A True Complete Diphalliahas Proven to Be a More Complex Case of Caudal Duplication Syndrome

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Abstract

Introduction and Objectives. Diphallia, also known as penile duplication, diphallicterrata or diphallasparatus, is an extremely rare congenital anomaly, frequently associated with other malformations. The prognosis and optimal management of this pathology depend on the type of diphallia and the associated anomalies.

Materials and Methods. This article presents the case of a newborn baby boy who was first admitted with complaints of abnormal appearance of the external genitalia and gluteal region and subsequently suffered two separate surgical interventions: one for the penile duplication and one for the gluteal and perineum assimetry.

Results. The second procedure was more challenging given that the diagnosis of meningocele was discovered intraoperative and changed the operation planning, thus reflecting the poor initial medical assesment of the patient and prooving once more the importance of complete imaging studies in case of patients with diphallia.

Conclusions. In spite of its rarity, penile duplication has a massive impact on both the patient's family and the medical staff. The treatment is complex and must be individualized depending on the form of diphallia and its associated anomalies, thus a complete imaging study should be performed.

Keywords: complete diphallia, sacral lipoma, meningocele, caudal duplication syndrome.

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Introduction and Objectives

Diphallia or penile duplication is an extremely rare congenital anomaly, estimated to occur once in every 5.5 million live births [1]. Approximately 100 cases were reported since Wecker first described it in 1609 in Italy [2]. Diphallia is frequently associated with other anomalies such as anorectal malformations, duplication of lower urinary tract, lower abdominal wall defects, renal malformations and neural tube defects [3]. We report a case of complete diphallia associated with sacral lipoma, meningocele, bifid scrotum, urethral duplication, vesicoureteral reflux, gluteal and perineum asymmetry. From our knowledge, this case has never been reported in Romania.

Materials and Methods

A newborn baby boy was brought in with complaints of an abnormal appearance of the external genitalia and gluteal region. His antenatal history was uneventful and there were no similar complaints in the patient's family.

The physical examination showed 2 well-developed penises laying one above the other, both with normal corporal bodies and normal urethral meatus (Figure 1). The patient was passing urine simultaneously through both penises in equal streams and had normal urinary continence until the age of 6 months. The scrotum consisted of three compartments and one fully descended testis was present in each lateral sac. Both gluteal region and the perineum were asymmetric mainly due to the presence of a soft, rubbery mass. The tumor was well defined, non tender, with free mobility of overlying skin. The clinical aspect was suggestive of a lipoma located in the left gluteal region with a dimple of the skin and hyperpigmentation of middline (Figure 2). There were no other notable anomalies on physical examination.

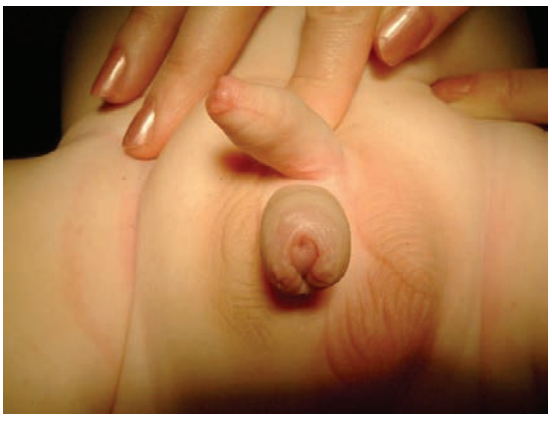

Figure 1: Diphallia and bifid scrotum

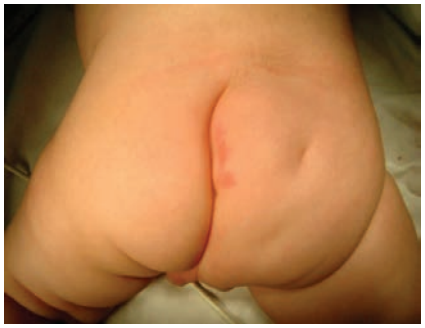

Figure 2: Gluteal region asymmetry due to the presence of a lipoma

Baseline blood tests and other routine investigations were within the normal range. The voiding cystourethrogram (VCUG) revealed complete duplication of the penile and bulbar urethra. The urethras merged along the way and ended up in a single bladder. This investigation also revealed right vesicoureteral reflux (Figure 4). The soft tissue ultrasonography did not reveal any information in addition to the clinical findings. Due to both technical difficulties in the radiology department of our hospital at the time of this patient's admissions and the family's social status, neither a CT scan nor an MRI were performed.

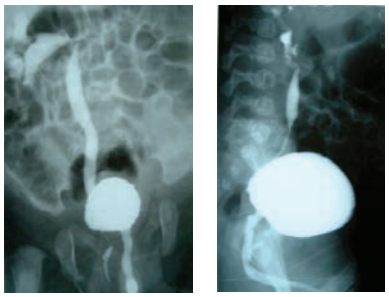

Figure 3: VCUG which revealed duplication of the penile and bulbar urethra and right vesicoureteral reflux

The patient was admitted to our pediatric surgery department 3 separate times. During his first admission basic investigations were performed and the newborn was diagnosed with penile and urethral duplication, bifid scrotum, subcutaneous sacral lipoma and grade III right vesicoureteral reflux. The anomaly wasn't considered life-threatening or with a potentially major impact on the child's future physical development. Thus, after the mutual agreement between the doctor and the family, no surgical treatment was performed and the newborn was discharged. At the age of 6 months, one of the urethras became, spontaneously, obstructed and therefore the decision of surgical management was taken. The first procedure was performed under general anesthesia with the patient in "frog-leg" supine position. Cystoscopy reveals a dimple-track on the middle line of the trigon and a STING was performed for the right VUR. An anal in-

cision at the base of the inferior perineum was made and the perineum was dissected up to the insertion of the corpora cavernosa on the right ischiopubic ramus, protecting the corresponding elements of the normal perineum (corpora cavernosa and urethra). The corpora of the supernumerary penis were ligated at the level of their insertion and resected using the electrocautery (Figure 4). The scrotum was closed with interrupted absorbable sutures. A Nelaton Fr 6 catheter was placed into the remaining urethra.

Results

The postoperative period was uneventful. At the short and medium term follow-up the result was considered satisfactory.

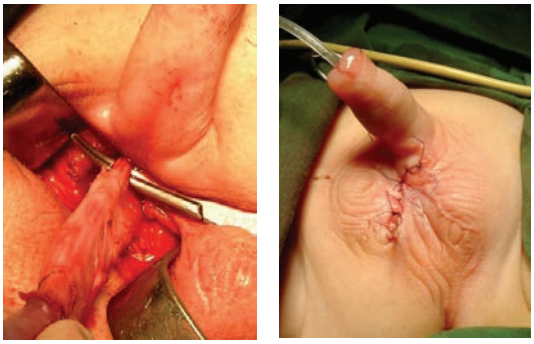

Figure4: First procedure: perineal dissection and excision of supernumerary penis

The third admission was at the age of 1 year and 1 month for the surgical treatment of the gluteal region and perineum asymmetry. This second intervention was also performed under general anesthesia but with the patient in prone position. The intervention started with a midline posterior sagital incision. During dissection of the subcutaneous fat, the leakage of a clear fluid was observed and therefore led to the discovery of a 3/5cm meningocele with perimeningoceal lipoma. The neural defect was dissected up to its neck which was inserted on a coccygeal defect. After the ligation with two transfixed sutures the meningocele was excised. The aponeurotic flaps were dissected and the coccygeal defect was closed with interrupted sutures. There were no complications during the postoperative period.

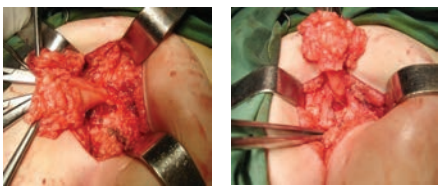

Figure 5: Careful dissection and removal of the lipoma and menigocele.

Discussions

Penile duplication is one of the very rare congenital anomalies of the urogenital system and it is believed that no two cases are identical. Therefore, before establishing a management plan one must make a thorough evaluation of the patient in order to identify any associated anomalies and properly classify the condition.

Schneider first divided diphallia into three groups: duplication of the glans alone, bifid diphallia and complete diphallia[4][5]. Villanova and Raventos later added a fourth category, pseudodiphallia. Currently, a widely accepted classification includes two main types of penile duplication: true diphallia and bifid phallus, each of them further divided into a partial or complete form [5,6,7]. In the case of true complete diphallia each penis has two corpora cavernosa and a corpus spongiosum. If the duplicated penis is smaller or rudimentary, then it corresponds to true partial diphallia. When only one corpus cavernosum is present in each penis, the term bifid phallus applies. This category of the anomaly is considered complete, if the separation is total or partial, if the separation is resumed to the glans alone. The term pseudodiphallia corresponds to true partial diphallia. Most patients with penile duplication have a single corpus cavernosum in each organ [8]. The penises can be equal or unequal in size and usually lie side by side but they can lie one above the other [9]. In the case of penile duplication, the urethra shows a range of variations, from fully functioning double urethras to complete absence of it in each penis [8]. The meatus can be normal, hypospadiac or epispadiac and the scrotum may be normal or bifid [8]. The testes can be normal, ectopic or undescended [5]. Penile duplication is frequently associated with other malformations such as: bladder and urethral duplication, exstrophy of the bladder, renal anomalies, anorectal malformations, colon and rectosigmoid duplication, ventral hernia, neural tube defects and cardiac anomalies [8]. This pathology may also be included in a rare congenital condition termed "caudal duplication syndrome" which describes the association between gastrointestinal, genitourinary and distal neural tube malformations [10]. In the presented case the patient had true complete diphallia with bifid scrotum, two normal testes and normal meatus. The urethral duplication was type III according to Effman's classification [11]. The associated anomalies were: right vesicoureteral reflux and lipomeningocele. The prognosis and optimal treatment option in the case of diphallia depend on the type of penile duplication and its associated malformations [7]. We acknowledge that because

the patient wasn't properly investigated during his first admission to our hospital, an important diagnosis that might have changed the prognosis in this case was omitted. Therefore, we consider that either a CT scan or an MRI is mandatory before any surgical treatment in the case of diphallia.

Management of diphallia poses a challenge in view of the medical, surgical and ethical decision-making. The treatment should be individualized and the anomalies that are potentially life-threatening should be solved first [2]. The surgical procedure, as applied in our case, usually includes excision of the duplicated penile structure and its urethra [2,12]. Priyadarshi and Djordjevic et al. reported a penis reconstruction by joining the corporal bodies in each penis in a patient with true complete diphallia [13].

Conclusions

In spite of its rarity, penile duplication has a massive impact on both the patient's family and the medical staff. The treatment is complex and must be individualized depending on the form of diphallia and its associated anomalies, thus a complete imaging study should be performed.

In our opinion, these patients should be treated as soon as possible with the goal of attaining a satisfactory functional and cosmetic result and without discarding the patient's opinion in the decision making.

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