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Pseudoexstrophy associated with penile duplication and hypospadias: A case report and literature review

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ABSTRACT

Bladder exstrophy is a rare developmental anomaly. Four principle variants of bladder exstrophy have been described and they themselves are rarer than the bladder exstrophy. Authors describe the management of a case of pseudoexstrophy type of variant in a 9 month old male child with penile duplication with torsion and coronal hypospadias. The rectal fascial defect was repaired without osteotomy as the distance between two pubic bones was <4 cm (3.1 cm). Genital reconstruction with excision of duplicate atrophic penile shaft and repair of coronal hypospadias with detorsion of the functional penile shaft could be accomplished. The patient had good outcome in terms of cosmesis and urinary stream. Total of 18 cases of the pseudoexstrophy have been described till date. Pseudoexstrophy of bladder is a very rare condition and can simultaneously present with other defects like omphalocele, anorectal malformations, pouch colon, multiple or solitary urogenital anomalies. The principles of correction though remain same with correction of abdominal wall defect with or without osteotomy depending upon severity of pubic diastasis. Other anomalies can undergo treatment as per standard protocol.

Key Words: Pseudoexstrophy; penile duplication; hypospadias; child.

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principle variants of bladder exstrophy have been described and they themselves are rarer than bladder exstrophy itself [2]. Authors describe the management of a case of pseudoexstrophy type of variant with penile duplication with torsion and coronal hypospadias along with review of literature.

Introduction

Exstrophy of the bladder is a malformation that occurs in 1/25000 to 1/40000 births [1]. Four

Case report

A 9 month old boy was brought by parents with chief complaints of an abnormal bulge in the

lower abdomen since birth with abnormal appearing external genitalia. Patient was diagnosed as a case of pseudoexstrophy of bladder with divergent pubic bones with interpubic distance of 3.1 cm at birth. Ultrasound of the upper urinary system did not reveal any abnormality. Micturating cystourethrogram showed a bilateral grade 2 vesicoureteral reflux. Patient was on chemoprophylaxis for the same and didn't suffer from urinary tract infection till now. He was continent with occasional dribbling on straining/crying with good stream directed cranially.

On examination, patient had midline defect in the rectus sheath starting from umbilicus with divergent pubic bones. Penis was buried and looked like having an epispadiac meatus [Fig. 1]. Bilateral testes were descended. Right sided inguinal hernia was present.

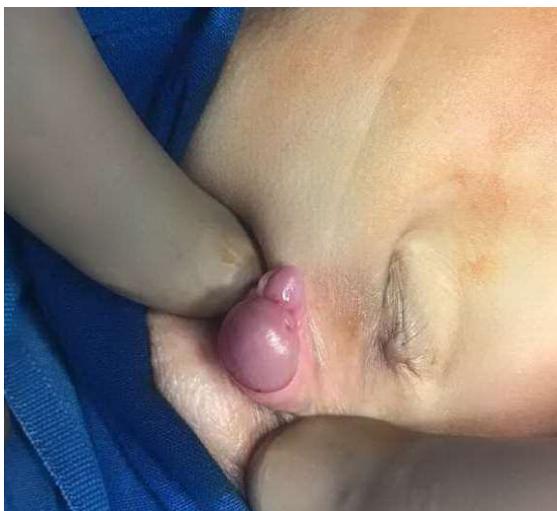


Fig. 1. Physical appearance of the defect.

Patient underwent corrective surgery under general anesthesia by a Pfannenstiel's incision. Divergent rectus sheaths were defined and mobilized and sutured with the interpubic band using Ethibond suture. It was possible to close the defect completely without osteotomy.

Right inguinal hernia repair was done through the same incision. Degloving of penis with a Firllet incision revealed penile duplication. One of the duplicated shafts was having torsion and hypospadiac coronal urethral meatus. The feeding tube could be passed through the same till urinary bladder. Other duplicated shaft was atrophic, incomplete and without any evidence of obvious urethral opening. It was attached to functional shaft at midpenile level, only with loose connective tissue [Fig. 2].

Atrophic penile duplication was excised. Functional shaft was straightened and Snodgrass repair was done for coronal hypospadias [Fig. 3].

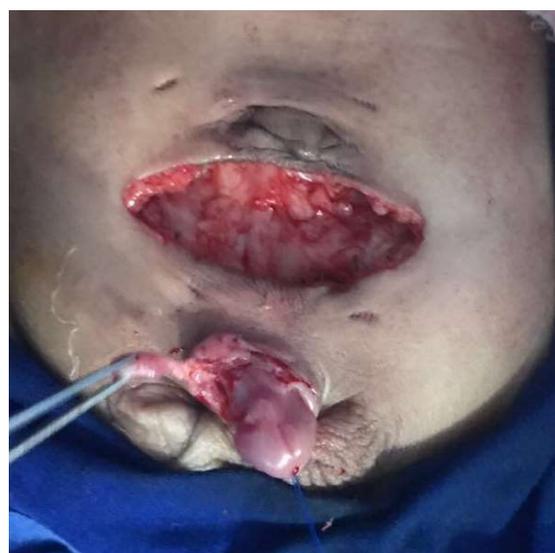


Fig. 2. Intraoperative photograph showing penile duplication with atrophic penile shaft and its attachment along with almost 180 degree penile torsion and coronal hypospadias.

Atrophic penile duplication was excised. Functional shaft was straightened and Snodgrass repair was done for coronal hypospadias. Patient was started orals after 4 hours. A 6 Fr Infant feeding tube was kept for urinary drainage across the repair and was removed after 48 hours. Patient was discharged

after 48 hours. Histopathological examination of the atrophic tissue confirmed the clinical diagnosis of penile duplication.



Fig. 3. Completed repair with detorsion and correction of hypospadias.

Discussion

The term pseudoexstrophy was first coined in 1954 by Hejtmancik et al [3]. The pseudoexstrophy is presence of characteristic muscular and skeletal defects of exstrophy without major defects in urinary tract as described by Marshall VF et al, 1962 [1]. The primary differentiating factor between covered exstrophy and pseudoexstrophy is that the former will generally have an ectopic bowel segment, commonly the colon present near the inferior abdominal wall [1,4,5].

A review of literature revealed 18 cases of pseudoexstrophy described till date, starting from 1954 by Hejtmancik et al [3]. Zivkovic et al [6], 1977 reported a newborn case of omphalocele with characteristic musculoskeletal defects and imperforate anus with anovulvar fistula. Mitchell et al [7], 1993 described a pseudoexstrophy and considered it as umbilical positional anomaly. Sözübir et al [8], 1997 reported a male pseudoexstrophy case with appearance of a low set umbilicus

associated with umbilical hernia and penile anomaly. Only penile reconstruction was done. Four cases of pseudoexstrophy with congenital pouch colon have been described till date. Pseudoexstrophy with right ectopic hemiscrotum with high anorectal malformation with type IV congenital pouch colon was described by Chadha et al in 1998 [9]. Herman et al [10], in the year 2000 reported a newborn girl with type II CPC and pseudoexstrophy associated with various spine, spinal cord, GIT, and genitourinary anomalies. Two newborn girls with single perineal opening (cloaca), and pseudoexstrophy in the form of divergent pubic bones and rectus muscles, and a low-set umbilicus were reported by Jhanwar et al in 2016. Both patients had a type II congenital pouch colon (CPC) with one hemiuterus and vagina on each side in the pelvis. In one patient, a Meckel's diverticulum was present 5 cm from the ileocecal junction. In both girls, a diverting proximal ileostomy was the initial surgery [11].

Apart from Zivkovic et al [6] in 1977, three more cases of pseudoexstrophy with omphalocele were described by Swana et al (1997) [12]. Meisheri et al [13] reported using a bilateral anterior pubic ramotomy for closure of suprapubic triangular defect. The case described by Otake et al [14], had wide pubic diastasis (greater >4cm) requiring two stage surgeries consisting of omphalocele repair at neonatal age and abdominoplasty with iliac osteotomy after the age of 6 months.

A one year old girl with Fraser Syndrome (the association of craniofacial abnormalities, syndactyly and cryptophthalmos) and multiple urogenital abnormalities including clitoromegaly, left renal agenesis and a unique urinary bladder exstrophy variant (psuedoexstrophy) with intact bladder which

herniated through the lower abdominal wall defect was reported by Daia et al in 2001 [15]. In the same year, Pandit et al [16] described a female pseudoexstrophy with bifid clitoris. She required only abdominal wall defect closure. A case of pseudoexstrophy with scaphoid megalourethra, and anorectal malformation was published by Devendra et al in 2002 [17]. Left-sided gastroschisis and pseudoexstrophy, a rare combination of anomalies was reported by Orpen et al, 2004 [18].

In 2005, Mahajan et al [19] described a case of pseudoexstrophy with epispadias. In this case, intact bladder was lined by the mucous membrane which later epithelized. Epispadias was repaired by penile disassembly technique. A female neonate with pseudoexstrophy with unilateral renal agenesis and having wide pubic diastasis with distance of separation being more than 4 cm was reported by Amouei et al, 2016. Patient underwent closure of abdominal wall and pubic symphysis by posterior osteotomy. Genital reconstruction was deferred for 4 to 6 months after first surgery [20].

In our case, we did repair of the fascial defect at 9 months of age, as child presented at that time. We could repair it without osteotomy, as the distance between two pubic bones was <4 cm (3.1 cm). Genital reconstruction with excision of duplicate atrophic penile shaft and repair of coronal hypospadias with detorsion of the functional penile shaft could be accomplished at the same sitting. The patient had good outcome in terms of cosmesis and urinary stream postoperatively.

We can safely say that pseudoexstrophy of bladder is a very rare condition and can simultaneously present with other defects like omphalocele, anorectal malformations, pouch colon, multiple or solitary urogenital

anomalies. The principles of correction though remain same with correction of abdominal wall defect with or without osteotomy depending upon severity of pubic diastasis. Other anomalies can undergo treatment as per standard protocol.

Compliance with ethical statements

Conflicts of Interest: None.

Financial disclosure: None.

Consent: All photos were taken with parental consent.

References

- [1] Marshall VF, Muecke EC. Variations in exstrophy of the bladder. *J Urol.* 1962;88:766–96
- [2] Lowentritt BH, Van Zijl PS, Frimberger D, Baird A, Lakshmanan Y, Gearhart JP. Variants of the exstrophy complex: a single institution experience. *J Urol.* 2005;173(5):1732–37.
- [3] Hejtmancik JH, King WB, Magid MA. Pseudo-exstrophy of bladder. *J Urol.* 1954;72(5):829–32.
- [4] Johnston JH. The genital aspects of exstrophy. *J Urol.* 1975;113(5):701–5.
- [5] Sheldon CA, McLorie GA, Khoury A, Churchill BM. Duplicate bladder exstrophy: a new variant of clinical and embryological significance. *J Urol.* 1990;144(2Pt 1):334–36.
- [6] Zivković SM. Variations in the bladder exstrophy complex associated with large omphalocele. *J Urol.* 1977;118(3):440–42.
- [7] Mitchell W, Venable D, Patel AJ. Pseudoexstrophy. *Urology.* 1993;41(2):134–36.
- [8] Sözübir S, Aritürk E, Rizalar R, Barış S, Bernay F, Gürses N. A pseudoexstrophy with penile anomaly. *Eur J Pediatr Surg.* 1997;7(1):55–56.

- [9]Chadha R, Sharma A, Bagga D, Mahajan JK. Pseudoexstrophy associated with congenital pouch colon. *J Pediatr Surg.* 1998;33(12):1831–33.
- [10]Herman TE, Coplen D, Skinner M. Congenital short colon with imperforate anus (pouch colon). Report of a case. *Pediatr Radiol.* 2000;30(4):243–6.
- [11]Jhanwar P, Shinde NK, Patel JN, Pant N, Chadha R. Congenital Pouch Colon associated with Pseudoexstrophy: Report of Two Cases. *APSP J Case Rep.* 2016;7(1):9.
- [12]Swana HS, Gallagher PG, Weiss RM. Pseudoexstrophy of the bladder: case report and literature review. *J Pediatr Surg.* 1997;32(10):1480-81.
- [13]Meisheri IV, Kasat LS, Bahety G, Sawant V, Kothari P, Kumar A. Pseudoexstrophy of the bladder: a rare variant. *Pediatr Surg Int.* 2001;17(2-3):224–25.
- [14]Otake K, Uchida K, Inoue M, Koike Y, Matsushita K, Oki S, et al. Staged management of pseudoexstrophy with omphalocele and wide pubic diastasis. *Journal of Pediatric Surgery Case Reports.* 2014;2(8):388–90.
- [15]Daia JA . Fraser syndrome with bladder pseudoexstrophy. *Saudi Med J.* 2001;22(5):455-56.
- [16]Pandit SK , Budhiraja S, Rattan KN, Gupta U, Solanki RS, Singh Y. Pseudoexstrophy in a female child. *Indian J Pediatr.* 2001;68(2):179-80.
- [17]Devendra K , Mahajan JK, Rao KL Pseudoexstrophy associated with megalourethra. *J Pediatr Surg.* 2002;37(9):E26
- [18]Orpen NM, Mathievathaniy M, Hitchcock R. Left-sided gastroschisis and pseudoexstrophy: a rare combination of anomalies. *Pediatr Surg Int.* 2004;20(7):551–52.
- [19]Mahajan JK, Ojha S, Rao KLN. Pseudoexstrophy with epispadias. *Urol Int.* 2005;74(4):365–57.
- [20]Amouei A, Yazdi MB, Ehsani F, Zarch MB. Pseudo-Exstrophy of Bladder with Unilateral Renal Agenesis: A Rare Combination of two Anomalies. *J Clin Diagn Res JCDR.* 2016;10(12):PD13-PD14.

