The congenital prepubic sinus is a rare type of fistula with a broad range of anatomical and histological diversity. We report the case of an 8-month-old boy with a congenital prepubic sinus where immuno-histochemistry studies showed only squamous epithelium and no transitional epithelium. The etiology of this disorder is still unclear and the most recent case reports suggest that the fistula might be a variation of a dorsal urethral duplication. We recommend performing immunohistochemical studies to further clarify the etiology, as the fistula may be caused by different developmental disorders.

The congenital prepubic sinus is a rare type of urogenital anomaly of an unknown and widely discussed etiology. First described and classified within the group of dorsal urethral duplications by Stephens [1], it is a fistula that starts with an opening in the midline prepubic area or dorsal side of the penis in men. Depending on the type it may join the urethra, end in the bladder, grow toward the symphysis or the umbilicus and end blindly. We present a case of an 8-month-old boy with a congenital prepubic sinus lined with squamous epithelium which ends blindly at the ventro-caudal side of his pubic symphysis.

1. Case report

Our patient is an 8-month-old boy who presented with an intermittently occurring clear discharge from a small opening above the base of his penis. He has not had any urinary tract infections and no other urogenital anomalies were noticed in previous examinations. His parents reported poor feeding and poor weight gain as well as decreased urine volume.

A voiding cystourethrogramy showed no vesicoureteral reflux and a normal urethra. Application of 5 ml of Ultravist® (Bayer Vital GmbH, Leverkusen, Germany) via a 4 Charrière catheter through the opening in the patients midline prepubic area showed a fistula ending on the ventral side of his symphysis (see Fig. 1). There were no signs of communication between this fistula and his bladder or urethra. This suggested a urethral duplication or a prepubic sinus as possible diagnoses.

Cystoscopy showed no signs of anomalies of the urethra and the bladder. Instillation of indigo carmine into the prepubic opening did not lead to blue-colored urine, confirming the assumption that the fistula was not communicating with his urogenital tract.

Surgical exploration via oval incision and dissection along the indigo-dyed structure showed a fistula ending at the caudal border of the patient’s pubic symphysis, where it was ligated and excised completely (see Fig. 2).

Histological findings showed a 4 cm long fistula lined with squamous epithelium. Surrounding tissue had signs of chronic infection with stromal fibrosis. Immunohistochemistry exhibited only squamous differentiation (CK-5/6 and focal CK-7 positivity) without signs of transitional epithelium (GATA-3 and CK-20 negative).

2. Discussion

The prepubic sinus usually presents as an opening of the skin in the midline prepubic area. Stephens [1] classified the congenital prepubic sinus as a subgroup of dorsal urethral duplications. Depending on the route of the fistula, he further classified them into three different groups (see Fig. 3). Type I is a sinus running alongside the urethra either ending blindly or leading into the urethra. Type II grows underneath the symphysis pubis where it ends blindly or enters the urethra or bladder. Type III is described as a fistula leading cranial towards the umbilicus [2].
There are several theories concerning the embryological etiology of the sinus including: a variant of dorsal urethral duplication [3], midline closure defects [4], fistulas of the primitive urogenital sinus [5], remnants of the cloaca [6] or a residual cloacal membrane and umbilicophallic groove [7]. The theory that an anomaly in urethral development leads to a variation of urethral duplication seems to be the most prevalent among recent case reports. To the best of our knowledge only two other groups have performed immunohistochemical studies on the excised sinus. Huang et al. [8] showed presence of transitional epithelium in the proximal area of the sinus in four cases, Balster et al. [9] presented a case with immunohistochemical verification of transitional epithelium within the distal sinus. Huang et al. [8] also reported a case which did not show immunohistochemical evidence of transitional epithelium; this particular sinus was lined with squamous epithelium only. The case of our congenital sinus showed the same cellular lining. These cases show that the embryological origin of the congenital prepubic sinus is more than just a variant of the dorsal urethral duplication. This leads to the conclusion that this disorder is either a clinically similar phenotype of different embryological defects, a fistula of the primitive urogenital sinus with secondary squamous metaplasia [5] or as proposed by Tsukamoto et al. [7], a residual cloacal membrane lined by a tubularized umbilicophallic groove.

Bearing in mind the possible anatomical diversity of this congenital disorder it is advisable to examine patients presenting with an apparent prepubic sinus using contrast-imaging of the fistula and voiding cystourethrography. Regardless of the clinical presentation and the severity of symptoms the treatment of choice is excision of the sinus [3]. The technique should be individualized according to anatomical variants. We found that preoperative dyeing of the sinus was helpful with the dissection. We recommend performing immunohistochemical studies on the excised sinus to further clarify the etiology of this disorder. Currently the few published cases do not allow assumptions on complications following operative treatment — individual follow-up plans are necessary for each case due to the diversity and rareness of this disorder.

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