Isolated Adult Congenital Anterior Urethrocutaneous Fistula: A Rare Anomaly and Review of Literature

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Abstract

Congenital anterior urethrocutaneous fistula of the male urethra is a rare deformity, frequently located at the subcoronal level. It is usually accompanied by anorectal and genitourinary malformations, but may present as a very rare, isolated anomaly along with a normal urethra and meatus. The adult type of isolated congenital anterior urethrocutaneous fistula is an extremely rare anomaly, with only two such cases previously reported in the English language literature. This case report presents an adult patient with this rare deformity.

Keywords: Adult, Isolated, Congenital, Anterior urethrocutaneous fistula.

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Introduction

Contrary to acquired urethrocutaneous fistula, which is a well-known complication of hypospadias repair, congenital anterior urethrocutaneous fistula (CAUF) of the male urethra is quite a rare anomaly of unknown etiology. It may present as a rare, isolated form or be associated with anorectal or several genitourinary pathologies such as distal hypospadias, penile chordee, deficient distal urethra, megalourethra, urethral duplication, and stenosed bulbar urethra.[1,2,3] Although the cause is still unclear, it reflects a focal defect in the urethral plate that prevents fusion of the urethral folds.[4] The surgical approach for repairing it depends on whether the fistula is isolated or is accompanied by another anomaly.

To the best of our knowledge to date, about fifty cases of CAUF have been described in the English language literature, two of them in adult patients. Chen et al. (2011) reported this anomaly in a 26-year-old male, and Jindal et al. (2011) reported it in a 19-year-old male.[5,6] We present a very rare case of an isolated adult congenital anterior urethrocutaneous fistula, discussing possible etiology, embryology, and treatment strategy.

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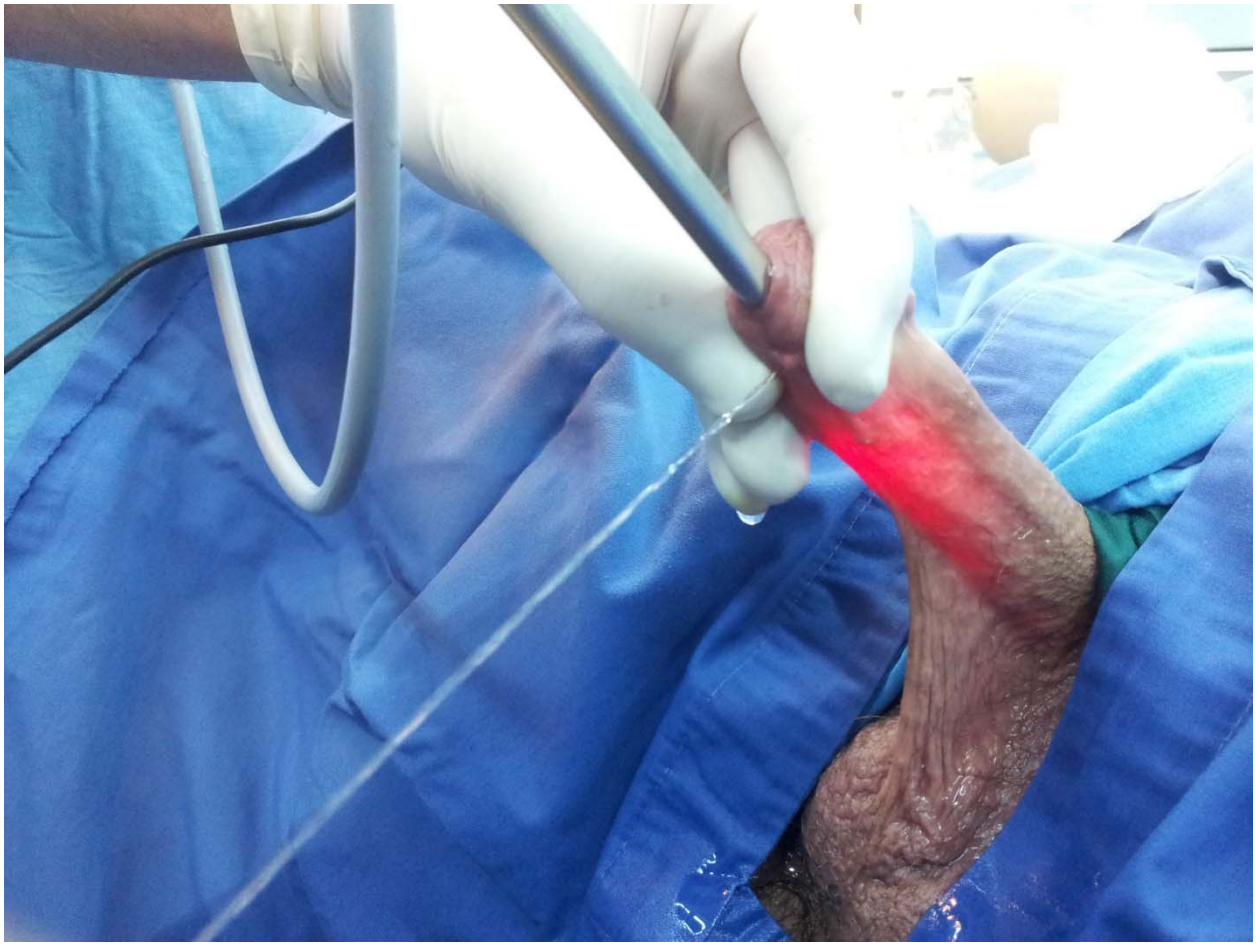
Case Report

A 25-year-old male presented with an abnormal extra opening on the ventral aspect of the penile shaft that had been present since birth, with a normal external urethral meatus distal to it. The patient stated that, since birth, most of his urine would be passed through the urethral opening and only a small amount through the fistula opening, but he did not care about it himself. The patient had normal erectile function and a normal sexual history. He denied previous history of trauma during or after birth, except an uncomplicated surgical circumcision at the age of 5 years that was done by a urologist. At the time of circumcision, he was planned for a repair of the fistula, but his parents decided to postpone it.



On examination, the glans was normal in size and shape, with a normally placed external urethral meatus and an absence of chordee, hypospadias or anorectal malformation. There was a small pinpoint opening 2 cm below the corona on the ventral aspect of the penile shaft, just lateral to the midline. There was no other local or systemic abnormality present. At the time of micturition, the patient voided mainly through the normally placed meatus, with dribbling of urine from the ventral opening.

The technique of repair depends on the appearance of the fistula and local findings, such as the skin and distal urethra. In our case, we started the procedure by a

cystourethroscopy evaluation, which showed the stream of irrigation coming through the fistula opening (Figure 1). Otherwise, the other parts of the urethra, bladder neck, and urinary bladder were normal. After cystourethroscopy, a 16 Fr urethral catheter was inserted from the external urethral meatus. The fistula tract was identified by insertion of a small caliber metallic probe and excised completely, and a double-layered repair of the defect was done with a 5/0 PDS suture (Figure 2).

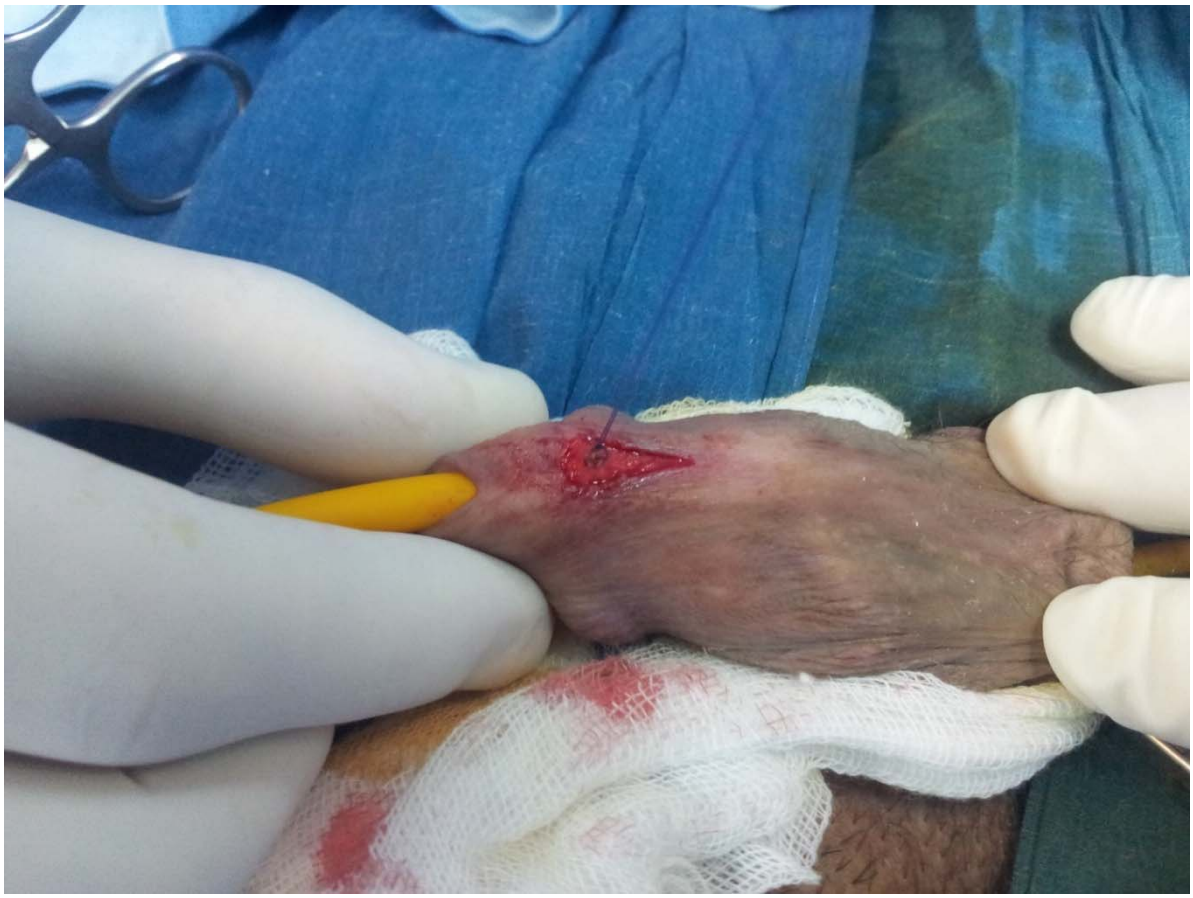


Figure 1: The anterior urethrocutaneous fistula after repair

On the second postoperative day, the patient was able to pass urine normally after removal of the urethral catheter, and the fistula was completely healed. After two years of follow-up, the patient reported normal voiding without recurrence of leakage.

Discussion

A congenital anterior urethrocutaneous fistula (CAUF) of the male urethra is a rare anomaly. It may be seen in an isolated form or be associated with other malformations, such as anorectal and genitourinary malformations.[1,2]

Having knowledge about the embryogenesis of the male genitourinary system is essential for understanding the genesis of this anomaly. Until 8 weeks of gestation, male and female external genitalia are similar.[7] Testosterone is secreted by Leydig cells of the fetal testis, under

the influence of human chorionic gonadotropin, and is converted to dihydrotestosterone, the main stimulus for masculinization to occur.[8,9] Elongation of the genital tubercle, which will become the penile shaft and glans, and an increase in the distance between the anus and genital structures are the early effects of testosterone.[10]

As the phallus enlarges and elongates to become the penis, the urethral fold gets pulled and forms the lateral wall of the urethral groove on the ventral surface of the penis. This groove has inner (endodermal) and outer (ectodermal) edges.[10,11] A urethral groove closed by fusion of the endodermal folds forms a tubular penile urethra.[9,12] The ectodermal edges then fuse in the median plane of the penis, forming the penile raphe and enclosing the penile urethra within the penis.[11]

Lastly, the distal or glanular urethra develops by one of two proposed mechanisms.[9] The classic theory implies that the distal portion of the urethra develops as an ingrowth of the ectoderm from the tip of the penis until it joins the proximal tubular urethra.[11,13] However, this theory for the development of the distal granular urethra has been challenged by the "endodermal differentiation theory." It has been suggested that the entire penile urethra might differentiate from the fusion of epithelial-mesenchymal interactions.[13]

The etiology of a urethrocutaneous fistula is unclear and considerably controversial, with some claiming that it is may be a result of urethral injury during circumcision.[4] In our case, however, the patient had reported that the urinary stream coming from the external urethral meatus with a few drops from the ventral aspect of the penis occurred before he underwent circumcision at the age of five. Therefore, the cause of his fistula was determined to be congenital rather than acquired.

There are several theories proposed to explain the embryologic mechanism of this anomaly. Glenister et al. (1954) suggested that the penile and glanular urethra develop from different sources, and a fistula can develop between them due to misalignment of the glanular and penile urethra.[14] According to Olbourne et al. (1976), the cause of a congenital urethrocutaneous fistula is due to a focal defect in the urethral plate, which prevents fusion of urethral folds, and leads to arrested distal migration of the urethral plate and localized deficiency of a portion of the plate.[4] Campbell (1951) mentioned that congenital urethral fistulas represent embryonal urethral blowouts behind a distal congenital obstruction.[15]

The type of fistula repair chosen depends upon position, size, appearance, and local findings, including the skin, distal urethra and presence of other abnormalities, such as an anorectal anomaly, chordee or hypospadias. Several surgical techniques for repair have been described, varying from simple to complex, including preputial bound skin flap, pedicle flap, modified Denis Browne urethroplasty or direct closure.[3,4,16] According to Alhazmi et al. (2014), primary repair was successful in 42 of 47 cases, with four patients requiring two operations and one patient requiring three closures.[3] In the present case, since the fistula was an isolated abnormality, and the urethra beyond the fistula was intact, primary closure was used successfully for repair.

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التشوه الخلقي المعزول عند البالغين بناسور بين الإحليل الأمامي والجلد، حالة مرضية نادرة ومراجعة للحالات

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الملخص

يعدّ التشوه الخلقي للإحليل الأمامي عند الرجال البالغين من التشوهات الخلقية النادرة، وعادة يكون مترافقا في نفس الوقت مع شذوذات في فتحة الشرج والمستقيم والجهاز البولي، او قد يكون بشكل منعزل؛ أي لا يترافق مع تشوهات أخرى في مجرى البول والفتحة البولية، وهذا النوع نادر الحدوث، وقد تم تسجيل حالتين من هذا النوع النادر جدا في الدراسات السابقة باللغة الإنجليزية. نقدم هذه الحالة من هذا التشوه الخلقي النادر مع مراجعة للدراسات السابقة.

الكلمات الدالة: البالغين، منعزل، خلقي، ناسور بين الإحليل الأمامي والجلد.