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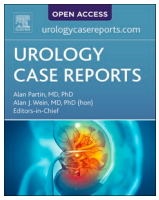

Pediatrics

Duplicate bladder exstrophy in a female infant: A case report

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ABSTRACT

Duplicate bladder exstrophy, a rare variant of the bladder exstrophy-epispadias complex, was diagnosed in a 5-month-old female infant presenting with a 4 × 4 cm lower abdominal mass. Physical examination revealed absent umbilical scar, wide symphysis pubis, and anteriorly displaced genitalia and anus. Imaging ruled out classic bladder exstrophy and vesicoureteral reflux. Surgical exploration confirmed a suprapubic exstrophic mucosal plaque without communication to a functional bladder. Management involved excision of the exstrophic plaque and primary abdominal wall closure. Post-operative course was normal intravenous pyelogram findings. This case emphasizes the importance of accurate diagnosis and tailored surgical approach for rare bladder exstrophy variants.

1. Introduction

Bladder exstrophy is a rare and complex congenital malformation affecting approximately 1 in 50,000 live births.[1] It is characterized by the failure of the anterior abdominal wall and bladder to close properly during embryonic development, resulting in an exposed bladder and associated anomalies of the genitourinary system and pelvic structures.[2] While classic bladder exstrophy presents with an open bladder visible on the abdominal surface, variants of this condition can manifest with varying degrees of severity and anatomical involvement.[3]

This case report describes an unusual presentation of a variant form of bladder exstrophy in a 5-month-old female infant. The case is particularly noteworthy due to its atypical presentation, with a closed but extrophied bladder segment, and the successful surgical management that preserved normal bladder function.

The manuscript was prepared following the CARE guidelines (https://www.carestatement.org).

2. Case report

A 5-month-old female infant, weighing 5 kg, presented with a lower abdominal mass present since birth. The mass, located in the suprapubic area, measured approximately 4 × 4 cm, with a red, moist appearance and normal surrounding skin.Upon applying pressure, it partially reduced, resembling a small cup before protruding again. Physical examination revealed an absent umbilical scar, wide symphysis pubis, and anterior displacement of the genital area and anal opening(Fig. 1). To rule out bladder exstrophy, a Foley catheter was inserted, and the bladder was filled with serum, showing no extravasation from the mass. An abdominal ultrasound was reported as normal. The patient was admitted and underwent surgery. The surgical procedure involved a lower midline incision from the expected umbilical area, extended around the mass (Fig. 2A). Dissection revealed the mass's closest attachment to the urinary bladder in the coronal plane, anterior to the bladder, with adhesions present up to the bladder neck (Fig. 2B and C). Careful dissection was performed without opening the bladder, which appeared normal in size with thin muscle at the attachment site. After removing the extrophied bladder tissue (Fig. 3), the abdomen was closed layer by layer, supported by fascia under tension due to symphysis pubis dehiscence.Post-operatively, the patient remained in the hospital for 3 days. A post-operative intravenous pyelogram (IVP) was performed, with results reported as normal. An orthopedic consultation was advised for management of the pelvic abnormality.

3. Discussion

Bladder exstrophy is a rare and complex congenital anomaly, occurring in approximately 1 in 30,000 to 50,000 live births.[4] This

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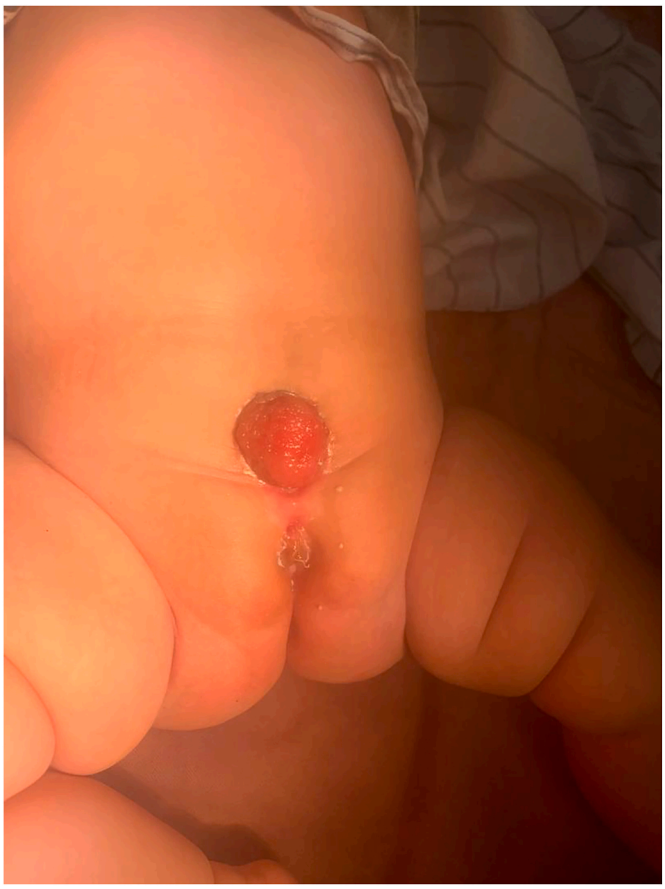


Fig. 1. Preoperative view of duplicate exstrophy. This image shows the patient's condition before surgical intervention, highlighting the duplicate exstrophy of the bladder.

condition demonstrates a notable male predominance, with male to female ratios ranging from 1.5:1 to 5:1. The risk of having a sibling with bladder exstrophy is about 1 %, indicating a potential genetic component.[5]

The etiology of bladder exstrophy is believed to stem from incomplete development of the infraumbilical portion of the anterior abdominal wall and the anterior bladder wall. This developmental anomaly is thought to result from delayed rupture of the cloacal membrane. The persistence of this membrane prevents normal medial mesenchymal ingrowth, causing the abdominal wall to remain lateral and exposing the posterior bladder wall to the external surface.[6]

Exstrophy variants, while rare, have been classified into four main subgroups based on their anatomical and embryological characteristics. These include **Superior Vesical Fissure**, which involves a minimal bladder eventration; Pseudoexstrophy, characterized by musculoskeletal defects without major bladder defects; **Duplicate Exstrophy**, where remnants of exstrophic bladder mucosa exist but the urinary tract remains intact; and **Superior Vesical Fistula,** which features a small communication between the bladder and the exterior.[7] A fifth subgroup, **Covered Exstrophy with Visceral Sequestration**, has also been identified, presenting additional complexities.[8]

Duplicate exstrophy, an extremely rare variant, has only few cases reported in the English literature.[9] Its embryological explanation suggests it may result from a superior vesical fissure that occurs but later fuses, leaving portions of bladder tissue outside while an intact bladder forms underneath the skin. This explains its rarity compared to superior vesical fissure.[10]

Duplicate exstrophy is divided into two groups: true duplication with classic findings of the exstrophy complex, and suprapubic exstrophic mucosal plaque with no communication to a covered normal functional bladder.[11] The latter group typically presents with well-formed external genitalia, urinary continence, and normal voiding pattern[12] same as our case.

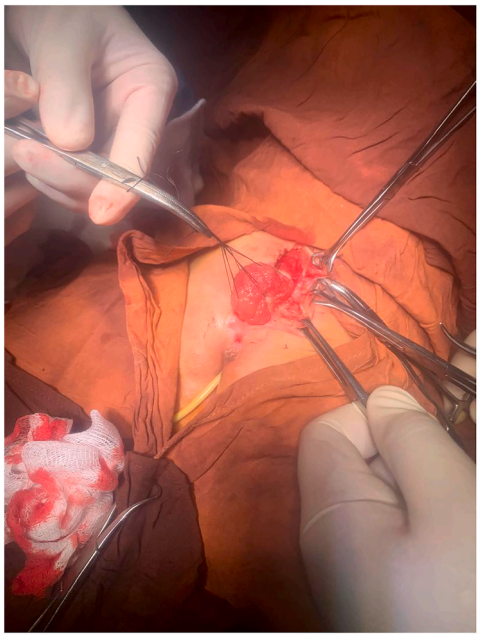
Surgical reconstruction for the second group involves excision of the exstrophic plaque and primary aesthetic closure of the abdominal wall defect. While immediate post-operative healing is often uncomplicated, long-term follow-up and possible scar revision are important to address potential psychological concerns in adolescence and young adulthood.[13]

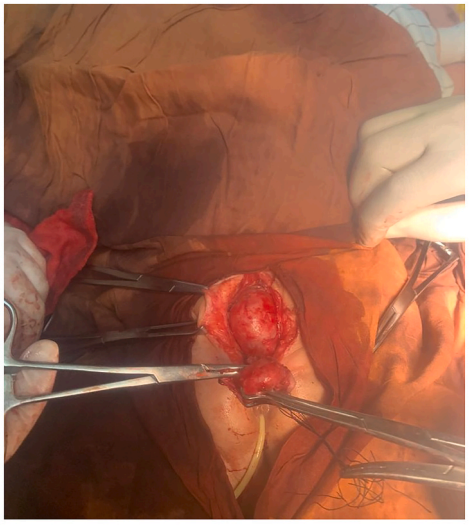
4. Conclusion

In conclusion, bladder exstrophy represents a significant challenge in pediatric urology and reconstructive surgery. Its successful management requires early diagnosis, careful surgical planning, and lifelong follow-up. Ongoing research into the genetic and embryological basis of this condition, as well as refinement of surgical techniques, continues to improve outcomes for affected individuals. The complex nature of bladder exstrophy underscores the importance of specialized, multidisciplinary care in centers experienced with this rare congenital anomaly.

CRediT authorship contribution statement

Yalda Obaidy: Writing – review & editing, Writing – original draft, Methodology, Conceptualization. **Ajmal Sherzad:** Writing – review & editing, Writing – original draft, Conceptualization. **Dunya Moghul:** Writing – review & editing, Writing – original draft, Methodology, Conceptualization.





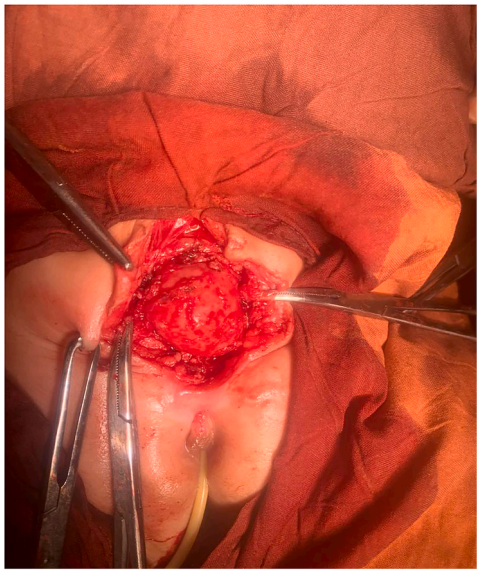


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Fig. 2. Surgical management of duplicate exstrophy.
(A) Dissection of duplicate exstrophy from the true bladder. This image demonstrates the careful separation of the duplicate exstrophy tissue from the normal bladder wall.
(B) Excision of duplicate bladder exstrophy from the neck of the actual bladder. This image demonstrates the surgical removal of the abnormal tissue from the junction with the normal bladder.
(C) Intact bladder wall after complete resection of duplicate exstrophy. This image shows the remaining healthy bladder tissue following the successful removal of the duplicate exstrophy.

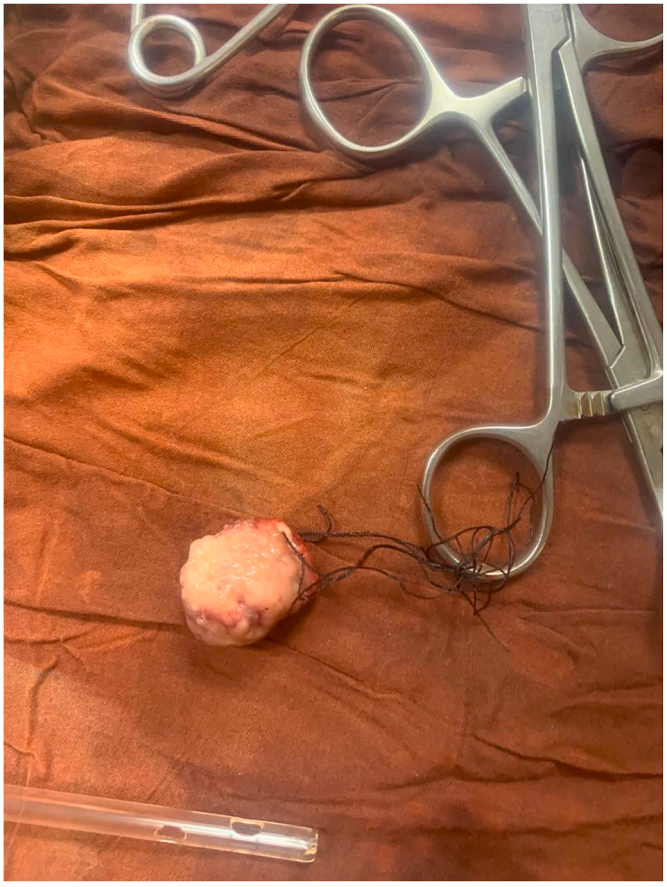

Fig. 3. Resected duplicate bladder exstrophy specimen. This image displays the excised duplicate exstrophy tissue after its complete removal from the patient.

Informed consent

Informed consent was obtained from the patient or guardian.

Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Declaration of interests

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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