Prenatal Diagnosis of Concomitant Accessory Scrotum and Diphallia in the Accessory Tissues: Case Report

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A case of accessory scrotum with duplicated penis (diphallia) in a male fetus is reported because of its rarity. This case is presented with proved negative androgen receptors in the accessory genitalia. The results of excisional surgery as well as immunostaining for androgen receptors in the resected specimens are presented as well. The outcomes of prenatal ultrasonography, clinical examination of the infant, and pathologic findings of the resected accessory genitalia are also discussed.
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An accessory scrotum or scrotal duplication has been considered as a very rare abnormality.[1] Duplication of penis (also known as diphallia or diphallasparatus) is also an extremely uncommon genital anomaly with frequency of 1 case per 5 million births.[2] Androgen receptor (AR) signaling plays a crucial role in the development of male-specific phenotypes in the period of embryogenesis, spermatogenesis, sexual behavior, and fertility in adulthood.[3] The exact causative factors for these anomalies are unknown. It has been considered that normal differentiation can be interfered with the teratogenic agents. Genital duplication may be part of a qaudal duplication syndrome to describe the association between genitourinary, gastrointestinal, and neural tube defects. The variability in the degree of duplication with additional anomalies results in diverse surgical approaches for each patient.

Here we report a case of pseudoduplication of external genitalia with proved negative ARs in the accessory genitalia.

CASE REPORT

The patient was a 34-year-old woman; gravida 5 with no previous history of medical diseases who was admitted to the pediatric urology clinic on April 2019. The second trimester fetal anomaly scan was performed which was completely normal except 2 external genitalia on the midsagittal plane in ultrasound scan (Fig. 1). Amniocentesis was performed aseptically under the guidance of ultrasonography and karyotype study showed normal 46, XY male karyotype.

The mother was reassured and underwent routine antenatal care for the rest of pregnancy. A full term male child weighing 3.2 kg with APGAR 8/10-9/10 was delivered by cesarean section. The infant was first examined when he was 2 months old. He was a healthy boy who had normal male genitalia and secondary dorsal external genitalia on his perineum with normal anus (Fig. 2).

The dorsal penis was shorter than the ventral one, with no urethral meatus on the dorsal penis. The accessory scrotal sac was soft and empty on palpation. He started voiding per ventral penis which had a urethra and meatus. No erections of the dorsal penis was demonstrated which was documented by parent reports. An ultrasound of the abdomen and pelvis was conducted which revealed single urinary bladder, apparently normal intra-abdominal organs, and the empty secondary dorsal scrotum. There was no other apparent congenital anomaly. There was no evidence of vesicoureteral reflux as well.

The infant underwent the surgery at 4 months of age. The parents were fully informed and consent was obtained. The dorsal external genitalia were excised under general anesthesia (Fig. 2) and circumcision of the ventral penis was also done at the same time. The baby was stabilized and discharged the next day with uneventful postoperative course.

Sections of scrotal specimen revealed fibroadipose tissue with no testicular tissue. Pathologic examination on the sections of penile-like structure demonstrated delicate small vessels and sinuses as well as fibroelastic tissue, smooth muscle, and neural structures resembling corpora cavernosum, but it had no tunica albuginea or urethral structure. Immunohistochemical staining of penile and scrotal tissues was negative for ARs in the presence of a positive control of seminal vesicle tissue (clone AR441, Diagostic Biosystems, CA) (Fig. 3). The result of voiding cystourethrography was normal during long-term follow-up. The appearance of the perineal region was satisfactory.

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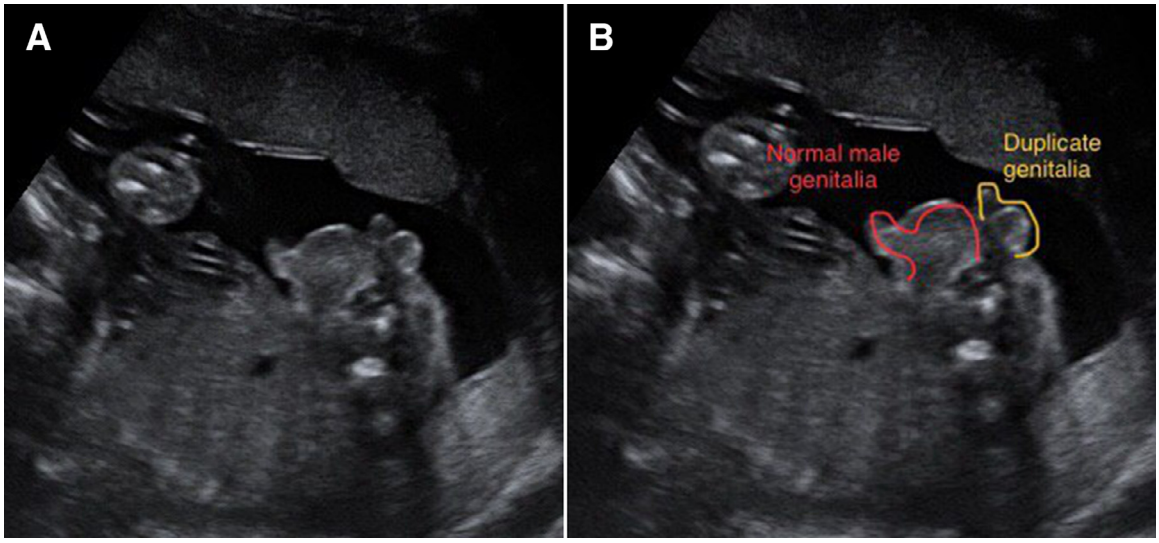

Figure 1. (**A-B**) Ultrasonography at 23 weeks of gestation shows male fetus with duplication of external genitalia. (Color version available online.)

DISCUSSION

Penile duplication is a very rare but a well-documented anomaly occurring once in 5-6 million births, which may be accompanied with wide range of congenital anomalies such as duplication of several parts of lower urinary tract. This maldevelopment is classified into 3 types including bifid diphallia, duplication of glans alone, and complete diphallia.[4] Pseudodiphallia, is another type in which a rudimentary atrophic penis exists independently of the normal penis. Accessory scrotum, a rare form of congenital anomaly, is often associated with genitourinary and gastrointestinal anomalies with few cases reported to date. This anomaly is characterized by ectopic scrotal tissue in the existence of a normal, orthotopic scrotum.[5] Combination of these 2 rare events has been seen in only a few case reports.[6,7] In patients with diphallia, the urethra shows a range of variations from functioning double urethras to complete absence of the urethra in each penis. In our case, histologic examination of the resected phallus did not demonstrate any urethral tissue.

Contrary to the current study, there is only one report in which it was demonstrated that the resected accessory

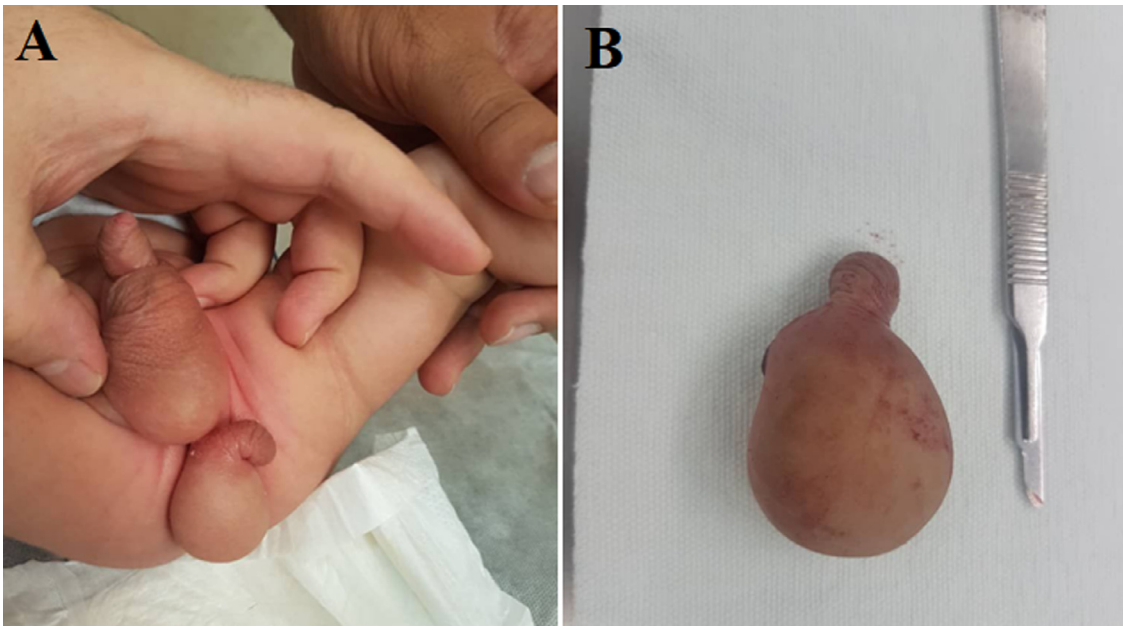

Figure 2. (**A**) Secondary smaller genitalia dorsal to the normal genitalia (**B**) The excised specimen. (Color version available online.)

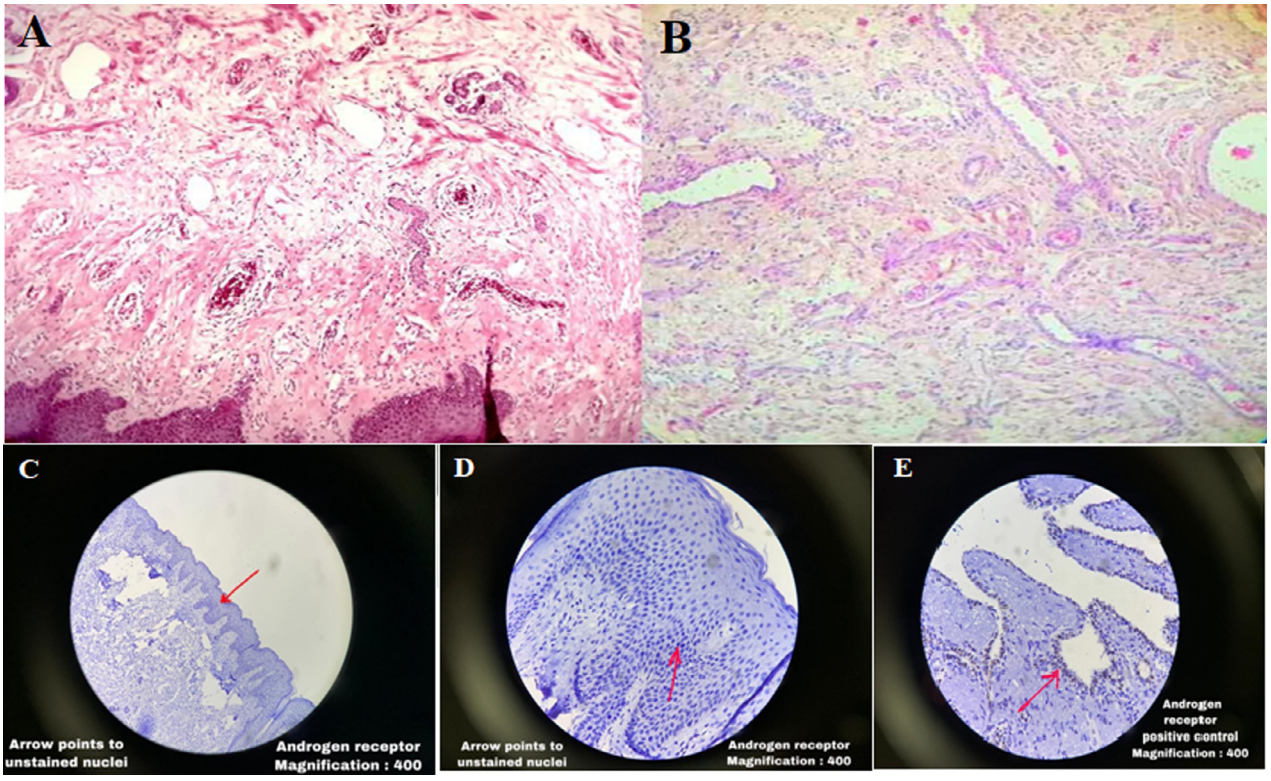

Figure 3. (**A**) Section of accessory scrotal tissue: fibroadipose tissue and smooth muscle bundles, (**B**) Histologic staining of pseudo phallus section: delicate small vessels, sinuses, fibroelastic tissue, smooth muscle, and nerve bundles (×100), (**C-D**) Negative result of immunostaining in accessory scrotum and pseudophallus for androgen receptor (×40) arrow points to unstained nuclei, (**E**) positive result of immunostaining in control seminal vesicle tissue (×40) arrow points to stained nuclei. (Color version available online.)

scrotal specimen was positive for ARs,[8] and the embryologic meaning of this finding is unclear. This finding in our patient may show lack of potential for future growth of this accessory organ after puberty, if not removed.

Although there is advancement in prenatal screening techniques, most cases of congenital perineal mass have been identified postnatally. Prenatal diagnosis of these anomalies has been reported very rarely. However, any genital malformation makes challenges in terms of medical, ethical, and aesthetic decision making. The differential diagnosis of a fetal perineal mass includes lipoma, lipoblastoma, infantile hemangioma, hamartoma, and choristoma. If a fetal perineal mass is detected during the antenatal period, it is of great importance to look for any associated congenital anomalies by using radiologic methods which help to identify the extent of the problem. The treatment of complete duplication of the urogenital system should be individualized. Separation of the urogenital and gastrointestinal tract as well as preservation of the continence and reconstruction of the external genital with satisfactory cosmetic results, are among the ultimate goals of surgical intervention.

The term pseudoduplication of external genitalia was first used in 2001 to describe this anomaly.[7] We have noticed that in some case reports the terms pseudodiphallia and accessory scrotum have been used alone to describe patients which are very similar to our case.[9,10] This seems to be an incomplete description of the anomaly.

CONCLUSION

In this study we present a rare case with pseudoduplication of external genitalia, absence of urethra, lack of explicit erectile tissue in the dorsal penis and more importantly, absence of ARs in the dorsal penile and accessory scrotal tissues. Surgical excision is the technique of choice for these patients.

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