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Congenital Megaprepuce

Konjenital Megaprepisyum

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ABSTRACT

Congenital megaprepuce is a rare condition leading to micturition problems with excessive ballooning in the postnatal period with unknown etiology. Congenital adrenal hyperplasia, cystic and tumoral diseases should be considered in the differential diagnosis of congenital megaprepuce. A 2-month-old male patient was admitted to our hospital with penile anomaly. Phimosis, penile oedema and megaprepuce were noticed on physical examination. Elective surgery was recommended by the pediatric urology department. We present an isolated congenital megaprepuce case that underwent successful reconstructive surgery.

Key Words: Congenital abnormalities, Foreskin, Penis, Surgery

ÖZET

Konjenital megaprepisyum doğum sonrası dönemde aşırı balonlaşma şeklinde işeme problemlerine neden olabilen, nedeni bilinmeyen, çok nadir görülen bir durumdur. Konjenital megaprepisyumun ayrıcı tanısında konjenital adrenal hiperplazi, kistik veya tümoral hastalıklar düşünülmelidir. İki aylık erkek hasta penis anomalisi şikayeti ile başvurdu. Fizik muayenesinde fimozis, penil ödem ve megaprepisyum tespit edildi. Pediatrik Üroloji bölümü tarafından elektif operasyon önerildi. Operayon sonrası klinik düzelme sağlanan izole konjenital megaprepisyumlu bir olgu sunuldu.

Anahtar Sözcükler: Konjenital anomali, Sünnet derisi, Penis, Cerrahi

INTRODUCTION

A non-retractable prepuce is commonly seen during infancy. Congenital Megaprepuce (CMP) is a rare condition of unknown etiology, and may cause micturition problems due to ballooning in the following months. Megaprepuce refers to a severely redundant inner foreskin covering a normal glans penis (1-5).

We present an isolated congenital megaprepuce case that underwent successful reconstructive surgery.

CASE REPORT

A 2-month-old male patient was admitted to our hospital with a penile anomaly that was noticed after birth. In the prenatal history, the mother did not have any chronic disease or drug usage. His birth weight was 3300 grams. He had no complaints except the penile anomaly. The parents had a second-degree consanguinity. On physical examination, the patient had a giant prepucial sac that measured 7.1 cm, a penis girth of 1,5 cm, and phimosis, hydrocele, and penile odema (Figure 1). The skin over the scrotum had thickened. Laboratory studies revealed a normal complete blood count, urogram, and kidney and liver function tests. Endocrine evaluation showed that normal cortisol, DHEA-S, testosterone, androstenedion and 17-OH progesterone levels. Abdomino-pelvic ultrasonography (USG) and retrograde cystouretrographyperformed to diagnose other congenital abnormalities were all normal. The patient was examined by a pediatric urologist that led to diagnoses of penile odema, phimosis, and megaprepuce being made and elective surgery being recommended. Surgery including correction of megaprepuce, degloving of the penis, and excision of the redundant skin was performed and findings were normal after surgery (Figure 2).

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Figure 1: Preoperative appearance of the genitalia.

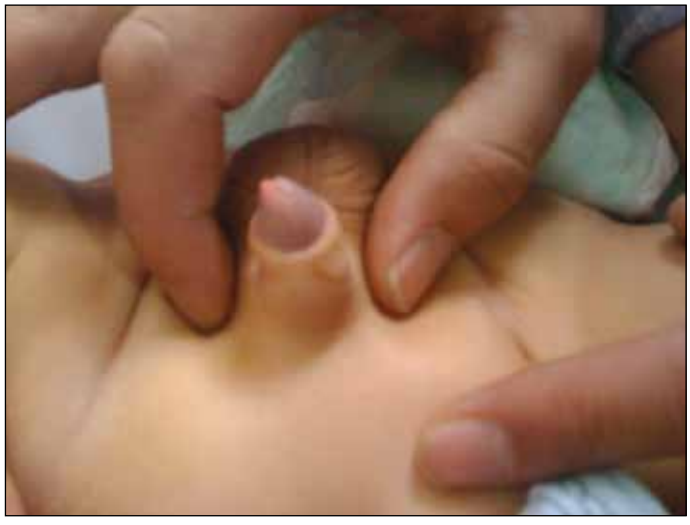

Figure 2: Postoperative appearance of the genitalia.

DISCUSSION

The prepuce is a tissue that differentiates from the genital tubercule during intrauterine growth. It starts to form as the epidermis beginning from the collus glandis, the conjunction point of the penile shaft and glans, extends distally on the 8th week of the embryological growth and completes its development after it fuses with the glans. The prepuce and penis need androgens and androgen receptors to be able to grow. The ventral part of the prepuce completes its development after the closure of the glandular urethra and completely covers the glans. During this period, the glans epithelium and the epithelium that covers the inner side of the prepuce are fused together. This fusion protects glans from chemical effects of the urine and external traumas. This fusion is subject to lysis and the prepuce becomes retractable behind the glans between 6 months and 14 years (2,3,6).

Congenital abnormalities of the prepuce are sporadic except hypospadias and epispadias. In CMP, the penis shaft is morphologically buried in the prepuce (2,5,6). CMP was first described by O'Brien et al in 1994 as a megaprepuce case that covered the penile shaft and glans and thus led to an appearance of being completely buried (1). Cascio et al reported 4 infants who had vesicoureteral reflux (VUR) at various degrees but none of the cases had other endocrine or morphological abnormalities (7). Although CMP may be isolated, cases should be investigated for any other abnormalities. Co-existence of CMP and ballooned phimosis has been reported in many cases (1,3,8). We performed voiding cystouretrography, abdominal USG, and endocrine evaluation and found no other abnormalities in our case. Summerton (4) and Philip (9) reported that the possibility of infections is higher in the cases with CMP who undergo surgery later after the diagnosis. Our case underwent surgery a short time after the diagnosis and did not have any preoperative infection.

CMP is a significant cause of anxiety for families, and early diagnosis and treatment of CMP are very important (10,11). No complication during and after early surgery has been reported (6,8). Similarly, our patient was successfully operated and no complications were observed during and after the operation. On his follow-up examination 2 months after the surgery, he remained in good health with no complications.

Although the phrenulum and glans were completely covered by the prepuce, the prepucial sac was measured 7.1 cm in our case with macrogenitosomia, as mostly seen in patients with congenital adrenal hyperplasia. No hyperpigmentation was present on the genital area.

In conclusion, CMP is a rare anomaly of penile tissue and cases with CMP should be investigated for possible accompanying abnormalities and endocrine diseases. Early diagnosis and appropriate surgical treatment are important issues in this patient group as urinary tract infections, obstructions, renal pathologies, and especially cosmetic problems may be bothersome for the patient and the family otherwise.

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