Case Report

Congenital penile urethrocutaneous fistula: A rare anomaly and review of literature

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Abstract

Congenital penile urethrocutaneous fistula is described as an unusual developmental anomaly in children who present with an abnormal opening on the ventral aspect of penis with a normal foreskin and an absence of chordee and hypospadias. The authors present a discussion on the etiology, embryology, and management of this entity along with a description of three cases. We emphasize meticulous clinical examination for the diagnosis and to rule out other associated anomalies.

Key Words: Anorectal malformations, hypospadias, urethrocutaneous abnormalities, urethrocutaneous fistula

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Received: 27.08.2010, **Accepted:** 03.11.2010

INTRODUCTION

Congenital urethral fistula and congenital chordee without hypospadias are best explained by the theory of deficient urethral plate.[1] This abnormality usually is an isolated deformity, but may be associated with imperforate anus, hypospadias, or ventral chordee.[2-5] The cause is unclear, but probably reflects a focal defect in the urethral plate that prevents fusion of the urethral folds.[5] Thirty-two cases of congenital urethral fistula were reported till 1999 that included cases with associated anomalies also.[6] Since then around half a dozen more such cases have been reported, most of them being isolated ones without any associated anomalies.[7-13] Here, we report three cases of congenital anterior penile urethrocutaneous fistula out of which two are isolated and one is associated with Anorectal malformation (ARM); management plans are discussed with review of literature.

Access this article online	
Quick Response Code:	Website: www.urologyannals.com
	DOI: 10.4103/0974-7796.84953

CASE REPORTS

Case 1

A 1-year and 6-month-old male baby presented with complaints of absence of anal opening since birth with status divided sigmoid colostomy done outside and an abnormal opening on the undersurface of penis, which had been present since birth. The patient was referred for definitive surgery for ARM from a district hospital. The patient was passing urine from both the terminal glandular meatus and the fistulous opening. There was no history of any trauma, strangulation, stone impaction, or surgery. There were no signs of inflammation. On examination of perineum, gluteal folds were well developed with a midline groove and an ovoid shape urethrocutaneous fistula with clear margins measuring 2 cm by 1 cm was present on the ventral aspect of penile urethra approximately 3 cm distal to penoscrotal junction [Figure 1]. The external urethral meatus appeared normal with normal foreskin and without any chordee or hypospadias. Distal colostogram showed intermediate anomaly with rectourethral fistula. Ultrasonography of abdomen, micturating cystourethrogram, Hemogram, blood urea, and serum creatinine were normal. After anterior sagittal anorectoplasty tubularized incised plate urethroplasty was planned with the aim to reconstruct a near-normal caliber neourethra. After applying stay sutures, proposed circumferential and racket-shaped incisions

incorporating the fistulous opening were marked. The penile shaft skin was degloved, fistulous margins were mobilized and urethral plate 'relaxing incision' was made in through fistulous opening. The margins of fistulous opening were tubularized on 6F Nelaton's catheter. The neourethral suture line was reinforced with dartos flap. The penile skin was rotated ventrally to provide skin coverage. The postoperative period remained uneventful, and presently patient is doing well for the last 2 years.

Case 2

A 3-month-old male presented with an abnormal opening on the ventral aspect of penile urethra, which had been present since birth. The patient was passing urine via normal meatus and the congenital fistula. There was no previous history of surgery, trauma, stone impaction, or strangulation. On examination, there was normal prepuce, glans, external urinary meatus, and penile shaft without chordee. A congenital urethrocutaneous fistula was present in the mid penile region measuring 1 cm × 0.5 cm in size [Figure 2] approximately 2.5 cms distal to penoscrotal junction with well-defined margins. There were no signs of inflammation and urethra distal to the fistula appeared to be normal on clinical examination. A catheter was passed through the fistula, which came out of the glandular meatus suggesting intact distal urethra. Routine blood tests, urine examination, micturating cystourethrogram, and abdominal ultrasonography were normal. At the age of 8 months, 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. A 6F Nelaton's catheter was left in the bladder for drainage. The postoperative period remained uneventful. After 12 months of follow-up, the patient is having normal voiding and there has been no recurrence.

Case 3

An 8-month-old baby was referred to us with complaint of passing urine from 2 openings, which had been present since birth. There was no previous history of surgery, trauma, stone impaction or strangulation. The patient used to pass most of the urine from the urethral opening and only a small amount through the fistula opening. On examination, a fistula of size 0.5 cms × 0.5 cms with well-defined margins was present in the penoscrotal region [Figure 3]. There were no signs of inflammation present and urethra distal to the fistula appeared to be normal clinically. The external urethral meatus was normal, with an absence of chordee or hypospadias. All basic investigations were normal. Surgery was performed by circumcising the fistula and then closing using local skin turn down flaps. The patient recovered satisfactorily and is on regular follow-up for the last 7 months and is doing well.

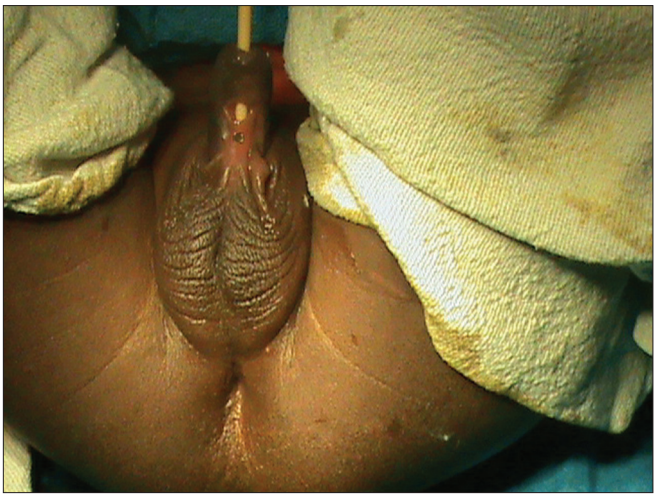

Figure 1: Congenital urethral fistula in a patient of anorectal malformation

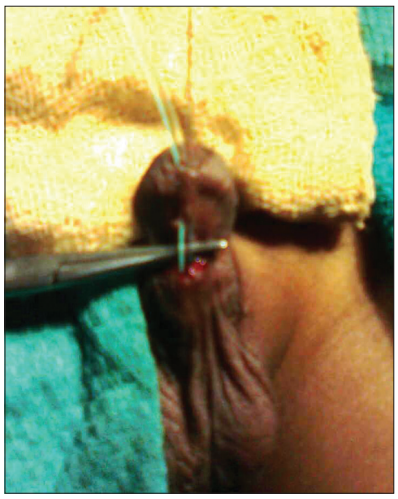

Figure 2: Isolated congenital urethral fistula

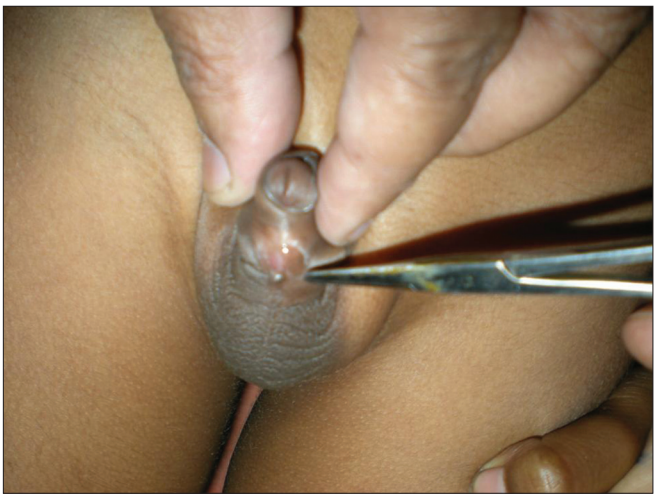

Figure 3: Fistula in penoscrotal junction

DISCUSSION

Congenital anterior urethrocutaneous fistula is not commonly

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.[14]

The etiopathogenesis of congenital penile urethrocutaneous fistula still remains unclear.[6,9,15,16] Although its etiopathogenesis has been explained by the theory of pressure necrosis of penile urethra by fetal parts, yet an absence of scarring in the present and a few reported cases supports the theory of focal developmental defect of urethral plate, which prevents the fusion of urethral folds, thus resulting in isolated congenital penile urethrocutaneous fistula.[4,9,15] Olbourne 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.[5] Similarly, the testosterone or androgen receptors may be at fault leading to the development of fistula.[1] The distal glanular penile urethra is normal as it develops from a surface ectodermal invagination.[4] 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. Distal type of fistula may be explained by misalignment of the glanular and penile urethra,[6] as seen in one of our cases.

Surgical approach to repair congenital anterior urethrocutaneous fistula depends upon the type of fistula. 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.[3] To conclude, congenital penile urethrocutaneous fistula is an extremely rare but easily manageable anomaly and a good clinical examination is needed for diagnosis and to rule out other associated anomalies.

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How to cite this article: Shukla RM, Mukhopadhyay B, Mandal KC, Barman SS. Congenital penile urethrocutaneous fistula: A rare anomaly and review of literature. Urol Ann 2011;3:161-3.

Source of Support: Nil, **Conflict of Interest:** None.