Urology Case Reports 14 (2017) 3–4

Contents lists available at ScienceDirect

Urology Case Reports

journal homepage: www.elsevier.com/locate/eucr

Pediatrics

Congenital Scaphoid Megalourethra: A Case Report

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ARTICLE INFO

Article history:
Received 17 April 2017
Received in revised form
1 May 2017
Accepted 5 May 2017

Keywords:
Congenital megalourethra
Scaphoid type

ABSTRACT

A congenital megalourethra is an enlargement of the pendulous urethra without evidence of distal obstruction. A 1-month-old boy presented to us with complaint of weak stream, ballooning of the penis before and during voiding and post voiding dribbling, since birth. Physical examination and cystourethroscope confirmed the diagnosis of congenital scaphoid megalourethra. He underwent reduction urethroplasty. During postoperative follow up, he had normal looking penis with good urinary stream.

Introduction

Congenital megalourethra is a rare congenital anomaly of the male anterior urethra and erectile tissue of penis.[1] The scaphoid type is caused by poor development of corpus spongiosum. In the more severe fusiform type is a deficiency of the corpus cavernosum, as well as the corpus spongiosum.[2] Megalourethra is often associated with other congenital anomalies of which genitourinary are most common.[3] We report a case of congenital scaphoid megalourethra without any other malformation.

A case report

A 1-month-old boy presented with the complaints of weak stream, ballooning of the penis before and during voiding and post voiding dribbling since birth. On examination, there was a scaphoid swelling on the ventral aspect of shaft of penis that ballooned markedly during voiding (Fig. 1). On compression of the swelling, urine dribbling from the normally-placed meatus was observed. Both testes were palpable in scrotum. Ultrasound showed normal kidneys and bladder with no postvoid residue. Intraoperative photographs are shown in Fig. 2. Cystourethroscopy showed wide dilatation of pendulous urethra without evidence of distal urethral obstruction. Proximal urethra and bladder were normal. Corpus spongiosum was poorly developed. Following cystourethroscopy, a reduction urethroplasty was done through circumcoronal incision. After degloving the scaphoid dilatation was noted. The redundant urethra with deficient corpus spongiosum was excised and the urethra was reconstructed over 8 Fr infant feeding tube using 7-0 polydioxanone suture. A protective dartos fascia flap is placed over the entire suture line as a reinforcing and waterproofing layer. The urethral catheter was removed after 10 days and the patient voided normally without any penile swelling (Fig. 3).

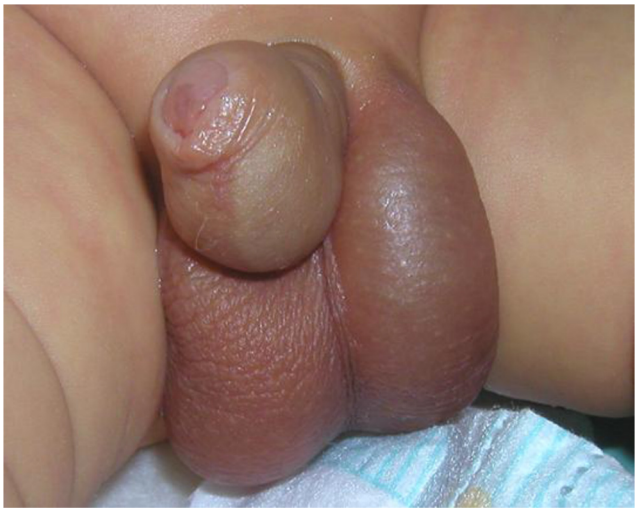

Figure 1. Congenital scaphoid megalourethra presenting as a swelling on the ventral side of the penis after voiding in a 1-month old boy.

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http://dx.doi.org/10.1016/j.eucr.2017.05.002

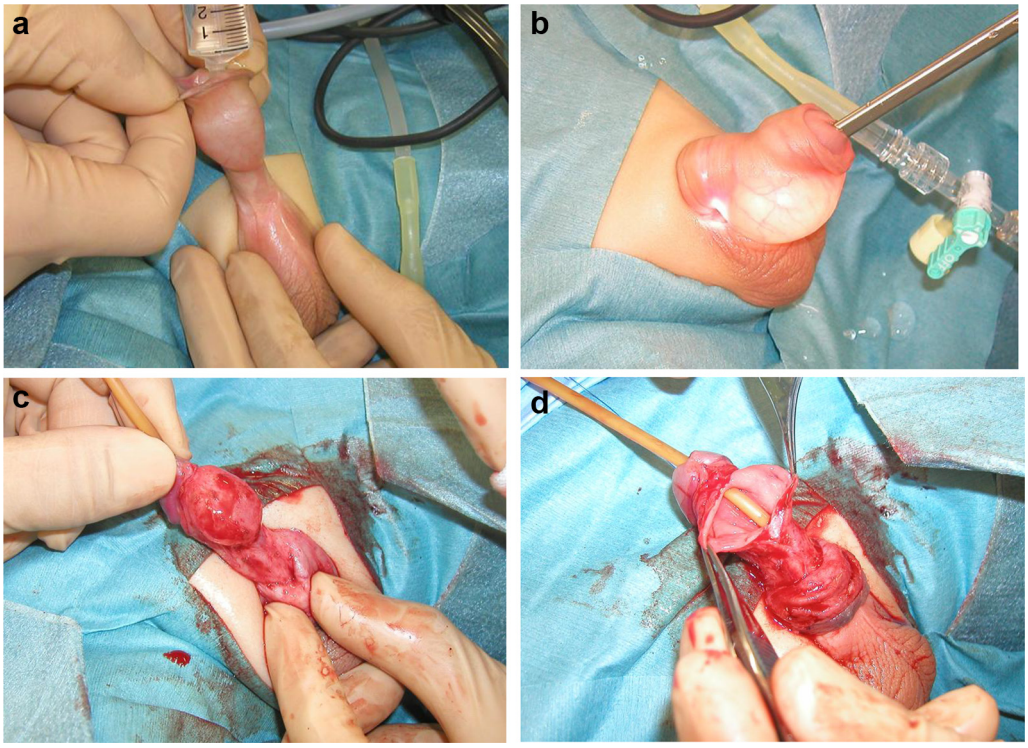

Figure 2. Megalourethra becomes remarkable after saline instillation from urethral meatus (a). The light of the cystourethroscope is observed through the thin urethral wall (b). Appearance of megalourethra after degloving of penile skin (c). Longitudinally opened urethral cavity (d).

Discussion

Congenital megalourethra is defined as diffuse dilatation of the anterior urethra due to lack of corpus spongiosum with or without corpora cavernosa. The etiology of megalourethra is not clearly understood. It is presumed that failure of the mesenchymal tissues of the phallus to differentiate and provide support to the urethral epithelium results in megalourethra.[4] Another assumption is that delayed or deficient canalization of the glanular urethra may be associated with maldevelopment of the corpus spongiosum and corpora cavernosa and the severity and the duration of the urethral anomaly determines the type of megalourethra.[5]

Megalourethra is known to be associated with other abnormalities of the genitourinary, gastrointestinal, or other systems, such as VACTERL (vertebral, anal atresia, cardiac, tracheaesophageal fistula, renal, and limb deformities) association. Abnormalities of genitourinary system include duplication of the urethra, renal dysplasia, polycystic kidney, hydronephrosis, hydroureter, vesicoureteral reflux, prune-belly syndrome, megacystis, hypospadias, posterior urethral valves and undescended testes.[3]

The treatment of megalourethra may be one stage or staged urethroplasty depending on the type of megalourethra. For isolated scaphoid type, a longitudinal reduction urethroplasty provides excellent results as seen in our case. However, for the fusiform type which has associated anomalies more frequent and more severe, the placement of a penile prosthesis in adulthood could be considered.

Figure 3. Penile appearance after reduction urethroplasty.

Conclusion

In isolated scaphoid type of megalourethra, reduction urethroplasty provides excellent functional and cosmetic results.

Conflict of interest

None.

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