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Accessory Scrotum with Bifid Scrotum and Hypospadias

Bifid Skrotum ve Hipospadiasın Eşlik Ettiği Aksesuar Skrotum

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ABSTRACT

Congenital abnormalities of the scrotum are extremely rare, and sometimes associated with other genitourinary tract abnormalities. In this report we presented a male neonate with multiple perineal anomalies including accessory scrotum, bifid scrotum, and hypospadias.

Key Words: Hypospadias, Male genitalia, Urogenital abnormality

ÖZET

Skrotumun doğumsal anomalileri bazen diğer genitoüriner sistem anomalileri ile birliktelik gösterir ve oldukça nadir görülür. Bu yazıda bifid skrotum, hipospadias ve aksesuar skrotumun bir arada olduğu çoklu perine anomalisi olan erkek bir yenidoğan olgusu sunuldu.

Anahtar Sözcükler: Hipospadias, Erkek genital, Ürogenital anomaliler

INTRODUCTION

Congenital anomalies of the scrotum are not common and include bifid scrotum, penoscrotal transposition, ectopic scrotum and accessory scrotum (AS) (1,2). The latter two, and especially accessory scrotum (AS), are very rare (2). Until now, approximately 35 cases of AS have been reported in the literature (3,4). Half of these cases were associated with other urogenital anomalies such as bifid scrotum and diphallia, hypospadias, Backer's nevus, ectopic scrotum and VATER association (5-7). Accessory perineal scrotum has been classified into two types, depending on its location: the mid-perineum type and the lateral type (1). Here we present an extremely rare phenotype of AS with bifid scrotum and penoscrotal hypospadias, of which only few cases with similar combination of anomalies have been reported in the English literature. Our aim on reporting this rare case was to signalize the importance of screening for additional anomalies and planning for appropriate surgical intervention in such cases.

CASE REPORT

A two-day-old boy was admitted to our Neonatal Unit for evaluation of perineal anomalies. He was the product of a normal vaginal delivery at 40 weeks of gestation with uneventful prenatal period and his birth weight was 1960 g (under 10 percentile) and length 47 cm (10-50 percentile) (disproportional small for gestational age). He was the product of healthy non-consanguineous parents. Neither notable family history nor exposures to teratogens, alcohol, or drugs were noted. During pregnancy his mother did not have routine follow-up examinations. His general condition was well, and vital signs were normal. He had perineal abnormalities. The scrotum was bifid in the normal position, and bilateral testes were fully descended in the well-developed scrotum (Figure 1A). In addition penoscrotal hypospadias and four blind dimples on the ventral side of the penis were present (Figure 1B). An approximately 2.5x2 cm in size soft, non-tender, mobile swelling was present posterior to the left side of the scrotum, up to the normally located anus and slightly left to the midline (Figure 1C). The overlying skin was rugated. It did not contain any testicular tissue. It was compatible with AS. No additional urogenital and anorectal malformations were detected by physical examination. The anus was normal in position and calibre. Urinary tract, cranial and abdominal ultrasonographic examinations were normal. Skeletal direct radiographs revealed no anomalies. Chromosome analysis revealed normal male

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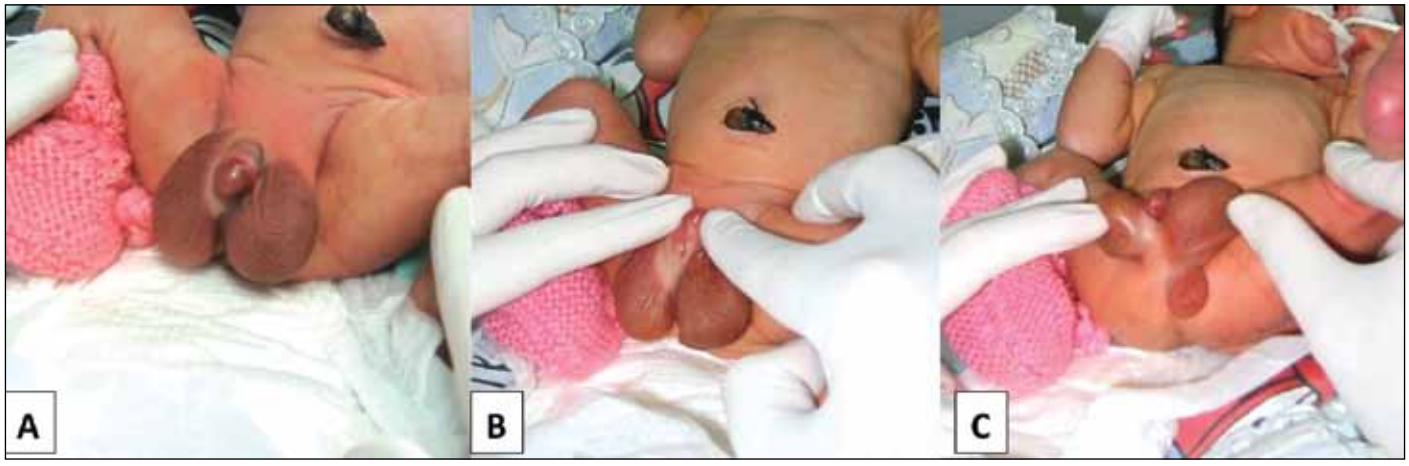

Figure 1: A) Bifid scrotum, **B)** Penoscrotal hypospadias, **C)** Accesory scrotum.

karyotype. Surgical excision of the AS was planned when the patient reaches an appropriate weight but the family did not bring back the child for surgery. The reason of this condition was thought to be the low socio-cultural structure of the family.

DISCUSSION

Accessory scrotum is an extremely rare condition defined as the occurrence of scrotal skin outside of its proper location with no testicular tissue. The presented infant had an unusual phenotype of accessory scrotum, in that both bifid scrotum and penoscrotal hypospadias coexisted in one patient. To our knowledge having all these infrequent scrotal malformations together is very unusual. In the English literature, we found only few cases similar to our case except that they had an associated anorectalanomaly (5,8,9). Similar to in our case, the AS always arises between the external genitalia and anus on the midline or laterally to it (9).

Scrotal development is closely related to penile development. At the fourth week of gestation, the phallus develops abruptly and forms the penis, while at the same time, genital swellings appear at both sides of the inguinal region and gradually move to the posterior site and form the labioscrotal swellings at 10-12 weeks gestation. This swelling migrates further to the caudal area of the penile site and forms the right and left parts of the scrotum (1,2). Although the etiology of scrotal anomalies remains unclear, the most accepted etiopathogenetic hypothesis for this condition is that of an abnormal migration of labioscrotal swelling mainly due to distention of an associated lipoma or compression by the heel in utero (1). Takayasu et al. (10) have hypothesized that a mid-perineum AS may result from a triple primordial anlage of the labioscrotal swelling or from a teratoid structure. Lamm et al. (11) suggested that the lateral type of AS, as in our case, might represent a duplication or division of the ipsilateral labioscrotal swelling. The anomaly represented in this case report, is a single one with no accompanying multiple organ malformations. The causative factor for this anomaly did not have any adverse effect on other systems differentiating at the same gestational period (12).

In the literature, none of the patients' mothers with AS had complicated pregnancies or history of harmful gestational exposure that might have predisposed to the development of such anomalies like our patients' mother (3,4,9).

Sule et al. suggested that it was useful to classify accessory labioscrotal folds by the presence or absence of an associated perineal lipoma, since those without a perineal lipoma might be associated with other genitourinary and anorectal anomalies that require diagnostic imaging (1). In our case, associated malformations in the external genitalia such as bifid scrotum and hypospadias were seen; however, other genitourinary and anorectal anomalies were not detected.

AS is treated with complete surgical excision. Histologic examination always shows the smooth muscle and stromal elements of normal scrotal tissue (8). The other urogenital anomalies would require additional corrective surgery. Surgical excision of the AS and corrective surgical interventions are planned for our patient. Eventually, these operations were planned for the near future, as they are not urgent. However, the family did not bring the patient back for operation. The planned surgery was a two-staged intervention; hypospadias repair followed by correction of bifid scrotum and excision of accessory scrotum. The patient had a penoscrotal hypospadias and the accessory scrotum may be preserved as a substitute for tissue replacement. This was the reason for the two-staged surgical intervention.

In conclusion, surgical planning should be performed after screening for other congenital anomalies in a patient with such a perineal anomaly.

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