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CASE REPORT

Successful surgical correction of an extreme form of ectopic penis

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Abstract Reported here is the successful multi-staged repair of a rare case of complete penoscrotal transposition with ectopic penis. The need to evaluate for associated anomalies is discussed.

Introduction

Complete penoscrotal transposition is a rare anomaly of the external genitalia whereby the scrotum is located cephal to the penis. This may be part of a wide spectrum of abnormalities; associated genitourinary tract and other organ system malformations are frequently present. We report the multi-staged repair of a case of complete penoscrotal transposition.

Case report

A neonate delivered at 35 weeks of gestation was noted to have a severe form of complete penoscrotal transposition with ectopic penis. After a vesicostomy on day 2 at the referral hospital, the infant was transferred to our department.

Physical examination showed a slightly ventrally located scrotum containing both testicles. A 2-cm hypoplastic phallus with hypospadias and chordee was ectopically placed in the perineum just cephal to the normally placed anus. Postnatal abdominal ultrasound scan revealed left crossed renal ectopy. Cystogram did not show vesicoureteral reflux. There was no associated cardiac anomaly and the karyotype was 46 XY. The patient underwent multi-staged surgeries for correction of his ectopic penis.

Stage 1. *Primary mobilization of the phallus at 8 months of age*: Following penile denudation, only one corpus cavernosum was noted; a chordectomy was performed and the penis moved caudal to the scrotum (Fig. 1a—d).

Stage 2. *Anteposition of the penis at the age of 1 year*: The penis was further mobilized and pulled through

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a button-hole to be placed cephal to the scrotum (Fig. 1e—h).

Stage 3. *Urethral replacement with buccal mucosa at 18 months of age*.

Stage 4. *Partial urethroplasty at the age of 2 years*.

Stage 5. *Further Thirsch—Duplay urethroplasty at the age of 4 years* (Fig. 1i—l).

Stage 6. *Closure of vesicostomy at the age of 6 years*.

A postoperative urethral stricture was managed by dilatation. At 7 years old, the patient has complete urinary continence and normal erection. Uroflowmetry showed a satisfactory single urine stream.

Discussion

There are two forms of penoscrotal transposition. In the incomplete form, which is more common, the penis lies mid-scrotum. In the complete form, the scrotum is located cephal to the penis, which emerges from the perineum [1].

The etiology of penoscrotal transposition is unknown. Several factors, such as a defect of the androgen receptor and chromosomal abnormalities, can lead to penoscrotal transposition. Patients with penoscrotal transposition often present with associated genital anomalies, including hypospadias and chordee. The rate of renal defects in these patients is as high as 100% of reported cases. Pinke et al. [2] in a series of 53 patients with penoscrotal transposition reported an incidence of 32% of other organ system anomalies, including cardiac anomalies, imperforate anus, and anomalies of the musculoskeletal and central nervous system. Renal ultrasound scan reveals associated renal defects. Most non-renal anomalies are clinically apparent. An increased awareness combined with thorough physical examination will identify these associated anomalies [2,3].

Correction of isolated penoscrotal transposition is usually performed as a single-stage procedure [4,5]. However, the severe form of complete penoscrotal transposition requires a multi-stage procedure. In our case, the ectopic penis with hypospadias and chordee was located in the perineum just cephal to the normally located anus. It required multi-stage procedures to step-by-step mobilize and advance the penis to its normal anatomical position. Postoperative urethral stricture was managed initially by

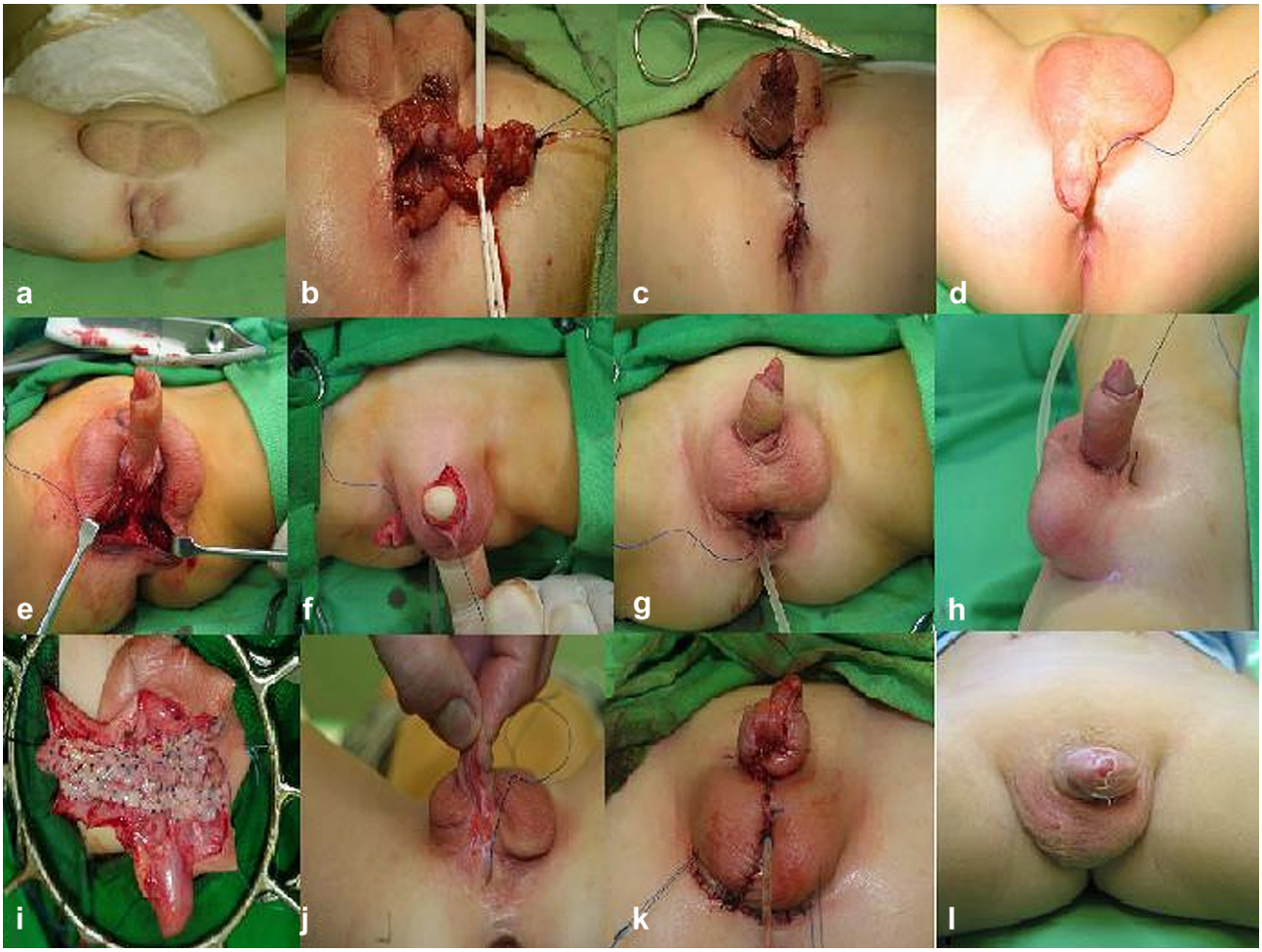

Figure 1 (a—d) Primary mobilization of the penis. (e—h) Anteposition of the penis. (i—l) Buccal mucosal free graft and urethroplasty.

dilatation under general anesthesia followed by home self dilatation.

Successful surgical correction of ectopic penis requires a multi-stage procedure. We were unable to find a report of surgical correction of a similarly severe form of ectopic penis in the literature. Following surgical repair, our patient had a straight penis, normal erection, and urinary continence with a satisfactory uroflow rate.

Conflict of interest/funding

None.

References

[1] Perovic PS. Penoscrotal transposition. 3rd ed. Adv Urologic Surg 2005:199—211
[2] Pinke LA, Rathbun SR, Husmann DA, Krame SA. Penoscrotal transposition: review of 53 patients. J Urol 2001;166:1865—8.
[3] Parida SK, Hall BD, Barton L, Fujimoto A. Penoscrotal transposition and associated anomalies: report of five new cases and review of the literature. Am J Med Genet 1995;59:68—75.
[4] Kolligian ME, Franco I, Reda EF. Correction of penoscrotal transposition: a novel approach. J Urol Part 2 2000;164:994—6.
[5] Perović S, Vukadinović V. Penoscrotal transposition with hypospadias: 1-stage repair. J Urol 1992;148:1510—3.