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Isolated bladder exstrophy with normal phallus and imperforated anus: "A case report"

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ABSTRACT

This case report presents a rare occurrence of isolated bladder exstrophy with a normal phallus in a one-day-old male newborn. The patient also presented with imperforated anus, congenital heart disease, and a left ectopic kidney. Surgical interventions included cut-back anoplasty, bilateral ureteral reimplantation, and primary bladder closure in a single operation, resulting in a successful outcome without complications. Three months of follow-up showed satisfactory results.

1. Introduction

The Exstrophy-Epispadias Complex (EEC) is a rare congenital malformation of the lower genitourinary tract, with a worldwide incidence of 1 per 46,000 live births.[1] While most cases are sporadic, genetic and environmental factors are implicated.

EEC comprises epispadias, bladder exstrophy (BE), and cloacal exstrophy (CE).[2] The prevailing theory attributes its pathogenesis to the premature rupture of the cloacal membrane due to the failure of mesenchymal migration between the ectodermal and endodermal layers to reinforce it. The timing and degree of premature rupture of the membrane are known to cause varying types of this spectrum, with cloacal exstrophy being the earliest defect and epispadias being a later insult.[3]

Isolated bladder exstrophy with a normal phallus is a particularly rare variant, with limited cases reported in the literature. This case report aims to contribute to the understanding of this rare condition by presenting a detailed analysis of a case encountered in our clinical practice.

2. Case report

A one-day-old male neonate, delivered vaginally without notable perinatal complications, was admitted to our hospital for surgical intervention presenting with a protruding abdominal mass with red discoloration on the inferior part of the abdomen (Fig. 1) and an imperforated anus. During palpation, protruding intestinal contents were identified, exerting pressure posteriorly on the exstrophied bladder. Additionally, the presence of ureteral orifices (Fig. 2) was confirmed by inserting an appropriately sized nasogastric (NG) tube into them. Notably, the penile anatomy appeared normal, devoid of associated epispadias (Fig. 3). On inspection of the perineum, an imperforated anus was confirmed, which was a low variety type with a rectoperineal fistula (Fig. 4).

Diagnostic imaging, including plain abdominopelvic radiography, echocardiography, and ultrasound, disclosed concomitant congenital anomalies, notably an approximately two-centimeter pubic diastasis, a patent ductus arteriosus (PDA), an atrial septal defect (ASD) measuring 3.2 mm, and a left ectopic (pelvic) kidney. Notably, there was no reported familial predisposition to similar malformations.

A comprehensive surgical approach was undertaken, encompassing cut-back anoplasty for the imperforated anus (Fig. 5), bilateral ureteral reimplantation using the Cohen cross-trigonal approach, and primary bladder closure. This was accompanied by the insertion of a 6Fr silicone Foley catheter through the patient's penis for a duration of 21 days. As the patient was a one-day-old newborn, osteotomy was not performed. Instead, the pubic bones were sutured with 1/0 PDS to approximate them, and the patient was immobilized using lower-extremity mummy wraps for 12 days. Following discharge from the hospital, the patient exhibited satisfactory progress during the three-month follow-up, with no postoperative complications or recurrence observed. Notably, normal voiding through his penis was consistently maintained throughout the follow-up period. Additionally, the parents received detailed counseling regarding potential long-term implications, including the risk of

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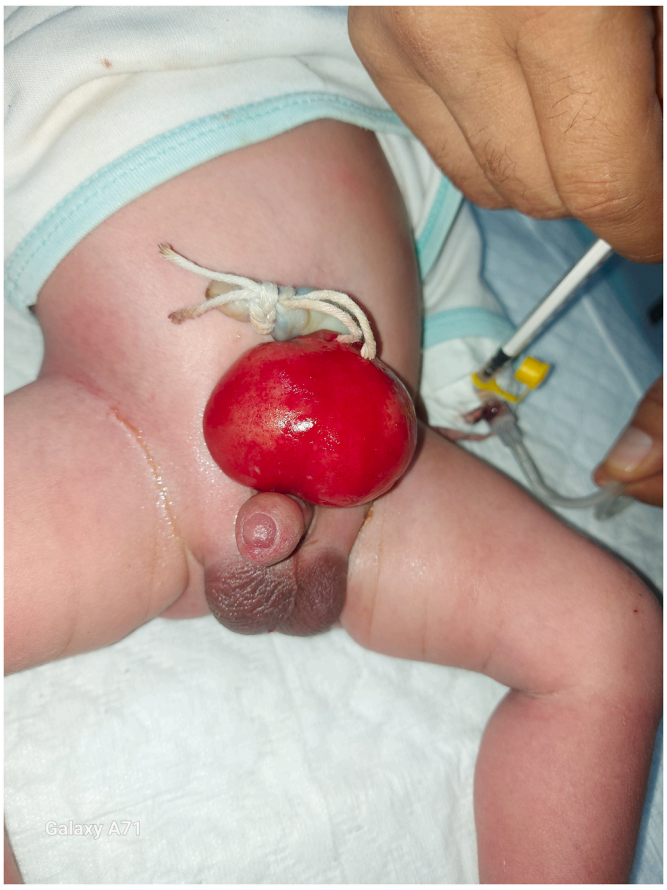

Fig. 1. A protruding abdominal mass with red discoloration on the inferior part of the abdomen. (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

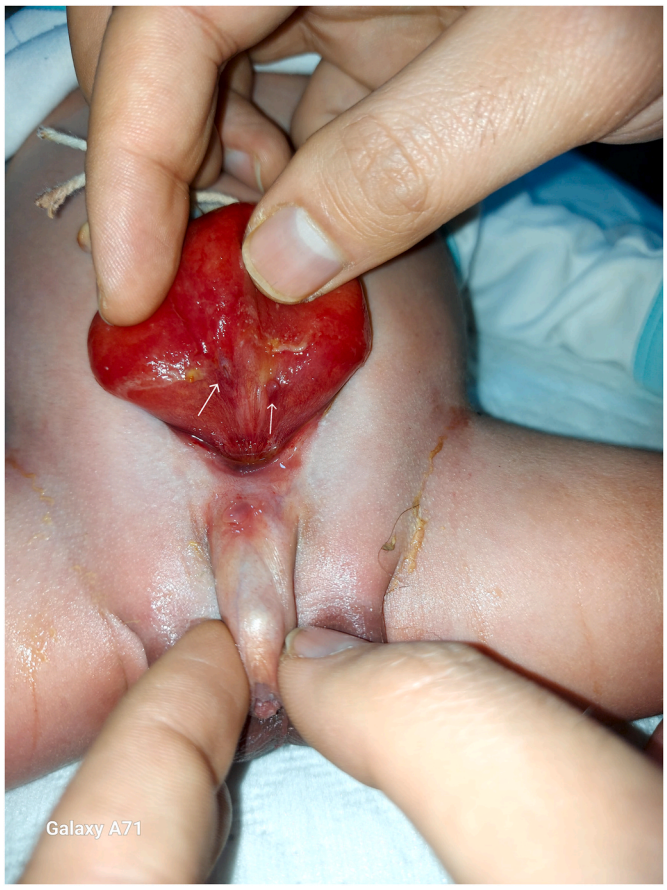

Fig. 2. Ureteral orifices (white arrows).

incontinence and the possibility of future surgical interventions.

3. Discussion

Exstrophy-Epispadias Complex (EEC) presents unique challenges in management due to its rarity and complexity, with isolated bladder exstrophy with a normal phallus being a rare variant. Surgical intervention typically involves a multidisciplinary approach, with careful consideration of associated anomalies and individual patient factors. Among the array of surgical interventions available, notable modalities include Modern Stage Repair of Exstrophy (MRSE), Complete Primary Repair for Classic Bladder Exstrophy (CPRE), and Radical Soft Tissue Mobilization (RSTM)/Kelly Repair.[1]

In this case, we employed a comprehensive surgical approach, which involved a cut-back anoplasty to repair the imperforated anus,[4] bilateral ureteral reimplantation is included in our EEC repairment protocol to mitigate the risk of future vesicoureteral reflux (VUR),[5] primary bladder closure,[6] and suturing to approximate the pubic bones,[7] given the less pronounced diastasis compared to other variants (in classic bladder exstrophy, the pubic symphysis is widely separated by an average of 4.8 cm).[1] Subsequently, the patient was immobilized using wraps,[8] leading to successful outcomes without complications. Long-term follow-up is crucial to monitor for potential complications and assess the patient's continence rate and quality of life.

4. Conclusion

This case report highlights the successful management of isolated bladder exstrophy with a normal phallus and imperforated anus in a one-day-old male newborn. The utilization of a comprehensive surgical approach yielded favorable outcomes without complications, at least during the follow-up period. However, continued follow-up is necessary to ensure long-term success. Further research and case studies are warranted to enhance our understanding of this rare condition and optimize management strategies for affected individuals.

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

Patient consent

Consent to publish the case report was not obtained because this report does not contain any personal information that could lead to the identification of the patient.

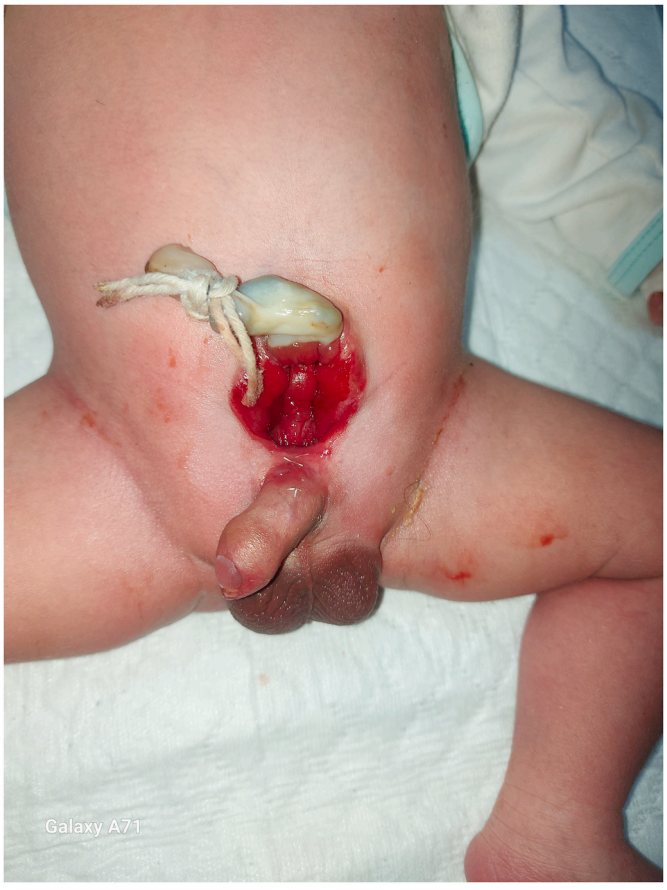

Fig. 3. Normal penis without epispadias.

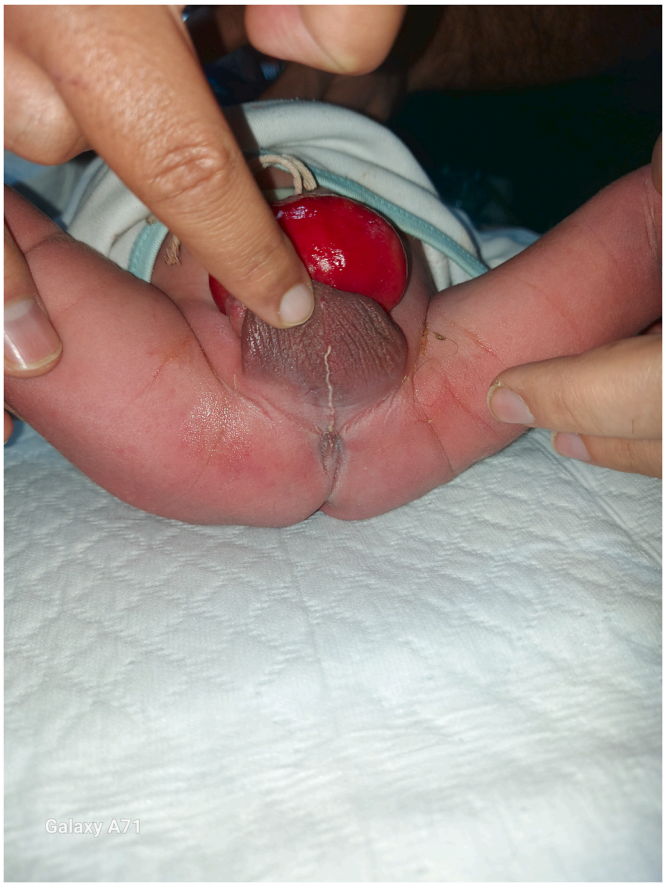

Fig. 4. Imperforated anus with rectoperineal fistula.

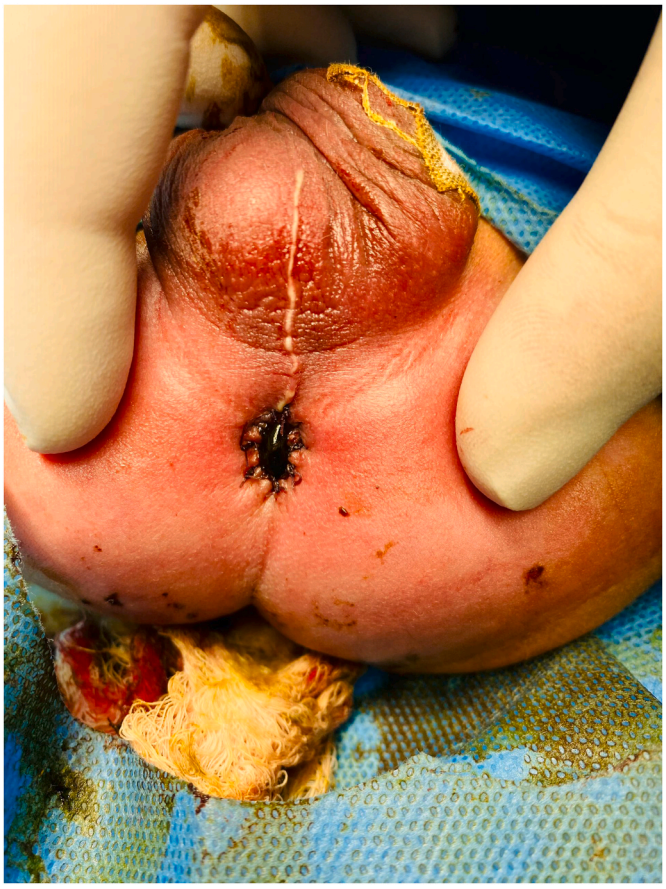

Fig. 5. Cut-back anoplasty for the imperforated anus.

Authorship

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

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CRediT authorship contribution statement

Ehsanullah Rasouli: Writing – review & editing, Writing – original draft. Abdullah Wahdat: reviewing – original draft. Mohammad Jawid Nazari: Writing – review & editing, Writing – original draft.

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