Scholars Journal of Medical Case Reports (SJMCR) **ISSN 2347-6559 (Online)**
Abbreviated Key Title: Sch. J. Med. Case Rep. **ISSN 2347-9507 (Print)**

A United of Scholars Academic and Scientific Society, India

Diphallia-About an Exceptional Case & Review of Literature

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Article History
Received: 22.10.2018
Accepted: 05.11.2018
Published:30.11.2018

DOI:
10.36347/sjmcr.2018.v06i11.004

Abstract: Diphallia is a rare anomaly, defined by a total or partial duplication of the male genitalia. This abnormality is rarely isolated, and fits most often in a table of varied malformation, determining management and prognosis. There are different forms Clinical and anatomical of this condition, which explains the variety and complexity of classifications suggested until now. However, the diagnosis is clinical, and paraclinical aims were used to determine the type of diphallia and detect any malformations. The therapeutic management is very variable: from the simple removal of supernumerary penis as in our case, to a complex reconstruction when associated with severe malformations. In our thesis, we reported a new case of isolated diphallia, and we have reviewed literature, from which the new epidemiological data have been established, we also reported the different etiopathogenic theories and pathological classifications, whose have been developed so far. Finally, we outlined the diagnosis and treatment management of this malformation.
Keywords: diphallia, penile duplication, bifid phallus, pseudodiphallia.

INTRODUCTION

Diphallia is a rare malformation of the male uro-genital, defined by total or partial duplication of the penis [1, 2]; having thus a double impact:

- Aesthetics: with subsequent psychic consequences, from where the interest of care at an early age.
- Functional: that is, the voiding and sexual function.

There are various more or less severe forms of this urogenital anomaly external, and can be isolated or most often fit into a poly-malformative syndrome more or less complex.

The therapeutic management varies according to the importance of the malformation and according to the overall diagnosis, and goes from the simple ablation of the supernumerary penis, to more complex reconstructions when the diphallia is associated with other malformations.

MATERIALS & METHODS

We will report in this work the case of an 8 year old patient who was referred in 2009, to the pediatric visceral surgery service "A" of the children's hospital in RABAT, for the management of its penile duplication. As a result, our patient underwent a series of clinical and para-clinical investigations, at the end of which he received an additional penis resection.

We will then present a review of the literature that we have realized about this, after doing a research on published studies concerning diphallia, using several resources bibliographies (electronic and printed), while emphasizing which have been published since 1987.

This is an 8-year-old child, the last of a 6-year-old fraternity, with no notion of consanguinity of the parents, resulting from a pregnancy not followed with home delivery, with no particular pathological antecedents. The symptomatology goes back to the birth by the finding of the parents of the presence of a polypoid formation at the level of the dorsal surface of the yard of the child, painless and which would be erectile according to the mother. On the other hand, the patient reports no concept of incontinence or urinary tract infection. This malformation has been neglected by his parents.

The clinical examination finds a child in good general condition, apyretic. Examination of the external genitalia finds a polyploid bud one centimeter in diameter, pinkish in color, of soft consistency, on the dorsal side of the penis with a collar. The urethral meatus is slightly apical and the testes are intraspecal (Figure 1). The patient has a good quality urine stream during urination. The patient has secondary sexual characteristics corresponding to the T2P2 stages according to the TANNER classification. The lumbar pits are free on palpation. The rest of the examination is peculiar, having found no other apparent clinical malformations.

Fig-1: Image showing the presence of a polyploid formation at the dorsal surface of the penis

An assessment was made, including an ultrasound of the urinary tract and of the penis that objectified normal kidneys, a single bladder and functional, and the presence of a penile tissue bud continuity with the corpora cavernosa of the penis, without extension towards the bladder.

Retrogressive urethrocystography ascending, was performed, and showed a single bladder of normal capacity, functional, without post-void residue.

The urethra is unique, presenting a normal morphology. On the other hand, there were no vesico-ureteral reflux or bone abnormalities (Figure 2).

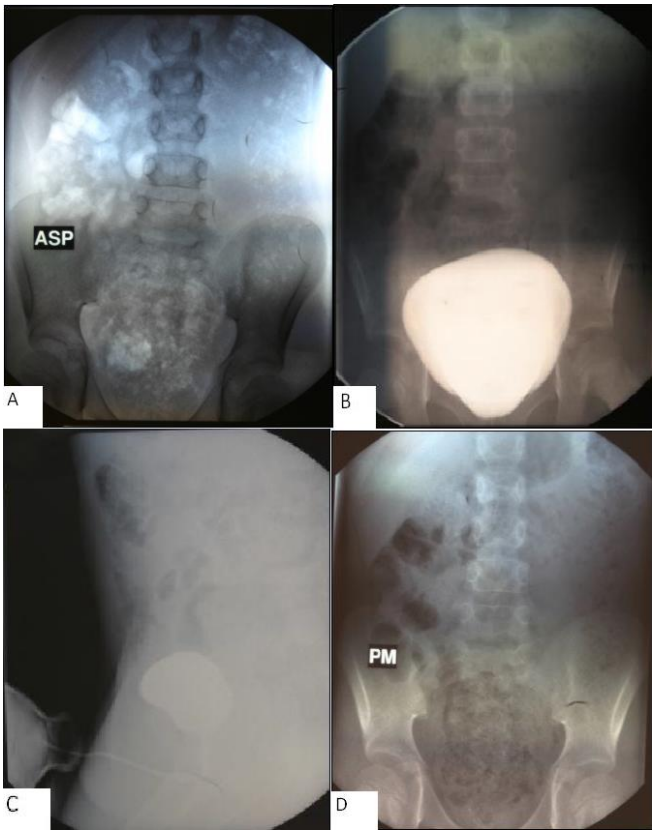

Fig-2: A- X-ray of the abdomen without preparation

B- Retrograde urethrocystography showing no bladder abnormalities at filling
C- Permictional image showing no reflux
D- Postmictional picture showing no residue

The child was operated on by performing a skin incision at the base of the accessory penis, followed by dissection of the vestiges of cavernous body, then the Ligature - section at the base.

Histopathological account of the resection room:

- ***Macroscopy:*** At the cut, aspect of a solid tube centered by a whitish cord, included in totality.

- ***Microscopy:*** Histologically, this formation shows a cutaneous tissue surrounding a beam of smooth muscle fibers of immature appearance and full of slits. We found in the body of this formation tubuliform structures lined sometimes with pseudostratified cylindrical epithelial lining. Structures are numerous at the level of the swollen end. The hypodermis is richly innervated and vascularized. There is a predominantly lymphoplasmocytic inflammatory infiltrate superficial dermal.
- ***Conclusion:*** This formation shows the appearance of an immature penile tissue

DISCUSSION

Diphallia is a very rare congenital malformation of the male child of varied clinical expression. There are several classifications of these external genital anomalies which testify to the difficulty encountered in defining these numeric penile anomalies.

According to SCHNEIDER, three anatomical forms can be individualized: the penile glandular diphallia, the bifid diphallia and the complete diphallia [3].

RAVENTOS and VILLANOVA complete this definition by introducing the term pseudo-diphallia [4]. ABDEL [5] takes over and simplifient the classification of SCHNEIDER by dividing the diphallias according to two main anatomical aspects: the bifid penis and the true diphallia.

In fact, these numerical anomalies of the penis can be grouped according to their external and histological morphological aspect in three categories:

- The complete and incomplete true diphallia
- Complete and partial bifid penis (glandular division)
- The pseudodiphallia.

This classification is based on the presence of cavernous and cancellous bodies in the duplicated organ and on the completeness or incompleteness of the duplication [2,6].

The bifid penis is characterized by the presence of à longitudinal furrow that reflects the bifurcation, a single urethra common to both "hemipenis" that compose it and the existence of a cavernous body present in each branch of bifidity. We distinguish anterior forms and posterior forms.

In earlier forms, the groove divides the penis into two "hemi-acorns". In the posterior forms, the division of the penis extends towards its root forming two well-individualized hemi-penis each possessing its cavernous body.

There are two very different forms of diphallia: incomplete diphallia or otherwise known as diphallia with bifid penis and complete diphallia or true diphallia.

In the incomplete diphallia (diphallia with penis bifidis), the urethra can be hypospade with a meatus opening in the balanopreputial furrow.

The two phalluses each have two cavernous bodies, a spongy body. In complete penile duplication or true diphallia, there are two separate penises, each with two cavernous body and an urethra, a scrotum more or less split. Each hemi-scrotum is located below penis corresponding with very often testicles abnormal, agenetic or cryptorchids. The bladder can be split or bilobed and each half receives a urethra while one of the urethra can be either epispade or hypospade [7], absent or atretic [8], but not causing a micturition disorder. Both penises can be side by side or above each other.

Pseudodiphallia, originally described en 1954 by VILLANOVA and RAVENTOS, have grouped forms of complete and incomplete diphallia. This term should be used exclusively to designate exceptional observations reporting an appendix that looks macroscopically to à penis whose histological structure contains erectile tissue without organization clean in spongy, cavernous body [4].

The embryological hypotheses that explain the occurrence of a diphallia are still very much discussed [9]. It's hard to understand the existence of the duplication of the penis while the genital tubercle is a unique structure throughout the development normal of the embryo. For CECIL [10], the diphallia result from a disorder of the fusion of the beads mesenchymal genital tubercle. In the diphallia true, it seems like the existence of two penises possessing each an urethra can be secondary to the division of the uro-genital membrane. After the implementation place of the cloacal membrane, two columns of mesenchyme progress in the ventral direction, and their fusion at the 6th week carries out the genital tubercle.

It should be admitted that the longitudinal duplication of the cloacal membrane and urogenital sinus allowed three or four columns of mesenchyme to grow and thus give rise to two genital tubercles.

Thus, there are two urethral drafts, lined by four mesenchymal loops at the origin of two genital tubercles. We thus understand the possible associations with a pubic diastasis, bladder duplication, colo-rectal or spinal [2]. The diphallia can be considered as a phylogenetic atavism (snakes and lizards), a monster-like teratoid structure double, a minor malformation in the same way as supernumerary fingers [9].

Diphallia is an abnormality that is not diagnosed in ante-natal period. All these pregnancies have benefited from precise ultrasound never allowed to discover this anomaly. The diagnosis Diphallia is neonatal and rests exclusively on the clinical examination. In literature, it is mentioned cases of diphallia discovered in boys more than two years old, which shows the difficulty and the specialization of this urogenital diagnosis.

Diphallia is a rare, sporadic anomaly. All some family forms have been reported [11]. Research of a mutation at the origin of this anomaly digital penis are in progress and requires for some authors carry out a karyotype [9, 12]. In diphallia, malformation syndrome is more complex and exceeds the urogenital sphere [5,9].

The malformation assessment must include

- Radiographic explorations that provide information on the condition of the lumbosacral spine, pubis (diastasis puberty), the type of anorectal malformation (high, low) or the existence of atresia of the esophagus.
- Retrograde and micturate urethrocystography functional and anatomical information from the urinary system.
- An ultrasound of both penis during the preoperative period to analyze at best penile duplication [12]. The ultrasound would show the number of bodies cavernous, spongy and complete information about the urogenital tract, the digestive system.
- Urethrocystoscopy occupies an important place in the preoperative assessment: the urethra, the bladder, the urethral meatus can be better described.
- M.R.I gives more information about the bodies cavernous, spongy, and their course. State, viability testicles, the erection of each penis are also evaluated. It allows the complete anatomical study of this malformation [13].

The treatment of diphallia is surgical. Every gesture starts with the correction of the associated malformations that are incompatible with life and that from the first hours of life. For example, the existence of a diphallia that associates with atresia of the esophagus or other malformation of the digestive or cardiovascular system.

The less developed penis (accessory) is resected for the benefit of the dominant penis. The associated anomalies of the urethra are corrected in the same operating time (genitoplasty, urethroplasty).

In the case of a diphallia with a bifid penis the correction passes by the union of the two cavernous bodies completed by a urethroplasty.

CONCLUSION

Diphallia is an extremely rare malformation, defined by a total or partial duplication of the penis. Most often this malformation integrates into a more complex polymalformative syndrome that varies from one patient to another.

His pathogenesis remains unknown. Several theories have been advanced without that no one can explain the anatomopthological polymorphism and the wide range of associated malformations.

Several classifications have been developed, including that of ALEEM, which is remained the most commonly used because of its simplicity.

The diagnosis of diphallia is exclusively clinical, however anatomopathological classification requires the use of certain para-clinics, and this is penile ultrasound and more recently the MRI that has gave better results.

The pre-operative genital assessment is fundamental, as is the assessment endoscopic urethra. It must be possible to determine which one is the dominant penis and decide later, which therapeutic conduct to undertake.

Treatment should always be individualized, and consists of resection from the least developed penis to the benefit of the dominant penis. In certain situations, a penile enlargement completed by a urethroplasty can be indicated. The ultimate goal is to achieve an aesthetic and functional result the closer to normal. The prognosis for this condition is generally excellent when it is care properly at an early age.

Conflict of interest

All the authors declare that they do not have any conflict of interest.

Author's contribution

All the authors have contributed to the redaction of this manuscript.

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