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Case report

Congenital anterior urethrocutaneous fistulas with chordee: A case report

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ABSTRACT

Congenital anterior urethrocutanous fistula is a rare genitourinary malformation with unknown etiology and may present as an isolated form or in association with hypospadias or chordee. We describe a case of congenital anterior urethrocutanous fistula with chordee variants in a 5-year-old boy. The patient successfully underwent chordectomy and urethroplasty with no adverse outcomes. We report a review of the literature about the etiology and surgical intervention especially with chordee variants. The congenital anterior urethrocutanous fistula is a rare anomaly with unkown etiopathogenesis.

1. Introduction and importance

Congenital anterior urethrocutanous fistula (CAUF) of the male urethra is a rare anomaly of unknown etiology [1]. CAUF is very rare with only 63 cases reported in the literature [2]. There are two variants of CAUF, one associated with hypoplastic distal urethra, chordee and hooded foreskin. This variant has strong association with anorectal malformation. The other is an isolated congenital fistula with normal foreskin, normal glans fusion, and normal urethral development proximal and distal to the fistula [2]. The cause of this problem is unclear but, a focal defect in the urethral plate can prevents fusion of the urethral folds [3,4]. We describe a case of CAUF with chordee variants discussing its etiology and surgical intervention as well as reviewing the literature in this case report. This case report has been reported in line with the SCARE Criteria [18].

2. Case presentation

A five-year-old boy referred to our pediatric urology department presented with passage urine from an abnormal opening in ventral surface of the penis. The patient had no history of trauma, strangulation, impacted stones, or surgery. There is no history of fever, pain, or signs of inflammation. The patient was the first child born at term by cesarean section with birth weight of 3 kg. History of growth and development is recognized according to age. The patient had not consumed or received any medication related to his symptom. History of allergy was denied. Upon inquiry, history of genitourinary malformation in family was denied. On physical examination, a distal patent urethra was found [Fig. 1] measuring 5 × 5 millimeters with chordee. Distal to the fistula, the glans is intact [Fig. 2] with normal wide urethral meatus at its tip. On catheterization an 8 Fr Nasogastric tube passed proximally with ease from the tip of the glans to the bladder. Laboratory findings were within normal limits. We decided to perform chordectomy and urethroplasty on this patient with grade I risk of complication according to Clavien-Dindo Classification. At operation, a formal tabularized incised urethroplasty (Snodgrass urethroplasty) was done. Closure of the fistula was done in double layer, continuous subepithelial manner using 6/0 Polydioxanone (PDS) sutures with a protective ventral dartos flap [Fig. 3]. The patient was discharged on the same day, as expected, while the catheter was removed 7 days later. The patient is behaving well after 6 months of follow up visits with no adverse outcomes.

3. Clinical discussion

CAUF has been named as criptospadias (a usual type of hypospadias) [1]. It is defined as a localized defect in penile urethra of congenital origin [2]. Many authors believe the etiology may be same as in hypospadias. But the exact etiology of congenital urethrocutaneous fistula is not clear yet, but some hypotheses have been proposed [1]. CAUF is an extremely rare condition [5]. The common site fistula location was subcoronal, followed by the mid-penile with proximal penile and penoscrotal being the least reported location [6]. Most of CAUF are coronal or subcoronal. The fistula was at the distal shaft in 24/47 patients, mid shaft in 13/47 patients, penoscrotal junction in 3/47 patients and non-specified in 7/47 patients [1].

Fusion failure of urethral folds leads on to various level of

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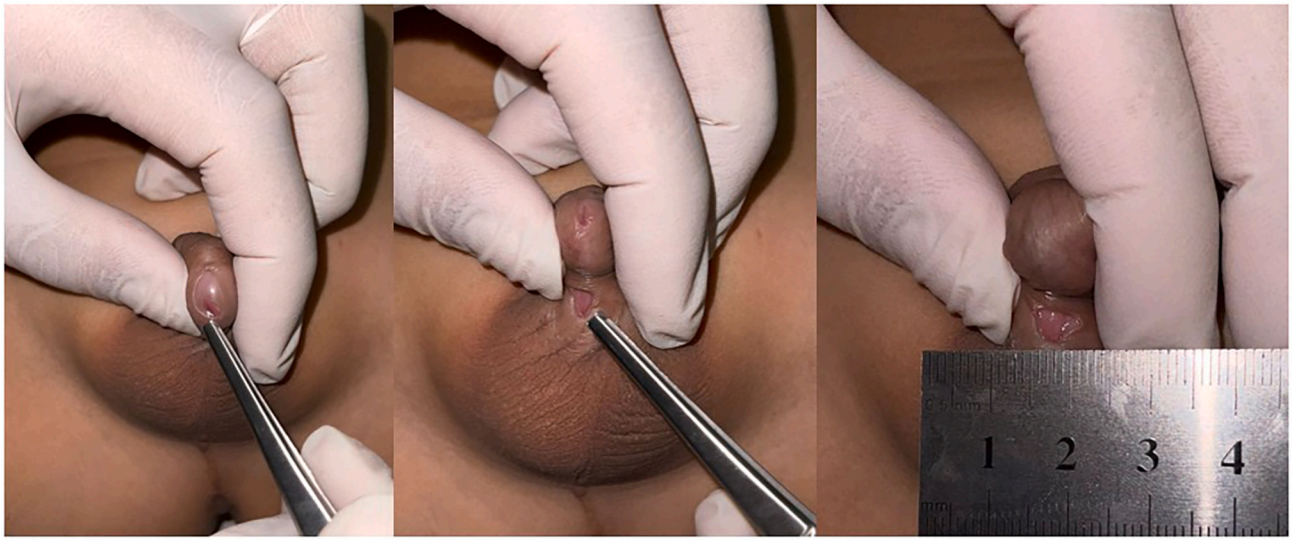

Fig. 1. A urethrocutaneous fistula in the part of the penis measuring 5 × 5 mm with normal urethral meatus at the tip of penis.

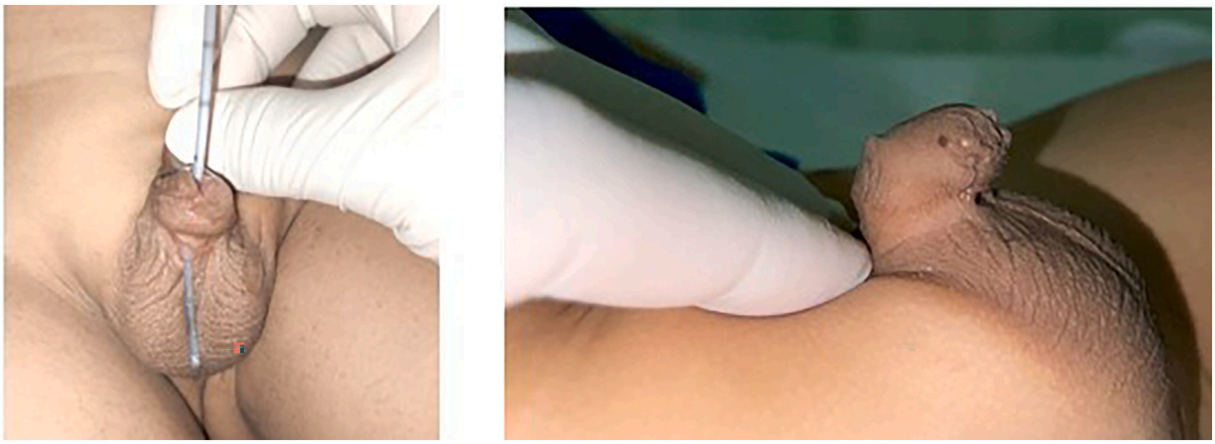

Fig. 2. Physical examination Urethrocutaneous fistula.

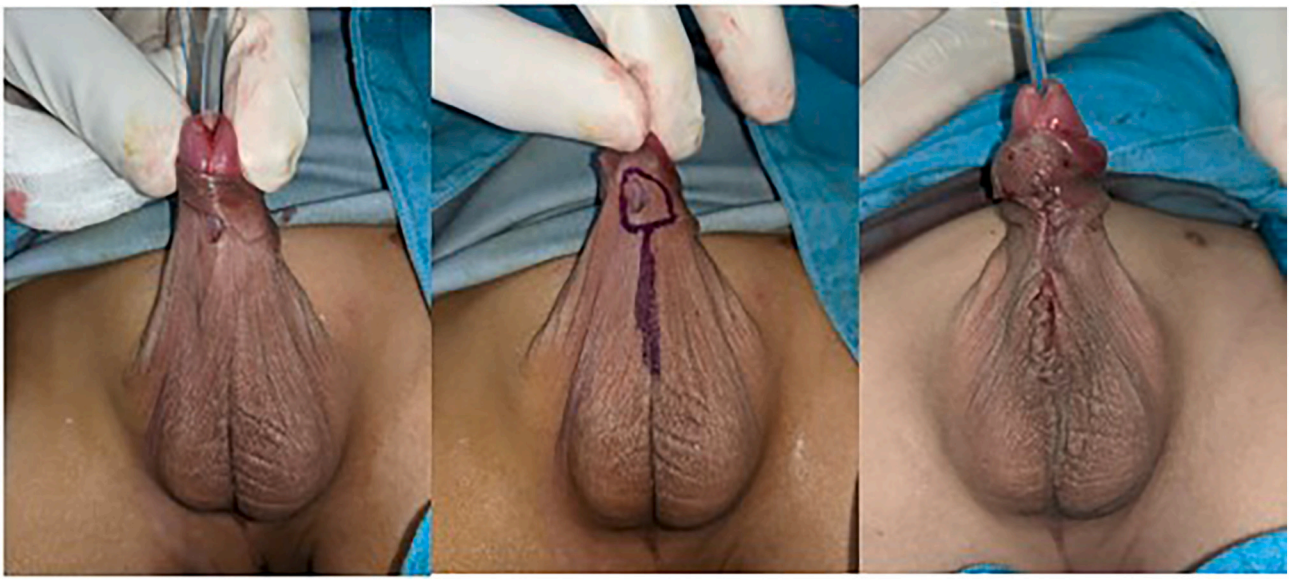

Fig. 3. Surgical procedure and post operation.

hypospadias or in extreme case may lead to congenital urethrocutaneous fistula. Lack of fusion may occur in any level from perineum to glans [7]. Goldstein also theorized that a transient deficiency in testicular evocator substance could produce congenital urethral fistula with chordee [8].

Theory from Olbourne suggested that fistula located in the penile shaft may reflect a focal or temporary defect in urethral plate function and this would result in a complete defect or a partial deficit of urethral fold fusion [10]. Karnak regarded that congenital urethrocutaenous fistulas (excluding those associated with anorectal malformations) as one set of anomalies [4].

CAUF also associated with other congenital anomalies, such as deficient distal urethra, distal hypospadias, ventral penile curvature, anorectal malformations, stenosed bulbar urethra, epispadic urethral duplication, and megalourethra [1]. Two types of CAUF have been described. The first type is the isolated urethrocutaneous fistula with intact distal urethra and spongiosum. This type has normal foreskin and without chordee or hypospadias and may be caused by the blowout phenomenon of a urethral diverticulum [10]. The second type, CAUF associated with hypospadias with chordee and dorsal hood with/without a distal spongiosal or urethral defect [11]. Our case is reported the second type of CAUF especially with chordee.

Patient usually present with frequent dribbling through the fistula despite a normal urethral meatus is present. Clinician must consider the history of the patient. Some of the crucial history such as, previous trauma, circumcision or surgical procedures can give the clue about the cause of the fistula. Other investigations may have a necessary role to evaluate other genitourinary anomalies [6].

Adjunct image such as ultrasound abdomen and cysthourethrogram can show the presence of double urinary flows, one from the tip and the other from the fistula [6]. Hasan et al. recommended a preoperative cystourethrogram for differentiating a congenital fistula from a urethral duplication [12]. If a Y duplication of the urethra is suspected, a preoperative micturating cystourethrogram can help to define the anatomy [13].

CAUF usually treated with surgical procedure. There are many methods of surgical repair have been described by many authors. The procedure includes local skin flap, preputial skin flap, Thiersch-Duplay Urethroplasty, Denis Brown urethroplasty, buccal mucosal graft, and tubularized incised plate urethroplasty with dartos flap and primary closure. The choice depends on many variables, e.g., the size of the fistula, the location of the fistula, other existing abnormalities, such as hypospadias or chordee and the condition of the distal urethra [14]. Isolated fistula abnormality with intact urethra may be treated with success primary closure. But, if there is a defective, this method is abandoned and the defect should be opened to converted and managed as hypospadias [11]. Surgical success rate are high (90 %) in isolated cases and fistula recurrence ratio was approximately 11 % [2].

Treatment usually consists of two techniques. For the isolated cases, the fistula can be circumscribed and then closed in simple multilayer fistula closure. As for the more complex cases, several procedures can be done, such as, straightening of the penis to close the fistula, opening the ventral glans through the distal urethra, reconstruction of the urethra, the used of Thiersch-Duplay tubularization, urethral plate technique incision, Denis Brown urethroplasty, and buccal mucosa urethroplasty. Endoscopic treatment of isolated congenital urethroperineal fistula had been reported. Primary repair was successful in 42/47 cases, four patients required two operations and one patient required three closures [15]. Small fistula under 0.5 cm can be treated with easily closed primarily after refreshing the edges and covered by skin [15]. Fistula with size more than 0.5 cm but less than 1.0 cm can be closed with turnover flap [8,17]. Larger fistulas with size more than 1.0 cm can be closed by tubularized incised plate urethroplasty using the Thiersch-Duplay technique [5,7].

The congenital anterior urethrocutaneous fistula (CAUF), especially the chordee variants as represent in our work, is an extremely rare malformation. The etiopathogenesis is unknown and the case we described is particularly interesting because a CAUF could be occurred without either previous related family history or any specific risk factors, thus, this opens an interesting and extensive discussion about the etiology of CAUF.

4. Conclusion

CAUF is extremely rare condition and correlates with the other congenital anomaly. Imaging investigation may help to rule out other associated anomalies before deciding surgical correction. There are many options for surgical correction, it depends on the fistula itself and other existing anomalies with 90 % success rate of the surgical procedure.

Data availability

The data used to support the findings of this study are included in the manuscript.

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Ethical approval

This study was approved by ethical committee of Faculty of Medicine Padjadjaran University and Hasan Sadikin Hospital, Bandung, Indonesia.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Safendra Siregar and Albert made contribution in the conception and design of the study, acquisition of data, analysis and interpretation of data, drafting the article, revising it, approve the final version to be submitted.

Guarantor

Safendra Siregar.

Declaration of competing interest

The authors declare that they have no potential conflicts of interest.

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