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Diphallus: A Rare Urological Anomaly - What to Do Next? Case Report and Literature Review

Difallus: Nadir Bir Ürolojik Anomali - Sırada Ne Yapılmalı? Olgu Sunumu ve Literatür İncelemesi

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Abstract

Diphallus is a very rare congenital anomaly usually accompanied by various congenital anomalies and can be classified according to the anatomical structure in which the anomaly develops. Generally, in cases with diphallus, the surgical approach is preferred, in that, the hypoplastic structure has been excised for esthetic and functional purposes and penile reconstruction is performed. In addition, urethroplasty has been also performed in the presence of any accompanying urethral abnormality. In the current case, we report a two-year-old boy who was diagnosed as having glandular diphallus.

Keywords: diphallus, diphallia, penile duplication, genitourinary anomaly, glans penis, reconstructive surgery

Öz

Difallus oldukça nadir rastlanılan konjenital bir anomalidir. Genellikle çeşitli konjenital anomalilerin eşlik ettiği difallus, anomalinin geliştiği anatomik yapıya göre sınıflandırılabilmektedir. Literatür incelendiğinde, genellikle cerrahi yaklaşım tercih edilen difallusta, estetik ve fonksiyonel amaçlarla hipoplastik yapının eksize edilerek penil rekonstrüksiyon işlemi uygulandığı, eşlik eden üretral anomali varlığında üretroplasti uygulandığı görüldü. Bu vaka sunumunda genitoüriner sistem muayenesi sonrası glans penisle sınırlı difallus tanısı koyulan 2 yaşındaki erkek çocuk sunulmaktadır.

Anahtar kelimeler: difallus, difalya, penis dublikasyonu, genitoüriner anomali, glans penis, rekonstrüktif cerrahi

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Introduction

Diphallus, which is also known as diphallia, is a very rare congenital anomaly encountered nearly one in 5.5 million population. Since the first case was presented by Wecker in 1609, only 120 cases have been reported in the literature so far [1]. Vast majority of cases with diphallus were accompanied by various congenital anomalies mainly genitourinary and anorectal malformations [1,2]. Diphallus can be classified as glandular diphallus, bifid diphallus, and complete diphallia according to the anatomical structure involved [2]. During the 12th week of the fetal development, a circular part originated from ectoderm is seen at the periphery of the glans penis, which gives rise to prepuce (foreskin), a skin part which covers the glans. The corpus cavernosum and corpus spongiosum of the penis develop from the mesenchyme in the phallus. Bilateral labioscrotal swellings elongate toward each other and fuse in the midline to form the scrotum. The fusion site is seen as scrotal raphe [3]. Bilateral urethral folds fuse in the midline to form the corpus spongiosum and cavernous urethra between the 12th and 14th weeks of the fetal development. It is estimated that a fusion defect in genital tubercle during the fetal development period results in the condition termed diphallia [4]. Surgical treatment involves penile reconstruction and restoration of accompanying malformations after excision of the hypoplastic structure, however, the number of patients who have undergone surgical interventions is relatively low. In our case report, we aimed to present a patient with glandular diphallus and the follow-up process.

Case

Urethral meatus was observed during the genitourinary examination of a two-year-old male patient without any complaint, who was admitted to the urology clinic by his parents for circumcision. The penile skin could not be retracted, and glans was wider than the normal size for his age, a finding which suggested the presence of a congenital genitourinary abnormality. Foreskin was retracted using a mosquito forceps **(Figure 1)**. While the corpus penis was single, the glans was distally separated from the midline and a glandular diphallus was observed. Mea was observed in distal parts of both glandes. The patient had been followed up for one year after birth with the diagnosis of patent foramen ovale. Apart from this, no additional pathological finding was detected in physical examination of the patient. In urinary USG examination, both kidneys were normal, bladder contours were smooth, and bladder wall thickness was within normal limits. Both meatuses were catheterized and cystourethrography was planned. A 6F feeding catheter was advanced through the right glans, however it could be inserted through the left glans only 2 cm. In the imaging performed a 1:1 diluted contrast fluid injected through the right meatus moved along the penile structure, reaching and filling the bladder. We have tried to inject the contrast material through the catheter that could be advanced distally only 2 cm through the mea of the left glans, but we failed due to an overwhelming resistance. The feeding tube was withdrawn, and the procedure was repeated, however the contrast material could not be injected further **(Figure 2)**.

We have concluded that the proximal part of the meatus was obliterated. Cavernosography could not be performed, because this infant was not able to cooperate. In physical examination, a connection was found between the glans and the corpus cavernosum. Multidisciplinary approach is very important for the cases with genitourinary abnormalities. For this reason, we have requested additional second opinions from plastic surgery and child psychiatry departments. Due to the complication risks of surgical procedures performed on glans penis and the very young age of the patient, we have decided to plan the operation in the following years upon the consent of the family.

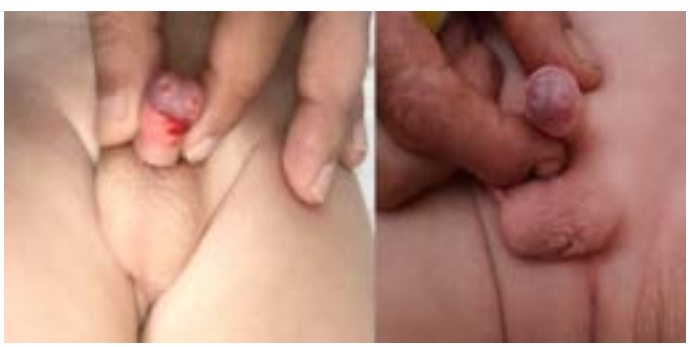

Figure 1. Bifid glans after foreskin retraction

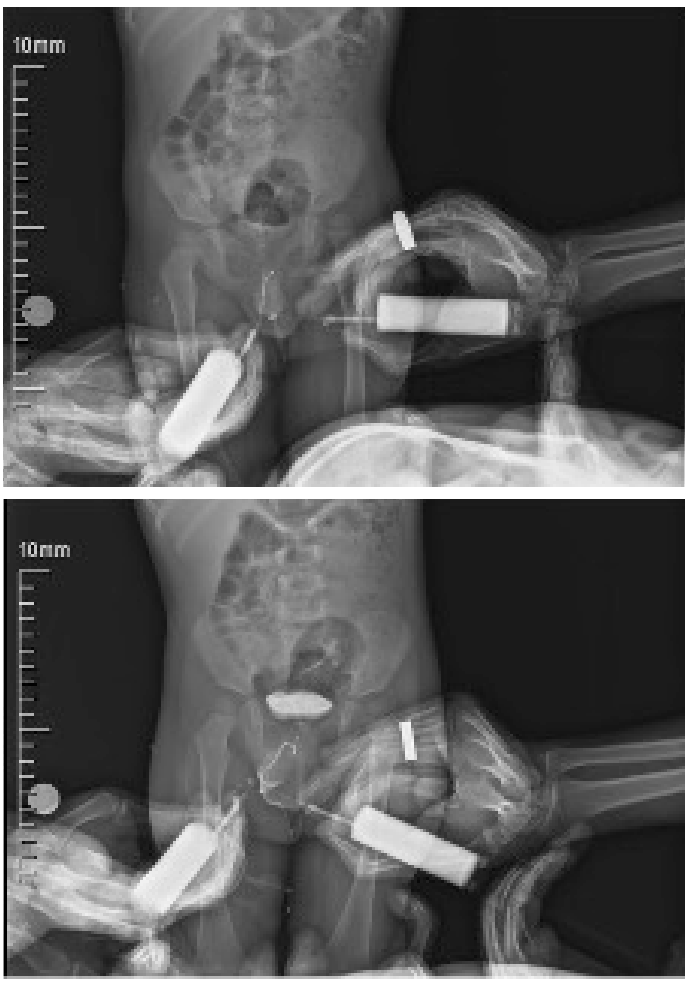

Figure 2. Cystourethrography performed after catheterization of both urethras with 6f feeding catheter

Discussion

Diphallus is a very rare anomaly that can be encountered associated with many other congenital malformations. Embryologically, a diphallus deformity can occur in two forms as pubic symphysis diastasis in which each phallus has the same set of corpora cavernosa and urethras, or cleavage of the pubic tubercle, in which each phallus has a unique set of corpora cavernosa and urethras. Diphallus has been classified in several types. Most cases of diphallus have the same corpora cavernosa in each penis [1,2]. It is important to perform a comprehensive examination in terms of congenital malformations that may present at the same time. It has been mentioned in various case reports that patients might have accompanying colon and bone anomalies, particularly genitourinary and anogenital anomalies [1,5]. Magnetic resonance imaging (MRI) could be performed instead of cavernosography to check the status of corpus cavernosum and external genitalia. But it could also be a challenging procedure considering relatively younger age of the patients and cooperation problems [6]. Multidisciplinary approach has a crucial importance for all cases with genitourinary abnormalities because of psychological impact of this condition on the patient, family, and quality of life in general. Parents should be informed about the process in detail, and should be included in the treatment process. A PubMed search was conducted with "diphallia" and "diphallus". Although 400 years

Table 1. A literature review of preoperative and postoperative datas of diphallus

	Age	Diphallus type	Associated anomaly	Methods of surgery
V. Deshpande- 2020	2 years	-Complete	-Bifid scrotum	-Phalloplasty, Scrotoplasty
Kundal et al- 2013	3 years	-Complete	-Hypospadias	-Phalloplasty, Urethroplasty
A. Tepeler et al- 2007	14 years	-Complete	-Bifid scrotum	-Phalloplasty, Scrotoplasty
Mirshemirani et al- 2010	2 days	-Complete	-Imperforate anus + colon, Bladder duplication, Hypospadias, Bifid scrotum	-Colostomy, Cystoplasty, Urethroplasty, Phalloplasty, Scrotoplasty, Colon resection + anastomosis
	4 years	-Complete	-Bladder duplication, Inguinal hernia	-Cysto-urethroplasty, Phalloplasty, Hernia repair
	12 years	-Complete	-Single kidney, Bifid scrotum, Hemi-vertebra, Bladder exstrophy	-Bladder extrophy repair, Ceco-vesical augmentation, Mitrofanoff
	1 year	-Complete	-Bladder duplication, Bifid scrotum	-Phalloplasty, Scrotoplasty, Cystoplasty
	14 years	-Bifid phallus	-Bifid scrotum, Hypospadias	-Phalloplasty, Scrotoplasty
	9 months	-Complete	-Bifid scrotum, Imperforate anus	-Colostomy, Phalloplasty, Scrotoplasty, PSARP
Elsawy et al- 2012	37 days	-Complete	-Inguinal hernia	-Phalloplasty + Penile anastomosis
Dunn et al- 2019	3 years	-Complete	-Bifid scrotum -Bladder duplication	-Phalloplasty, Scrotoplasty, Cystoplasty
Tirtayasa et al- 2013	12 years	-Complete	-Bifid scrotum	-Phalloplasty, Scrotoplasty
Karagözlü Akgül et al- 2018	4 years	-Complete	-Rectovesical fistula, Bladder duplication, Anal atresia, Colon and Rectum duplication	-Urethroplasty, Phalloplasty, Abdominoperineal pull-through, Colon resection
Zhang et al- 2020	23 years	-Pseudodiphallia	-	-Phalloplasty

have passed since the first case report and about 120 cases have been reported in the publications, there is still not enough information about the surgical procedure to be applied in most of them. After review of the literature, case reports with sufficient preoperative and postoperative data are summarized in **Table 1**.

Many studies apparently have not longer follow-up periods. In a comprehensive study with six cases Mirshemirani et al., stated that additional anomalies were observed in all cases that required additional surgical procedures [1]. In case reports presented by Deshpande [7], Kundal [8], and Tepeler [2], Elsawy [9], Dunn [10], Tirtayasa [11], and Karagözlü Akgül [12], phalloplasty plus scrotoplasty was performed for the correction of an additional anomaly. In only one case presented by Zhang [13], any additional anomaly was not reported. There is not enough information about the long-term complication rates of the surgical procedures performed for accompanying urethral malformations, and there is also no clear data on the effect of surgical procedures on the improvement of erectile dysfunction, which is associated with the congenital defects of corpus cavernosum. More comprehensive and prospective studies with longer follow-up periods are needed to get more insight into diphallus and other associated conditions.

Ethics Committee Approval: N / A.
Informed Consent: An informed consent was obtained from the patient.
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Authorship Contributions: Any contribution was not made by any individual not listed as an author. Concept – H.I.I.; Design – H.I.I.; Supervision – B.A.; Resources – B.A.; Materials – I.G.K.; Data Collection and/or Processing – O.Y.S.; Analysis and/or Interpretation – I.G.K.; Literature Search – A.K.; Writing Manuscript – H.I.I.; Critical Review – B.A.
Conflict of Interest: The authors declare that they have no conflict of interest.
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