

Case report**PSEUDODIPHALLIA WITH DUPLICATION OF URETHRA****Prakash Billakanti Babu, Ramachandra Bhat K***Kasturba Medical College, Manipal University, Manipal, Karnataka, India***RESUMEN**

Se presenta un caso raro de "Pseudofalia" en un adulto de 50-60 años de edad, cuyo cuerpo fue donado al departamento de la anatomía del Hospital Universitario Kasturba, Manipal. El sujeto presentaba un pene verdadero, de tamaño normal y otro en miniatura junto a la zona ventral de estructura principal cercana al glande. El glande del pene verdadero no estaba cubierto por el prepucio. El pene accesorio estaba plenamente recubierto por piel y en la punta una depresión. La observación cercana mostró dos aberturas que indicaban conexión con la uretra. No había más prolongaciones indicando la presencia de glande en este apéndice. El escroto tenía apariencia normal con los testículos en el lugar. Las arterias y los nervios observados en el pene accesorio eran desviaciones del pene principal. Sin embargo las venas mostraban variaciones. La vena dorsal superficial derecha se originaba en el pene accesorio, mientras que la vena dorsal izquierda estaba formada por la unión de dos venas separadas procedentes del pene principal y el accesorio. Una parte pequeña del órgano accesorio fue para observaciones microscópicas, mostrando un cuerpo esponjoso como una extensión del pene verdadero. El corte mostraba dos canales uretrales rodeados por tejido esponjoso con espacios cavernosos. Los nervios y los vasos sanguíneos podían verse entre el tejido esponjoso. El epitelio parecía ser una clase de estratificado escamoso (no queratinado). No se observaron anomalías en el sistema urinario.

Palabras clave: *Pseudodifalia, duplicación de uretra, escroto normal*

ABSTRACT

We present a case of pseudodiphallia in a person whose body was donated to the anatomy department of Kasturba Medical College, Manipal. The age of the

individual was approximately 50-60 years. There was the presence of true penis of normal size and miniature penis attached to the ventral aspect of main structure close to glans. The glans of the true penis was not covered by the prepuce. The accessory penis had full covering of skin and at the tip a depression. Close observation of this showed two openings indicating openings of the urethra. There was no enlargement to indicate the presence of glans in this appendage. The scrotum had normal appearance with the testes in place. Arteries and nerves observed on the accessory penis were derived from the main penis. However veins showed some variations. The superficial dorsal vein on the right side was originating from the accessory penis. Whereas, the left superficial dorsal vein was formed by the union of two veins arising separately from the accessory and main penis. A small piece of the accessory organ was processed for microscopic observations, which showed the presence of corpus spongiosum only, as an extension from the true penis. The section showed two urethral channels surrounded by spongy tissue with cavernous spaces. Nerves and blood vessels could be seen among the spongy tissue. The epithelium appeared to be stratified squamous (non-keratinizing) type. No abnormalities were seen in the Urinary system.

Key words: *Pseudodiphallia, duplication of urethra, normal scrotum*

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INTRODUCTION

Diphallus or penile duplication (PD) is a rare congenital anomaly that occurs once in every five million live births (Adair and Lewis, 1960; Pendino, 1950). PD may be considered a subset of the more common urethral duplication with one penis, of which there are over 200 cases reported in the literature. PD is rarely reported in isolation, and can be associated with other anomalies in kidneys, ureters, urinary bladder, urethra, spina bifida, exomphalos, separation of the symphysis pubis and rectus abdominis musculature, vertebral and anorectal anomalies including hindgut duplication (Kode, 1991; Azmy, 1990). The term caudal duplication syndrome was proposed to describe the association between genitourinary (GU), gastrointestinal (GI) and distal neural tube defects; it has been suggested that this may result from an insult to the caudal cell mass at approximately the 23rd to 25th day of gestation as reported by Hallowell et al (1977). The form of PD can be varying from small accessory penis or duplication of glans to complete duplication (Fujita et al 1979; Mizogushi et al 1984; Melekos et al 1986). Here we report a case of accessory penis (pseudophallus) with double urethra in it. This case of PD illustrates the complex cluster of associated anomalies that occurred to a greater or lesser degree, with almost all of the few reported cases. It also highlights the importance of thorough investigation of the GU system to delineate the full extent of duplications and other anomalies.

CASE REPORT

A rare case of pseudodiphallia was seen in an adult, 50-60 years old, donated to the anatomy department of Kasturba Medical College, Manipal. The case presented a true penis of normal size and a miniature penis attached to its ventral aspect close to glans (Fig.1). The glans of the true penis was not covered by the prepuce. Its absence could not be assigned whether it was a case of circumcision or a malformation. Glans had the opening of the spongy urethra at its tip. The accessory penis was covered by skin and a depression at its tip. On close observation it showed two openings indicating openings of the urethra. There was no glans in this appendage. The scrotum was normal with the testis in place. When a thin probe was passed through the opening in the accessory penis, it reached the ventral part of spongy urethra of the true penis

showing its continuity with it. Arteries and nerves of the accessory penis were derived from the main penis. However veins showed some variations. The superficial dorsal vein on the right side was originating from the accessory organ, whereas, the left superficial dorsal vein was formed by the union of two veins arising from the accessory and true penis (Fig.2). Microscopic observation of the accessory penis showed the presence of corpus spongiosum which was an extension from true penis. There were two urethral channels surrounded by spongy tissue with cavernous spaces (Fig.3). Nerves and blood vessels could be seen among the spongy tissue. The epithelium appeared to be stratified squamous (non-keratinizing) type (Fig.3). On dissection of the abdomen, no abnormalities were seen in the urinary system.

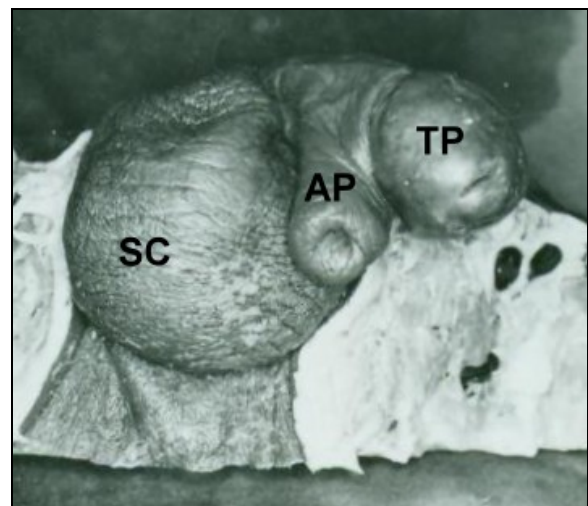


Figure1: Photograph of anterior view of well formed penis and unseparated smaller accessory penis. TP: True penis; AP: Accessory penis; SC: Scrotum.

DISCUSSION

Diphallus is a rare congenital anomaly that occurs once in every five million live births. The first case was reported in 1609 by Wecker. Neugebauer (1898) and Nesbit and Bromme (1933) have reviewed such cases of diphallus. PD has been classified in different ways, such as glandular, bifid, concealed, complete, hemidiphallus and triple penis as reported by (Ravi et al 1987; Wilson et al 1973). Scheneider (1969) classified diphallus in three groups; diphallus of glans alone, bifid diphallus and complete

diphallus. Vilanova and Raventos (1954) have added fourth category pseudodiphallus. Azmy (1993) reported a male infant of unspecified karyotype with a complex group of anomalies including diphallus. The phalli had corpora cavernosa with urethra, which conveyed urine to the exterior. Duplication of the bladder with vesicoureteric reflux (VUR) into the left renal moiety of a horseshoe kidney was noted. The child was surgically managed by joining the two bladders side to side, performing a right orchidopexy and removing the left penis. A case of complete diphallus with many of the anomalies

seen in the other cases was reported by Huang W et al (1994): a 28 years old Chinese 46 XY male with divarication of the recti, duplication of the symphysis pubis, pelvic asymmetry and a history of imperforate anus with a double gluteal cleft. He had two small penises with normal erectile function, the right having a normal urethra with poor urine flow and the left a perineal hypospadiac urethra, which was the dominant voiding route. The scrotum was bifid and the left compartment contained the small solitary testis. Azoospermia was noted on seminal fluid analysis.

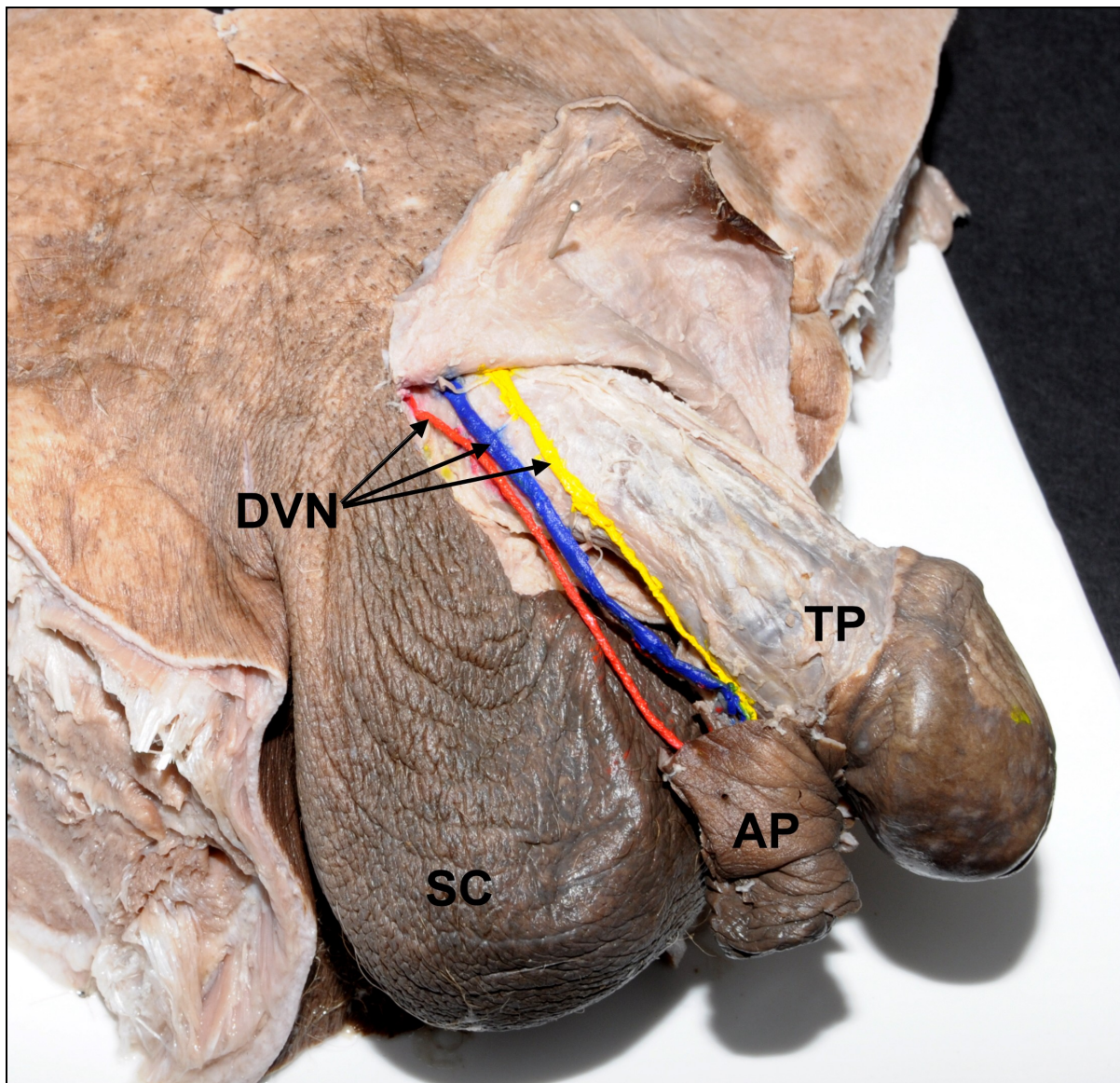


Figure 2: Photograph showing vessels and nerves of accessory penis. TP: True penis; AP: Accessory penis; SC: Scrotum. DVN: Dorsal vessels and nerves seen on accessory penis.

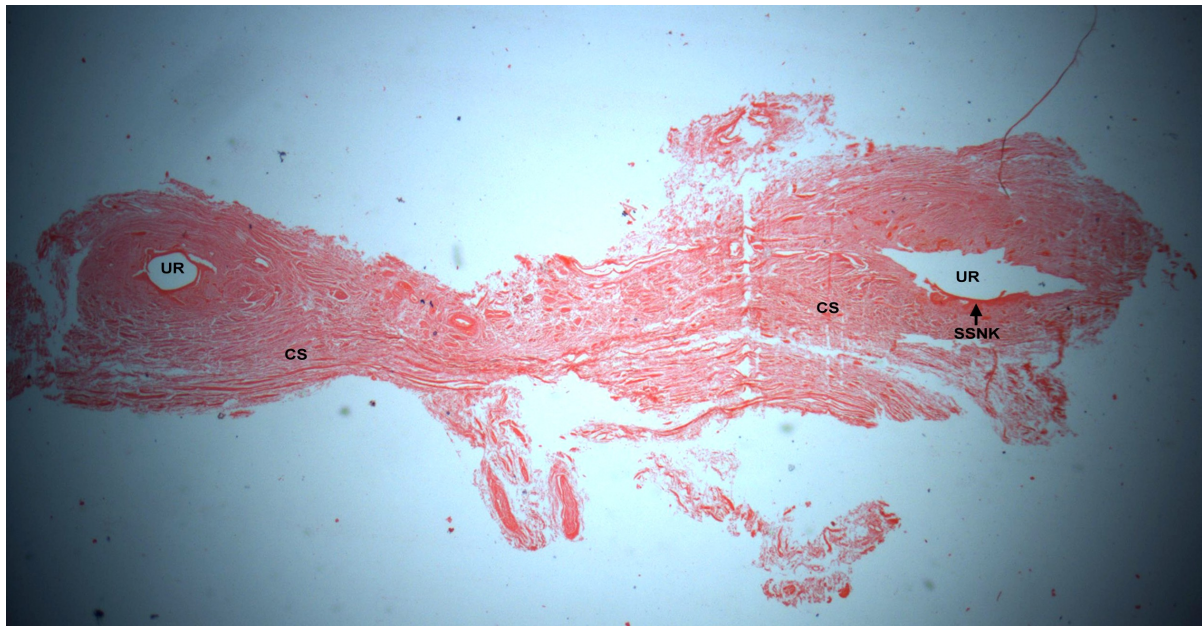


Figure 3: Histological picture of accessory penis showing double urethra. UR: Urethra; CS: Corpus spongiosum; SSNK: Stratified squamous non-keratinized epithelium.

Surgical reconstruction involved resection and reimplantation of the right penis to elongate the left. The left glans was resected and the corpora cavernosa and neurovasculature of the two phalli joined. A radical free forearm flap was used to reconstruct the urethra and scrotum. Erectile function had returned by two months postoperatively and appropriate sensation had returned by six months. The case described by Kode (1991) revealed an otherwise morphologically normal male infant with a perfectly formed micro-glans penis projecting from the dorsum of the normal glans at the corona. There was a rudimentary, blind ending urethra through which no urine passed. The rest of the external genitalia were normal, as was a plain radiograph of the pelvis and a micturating cystourethrogram. This accessory glans was excised and histology confirmed duplication with normal glans and urethral tissue. There were no corporal structures, and this was considered a duplication of the glans rather than the phallus.

Karna and Kapur (1994) reported the only documented case associated with an abnormal karyotype; a male 46, XY infant with complete diphallus, multiple anomalies from the associated cluster, and a balance translocation t (1:14) (p36.3; q24.3). The omphalocele and chromosome translocation were diagnosed antenatally and, at birth, the child was found to have multiple anomalies including an imperforate anus; tracheoesophageal fistula; two well formed

penises, both passing urine via urethral orifices; a bifid scrotum without palpable testes. The child died of respiratory complication in the neonatal period prior to definitive corrective surgery. Alireza et al (2010) reported a case in a two year old infant with imperforate anus, complete rectosigmoid duplication, rectal pouch, doubling bladder, functioning double urethra, and hypospadias on the left side urethra, complete diphallus and bifid scrotum.

Sarmentero et al (1990) reported a case of diphallus with urethral duplications in a 46, XY male with multiple malformations including rectus abdominis diastasis; membranous anal atresia; right sided cryptorchidism, VUR and diphallus with an ectopic penis on the upper right thigh. Both phalli had a functional urethra. Definitive surgery was performed which involved excision of the ectopic right penis, including complete urethral excision, via a combined intra and extravesical approach. A right orchidopexy was also performed and the vesicoureteric reflux (VUR) was managed conservatively. Sharma et al (1996) reported a case of diphallus in an infant of unspecified karyotype with ileal atresia and tubular duplication of the colon.

The above reports indicate the wide variety of anomalies associated with diphallus. Imperforate anus and other hindgut anomalies including duplication, vertebral anomalies including spina bifida, renal and urinary tract anomalies with external genital anomalies are all common in this

rare condition. This may be due to duplication of the caudal body elements of mesodermal origin (Hallowell et al 1977). Abdulkadir (2007) has reported in a 14-year old male well formed penis with normally located and functioning urethra and an unseparated accessory smaller ventral penis and a rotated glans with a blind depression at its tip. There were two penile shafts located one above the other with two corpora cavernosa. Abdominal and other physical examination were normal. Voiding-cystourethrography revealed a normal single bladder and urethra and no vesicoureteric reflux. Between 3 and 5 weeks of gestation mesoderm migrates caudally, separating the urogenital sinus from the rectum. The genital tubercle, which later forms the phallus, results from merging of the paired columns of mesoderm around the lateral margins of the cloacal plate (Hallowell et al 1977). They also proposed that diphallus may be the result of a defect in migration of mesoderm through the cloacal membrane in early embryogenesis. This hypothesis is supported by the associated defects arising predominantly in tissue of mesodermal origin. Thus, diphallus may be considered as a part of the more encompassing caudal duplication syndrome. Most of the cases had a normal karyotype except one described by Karna and Kapur (1994). They suggested that defects in homeobox genes, which are thought to be master controller genes of differentiation, may be involved in caudal duplication, including diphallus (Karna and Kapur 1994). Testicular biopsy (Hallowell et al 1977) and seminal analysis (Nesbit et al 1933) indicate fertility is being compromised in some cases of complete diphallus. However, long term follow ups have suggested normal physical growth and development (Hallowell et al 1977, Nesbit et al 1933). Diphallus, although an exceedingly rare abnormality, it requires a detailed clinical, preoperative investigation in the management of these anomalies.

In the present case, accessory penis attached to the ventral aspect of the true penis near the glans. Such an accessory penis may be referred as bifid penis or pseudophallus. On dissection this accessory penis was observed to be the continuation of the corpus spongiosum of the true penis without corpora cavernosa which was confirmed by our histological findings. Complete duplication of penis, duplication of glans into right and left, have been described earlier. Penile duplication is commonly associated with anomalies in the urinary system and the hindgut. No abnormalities were observed in these organs in the present case. Microscopic observations of the accessory penis showed the presence of two

urethral channels. They were continuous with the spongy urethra of the true penis. The lining epithelium of these canals was stratified squamous (Non-Keratinizing) type. Hence these two channels formed additional passages for the urine. Duplication of the urethra in various forms has been reported. Duplication of the urethra in an accessory penis probably has been not documented in the available literature.

Embryological aspects

The normal development of phallus begins with the coalescence of bilateral genital tubercles at the anterior end of the phallic of the urogenital sinus (Stephens et al 1996). Columns of mesoderm growing rapidly around lateral margins of the cloacal plate form the genital tubercle. Failure of migration or fusion of mesoderm in the early embryonic development is a logical explanation for diphallus. This theory is supported by the fact that most penile duplications are composed of one corpus cavernosum (Aleem 1972).

The bladder, prostate and associated structures develop from the fused terminal ends of mesonephric ducts; failure of fusion in this area will lead to duplication of genitourinary tract.

To summarize, the entire urogenital tract is derived from bilateral primordia which fuse to produce a single end product. Therefore, PD is usually the result of incompletely fused primordia.

Conclusion

Complete duplication of the urogenital system is a rare anomaly and treatment should always be individualized, Associated malformations should be treated first, and the ultimate goal of surgical intervention should include separation of the urogenital and gastrointestinal tract, preservation of continence, eradications of infections and finally, reconstruction of the external genitalia in a functional and aesthetic fashion. Treatment of diphallus usually includes excision of the duplicated penile structure and its urethra. Therefore, all the patients with diphallus have to be examined carefully because of the high incidence of other systemic anomalies as cited by Hallowell et al (1977).

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