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Congenital perineal urethrocutaneous fistula without rupture in a neonate

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ABSTRACT

Congenital anterior urethrocutaneous fistula (CAUF) is a rare anomaly, commonly observed in the mid-shaft or more distal position of the penis. It can be seen in the perineum in anorectal anomalies or in cases of duplicated urethra. However, isolated CAUF opening into the perineum has not been reported. Herein, we present a case of congenital anterior urethral diverticulum and possible perineal urethrocutaneous fistula of an otherwise healthy neonate.

Introduction

Urethrocutaneous fistula is defined as a tract lined with epithelium that leads from the urethra to the skin. Congenital urethrocutaneous fistula can be seen in the perineum with respect to anorectal anomalies or in cases of urethral duplication anomalies.[1] Such fistulas arising from the posterior urethra may be open in the perineum. However, isolated congenital anterior urethrocutaneous fistula (CAUF) opening in the perineum has not been reported. We present a case of CAUF found in the perineum of a neonate without complication.

Case presentation

A 6-day-old boy was referred to our department for evaluation of genital anomalies. He was born at 36 weeks' gestational age by vaginal delivery, and his birth weight was 3022 g. Both his antenatal and family histories were unremarkable. Physical examination revealed a dimple associated with cystic mass, measuring 1 cm in diameter, in the median raphe of the perineum (Fig. 1). The penile shaft looked normal without curvature, and bilateral testes were present in the scrotum. There was no abnormal finding of the anus. A cystic dilation of the urethra was suspected by ultrasonography. Voiding cystourethrography revealed a diverticulum just distal to the bulbous urethra (Fig. 2). No signs of bladder outlet obstruction nor vesicoureteral reflux were observed. The patient voided with a single urinary stream from tip of the glans.

As the dilation of the diverticulum gradually progressed, surgical correction was performed at the age of 10 months. Under general anesthesia, urethroscopy prior to surgery detected no abnormalities such as stricture or valve-like structure except for a diverticulum. A midline incision was made over the diverticulum, and dissection was extended to the normal urethra. The urethra was opened at the dimple (Fig. 3) where no subcutaneous tissue existed. After excision of the fistulous dimple and diverticulum, urethroplasty was performed over the 8F transurethral catheter in layers. Histopathological examination showed no urethral spongiosum in the ventral wall of the diverticulum. The patient's postoperative course was uneventful. He was doing well with normal voiding at 5 years follow-up.

Discussion

Although urethrocutaneous fistula in the male most commonly presents as sequelae of hypospadias repair, urethrocutaneous fistula of congenital origin is very rare. To date, 65 cases of CAUF have been reported in the literature.[2–4] While CAUF is usually seen in the mid-shaft or more distal position of the penis, there is no report of isolated CAUF opening into the perineum. In a systematic review by Lin et al.,[3] subcoronal fistula was detected in 29 of 63 patients with CAUF, mid-penile shaft in 24, proximal penile to subcoronal in 4, and penoscrotal in 1. The location of fistulous opening in our patient is the most proximal in isolated cases of CAUF.

CAUF has multifactorial etiology. In the largest series of CAUF by Caldamone et al.,[4] isolated CAUFs were divided into two types based on the presence of normal prepuce or absence of chordee: ruptured urethral diverticulum and variant of hypospadias. The former type may be caused due to the blowout phenomenon of a urethral diverticulum.[5] Although no urinary leakage was observed in our case, there was a definitive point, with no subcutaneous tissue in the urethral diverticulum, suspecting a fistulous formation.

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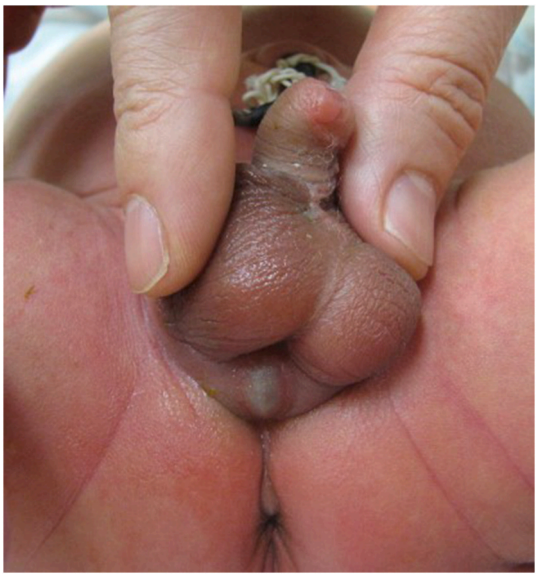

Fig. 1. Appearance of external genitalia at presentation.

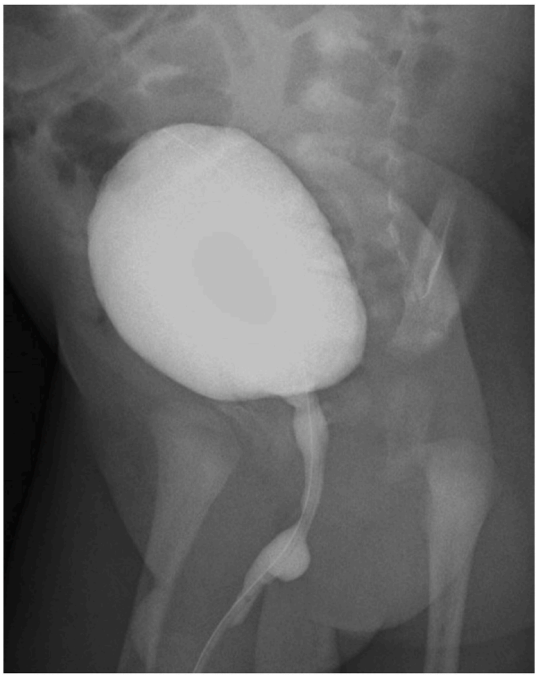

Fig. 2. Voiding cystourethroscopy showed a diverticulum just distal to the bulbous urethra.

Since most of the CAUFs are symptomatic, they should be treated surgically. Surgical approach depends on the complications and the type of CAUF. The success rates are high in isolated cases and fistula recurrence ratio was approximately 11%.[3]

Conclusion

We report a case of congenital anterior urethral diverticulum and possible perineal urethrocutaneous fistula in a neonate. To the best of our knowledge, the location of fistulous opening in this boy is the most proximal in isolated cases of CAUF.

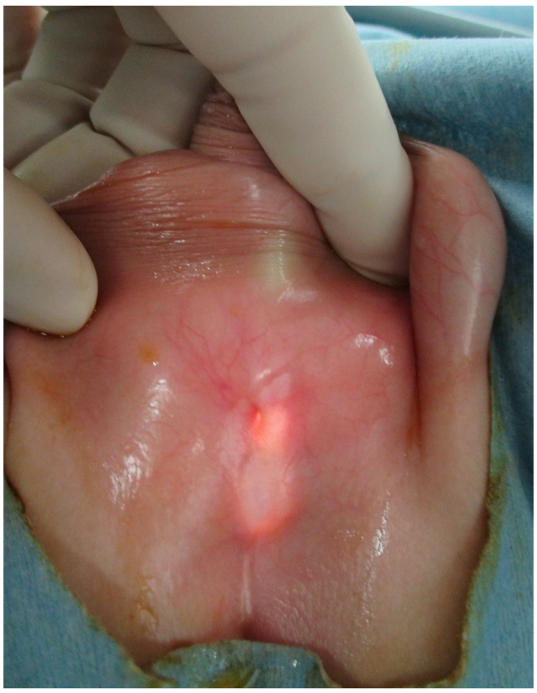

Fig. 3. Congenital urethrocutaneous fistula associated with urethral diverticulum lightened with urethroscopy.

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Credit author statement

Fumi Matsumoto: Investigation, Data curation, Visualization, Writing- Original draft preparation. Satoko Matsuyama: Writing- Reviewing and Editing. Futoshi Matsui: Writing- Reviewing and Editing. Koji Yazawa: Writing- Reviewing and Editing.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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