Anteroposterior Duplicated Exstrophy: A Case Report

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Duplicated bladder exstrophy is an extremely rare variant of the exstrophy/epispadias complex. Duplicated exstrophy defines an exstrophic mucosal plate in hypogastric area with a normal closed bladder. We present a unique case of an anteroposterior duplicated exstrophy in a female newborn. UROLOGY 131: 220–222, 2019. © 2019 Elsevier Inc.

Variants of the bladder exstrophy are very rare birth defects. Bladder exstrophy incidence is approximately 1 per 50,000 live births, while the variant presentations are seen approximately 10 times less frequent. Even though there are various nomenclatures in the literature, these highly rare exstrophy variant presentations are generally categorized as pseudoxestrophy, superior vesical fissure, duplicated exstrophy, and covered exstrophy. We present a unique case of variant exstrophy with a final diagnosis of duplicated exstrophy.

CASE REPORT

A 45-day-old girl was referred to our institution because of an infraumbilical patch of dry exstrophic mucosa. She was born to 26-year-old mother by normal delivery with a gestational age of 40 weeks. There was no history of maternal drug use and radiation exposure during the pregnancy. Physical examination revealed a 2.5 × 2 cm patch of exstrophic mucosa between the umbilicus and symphysis (Fig. 1). The appearance of clitoris, vaginal introitus, urethral meatus, labia minora and majora and anus were normal on external genitalia examination. The umbilicus was involved within the skin lesion. The exstrophic mucosa did not have any ureteral opening, and also the rectus muscles were separated. The girl was continent, had normal urinary tract on abdominal ultrasonography and voiding cystourethrography. There was the diastasis of symphysis pubis (Fig. 2). The patient underwent surgery on the 52 day of her life. Initially, the cystoscopy and vaginoscopy were performed and normal urethra, bladder neck and ureteral orifices were seen on cystoscopy, and normal vagina and cervix were seen on vaginoscopy. Then the exstrophic mucosa and the umbilicus were excised, the rectus fascia and muscles were defective in the midline, and the normal bladder was seen under the exstrophic plate. Level of fascial defect was not extending over umbilical level and there was not an umbilical hernia. No bowel segment was associated with the exstrophy. The rectus structures were approximated with interrupted absorbable sutures. There was no intraoperative complication. Postoperative period was uneventful. Histopathologic examination revealed stratified squamous epithelium, transitional epithelium, and muscle layer (Fig. 3). Physical examination at 7 months of age was normal and there were no symptoms of urinary tract or other organ systems. Urinary ultrasonography and urinalysis were normal.

DISCUSSION

Variants of the bladder exstrophy are very rare, it is seen approximately 1/500,000 live births. Exstrophy anomalies are thought to result from incomplete closure of the infraumbilical abdominal wall. Separations of the pubic rami and low-set umbilicus with or without epispadias have been suggested to result from the abnormal persistence of the cloacal membrane. The variants exstrophy anomalies are thought to result from incomplete rupture or the abnormal persistence of the cloacal membrane. According to the literature, bladder exstrophy is usually seen in boys; however the variant presentations are mostly seen in girls. Our patient is similar to the literature regarding the association of gender.

In a child with these aforementioned findings as in our case, differential diagnosis should be done amongst several very rare pathologies. These exstrophy variants are:

Pseudoxestrophy refers to a characteristic musculoskeletal defect of the exstrophy anomaly with no major defect in the urinary tract. Major findings are elongated, low-set umbilicus and divergent rectus muscles those attach to the separated pubic bones. In our case, there was an ectopic dry mucosal tissue below the umbilicus which was excluding pseudoxestrophy diagnosis.
Superior vesical fissure defines a congenital anomaly that the upper bladder is open near the umbilicus in association with musculoskeletal defects. The absence of any vesical fissure appearance was negating this diagnosis.

Covered extrophy (also called as split symphysis) variant is the presence of musculoskeletal defect associated with classical extrophy. The urinary tract is normal, and bladder is covered by a translucent membrane of skin without muscle or a fascial layer. In our patient, there was no translucent membrane, the skin had a dry appearance that was giving the impression of keratinized mucosa. Therefore, our diagnosis was not compatible with covered extrophy.

Duplicated extrophy describes an extrophic mucosal plate in hypogastric area with a normal closed bladder and diastasis of the symphysis pubis. There are 2 different forms of bladder duplication: side-by-side and antero-posterior. Side-by-side duplicated cases have 2 separately formed bladder halves in a left-right orientation with a muscle containing midline septum between the bladders. Each bladder has its own ureter and intact sphincter. On the other hand, anteroposterior duplicated extrophy is characterized with a patch of everted bladder mucosa on the anterior abdominal wall and a second bladder lying in the pelvis. The ureters attach to this posterior closed bladder, rendering the superficial mucosa dry. In the presented case, the appearance at the first look was like a bladder extrophy, however as the detailed inspection revealed no ureteral orifice or urine. The mucosa was dry and the fascial defect could be palpated. The ultrasonography and voiding cystourethrography revealed normal urinary tract and diastasis of symphysis pubis. Intraoperative cystoscopy confirmed normal bladder neck and external genitalia. During the operation, we detected defective rectus fascia and muscles anteriorly and a normal bladder was located beneath these defective structures. Histopathologic examination of excised skin...
lesion revealed the presence of stratified squamous epithelium, transitional epithelium, and muscle layer (Fig. 3). All these findings let us to make the diagnosis of anteroposterior duplicated exstrophy. The excision of the ectopic mucosa and closure of the abdominal wall defect was sufficient.

CONCLUSION

The bladder exstrophy is a rare congenital abnormality that the incidence of variants is distinctively more rare. Preoperative imaging techniques and interpretation of physical examination findings are important to make differential diagnosis. We presented a case of anteroposterior duplicated bladder exstrophy in a newborn girl. Despite, being a very rare entity, the abnormality is easily correctable and has an excellent prognosis when compared to the classical exstrophy patients.

References