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A Case Report

ANTERIOR URETHROCUTANEOUS FISTULA CONGENITAL ANTERIOR URETHROCUTANEOUS FISTULA WITH CHORDEE: A CASE REPORT AND LITERATURE REVIEW

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ABSTRACT:

Background: Congenital anterior urethrocutaneous fistula is a rare genitourinary anomaly involving an abnormal connection between the urethra and the skin. It may present alone or with other conditions like hypospadias and chordee. The exact cause is unclear, but it's believed to result from incomplete urethral fold closure during embryogenesis. The presence of chordee complicates diagnosis and treatment, especially when not associated with hypospadias. Due to its rarity, standardized treatment protocols are lacking. This case report adds to the limited literature and discusses surgical management strategies.

Objective: To describe the congenital variant of anterior urethrocutaneous fistula with chordee and review the literature for its etiology and surgical management.

Study design: A Case Report

Place and duration of study: Department of Urological Surgery and Transplantation, Jinnah Postgraduate Medical Center, Karachi, Pakistan from Jan 2022 to Jan 2023

Methods: A comprehensive literature review was performed to collect information about the etiology and surgical management of a congenital anterior urethrocutaneous fistula, especially with respect to chordee variants.

Results: Congenital anterior urethrocutaneous fistula is a rare entity and only 67 cases had been reported worldwide. It may be an isolated anomaly or may exist along with other conditions such as hypospadias and chordee.

Case Presentation: A 14 year-old boy who presented with chordee was diagnosed as having this rare condition. The chordee did not prevent the surgical correction and urethroplasty with good postoperative results. The patient was observed in outpatient clinic and is doing well in follow up.

Keywords: Congenital Urethrocutaneous fistula, Chordee, Urethroplasty

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INTRODUCTION

Congenital urethrocutaneous fistula of the anterior urethra is an extremely rare anomaly. It may present in varying forms; sometimes isolated and sometimes associated with other anomalies like chordee, hypospadias, and at times, anorectal malformations. Exact cause of this malformation is unknown but it is hypothesized to be caused by localized defect in epithelial lining of ventral urethra.[1] In comparison to posterior urethrocutaneous fistula usually classified as urethral duplication or accessory urethra, Congenital Anterior Urethrocutaneous Fistula or CAUF is very uncommon and till date only 67 cases have been reported. Effect of this deformity on future potency and fertility is not known yet. Surgical approach for the treatment of CAUF is individualized and is based on a number of features like size and site of fistula, patency of the distal urethra, and other associated anomalies. Here, we report a case of CAUF associated with chordee in a 14 year old boy and discuss the management of this uncommon deformity.

Case Presentation

A 14-year-old child complained to us that he had been leaking urine from two apertures since birth. Family history of comparable defect was not there. The youngster had a typical mental and physical make-up. He showed no signs of endocrine malfunction and had not acquired secondary sex traits. There was not another congenital abnormality that affected him. He was able to erect his penis normally. Upon local inspection, the gonads were in their usual position, the scrotum and corpora cavernosa were developing normally for his age, and the phallus was well- circumcised and of normal size. There was palpable corpus spongiosum up to the penoscrotal junction, distal to which it was splayed. (Figure 1)

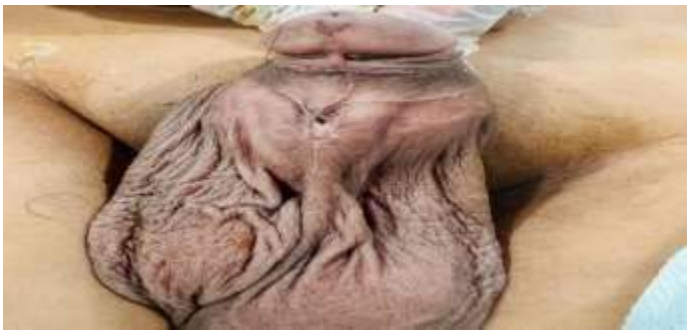


Figure 1: normal sized phallus a normal position of the gonads and normal development of the scrotum and the corpora cavernosa with distal splayed corpus spongiosum

An opening of 3mm×3mm with well-defined margins was present on the ventral aspect of the midshaft of the penis with type 2 chordee. (Figure 2)

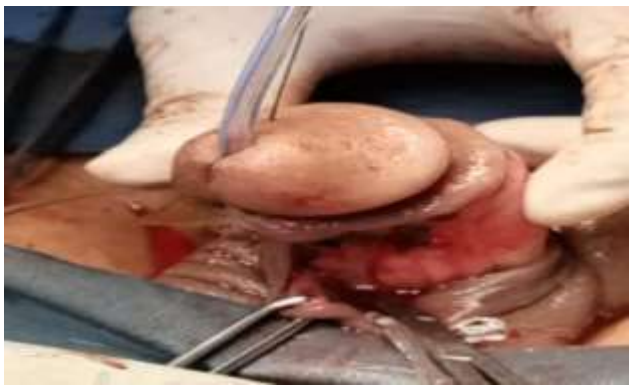
Figure 2: a fistula of 3mm×3mm with well-defined margins was present on the ventral aspect of the midshaft of the penis

No signs of inflammation were presentand urethra distalto the fistula appeared to be normal clinically with adequate external urethral meatus (Figure 3).

Figure 3: adequate external urethral meatus

No prior history of trauma, stone impaction, surgery, or strangulation existed. The patient used to pass most of the urine from the normal urethral opening and only a small amount dribbles through the abnormal fistulous opening. Normal baseline examinations were conducted. To rule out urethral duplication, a micturating cystouretrogram was performed; the results were unremarkable. Under general anaesthesia, the patient had the defect surgically repaired. Using aurethral dilator, distal urethral patency was examined till 14Fr, indicating the lack of stricture (Figure 4).

Figure 4: Distal urethral patency was checked with a urethral dilator until 14Fr



An artificial erection confirmed the extent of chordee (Figure 5).

Figure 5: Gittes Test

The ureterorenoscope was used to investigate the fistula's connection with the urethra; however, since the fistula has a pin-point lumen,contact can not be established. A longitudinal incision around the ventral fistula was made. Ventrally, the penile shaft skin was degloved. Few subcutaneous fibrotic bands were encountered which

were surgically removed. The corpus spongiosum encircled a normal proximal urethra, and the distal urethra surrounding the fistula likewise seemed normal, but the corpus spongiosum was splayed ventrally. There was good urethral plate development. After the fistula was outlined, it was layer-closed using a continuous, resorbable polydioxanone suture while the urethra was inverted. There was enough tissue for two more layers to be formed before the skin closed. After that, the corpus spongiosum was dissected to conceal the urethroplasty during spongioplasty. In order to reduce the possibility of a fistula recurrence, sutures were subsequently covered with the subcutaneous tissue as an extra layer. To verify chordee repair, an artificial erection was once again generated. (Figure 6)

Figure 6: Post repair Gittes Test

The skin was finally closed longitudinally. For drainage, a tiny silicon catheter was left with in the bladder. Day 2 saw the patient's release, and Day 6 saw the outpatient removal of the Foley catheter. The patient made a full recovery, is still receiving frequent follow-up care, and is doing well. There has also been a noticeable improvement in chordee.

DISCUSSION

Etiopathogenesis of congenital penile urethrocutaneous fistula is still not well understood. Several factors have been believed to be cause of this defect including genetic factors and abnormalities in androgenic function. The Absence of scarring in all reported cases supports the theory of focal developmental defect of urethral plate preventing the fusion of urethral folds resulting in isolated congenital penile urethrocutaneous fistula. [1] Spongiosum may be deficient but distal glanular penile urethra is normal with complete canalization as it develops from a surface ectodermal invagination. Distal type of fistula may be explained by misalignment of the glanular and penile urethra.[2] Although our patient as no associated anorectal abnormality, different cases have been reported in literature of congenital penile urethral fistula with associated imperforate anus, thus it is also postulated that congenital fistula may be a result of asymmetric closure of the urorectal septum. The differentiation of the urethral plate is induced by male hormones. [3] Our patient had a well developed distal penile urethra with chordee, but androgen abnormality was not present as ruled out by normal serum testosterone, follicular stimulating and luteinizing hormone levels. Apart from single penile anterior urethrocutaneous fistula, patient did not have any other anomaly.Out of 67 patients reported in past, chordee was present in 8 patients (14.5%), associated genitourinary anomalies were present in 19 patients (30.2%) and mostly fistulas were found in sub coronal region, (46%). Similar to our case, usually urethra distal to fistula is intact and success rates are good in repairing fistulas. In past cases reported, recurrence of fistulas was noted in 11.3% of patients out of which 50% were closed spontaneously without any further intervention [4].Different methods have been employed in past for treatment of CAUF. In isolated subcoronal CAUF cases Gupta and Olbourne used Denis Browne Procedure [3]; Tiersch-Duplay technique has been employed in repair of fistula by Caldamone[5], Tennenbaum[6] and Rashid[7], Karnak[8] used proximal based skin flap, Biswas[9] and Alhazmi[10] used used Bayer's and dartos flaps respectively, whereas, Jindal[11] and Bhatnagar[12] used 2-layer closure technique in the management of the mention as we did in our case. Difference is that they used in treating subcoronal fistula whereas our patient had fistula in mid penile region.In treating midpenile isolated CAUF cases, majority of the cases previously reported used transverse preputial island flap. Barwell [13] used split preputial flap, Shukla [14] used tubularized incised plate urethroplasty in one case with anorectal malformation , in rest of the cases, fistula was circumcised and then closed using local skin turn down flaps as we did in our case; others also used similar techniques to close the defect in 3-layers. Similar to our case, Islam et al[15] closed midpenile isolated CAUF using 2-layer technique with aid of local skin flaps. Before proceeding with surgical intervention, the type of congenital urethral fistula must be well established. Urethral duplication or 'Y' type fistula must be excluded. Unremarkable voiding cystourethrogram and probing the fistula to demonstrate continuity with the distal penile urethra was sufficient to confirm the diagnosis in our case. Treatment of congenital fistula depends on the associated penile abnormalities and fistula location. Usually the urethra distal to fistula is deficient and simple closure is likely to be followed by recurrence. To our knowledge, fistulas were closed successfully in 1 stage in a number of cases previously reported. Hence, primary closure can readily be performed if there is healthy spongiosum or dartos tissue. In cases of associated chordee or a thin distal urethral segment excision and replacement of the segment distal to the urethra may be necessary. Staged repair probably would not be needed even under these circumstances because replacement of the distal urethra could be managed with a vascularized preputial flap. 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.

CONCLUSION:

The uncommon genitourinary defect known as congenital anterior urethrocutaneous fistula (CAUF) is sometimes accompanied with chordee, hypospadias, or other congenital abnormalities. The precise cause of CAUF is still unclear despite its rarity, while localised abnormalities in the ventral urethral lining's epithelium are thought to be responsible. Individualised surgical carefor CAUF takes into account the location, size, and accompanying abnormalities of the fistula. Our 14-year- old boy's case report demonstrates the effective surgical repair of both chordee and CAUF.By using multilayered closure procedures and careful dissection, we were able to obtain positive results without any negative repercussions.

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Authors'Contribution:

1-Concept & Design Of Study: AnumAnsari[12]-

Drafting:. Anum Ansari[1]

3-Data Analysis: NareshKumar[24]-

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5-Final Approval Of Version: All Mantion Above.

REFERENCES

1. Glenister TW. The origin and fate of the urethral plate in man. J Anat. 2019;88(3):413-25. PMID: 13192029; PMCID: PMC1244688.

2. Gupta SC. An unusual type of hypospadias. Br J Plast Surg. 2016;15:191-93. doi:10.1016/S0007-1226(62)80024-1.

3. Burns RC, Fairbanks TJ, Sala F, De Langhe S, Mailleux A, Thiery JP, et al. Requirement for fibroblast growth factor 10 or fibroblast growth factor receptor 2-IIIb signaling for cecal development in mice. Dev Biol. 2020;265(1):61-74. doi:10.1016/j.ydbio.2020.09.021. PMID: 14697353.

4. Lin Y, Deng C, Peng Q. Congenital anterior urethrocutaneous fistula: A systematic review. Afr J Paediatr Surg. 2018;15(2):63-68. doi:10.4103/ajps.AJPS_97_17. PMID: 31290465; PMCID: PMC6615009.

5. Caldamone AA, Chen SC, Elder JS, Ritchey ML, Diamond DA, Koyle MA. Congenital anterior urethrocutaneous fistula. J Urol. 2019;162(4):1430-32. PMID: 10492230.

6. Tennenbaum SY, Palmar SL. Congenital urethrocutaneous fistula. J Paediatr Urol. 2022;43(1):98-99. doi:10.1016/S0090-4295(94)80275-0.

7. Rashid KA, Kureel SN, Tandon RK. Congenital anterior penile isolated urethrocutaneous fistula: A case report. Afr J Paediatr Surg. 2021;5(1):52-53. doi:10.4103/0189-6725.41640. PMID: 19858667.

8. Karnak I, Tanyel FC, Hiçsönmez A. Congenital urethrocutaneous fistula: A case report and literature review, with a nomenclature proposal. J Pediatr Surg. 2022;30(10):1504-05. doi:10.1016/0022-3468(95)90422-0. PMID: 8786504.

9. Biswas S, Ghosh D, Das S. Congenital urethrocutaneous fistula: Our experience with nine cases. Indian J Surg. 2020;76(2):156-58. doi:10.1007/s12262-012-0763-6. PMID: 24891786; PMCID: PMC4039680.

10. Alhazmi HH. Congenital anterior urethrocutaneous fistula: Two case reports and review of literature. Urol Ann. 2022;6(3):239-41. doi:10.4103/0974-7796.134285. PMID: 25125898; PMCID: PMC4127862.

11. Jindal T, Kamal MR, Mandal SN, Karmakar D. Isolated congenital urethrocutaneous fistula of the anterior urethra. Korean J Urol. 2019;52(5):368-70. doi:10.4111/kju.2011.52.5.368. PMID: 21687400; PMCID: PMC3106173.

12. Bhatnagar A, Upadhyaya VD, Kumar B. Congenital urethrocutaneous fistula: Case report with review of literature. Indian J Plast Surg. 2019;45(3):563-65. doi:10.4103/0970-0358.105979. PMID: 23450670; PMCID: PMC3580363.

13. Barwell J, Harris D. Case report: congenital urethrocutaneous fistula. J Anat. 2020;190(Pt 1):155-56. doi:10.1046/j.1469-7580.1997.19010155.x. PMID: 9034892; PMCID: PMC1467594.

14. Shukla RM, Mukhopadhyay B, Mandal KC, Barman SS. Congenital penile urethrocutaneous fistula: A rare anomaly and review of literature. Urol Ann. 2022;3(3):161-63. doi:10.4103/0974-7796.84953. PMID: 21976932; PMCID: PMC3183711.

15. Islam MK. Congenital Penile Urethrocutaneous Fistula. Indian J Pediatr. 2019;68(8):785-86. doi:10.1007/BF02752424. PMID: 11563258.