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Case Report

Congenital Anterior Urethrocutaneous Fistula: A Case Report and Review of Literature

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Abstract: *Introduction*: Congenital anterior urethro-cutaneous fistula (CAUF) is an extremely rare abnormality that can register as a malformative entity with hypospadias or cord, or present in an isolated form. *Purpose*: The purpose of our observation is to present our surgical management of a case of CAUF. *Observation*: We describe the clinical case of a 10-year-old patient, who presented with CAUF in the ventral mid-penile position, diagnosed at birth. He underwent urethroplasty using Mathieu's technique with simple consequences. The uretral catheter removed at 21 days post-surgery. No complications at 30 days post-operatively. *Discussion*: CAUF is an extremely rare condition. The etiology is unknown, and several pathogenetic theories have been used to explain its causes.The clinical characteristics are not well defined. An overall assessment of the disease can be carried out in search of other associated abnormalities. 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. Abdominal ultrasound and cystoscopy and/or cystography may be performed Therapeutically, surgery remains the only treatment, it includes many methods of surgical repair. Mathieu's technique is the most practiced and gives excellent results. *Conclusion*: CAUF is a rare anomaly, which must be diagnosed in order to be treated. Surgical treatment offers satisfactory results.

Keywords: Fistula, Urethrocutaneous, Congenital, Urethroplasty

1. Introduction

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]. It was first described by Gupta in 1962 and the largest series was conducted by Caldamone et al. on 14 cases 1 There

are two variants of CAUF, one associated with hypoplastic distal urethra, chordee and hooded foreskin [2]. 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 management in our hospital.

2. Clinical Case

Ten-year-old patient with an isolated congenital urethro-cutaneous fistula, diagnosed at birth by the pediatrician. There are no other malformative abnormalities clinically. In the absence of financial means, surgical management was not done in the neonatal period. The clinical examination found a ventral, mid-penile urethro-cutaneous fistula, with foul-smelling urine, purulent appearance. Circumcision had already been done. The glans is intact with a large normal meatus. ECBU was sterile. the imaging assessment, in particular the ultrasound, was performed to rule out an associated malformation. After a standard pre-operative assessment, we carried out under general anesthesia, a repair according to the technique of Mathieu.

The fistula is first circumscribed (figure 1), after urinary catheterization, then a local skin sliding flap will close the fistula step by step (figure 2, 3, 4). This first step was reinforced by skin closure (figure 5). The urinary catheter was removed on D14. Control consultations were made on D30. No recurrence, no complications at the various follow-up consultations were noted.

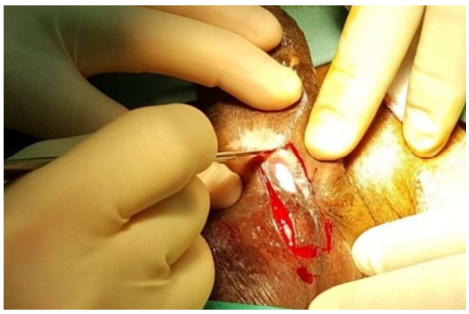

Figure 1. *Individualization of CAUF.*

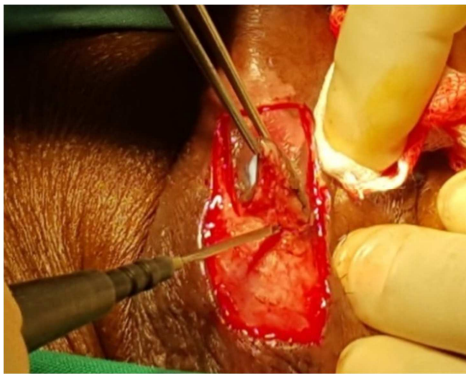

Figure 2. *Onset of local skin flap detachment.*

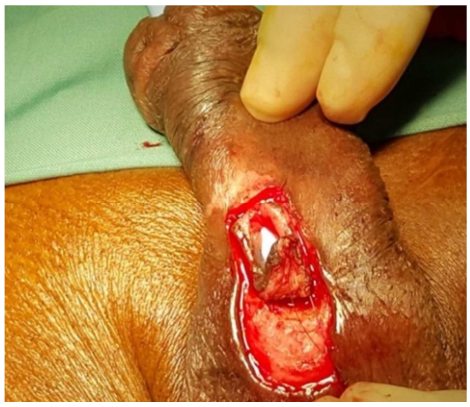

Figure 3. *Star of closure by application of the flap.*

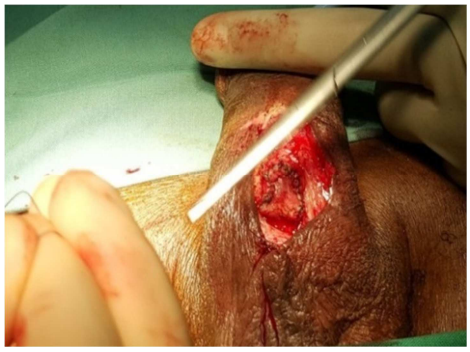

Figure 4. *Fistula closure.*

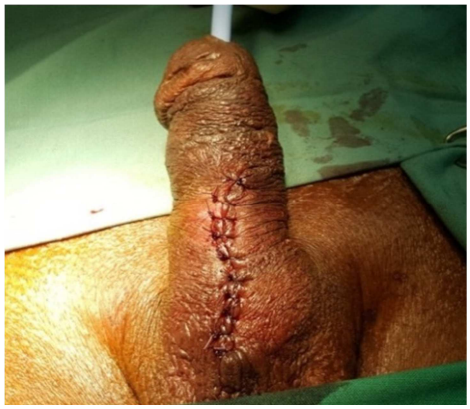

Figure 5. *Skin closure.*

3. Discussion

CAUF is an extremely rare condition. Nearly sixty cases have been described today, the largest of which is that of Caldamone et al. [5, 6]. It is usually isolated in nearly 50% of cases, or associated with genito-urinary abnormalities, or other penile abnormalities [7, 8]. The clinical characteristics are not well defined [8, 9].

The etiology is unknown, and several pathogenetic theories have been used to explain its causes. 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 [1]. Karnak regarded that congenital urethrocutaenous fistulas (excluding those associated with

anorectal malformations) as one set of anomalies [1, 10]. Subcoronal positioning is the most frequent site, followed by the middle penis, the proximal penis and the penoscrotal [7, 11].

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 an isolated congenital fistula with normal foreskin, normal glans fusion, and normal urethral development proximal and distal to the fistula.

Clinically, the diagnosis is made in the perinatal period by the pediatrician or even the mother who sees urine escape through an accessory orifice. Our patient's CAUF was diagnosed 13 months after circumcision. The management of these abnormalities requires a clear history of the child, especially those related to previous trauma, circumcision, surgery. In general, no additional examination is necessary for a positive diagnosis [3, 8].

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]. Our patient presented an isolated CAUF, so the clinical examination suggests an absence of other abnormalities.

Therapeutically, surgery remains the only treatment, it includes many methods of surgical repair [3, 7, 8]. These various procedures include preputial skin flap, Mathieu urethroplasty, Thiersch-Duplay urethroplasty, Denis Brown urethroplasty, oral mucosal graft and tubular plate urethroplasty with dartos flap, and primary closure [3, 5, 9]. The choice of technique depends on the anatomical variant of the fistula, taking into account the history [10, 11]. For financial concerns, and of adequate structure in certain regions of our country, the patient was operated on at the age of 10. However, there is no publication on the ideal time frame for these interventions. Our patient benefited from urethroplasty according to the Mathieu technique, with the same process as for the treatment of hypospadias. The post-operative follow-up was simple [8, 13, 14]. It consists in verifying the presence of a recurrence, which will have to be quickly diagnosed from the first month in order to program a recovery [14, 15]. In the event of a recurrence, the mathieu technique can be redone again. Some authors prefer flap urethroplasty [1, 15]. The urinary catheter was removed on D 21. There is no recurrence at one month.

4. Conclusion

CAUF is a rare anomaly, which can be isolated or associated with other malformations that must imperatively be sought at the time of the assessment of the disease before surgical management. The complex and variable anatomy and embryology of CAUF are not yet fully understood. The treatment is surgical, it offers satisfaction rates close to 90%.

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