CASE REPORT

SUPRAPUBIC DISCHARGING SINUS ASSOCIATED WITH CLITORAL CLEFT AND PUBIC DIASTASIS: A VARIANT OF BLADDER EXSTROPHY

YAQOOT JAHAN

ABSTRACT

A variant of the exstrophic urinary bladder (duplicate bladder exstrophy) presenting with a discharging sinus in suprapubic region in a seven months old continent girl is being reported. The infant also had cleft mons pubis, cleft clitoris and pubic diastasis. VCUG was normal. Contrast x-ray through discharging sinus showed a blind tract which was excised at surgery. It extended through the rectus sheath and ended blindly at bladder surface, which itself was intact. Histopathology showed transitional epithelium with fibromuscular layer.

Key words *Bladder exstrophy, Exstrophy variants, Duplicate bladder exstrophy.*

INTRODUCTION:

Variants of classic bladder exstrophy are extremely rare, making up 8% of all exstrophy/epispadias complex.[1] Another name for these lesion is "split symphysis" variants.[2] These have all the usual musculoskeletal findings of classic exstrophy, however the urinary bladder is closed with varying degrees of skin and subcutaneous cover, and the urethra and sphincter mechanism may be intact.[1,3] Three main variants are superior vesical fissure, covered exstrophy and duplicate exstrophy.

CASE REPORT:

Seven months old girl presented in a private clinic with complaint of small opening in the suprapubic area which was noted at birth. Occasionally pus discharged from the opening. She voided from the normal urethral opening with a normal stream. On examination there was a moist, soft mucosal area about 2-cm in diameter in the suprapubic area. Below it was cleft mons pubis and cleft clitoris. Umbilicus was normally placed (Figure Ia & b).

A pelvic x-ray showed a 4-cm pubic symphysis diastsis. A voiding cystourethrogram demonstrated a normal bladder and urethra without reflux or any outside communication (Figure II). Contrast x-ray through the opening showed irregular shaped tubular structure, not communicating with bladder (Figure III). Cystoscopy could not be done. Surgical excision of the sinus was done through an elliptical incision. The tract extended through the rectus sheath and found lying blindly over the bladder surface .There was no communication with bladder. Bladder was accidentally opened and repaired. Postoperative recovery was smooth.

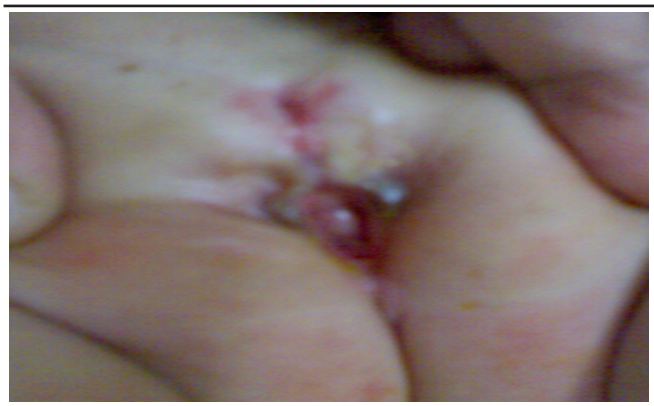
Histopathologic examination of the exstrophic mucosal area revealed transitional epithelium with

focal chronic inflammatory infiltrate and fibromuscular layer with frequent nerve bundles and adipose tissue consistent with soft tissue of urinary bladder.

DISCUSSION:

Duplicate exstrophy is one of the rarest variant and only 23 cases have been reported in the English literature till 2009.[4] Duplicate bladder exstrophy consists of an exposed patch of exstrophic bladder with a normal or smaller than normal intact bladder deep to the exstrophic bladder, a musculoskeletal defect, and an occasional epispadias. There is no urinary communication to the exstrophied component.[3] The possible embryological cause behind exstrophy anomalies is thougth to be incomplete closure of the infraumbilical abdominal wall. A low lying umbilicus, separated pubic rami, extroversion and nonclosure of the bladder, with or without epispadias, are proposed to result from abnormal persistence of cloacal membrane.[5]

Correspondence:
Dr. Yaqoot Jahan
Department of Paediatric Surgery
Civil Hospital and DUHS,
Karachi



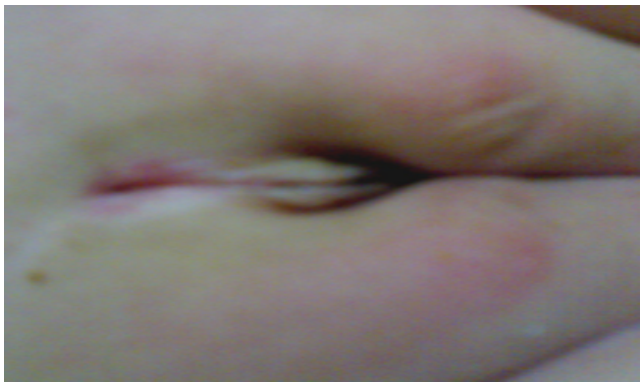


Fig I a & b: Cleft mons pubis and cleft clitons with exstrophied bladder mucosa.

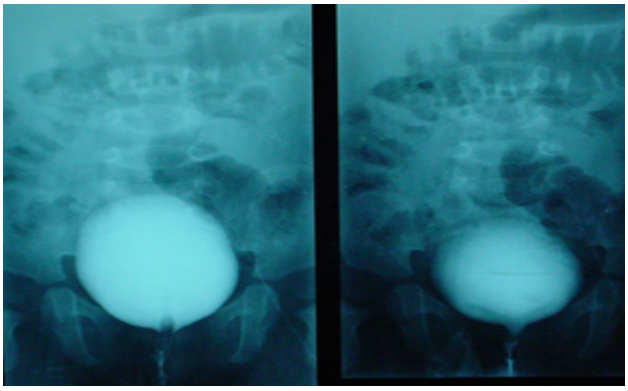


Fig II: Normal VCUG with pubic diastasis.

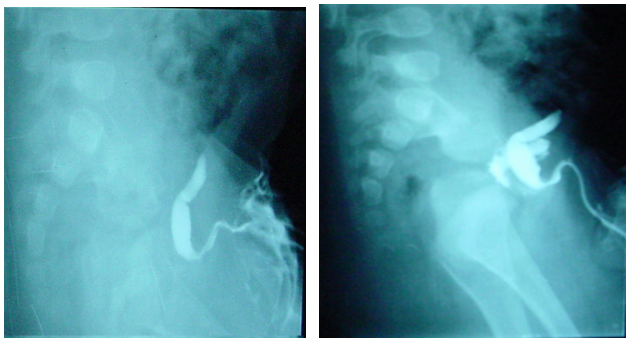


Fig III: Sinogram showing blind tract.

This infraumbilical cloacal membrane acts as a wedge and prevents the lateral mesoderm from progressing medially between its ectodermal and endodermal layers. Rupture of this unstable membrane results in an absent lower abdominal wall and an exposed bladder with epispadias. Exstrophy variants are explained by incomplete rupture or persistence of the abnormal cloacal membrane.[6]

Treatment is gratifying, because the patients have normal internal urogenital structures. The difficult problems with urinary continence with bladder exstrophy may not exist with variants of the exstrophy complex.[7] Careful examination with fluoroscopy and cystoscopy is helpful in defining the anatomy in these variants.

REFERENCES:

1. Turner WR, Ransley PG, Bloom DA, et al. Variants of the exstrophy complex. Urol Clin North Am 1980;7:493-501.

2. Williams DI. Split symphysis variants, in Williams DI (Ed): Encyclopedia of Urology., Ch 15, New York, NY, Springer Verlag, 1974, p 2779.

3. Marshall VF, Muecke EC. Variations in exstrophy of the bladder. J Urol 1962;88:766-96.

4. Kumar B, Sherma C, Sinha DD. True duplicate bladder exstrophy: A rare lesion. Indian J Pediatr 2009;76;852-3.

5. Jeffs RD: Exstrophy of the urinary bladder, in Welch KJ, Randolph JG, Ravitch MM, et al(eds):Pediatric Surgery. Chicago, IL, Year Book Medical 1986, pp 1216-41.

6. Boemers TML, de Jong TPVM, Rovekamp MH, et al. Covered exstrophy associated with an anorectal malformation; A rare variant oF classic bladder exstrophy. Pediat Surg Int 1994; 9:438-40.

7. Gearhart JP, Mathews R. Exstrophy-epispadias complex. In: Wein AJ, Kavoussi LR, Novick AC, et al, (editors), Campbell-Walsh urology. 9th ed. Philadelphia: Saunders Elsevier; 2007.pp 3348-50.