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Congenital scaphoid megalourethra

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ABSTRACT

The congenital megalo-urethra is a rare malformation of the urogenital tract characterized by the dilation of the anterior urethra due to an absence of erectile tissue within the penis. The objective of present paper is to report the management of 02 cases and performed a literature review. 5-day-old and 5-months boys were admitted in our institution for a scaphoid penile dilation. The physical examination and the urethroscopy confirmed the diagnosis. No associated malformations were found. The patients underwent successful urethroplasty and no adverse outcomes were encountered at 1 year and 3 years post-operative respectively. The congenital scaphoid megalo-urethra is curable surgically; however, the pronostic is dependent of the associated malformations.

1. Introduction

The congenital megalo-urethra is a rare malformation due to a total or partial absence of erectile body in the penis leading to a dilation of the anterior urethra. Initially described by Nesbitt et al., in 1955, the literature reports less than a hundred published cases. To date, the embryological etiology is not clearly understood. Herein, we reported our two first cases and discuss the literature review.

2. Case reports

2.1. Case 1

A 5-day-old boy was referred to the consultation for a penile malformation. It was born from a well-followed, full-term pregnancy with a birth weight of 3.5 kg. He had a weak urinary stream with drip-like urine. During micturition, the parent noted an increasing of swelling of distal penis.

On clinical examination the distal penis had a sacciform enlargement with a urethral meatus in apical position (Fig. 2). The both testes were palpable in the scrotum. No other clinical abnormalities were associated.

The blood cell count, serum urea and serum creatinine were normal. The ultrasound examination of the urinary tract didn't reveal any associated malformations. The urine sample was normal. Retrograde cystourethrogram (RCU) was not performed.

The Urethrocystoscopy was performed to establish the diagnosis of scaphoid megalourethra and to verify the absence of associated malformations. An urethroplasty was performed at 29 days, at the same time as urethrocystoscopy (Fig. 3).

The surgical technique was as follows:

- Under general anesthesia, rigorous asepsis was mad
- Urethra catheter (Foley Probe) CH 10 was set up and the foreskin was retracted (Fig. 1a)
- Traction with vicryl 4/0 at the top of the glans was realized
- An encircling incision was made one-half centimeter below the balano-preputial groove (Fig. 1a)
- The penis was degloved completely, but the dilated urethra, covered only by a fine subcutaneous tissue,
- Vertical and medial incision of the urethra at the ventral surface (from one centimeter below the urethral meatus to 0.5 cm below the swollen area) (Fig. 1b)
- The redundant flaps of lateral tissue (Fig. 1c) were excised leaving a strip of normal urethra
- Continuous sutures of PDS 6/0 over urethra catheter CH10 is

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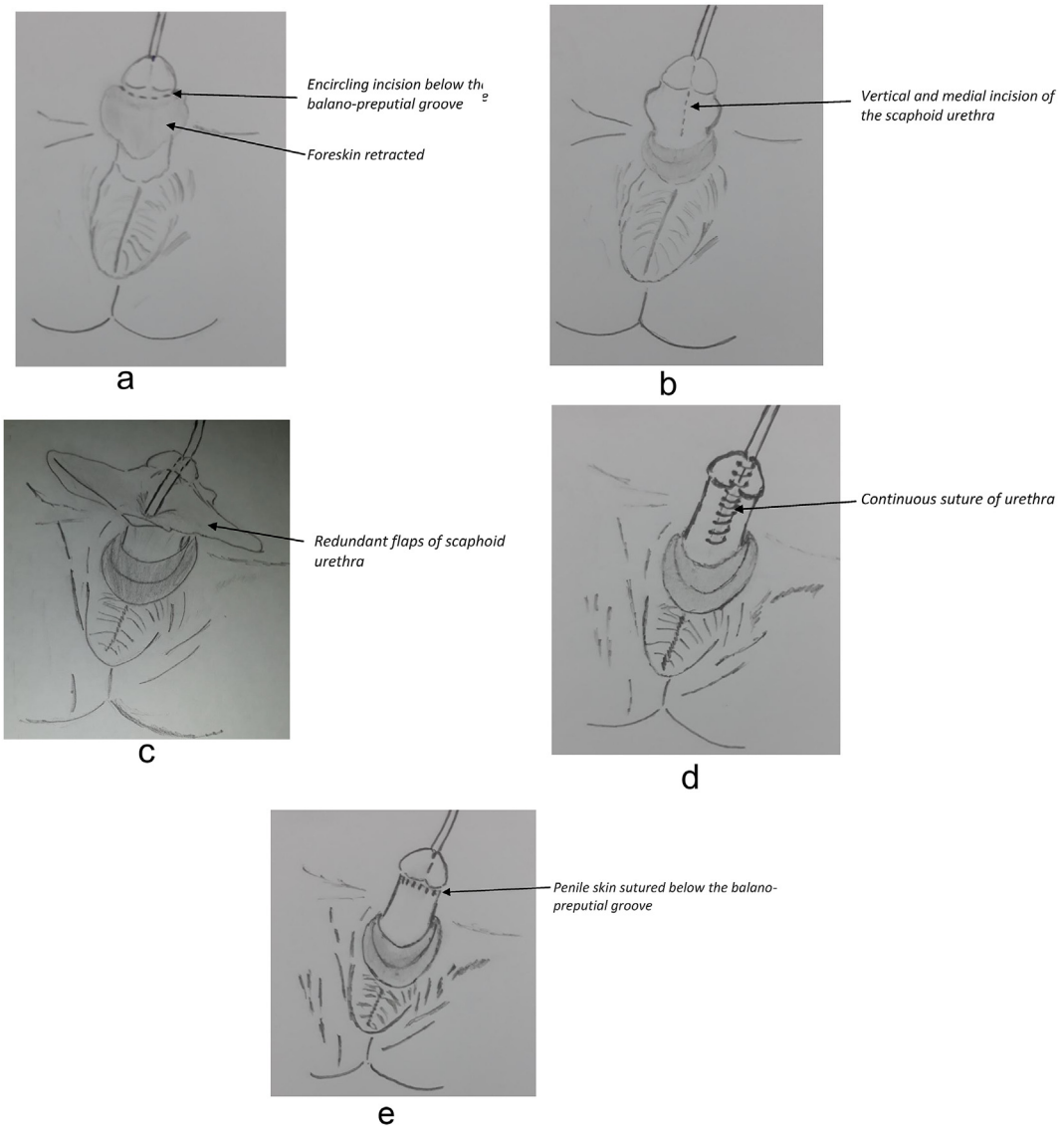

Fig. 1. a: Retraction of foreskin and encircling incision. b: Incision of the urethra. c: After incision of urethra. d: urethroplasty after excision of redundant tissue. e: suture of the penile skin after excision of the excessive part.

achieved and then reinforced by another continuous suture (Fig. 1d),
- The penile skin was rolled up on the penis shaft and after simple excision of the excessive part (with the foreskin), the free margin was sutured (Fig. 1e),
- Fixing the urethra catheter with the traction wire placed at the top of the glans

The urethra catheter remained in place for 10 days. At ablation, urination was normal with a good urinary stream.

A stenosis of the urethral meatus occurs one-month post-surgery and was treated by instrumental dilation. The follow-up until 3 years later was uneventful.

2.2. Case 2

A 5-month-old boy was admitted for a dysuria and swelling of the penis. The prenatal pregnancy follow-up was uneventful, and the obstetric ultrasound failed to diagnose any malformations. The baby was delivered at full term without complications. The mother reported a swelling that increases during urination and is associated with a weak urinary stream. On physical examination, the penis was enlarged, especially at the distal part (Fig. 4). The urethral meatus was at an apical position. The both testes were normally located.

The blood cell count, serum urea and serum creatinine and ECBU were normal. The ultrasound of the urinary shaft did not objectify associated malformations. Although the RCU was not been performed, the diagnosis was made on the clinical examination.

An urethroplasty was performed at 15 months of life (Figs. 5 and 6). The technique used was identical to the first case.

Regular post-operative visits up to 1 year showed normal voiding pattern without urethral stenosis. The bladder and upper urinary tract were normal at the ultrasound. No complications occurred later.

3. Discussion

Megalourethra, malformation of the anterior urethra, is the consequence of aplasia of erectile bodies. This malformation, of unknown cause, causes a stasis of urine in the dilated segment responsible for a functional obstruction. No cause of mechanical obstruction was detected [3].

The exact embryological cause isn't exactly elucidated. It is due to a