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CASE REPORT

A case of penile duplication with neonatal teratoma and bladder neck incompetence

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ABSTRACT

An 8-year-old boy presented with a duplicated penis and urinary incontinence. He had a history of a perineal teratoma which was removed during his first week of life. Examination revealed a large prepuce, 90-degree counter clockwise rotation of the penis, an orthotopic megalomeatus and an additional smaller glans dorsally. Cystourethroscopy and artificial erection showed a wide-open bladder neck and deviation of the penis(es) to the right. There were two cavernosae in the orthotopic penis and one in the duplicated rudimentary penis. The patient was subjected to Young-Dees bladder neck reconstruction and two years later, excision of the rudimentary penis. A satisfactory cosmetic result was achieved, the patient is voiding normally, and urinary incontinence improved. Penile duplication is a rare anomaly, which presents differently in each patient. Therefore, treatment should be individualized, and the goal of surgery being to achieve as a near normal cosmetic and functional result as possible.

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Introduction

Penile duplication or diphallia is a rare anomaly, with about 100 cases reported in the literature. The incidence is estimated to be 1 per 5–6 million live births, and often no cases are identical [1–4]. The first case was reported in 1609 by Wecker in Bologna, Italy [5].

The extent of duplication and the number of associated anomalies vary greatly, ranging from glans duplication arising from a common shaft to complete duplication of the penis associated with urinary tract and extra-genitourinary anomalies [6]. The duplication can be orthotopic or ectopic, sagittal or coronal, symmetric or asymmetric, and of varying sizes and shapes [6,7].

Here, we report a case of penile duplication accompanied by a neonatal large perineal teratoma and bladder neck insufficiency.

Case report

An 8-year-old boy was referred to our Department with urinary incontinence for evaluation of a genital anomaly. Antenatal, gastroschisis was suspected at the routine antenatal ultrasound scans, but was subsequently ruled out by fetal MRI. A large ventral hernia of the lower abdomen was however noted. At birth physical examination revealed a bifid scrotum, a redundant prepuce and a large $20 \times 30 \times 40$ mm skin covered tumour in the region between the scrotum and penis. An MRI scan of the lower abdomen showed bilateral descended testes and a heterogeneous mass adjacent to the right testis, without connection to the peritoneal cavity, but with an extension to the perineum.

Histological examination of the removed mass revealed a pleomorphic tumour with elements from all primitive embryological germ cell layers – a prepubertal teratoma with components of immature smooth- and striated muscle cells, neural tissue, glandular structures, colonic epithelium, and mature neuroectodermal components. There was no sign of malignancy or gonadal tissue in the removed mass.

Physical examination revealed a redundant prepuce, 90-degree counter clockwise rotation of the penis, an orthotopic megalomeatus and an additional smaller glans dorsally on the right side of the orthotopic penis (Figure 1). Urine was passed exclusively from the orthotopic penis' urethra. A cystogram with antegrade filling of the bladder *via* a suprapubic catheter, demonstrated a small bladder with a single urethra during voiding (Figure 2). Urodynamic studies displayed a bladder capacity between 160 and 180 ml, with leakage at a capacity of 100 ml, normal compliance, prolonged urinary flow, and low peak flow rate. Additionally, an MRI scan of the spine was normal and showed no signs of spinal dysraphism.

Examination under anaesthesia including artificial erection showed that the penis(es) deviated to the right, and with two carvernosal bodies in the orthotopic penis and one in the duplicated rudimentary penis, the latter without any

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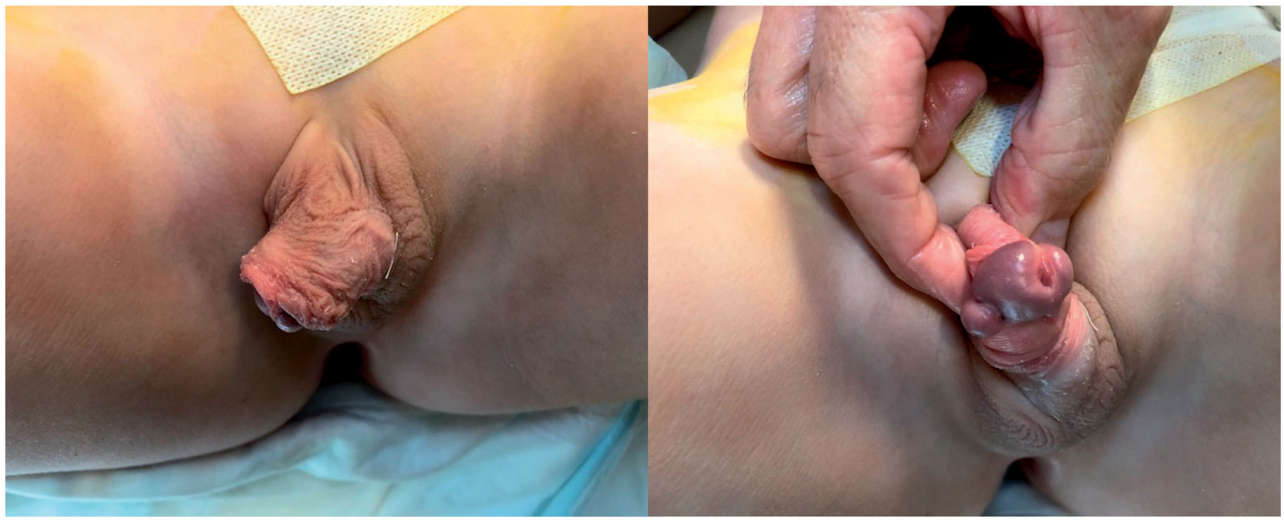

Figure 1. Physical examination at referral. The boy was 8 years old and urinated from the ventrally orthotopic penis only.

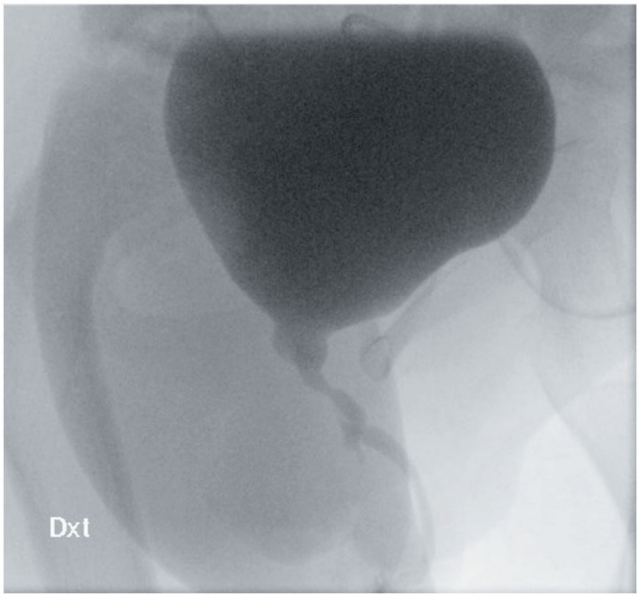

Figure 2. Voiding Cystourethrogram demonstrating a dominant normal (orthotopic) urethra.

significant tumescence. Cystourethroscopy of the orthotopic penis revealed a short posterior urethra, with an almost intravesical verumontanum and a wide-open bladder neck.

Subsequently, at the age of nine, open Young-Dees bladder neck reconstruction was performed in order to achieve urinary continence.

At the age of ten, surgical correction of the penis was performed in the following manner: penile degloving including resection of chordée, dissection of the hypoplastic rudimentary penis off the main penis proximally to the ischiopubic region, where after the hypoplastic penis was excised, correction of penoscrotal fusion and circumcision (Figure 3). Examination of the excised penis revealed a hypoplastic urethra with a calcified formation (Figure 3(c)). The postoperative period was uneventful.

Histological analysis of the rudimentary penis showed one well-developed corpus cavernosum and one less developed corpus spongiosum with lack of urethral tissue (Figure 4). At 6 months follow up (Figure 5), the patient was voiding normally without any residual urine. He still had episodes of urinary incontinence, which had slightly improved.

Discussion

Penile duplication is rare, and the embryologic cause is not entirely understood. However, most researchers agree that a defect of the genital tubercle occurs between 23th and 25th day of gestation. The theory holds that the caudal fetal mass of mesoderm is deeply disturbed by trauma, drugs or infection [3,8–10]. Penile duplication is therefore, often accompanied by other anomalies within the urogenital tract (hypospadias, vesicoureteral reflux, hydronephrosis, ectopic kidneys, duplex kidneys, bladder and cloacal extrophy). Additionally, extra-genitourinary abnormalities may also coexist (colon duplication, vertebral deformities, imperforate anus, ventricular septal defect, polydactyly) [3,7,8].

Congenital teratomas are usually extragonadal [11], in our case the tumour infiltrated the scrotal sac but no the nearby testis. A review of the neonatal MRI confirmed that the teratoma had its base in the perineum and was not classified as sacrococcygeal, which is the most common (35–60%) neonatal teratoma [11].

Many classifications have been proposed to categorize the various forms of diphallia. Aleem and colleagues [5] divided the malformation into two groups: True diphallia and bifid phallus. These two groups are further divided into partial or complete duplication. In case of true, complete diphallia, each penis is identical, and each has two corpora cavernosae and a corpus spongiosum. When the duplicated penis is smaller or rudimentary, it is classified as true partial diphallia. In cases where only one corpus cavernosum is present in each penis, the term bifid phallus applies. Complete and partial bifid phallus are recognised when the degree of

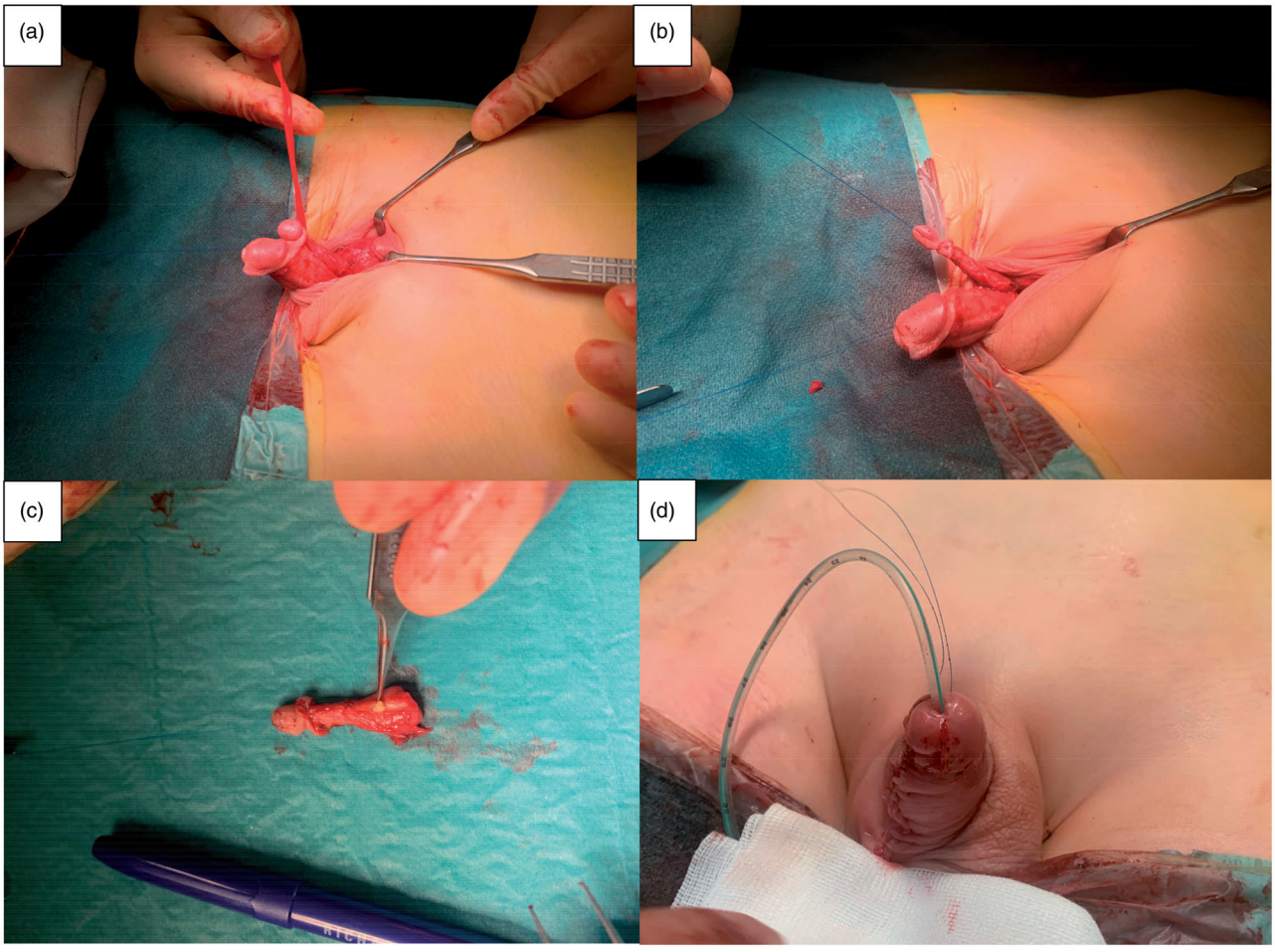

Figure 3. Peroperative pictures. (a) After degloving the two penises. (b) Dissection of the rudimentary penis from the orthotopic penis proximally to the ishiopubic region. (c) Amputation of the rudimentary penis. (d) Correction of penoscrotal fusion and circumcision.

separation is complete to the base of the shaft or confined to the glans, respectively. Schneider [6] added a third category: diphallia of the glans alone and a fourth category was added by Vilanova and Raventos [12], named pseudodiphallia, in which there is a rudimentary atrophic penis that is anatomically independent of the normal penis. The overall rarity of the malformation renders current classification systems suboptimal with increasing subcategories added on in tandem with new reports.

In the current case, pseudodiphallia was present, with a rudimentary atrophic penis with only one well-developed corpus cavernosum and one less developed corpus spongiosum and lack of urethral tissue and hence, no connection to the bladder. The patient voided through the orthotopic penis which had two corpora cavernosae and a corpus spongiosum with normal erectile function. Cystourethroscopy of the orthotopic penis revealed a normal distal urethra, but a short posterior urethra and a wide-open bladder neck. The latter has been described before in patients with penile duplication [9]. Some of these cases, moreover have other characteristics of exstrophy-epispadias complex [8], including variants of covered exstrophy, pseudoextrophy and cloacal exstrophy with concomitant penile duplication [13–14]. In the present case, no other characteristics of exstrophy-epispadias complex were noticed.

The surgical correction of penile duplication is a challenging subject, not only because of its medical peculiarities but also because of ethical and aesthetic considerations. The management is individualized and based on the assessment of the corporal and urethral anatomy. Thorough evaluation is mandatory with the goal of surgery being to achieve as a near normal cosmetic and functional result as possible. This may be achieved in a one-stage procedure, as in our case, or multiple-stages performed over time to correct associated anomalies [4].

Conclusion

Penile duplication is a rare congenital anomaly, usually associated with other malformations. Because of its rarity and variation in presentation, no standardized management can be recommended. Therefore, treatment should always be individualized with the main concern being to manage the more complicated malformations first. In our case, diphallia was diagnosed in association with a perineal teratoma which infiltrated the scrotal sac in addition to bladder neck incompetence. To the best of our knowledge this combination has not be described before in the literature [12,15,16].

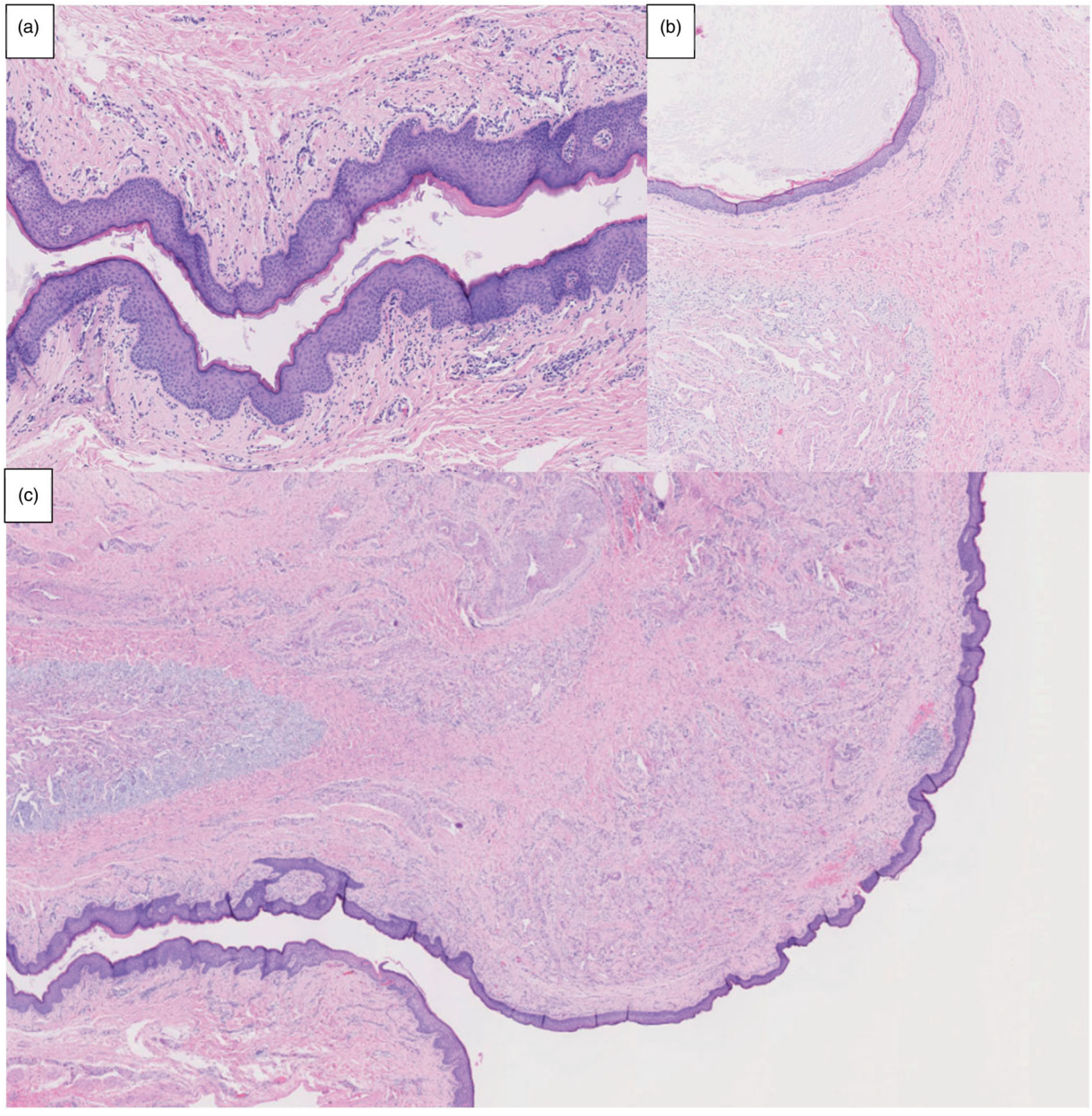

Figure 4. Histopathological images. (a) The urethra was lined by a keratinized stratified squamous epithelium, and no urothelium was present. Around the urethra there is an underdeveloped corpus spongiosum consisting of small blood vessels (longitudinal section). (b) There was only one corpus cavernosum present (cross section). (c) The underdeveloped corpus spongiosum was present around the urethra throughout the longitudinal section of the penis and at the glans penis. The tip of the single corpus cavernosum is visible to the left.

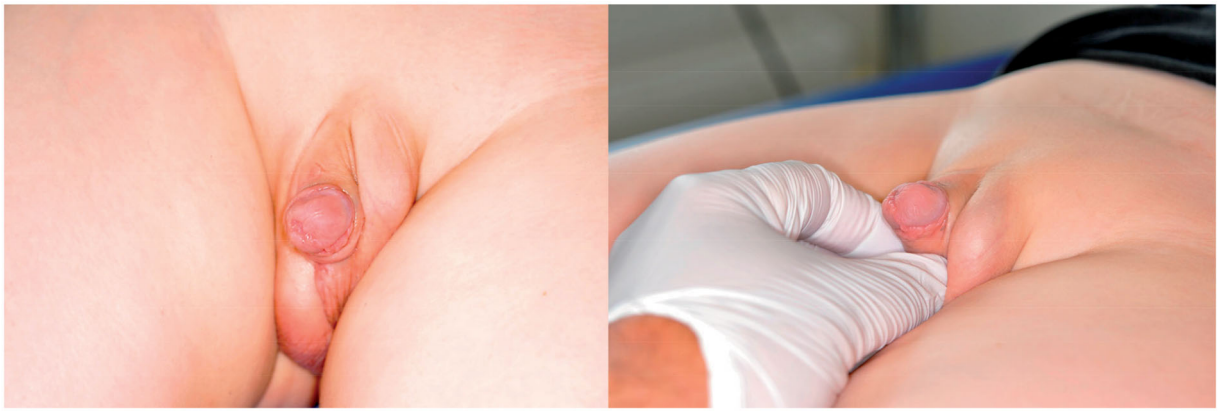

Figure 5. Cosmetic result 6 months after surgery.

Disclosure statement

The authors report no conflicts of interest. The authors alone are responsible for the content and writing the paper.

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