

Case Report

Congenital anterior urethrocutaneous fistula revisited

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ABSTRACT

Congenital anterior urethrocutaneous fistulas are rare. A case of this unusual anomaly is presented with review of literature, discussion of the possible etiology and management strategy.

KEY WORDS

Congenital, fistula, urethrocutaneous

INTRODUCTION

In contrast to acquired urethrocutaneous fistula congenital anterior urethrocutaneous fistula is a rare anomaly that may present in an isolated fashion or in association with hypospadias or chordee. In the literature, it has been described as unusual type of hypospadias, cryptospadias and congenital urethrocutaneous fistula.^{1,2} Till now only 32 cases have been reported in the English literature.¹⁻⁷ It is a different entity from posterior urethrocutaneous fistula which usually represents Y type duplication of the urethra with a proximal ventral limb and associated anorectal atretic malformations. There is considerable controversy regarding existence of this entity and some claim that it is a result of injury following circumcision.⁶ However, urethral epithelium was not found in the excised skin after circumcision and few previously reported cases were un-circumcised. This firmly established this condition as a distinct entity⁷.

CASE REPORT

A 5 years old boy presented with an additional opening

on the ventral side of the penis, which was present since birth. He was voiding through both the openings. There was no previous history of circumcision, trauma, stone impaction or strangulation. On examination, there was normal prepuce, glans, external urinary meatus and penile shaft without chordee. Distal urethra was patent and surrounded by spongiosum. A congenital urethrocutaneous fistula was present in the proximal penile region measuring 1 x 0.5 cm in size. A catheter was passed through the fistula, which came out of the glandular meatus suggesting intact distal urethra. Anorectum was normal. No other local or systemic abnormality was present. Investigations like urine examination, intravenous pyelography, micturating cystourethrogram and abdominal Sonography were normal. Patient underwent surgical correction under general anaesthesia; artificial erection test confirmed the absence of chordee. Fistula was circumcised and then closed using local skin turn down flaps. An additional layer of local soft tissue was used to re-enforce the repair before final closure of fistula by local skin rotation flap.⁹ A feeding tube was left in the bladder for drainage. On removal of catheter on the 10th day the fistula healed completely. After 1 year

of follow up the patient is having normal voiding and there has been no recurrence.

DISCUSSION

Congenital anterior urethrocutaneous fistula is uncommon as compared to posterior urethrocutaneous fistula, which usually represents Y type duplication of the urethra with anorectal atretic malformations. In the normal process of penile urethral development, external genitalia are represented by a genital tubercle and urethral plate before 9 weeks of gestation. After 9 weeks of gestation, external virilization occurs under the influence of testosterone and dihydrotestosterone. At the end of 12 weeks, the urethral plate folds inwards to fuse in the mid line, forming a tubularized urethra. The urethral plate is made of lamellar cords of cells, which represent the anterior surface of the urogenital sinus. Closure of the open urethral groove occurs following mesenchymal proliferation under the urethral plate epithelium and approximation and fusion of the epithelium in a proximal to distal progression. Glanular urethra formed by proximal closure of the urethral folds, distal epithelium ingrowth and subsequent canalization.⁸⁻¹⁰

The etiology of congenital urethrocutaneous fistula is not well established. Campbell stated that congenital anterior urethrocutaneous fistula represents embryonal urethral blow-out behind a distal congenital obstruction.¹¹ Olbourne theorized that a focal defect in the urethral plate results in arrested distal migration of the urethral plate or localized deficiency of a portion of the plate.¹² Similarly, the testosterone or androgen receptors may be at fault leading to the development of fistula.¹³ 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.¹⁴ Cook and Stephens suggested an alternative mechanism namely pressure atrophy from the heel of the babies' foot, leading to the pressure necrosis.¹⁵ These fistulas may also result from development of cysts along the mediogenital raphae at the frenulum during prenatal period.¹⁶

There are two varieties of congenital anterior urethrocutaneous fistula. One is isolated fistula, which is associated with normal foreskin, no chordee or hypospadias and an intact distal urethra and spongiosum. The other type is associated with hypospadias like characteristics i.e. chordee, a dorsal hood with or without distal urethra or spongiosal defect.

Congenital anterior urethrocutaneous fistula is not associated with imperforate anus; this association is more common in posterior urethrocutaneous fistula, which may represent Y type urethral duplication. Urethra develops from urogenital membrane and this urogenital membrane is the anterior portion of cloacal membrane after it has been divided by urorectal septum. Therefore, a primary defect in the urogenital membrane and cloacal membrane may explain associated imperforate anus and congenital urethrocutaneous fistula.¹⁷

Surgical approach to repair congenital anterior urethrocutaneous fistula depends upon the type of fistula. It is important to exclude the urethral duplication, which produces the Y type of fistula and associated with anorectal malformations. Probing the fistula, radiographic dye study or cystourethroscopic examination may be required to corroborate the diagnosis. In cases of an isolated fistula with intact spongiosum, repair with local flaps are sufficient but if they are associated with deficient distal urethra or spongiosum, associated chordee or hypospadias then formal hypospadias repair is recommended.¹⁸ Various techniques are proposed to repair these fistulas including primary closure via Thiersch-Dupley urethroplasty, turned down flap urethroplasty and pedicled island tube or onlay urethroplasty.

SUMMARY

A case of congenital anterior urethrocutaneous fistula without any other associated anomaly is presented. Principles for repair of these defects are the same as for correcting hypospadias or fistula following hypospadias repair. Etiology of this defect is not clear but appears to be similar to hypospadias.

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