A Case Report on A Rare Variant of a Common Anomaly – Covered Bladder Exstrophy

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Abstract

Variants of bladder anomalies are rare and often associated with other abnormalities of the genitourinary system. Covered bladder exstrophy, a rare variant within the exstrophy-epispadias complex, is one such example.

We report a case of a 2-year-old female presenting with covered exstrophy of the bladder. The patient exhibited intermittent swelling over the anterior abdominal wall since birth, which increased in size during urination. Examination revealed a midline abdominal wall defect covered by skin in the hypogastric region. Diagnostic imaging, including a micturating cystourethrogram and ultrasound, confirmed the presence of an anterior abdominal wall defect with diastasis of the pyramidalis muscle and a focal bladder mucosal defect with intact detrusor muscle continuity. Surgical exploration revealed a smooth, globular structure with thinned walls and a vascular appearance herniating through the defect, confirmed to be the urinary bladder. Primary repair was performed, augmented with an onlay mesh hernioplasty using a polypropylene mesh.

The intraoperative period was uneventful, and the patient was alert and active postoperatively. Follow-up showed no complications, and the patient exhibited normal urinary function.

This case highlights the importance of early diagnosis and surgical intervention in managing covered bladder exstrophy. The successful repair of the defect and the favourable postoperative outcome underscore the efficacy of the surgical approach used.

Keywords

- Bladder exstrophy
- Pediatric bladder anomalies
- Anomalies ultrasound

Introduction

Variants of bladder anomalies are rare and often associated with other abnormalities of the genitourinary system. Covered bladder exstrophy, a rare variant within the exstrophy-epispadias complex, is one such example.[1] Such cases frequently present with additional complications, including vaginal duplication, bicornuate uterus, anorectal malformations, renal agenesis, and urethral duplication.[1-3] We present a

case of a 2-year-old female with covered exstrophy of the bladder, characterized by an intact detrusor muscle and a bladder mucosal defect.

Case presentation

A 2-year-old child presented to the pediatric surgery outpatient department with a history of intermittent swelling over the anterior abdominal wall since birth. There was no history of urinary leakage, other abdominal swellings, difficulty urinating, defecating, or incontinence. On examination, the swelling noticeably increased in size during urination. A midline abdominal wall defect, covered by skin, was noted in the hypogastric region and protruded with coughing and crying.

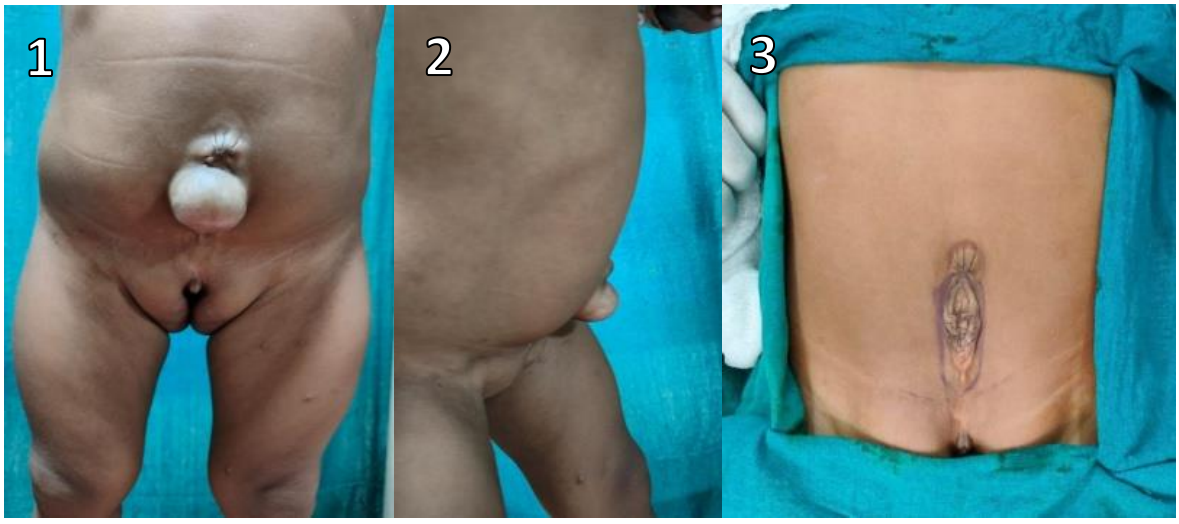

Figure 1, 2, 3: Figure 1 and 2 show frontal and lateral clinical images of a 3 x 3 cm soft, cystic, expansile, midline, infraumbilical swelling. The umbilicus was visibly separate from the swelling when the baby was crying and standing. **Figure 3** is a preoperative image demonstrating redundant skin after the swelling spontaneously decreased.

A micturating cystourethrogram

A micturating cystourethrogram revealed normal bladder opacification, normal anterior urethral contour, and no vesicoureteral reflux.

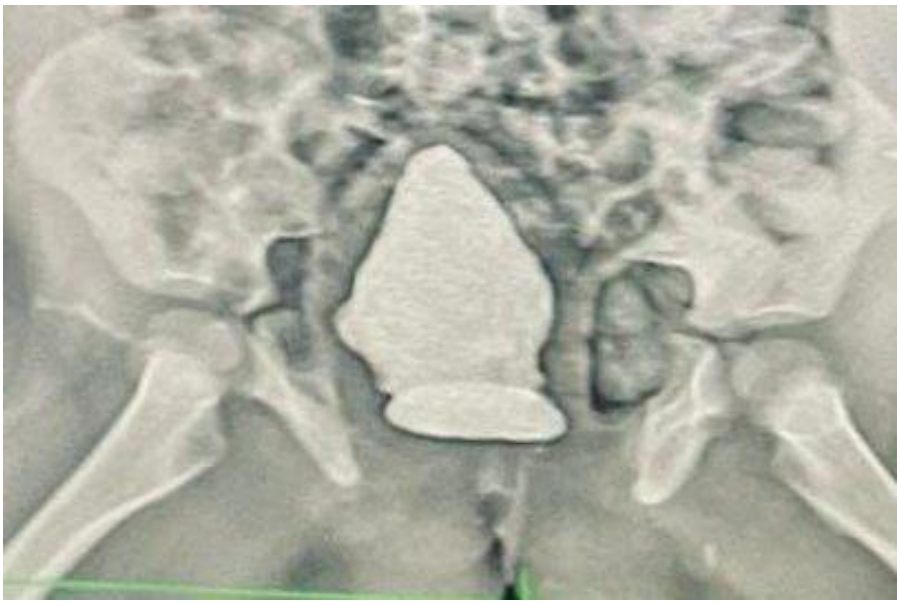

Figure 4: Figure 4 shows an MCU image of a well-distended, smooth-walled urinary bladder without any diverticula.

Diagnostic ultrasound imaging confirmed an anterior abdominal wall defect with diastasis of the pyramidalis muscle, through which portions of the urinary bladder and small bowel protruded. High-frequency linear probe interrogation of the urinary bladder revealed a focal bladder mucosal defect with intact detrusor muscle continuity at the superior margin. These findings supported the diagnosis of covered bladder exstrophy.

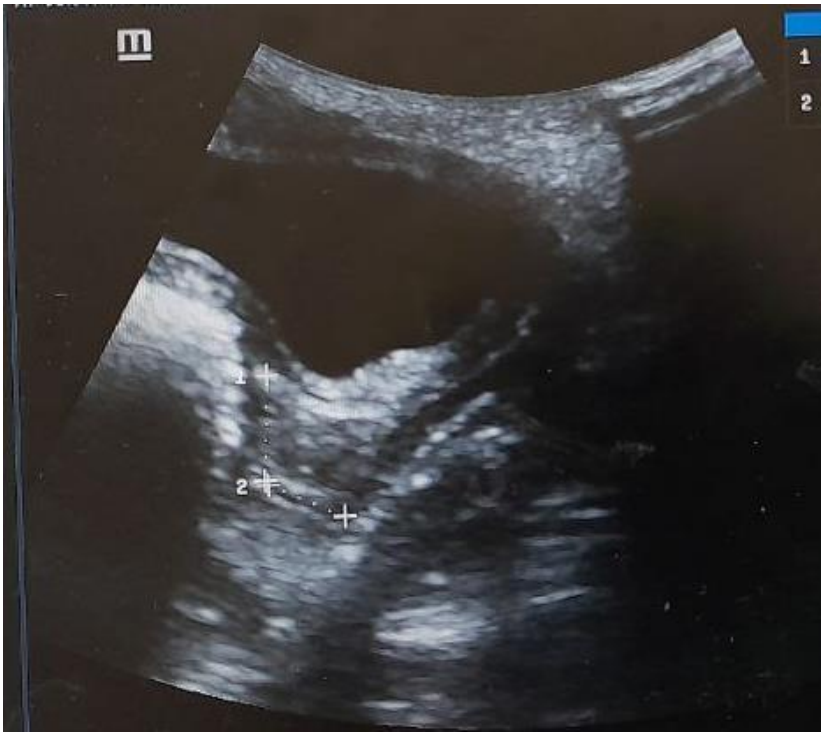

Figure 5: A grayscale ultrasound image demonstrating the urinary bladder wall herniating through the anterior abdominal wall defect, indicated by the arrow.

Local examination prior to cystoscopy revealed labial swellings and a single opening in the vestibule. Cystoscopy revealed a common channel approximately 3 cm in length with intact bladder wall trabeculae. Bladder capacity was adequate at approximately 150 ml. A bladder mucosal wall defect was observed in the anterosuperior border as an outward bulge, confirmed by external pressure.

Both ureteric orifices appeared normal. Intraoperatively, an incision was made just above the swelling, and skin and subcutaneous flaps were elevated. The rectus sheath was intact but attenuated over a 2.5 x 2 cm area in the infraumbilical region, through which a smooth, globular structure with thinned walls and a vascular appearance herniated, as shown in **Figure 6.**

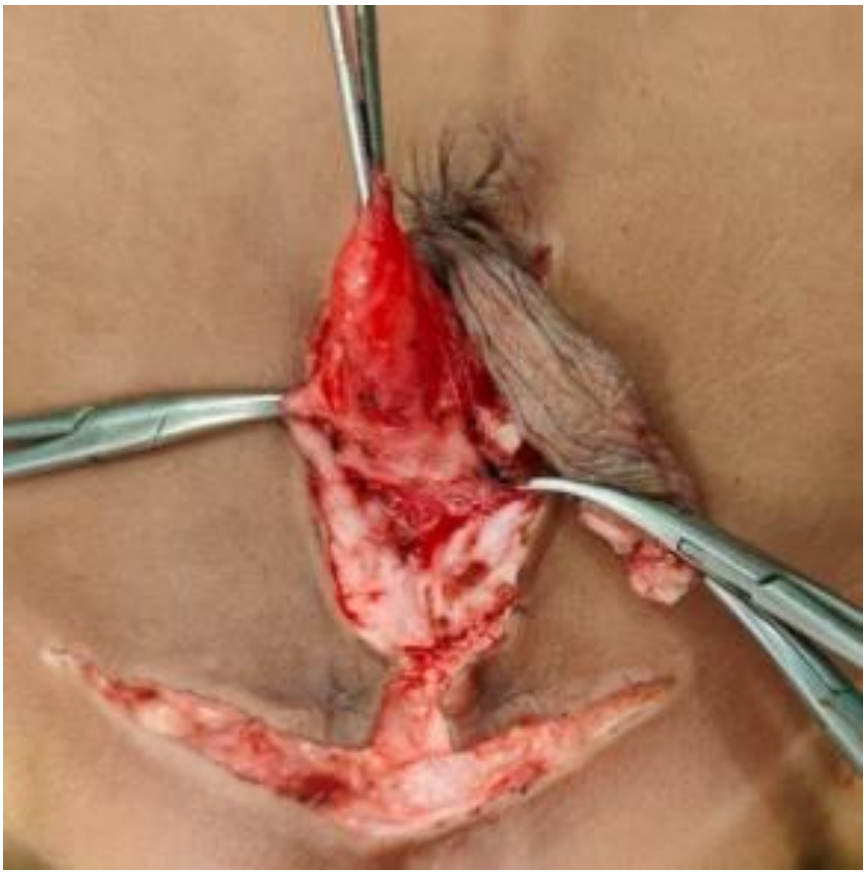

Figure 6:

The structure was confirmed to be the urinary bladder by inflating it with a betadine and saline solution per urethrally via a feeding tube. This solution was aspirated from the smooth swelling, confirming the diagnosis as shown in **Figure 7**. The bladder wall at the site of diastasis was thin but intact.

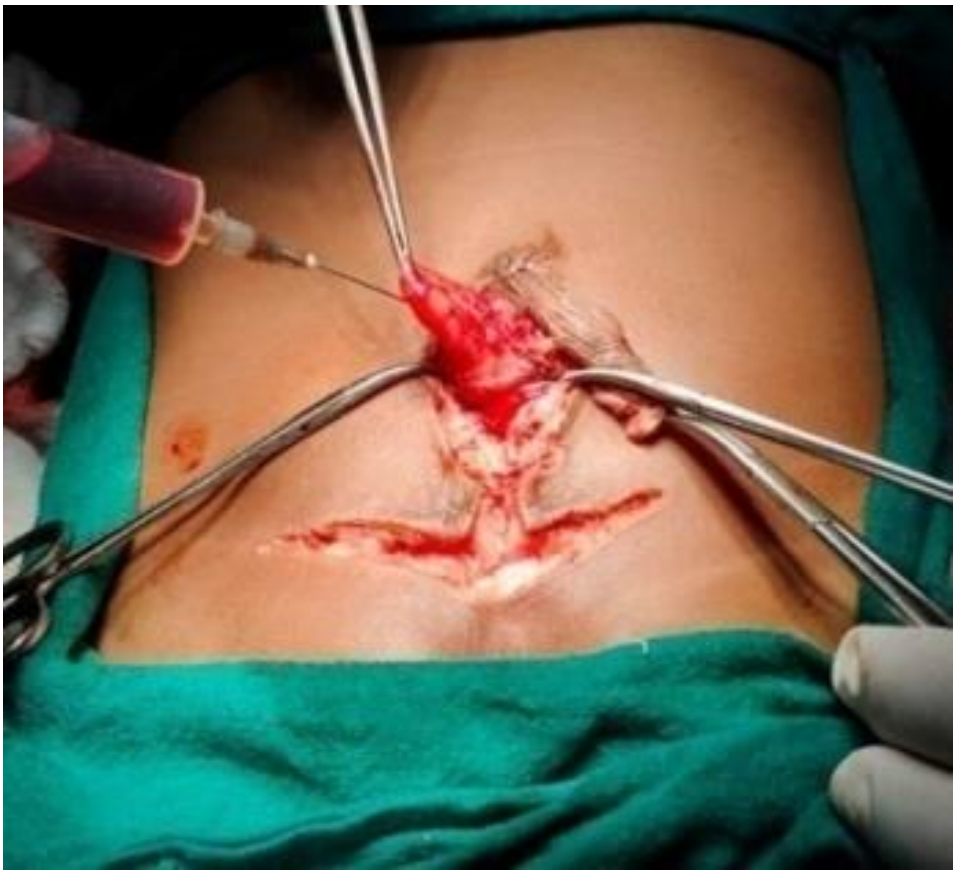

Figure 7:

As the defect was small, bilateral rectus sheath flaps were raised, followed by primary repair. This primary repair was augmented with an onlay mesh hernioplasty using a polypropylene mesh secured with Prolene sutures. A mini suction drain was placed, and the skin was closed as shown in **Figure 8.**

The intraoperative period was uneventful, and the child was alert and active postoperatively.

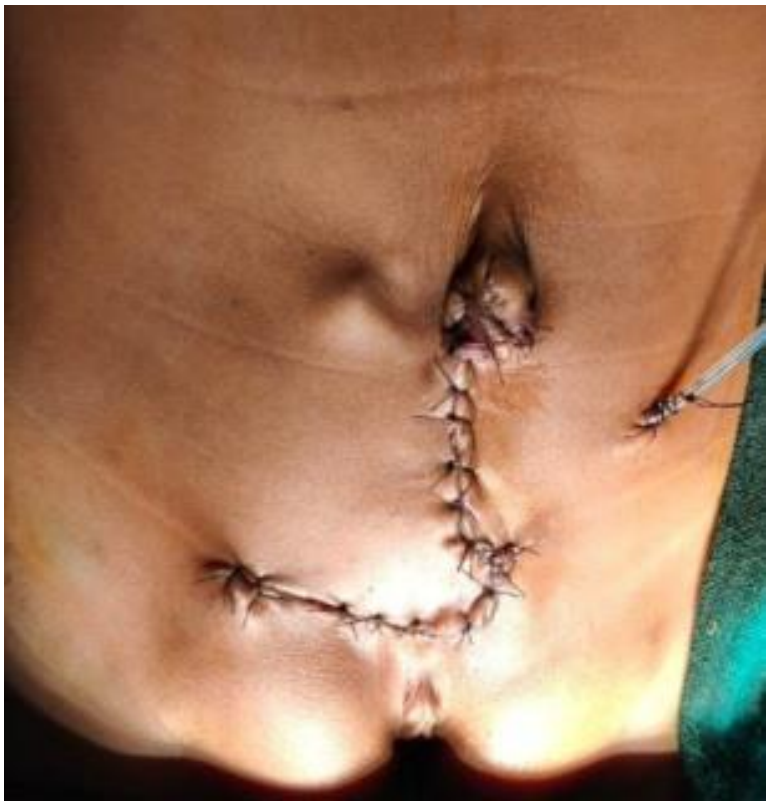

Figure 8:

Discussion

Bladder exstrophy complex, also known as "Split Symphysis Variants," shares similarities with classical bladder exstrophy but varies in degrees of skin closure, with sphincter mechanism integrity being inconsistent. These variants are categorized as pseudoexstrophy, superior vesical fissure, duplicate exstrophy, and covered exstrophy. In classical bladder exstrophy, the urinary bladder is exposed outside the abdominal cavity with a poorly formed umbilicus and urinary incontinence. In superior vesical fissure, the upper bladder is open near the anterior abdominal wall. Covered exstrophy differs from pseudoexstrophy by the presence of an isolated ectopic bowel segment on the inferior abdominal wall

near the genital area, which is colonic and lacks communication with the remaining gastrointestinal tract.[1-9]

An exhaustive PubMed search using the MeSH terms “covered,” “bladder,” and “exstrophy” yielded nine results. Case reports predominantly linked covered exstrophy to duplications in lowerurogenital tract structures, including the bladder, scrotum, vagina, clitoris, uterus, and, less commonly, the kidneys.[1-7,12] A rare association with sarcomatoid urothelial carcinoma was also reported.[8] The exact etiology of this rare variant remains unclear, but embryological theories propose an abnormal infraumbilical cloacal membrane hindering lateral mesoderm migration[9] or a body stalk abnormality with caudal insertion.[10]

Treatment encompasses genital reconstruction and abdominal wall repair. Many cases achieve good continence, often negating the need for sphincter repair. Unlike classical exstrophy, surgical intervention is typically simpler, and associated genitourinary reconstruction is relatively straightforward.

Conclusion

This case highlights the importance of early diagnosis and surgical intervention in managing covered bladder exstrophy. The successful repair of the defect and the favorable postoperative outcome underscore the efficacy of the surgical
approach used.

Ethical Consideration

This study was conducted in accordance with the ethical standards of the institutional and national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. Informed consent was obtained from all individual participants included in the study. (792/IEC/2022/IGIMS- 15/11/2022).

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Conflict of interests

There is no conflict of interest

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