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True Duplicate Bladder Exstrophy

Abstract

A three months male child was admitted in our department as a rare variant of bladder exstrophy (true duplicate bladder exstrophy). There are only 8 cases of this variant reported in the literature so far, we report an additional case to add the scarce literature.

Keywords: True duplicate exstrophy; Bladder exstrophy; Exstrophy variants

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Introduction

Bladder exstrophy is a rare congenital anomaly and classical bladder exstrophy is the most common. Rare variants of exstrophy bladder constitutes only 10% of cases and duplicate exstrophy is one of the rarest variant of which only 24 cases have been reported [1]. Patients present with patch of exstrophic bladder just below the umbilicus with either a normal or smaller than normal complete bladder underneath and diastasis of symphysis pubis. The duplicated extroverted patch of bladder mucosa is dry and does not receive ureters. True duplicate exstrophy is very rare variant of bladder exstrophy and only 8 cases have been reported, we present another case to add to the literature.

Case Report

Three months full term male child was admitted in our department as a case of bladder exstrophy. On examination, dry bladder mucosa was exposed just below the umbilicus with separation of rectus. Penile length was normal with orthotopic meatus but poorly developed corporal bodies (**Figure 1**). Child was passing urine normally. Both the testicles were palpable in scrotum and there was no other anomaly present. Widening of symphysis pubis was seen on X-ray. All baseline investigations were normal.

Dry exstrophied bladder mucosa was excised and the abdominal wall defect was closed (**Figure 2**). Postoperative period was uneventful and baby was discharged on third postoperative day. HPE revealed transitional cell lining of the respected specimen.

Discussion

Marshall and Muecke defined the true bladder duplicate exstrophy as a suprapubic exstrophic mucosal plate with subjacent normal

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or near normal bladder with relatively normal phallus. Epispadias is absent and exstrophic plate does not receive ureters [2]. Embryologically if the superior vesical fissure fuses, an overfilling bladder mucosal plate presents as duplication of bladder [3].

Duplication exstrophy is of two types, the first is suprapubic exstrophic mucosal plate and a covered bladder as in our case. The second type is associated with classical findings of exstrophy complex [4]. The embryological explanation of the numerous variations in the exstrophy complex suggests that the urogenital part of the cloacal membrane occupies cranial position to the genital primordial. During development the cloacal membrane prevents in growth of mesoderm towards the midline and preventing its insertion between the ectodermal and endodermal layers. Thus, number of variants of exstrophy complex can develop. Bladder duplication may have associated duplication of ureters, vagina, bifid clitoris, caecum, double appendix, ARM, and spinal dysraphism [5].

The operative procedure is simple in such cases. In our case, excision of dry exstrophied bladder mucosa with closure of abdominal wall defect was done. Postoperative period was uneventful and patient is doing well in follow up. Our case was identical to cases reported by other authors [2,3,6].



with separation of rectus.



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