape was normal with external urethra meatus normally placed at its tip. The child was voiding through the additional opening with a downward and forward stream and would dribble through the meatus [Figure 1]. He would void through the meatus with stream only when the abnormal opening was blocked with his thumb. Patency of the urethral segment distal to fistulous opening could be confirmed by calibration with an 8-Fr infant feeding tube. The tube could easily be passed down the urethra into the bladder, remaining visible through the fistulous opening [Figure 2]. There was no clinical or radiological evidence of any associated anomaly. A diagnosis of congenital urethrocutaneous fistula was made and Thiersch-Duplay repair was done. Patient is voiding normally after 18 months follow-up.

DISCUSSION

Formation of urethrocutaneous fistulae after hypospadias repair is a common complication, but congenital ones are a rarity.[1-11] Even when they occur, congenital urethral fistulas are usually posteriorly positioned, associated with anorectal malformations, and often represent the ventral functional limb of a Y-type urethral



Figure 1: Patient voiding with a forward and downward stream



Figure 2: Infant feeding tube visible through the fistula

duplication.[3,4,9] Congenital anterior urethrocutaneous fistula is most of the times an isolated deformity, but may be associated wit ventral chordee or imperforate anus.[1,3,4,9] In a series of 14 boys with a congenital urethral fistula by Caldamone *et al*.,[5] the largest series so far, four had distal hypospadias and two had chordee. Although variedly postulated, the cause of congenital urethral fistula is yet to be clearly understood. Some authors believe that the etiology may be same as in hypospadias.[2] Olbourne[4] suggested that the fistula probably reflects a focal or temporary defect in the urethral plate function that results in a complete defect or a partial deficiency of urethral fold fusion. A deficiency of spongiosum with complete canalization of glanular urethra may represent an abnormality of the anlage of corpus spongiosum which is derived from the inner genital folds. Coronal type of fistula may be explained by misalignment of the glanular and penile urethra.[5] Some patients, as in our case, have undergone circumcision before diagnosis that raises the suspicion of fistula being iatrogenic rather than congenital in some of these cases. But, the fact that most of the reported patients were uncircumcised (9 of our 14 in a single-reported series) confirms the congenital nature of this lesion.[5] One patient has 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.[5] Surgical technique must be individualized to fit the defect.[1-11] Treatment usually consists of one of two techniques: (i) the fistula can be circumscribed and then closed in multiple layers, similar to a urethrocutaneous fistula after hypospadias repair, or (ii) if the glans bridge is thin, the ventral glans can be opened through the distal urethra, the distal urethra then being closed by a Thiersch-Duplay tubularization or tubularization of incised urethral plate technique.[1,3,5,9] In cases of deficient distal urethra or spongiosum, associated chordee or hypospadias, the formal hypospadias repair is recommended.[2] Pedicled island preputial tube or onlay urethroplasty can be used to replace distal hypoplastic urethra.[2,9] Before deciding about the surgical correction, it is important to rule out the urethral duplication and association with anorectal malformations. Probing the fistula, radiographic dye study or cystourethroscopic examination may be required to corroborate the diagnosis.[9] We believe that a meticulous clinical examination at presentation is the most useful way of correct diagnosis in case of a congenital anterior penile isolated urethrocutaneous fistula.

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