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Pediatrics

CONGENITAL PREPUBIC SINUS

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ABSTRACT Congenital prepubic sinus (CPS) is an extremely rare congenital anomaly with less than 30 cases reported so far1.It is a disorder of unknown etiology. We report the management of 8 month old boy with congenital prepubic sinus. Also a review of the available cases in the literature is presented.

KEYWORDS : congenital; prepubic sinus; urethral duplication

Introduction

Congenital prepubic sinus (CPS) is a rare congenital anomaly which is associated with discharge from an opening overlying the pubis symphysis. Various names including congenital prepubic sinus, subpubic fistula, prepubic dermoid sinus, and suprapubic dermoid sinus, were used to identify this lesion. This variant of urethral duplications deserves special attention as none of classification clearly describes this entity. Two etiological theories have been proposed, including an anomalous anterior abdominal wall closure or variant of dorsal urethral duplication.[1-4]

Case Report

An 8 month old baby presented with a discharging sinus in the prepubic region since birth. A tiny opening was located in the midline prepubic area around 2 cm above the root of penis (Figure 1). He had no other signs and symptoms related to the genitourinary tract. Laboratory findings were normal. Sinography showed a non-communicating fistulous tract going towards the bladder neck (Figure 2). Micturating cystourethrogram was performed which showed no communication with the tract and no other urological anomaly. Surgical exploration was carried out taking an elliptical incision surrounding the orifice. The tract was found to be going under the pubic symphysis and communicating with the anterior wall of bladder through a very small opening. The tract was completely excised along with a cuff of the bladder (Figure 3). Patient was catheterized for 5 days. Histopathology with H&E staining demonstrated presence of transitional epithelium proximally and squamous epithelium distally lining the sinus.

Discussion

CPS is an extremely rare congenital anomaly with only around 30 cases reported so far[1]. Patients with a congenital prepubic sinus come to medical attention because of the opening or persistent discharge.

Prepubic sinus often mimics the congenital prepubic sinus arising due to failure of midline fusion of anterior abdominal wall but later has abdominal wall defects and tract mostly lined by squamous epithelium while the former is lined by transitional epithelium.[5,6] The etiology is still unknown. Two theories have been proposed. Campbell et al1 proposed that CPS may be a localized failure of midline fusion of anterior abdominal wall. The other theory is that CPS is a variant of dorsal urethral duplication. [2-4] The epithelium of the entire female urethra and most of the male urethra develops from the endoderm of the urogenital sinus, which gives rise to transitional epithelium. However, the distal portion of the male urethra is derived from the ectodermic glandular plate, which explains the squamous epithelium[2].
A Van der Putte[7] considered that interruption of the ventral part of cloacal membrane is the explanation of double urethral orifice. This defect does not allow the complete and correct replacement of ventral cloacal membrane by lateral mesodermic folds ,creating a fistulous tract. Stephens described 3 types of dorsal urethral duplication according to the anatomy.[8] Type 1 is a complete or incomplete tandem channel that runs parallel to the normal urethra from the glans to the bladder, which joins the urethra or ends blindly. Type 2 is an epispadiac type of channel from the dorsum of the penis to the bladder or one that joins the urethra at some point. Type 3 is a dermoid sinus that simulates an accessory urethra but tracks from the base of the penis in front of the pelvic urethra and bladder behind the pubic symphysis to or towards the umbilicus. The anatomy of our case is similar with the type 3 variant of the Stephens classification.

Incomplete urethral duplication are generally asymptomatic or may present with infected tracts. Indications of surgery include cosmesis, annoying symptoms such as double stream, recurrent urinary infections and persistent discharge. Complete excision is the only accepted standard management which yields gratifying results.

Some particular characteristics of importance are 1) all fistula are in the midline independently below ,through or above the pubic symphysis 2) epithelial lining is usually stratified squamous but transitional epithelium and smooth muscle fibres may be present in some cases .

Conclusion

This rare type of urethral duplication can be successfully treated by complete excision. Whatever the etiology, in patients with a congenital prepubic sinus the functional and cosmetic outcome in all types of duplication can be really good.

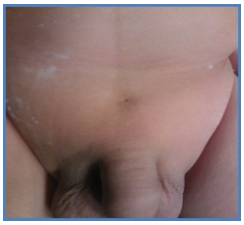

Fig 1—Prepubic sinus

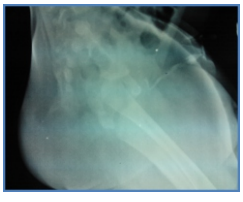

Fig 2: Sinogram

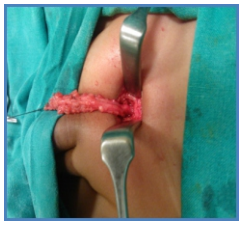

Fig3: Intra operative dissected sinus tract

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