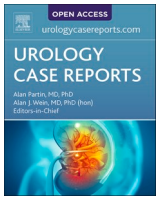
Urology Case Reports 52 (2024) 102643

Contents lists available at ScienceDirect

Urology Case Reports

journal homepage: www.elsevier.com/locate/eucr



Pediatrics

Inferior vesical fistula with solitary kidney and ureterocele - Rare exstrophy variant

Maitreyee Save [a,*], Aditi Dalvi [a], Dhvani Shah [b], Abhaya Gupta [a], Shahaji Deshmukh [a], Paras Kothari [a]

[a] Dept of Pediatric Surgery, LTMGH, Sion, Mumbai, India
[b] Dept Of Pediatric Surgery, BJMC, Pune, India

A B S T R A C T

Exstrophy variants are uncommon developmental anomalies, with the inferior vesical variant being the rarest among them. We present the case of a 1-year-old continent boy with an inferior vesical fistula (IVF) with solitary kidney and ureterocele where simple closure was done followed by a normal micturition pattern. Only two cases of IVF have been reported yet, ours being the first with solitary kidney and ureterocele.

1. Introduction

Exstrophy of the bladder is a rare anomaly with an incidence of 1 in 30–50,000 live births, the inferior vesical variant constituting about 8 % of the total exstrophies.[1,2]

Anomalies involving the bladder neck as IVF are the rarest and only 2 cases have been reported. We present the first case of IVF with solitary kidney and ureterocele.

2. Case report

A 1-month-old boy presented with the complaint of passing urine from an abnormal opening above the penis since birth. He was passing urine in a good stream per urethra with continuous dribbling of urine from the opening. A small opening with a mucosal plate was present just above the penis (Fig. 1). Meatus, penis, and hernia sites were normal. Both testes were palpable in the scrotum. Pubic diastasis of 2 cm was found, umbilicus was central.

Ultrasonography revealed left kidney agenesis with a normal right system with no other abnormality. Micturating cystourethrogram (MCU) showed an abnormal opening at the bladder neck with a capacity of 20 cc and no reflux (Fig. 2).

All blood investigations were within normal limits. Excoriation around fistula was treated and patient was planned for diagnostic cystoscopy at age of 6 months.

During diagnostic cystoscopy, infant feeding tube inserted through the fistula was seen coming out from an opening above the bladder neck confirming the diagnosis of IVF. Left ureteric orifice not visualised. Right ureterocele was noted, there was no free flow of urine from right ureteric orifice. Cystoscopic ureterocele incision was done. Perimucosal incision was taken and tract dissected, ligated at bladder neck and bladder closed in two layers. (Fig. 3). Patient managed with Foley's catheter post operatively which was removed after 10 days. Patient was continent and passed urine in single good stream (Fig. 4). Micturating cystourethrogram done at 3 month post operatively showed no reflux. Patient is currently on regular follow up with no urinary tract infection or any other symptoms.

3. Discussion

Exstrophy bladder variants constituting 8 % of total exstrophies.[1] Embryologically, persistence or overdevelopment of cloacal membrane preventing migration of the mesenchymal tissue to the midline results in defects.[2] The superficial cloacal membrane due to poor penetration by the mesenchyme is prone to rupture. The size, site and timing of rupture decides type of exstrophy variant. Many modern theories have been proposed yet exact etiology is not clearly defined. Unlike bladder exstrophy, variants are more common in females.[3] I. D. Williams classified variants for the first time as superior vesical fissure, vesical fistula and duplex bladder.[4] Other authors subsequently elaborated rest of variants. They have been classified into sub groups: pseudoexstrophy, covered exstrophy, superior vesical fissure, or fistula, inferior vesical fissure and fistula and duplicate exstrophy. Musculoskeletal defects like divergent recti, pubic diastasis and a low-set umbilicus are present in variants of

* Corresponding author.
E-mail address: save.maitreyee@gmail.com (M. Save).

https://doi.org/10.1016/j.eucr.2023.102643
Received 16 November 2023; Received in revised form 9 December 2023; Accepted 22 December 2023
Available online 27 December 2023

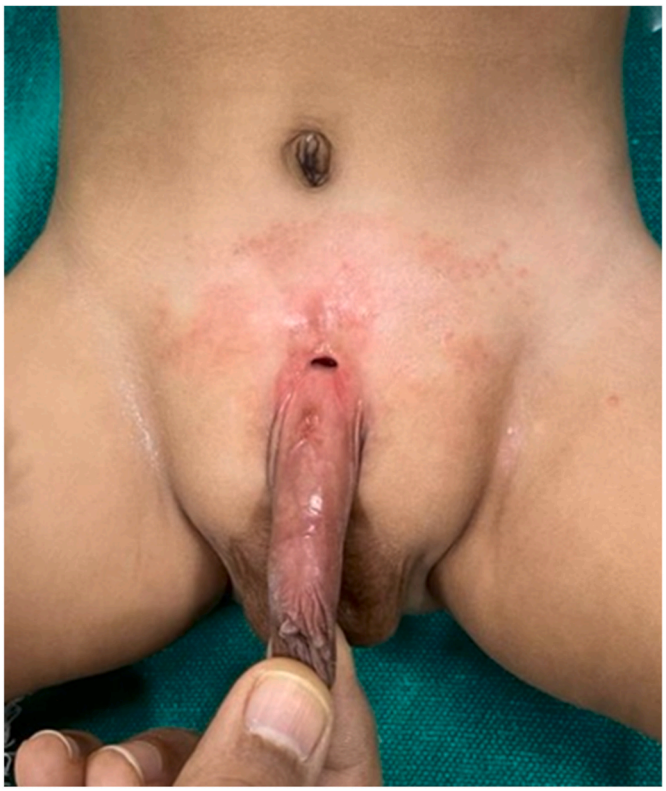


Fig. 1. Clinical picture.

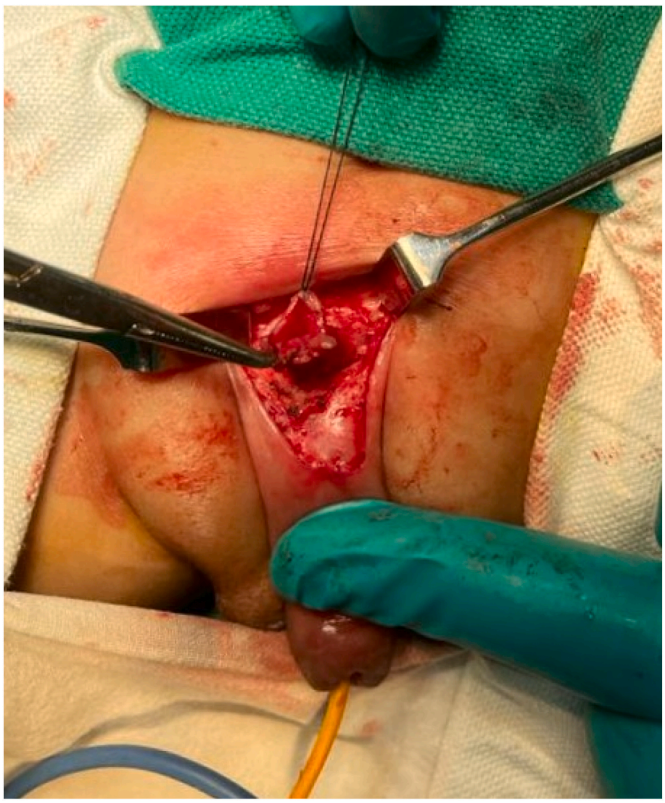


Fig. 3. Intraoperative picture.

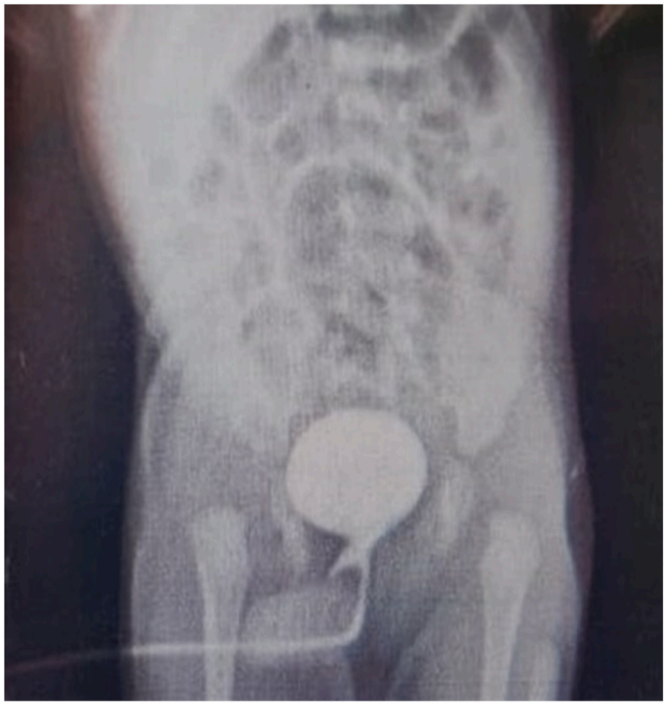


Fig. 2. Micturating cystourethrogram.

Fig. 4. Postoperative picture.

bladder exstrophy.

Inferior vesical variants are rarest among all the other variants. They involve the lower part of bladder above the bladder neck. It can be a fistula which is communication between bladder and skin or a fissure which is eventration of bladder.[5] Musculoskeletal component with defect being above bladder neck differentiates it from urethral

duplication.

A review of the literature revealed two cases conforming to the definition of inferior vesical fissure, first by Marshall and Muecke's review of exstrophy variants[6] and other by Johnson et al. which required bladder augmentation.[7] Mahajan et al. described a case of inferior vesical fistula in a 20 month old female where simple closure was done,[5] Vini Joseph et al. reported similar case in 2 year old boy. We are the first one to report case of inferior vesical fistula with solitary kidney and a ureterocele.

4. Conclusion

Treatment and prognosis is different for different variants hence diagnosing rare exstrophy variants is important. Simple closure is treatment for inferior variants. In our case, good bladder capacity and normal upper tract has resulted in good post-operative prognosis with continence though long term follow up will be needed.

Funding source

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

CRediT authorship contribution statement

Maitreyee Save: Writing – original draft. **Aditi Dalvi:** Investigation. **Dhvani Shah:** Supervision. **Abhaya Gupta:** Writing – review & editing. **Shahaji Deshmukh:** Validation. **Paras Kothari:** Resources.

References

1. Khemchandani SI. Duplicate bladder exstrophy with superior vesical fistula in a male infant, a rare variant. *SM J Pediatr Surg.* 2016;2(1):1008.
2. Chadha R, Sharma A, Bagga D, Mahajan JK. Covered exstrophy with incomplete duplication of the bladder. *Pediatr Surg Int.* 1999 Jul;15(5):422–424.
3. Valioulis I, Levard G, Lottmann H, Hameury F, Cendron J. A rare complex bladder exstrophy variant. *BJU Int.* 2000 Aug;86(3):398–400.
4. Turner William R, Ransley Philip G, Bloom DA, Williams DI. Variants of the exstrophic complex. *Urol Clin.* 1980 June;7(2):493–501.
5. Mahajan JK, Bagga D, Chadha R, Kumar A. Inferior vesical fistula. *Pediatr Surg Int.* 1999 Sep;15(7):517–518.
6. Marshall VF, Muecke EC. Variations in exstrophy of the bladder. *J Urol.* 1962 Dec 1; 88(6):766–796.
7. Johnson P, Sarin YK, Kapoor R, Mammen KJ. Inferior vesical fissure. *J Urol.* 1995 Oct; 154(4):1478.