Proximal isolated congenital anterior urethrocutaneous fistula resulting in a urethral “skip” lesion

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ABSTRACT

Urethrocutaneous fistula is an unfortunate but well recognized complication of hypospadias repair surgery and traumatic injury to the penis. Congenital anterior urethrocutaneous fistula of the male urethra is an exceedingly rare phenomenon, with approximately 50 cases being reported in the literature. We report a case of proximal isolated congenital anterior urethrocutaneous fistula at the penoscrotal junction.

1. Introduction

Congenital anterior urethrocutaneous fistula is a rare entity, with approximately 50 cases described in the literature. More commonly these are associated with chordee, dorsal hood, distal hypospadias with or without a distal urethral or spongiosal defect, and anorectal anomalies. Less commonly, they occur in an isolated fashion with no chordee or hypospadias and an intact distal urethra and spongiosum. To our knowledge, only 3 have been reported at the penoscrotal location. We report a 15-month-old male with proximal isolated congenital anterior urethrocutaneous fistula.

2. Case presentation

A 15-month-old boy presented for evaluation after referral for a penile abnormality present since birth. On physical exam, patient was noted to be Tanner stage I with bilaterally descended testes. He had a normally formed scrotum and penis. He was uncircumcised with complete foreskin with apparent orthotopic meatus. At the penoscrotal junction, there was a large opening appearing to be urethral. The parents could not describe whether urine came from the penoscrotal junction or the end of the penis. A catheter was placed through the penoscrotal opening into the bladder to obtain a voiding cystourethrogram. The posterior urethra appeared normal and nondilated with no additional accessory urethral tracts visualized. No definite excretion during voiding was seen along the expected normal location of the urethral meatus (Fig. 1).

Indications for surgical intervention were discussed with the family.

Fig. 1. Voiding cystourethrogram demonstrating urine extravasation at the penoscrotal junction. No contrast excretion is seen within the expected course of the penile urethra.
who wished to proceed. The patient underwent diagnostic cystoscopy through the orthotopic meatus and fistula site which revealed a normal anterior and posterior urethra. A 6Fr kendall stent was passed through the orthotopic meatus and exited through the defect at the penoscrotal junction confirming the diagnosis of proximal isolated congenital urethrocutaneous fistula (Fig. 2). An incision was made around the fistula and the tract was excised. The urethra and spongiosal tissue appeared healthy and were closed in separate layers. The skin incision was extended slightly into the scrotum in order to harvest a pedicled dartos flap for another layer of closure. A catheter was left in place for 11 days post-operatively. He had 5mm of superficial skin separation initially but at most recent follow up his wound is well healed with no evidence of urine extravasation by exam nor by parent report.

3. Discussion

Urethrocutaneous fistula is a well-known entity of pediatric penile urethral surgery. While the vast majority of fistulas are post-procedural, a small number of congenital anterior urethrocutaneous fistulas have been described. Most of these reported rare cases occur in the mid to distal penile shaft and are more commonly associated with hypospadiac manifestations. The urethral groove is thought to fuse in a proximal to distal fashion to form the tubular urethra. While hypospadias can be explained in part by disruption of the canalization or fusion process of the urethral plate, it is currently unclear embryologically how a “skip” lesion in the anterior urethra might occur in the setting of healthy and normal urethra and spongiosum. Tenets of penile urethral fistula repair such as multi-layer closure, non-overlapping suture lines, and catheter drainage hold true regardless of etiology.

4. Conclusion

Congenital anterior urethrocutaneous fistula is a rare anomaly that can present anywhere along the anterior male urethra. Fortunately, this condition can be easily and effectively managed with proper diagnosis and surgical intervention.

Declaration of competing interest

The authors declare that there are no conflicts of interest regarding the publication of this article.

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