CASE REPORT

Congenital prepubic sinus (an epispadiac variant of dorsal urethral duplication) with dorsal penile curvature in an adult man: a rare association

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SUMMARY

Urethral duplication is among a very rare congenital disorder with multiple anatomical variants reported. Urethral duplication of complete type is usually diagnosed during childhood with urinary incontinence or double urinary stream as common presentation. However, patients with incomplete urethral duplication usually present with intermittent mucous discharge from the accessory urethral opening. A 20-year-old man presented to us with intermittent mucous discharge from the accessory opening along with dorsal penile curvature making sexual intercourse very difficult. The epispadiac urethral tract with proximal fibrous tract was excised, and the dorsal penile curvature was corrected by ventral plication, implicating that the fibrous cord may be the causative factor for the development of dorsal curvature.

BACKGROUND

Urethral duplication is a rare congenital anomaly and includes a spectrum of anatomical variants among which incomplete urethral duplication is more common than complete type.^{1,2} Contrary to complete urethral duplication which is diagnosed early in life due to urinary dribbling or double urinary stream, incomplete urethral duplication is either asymptomatic or present late with intermittent mucopurulent discharge, thus delaying the diagnosis.³ Due to late presentation, asymmetrical penile development may lead to penile curvature, and only two such cases of urethral duplication

associated with dorsal penile curvature have been reported in the literature.^{2,4} We present a unique case of a 20-year-old man with an epispadiac variant of urethral duplication who presented with intermittent mucous discharge and difficulty during sexual intercourse due to dorsal penile curvature since adolescence.

CASE PRESENTATION

A 20-year-old young man presented with intermittent mucous discharge since birth from an opening located in the midline over the dorsum of the proximal penile shaft with a penile curvature with no history of voiding symptoms or urinary tract infections. During adolescence, the patient noticed the abnormal penile curvature which used to accentuate on erection. Physical examination revealed bilateral normal testis and well-developed penis with dorsally deficient preputial skin. There was an accessory crescent-shaped opening located dorsally at the proximal penile shaft with a rudimentary groove located in the midline of the dorsum of the glans penis (figure 1) and severe degree of dorsal penile curvature was noted on penile tumescence (figure 2).

INVESTIGATIONS

Laboratory examinations were within normal range. Sinogram done through accessory meatus revealed the absence of communication either with

Figure 1 (A) An accessory crescent-shaped opening located dorsally at the proximal penile shaft with a rudimentary groove located on the dorsum of the glans penis. (B) A partial septum in the accessory opening giving the appearance of two tracts.

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Figure 2 (A) Preoperative photograph showing severe dorsal penile curvature with erect penis lying completely on the abdomen. (B) Postoperative image showing minimal residual curvature.

the ventrally located normal urethra or the bladder, and it was ending blindly at the pubic symphysis region (figure 3A). Retrograde urethrogram through orthotopic meatus and voiding cystourethrography revealed a widened symphysis pubis with normal single urethra with normal bladder neck and no feature of the epispadias–exstrophy complex (figure 3B). Flexible cystoscopy was also performed to exclude any connection with the dorsal tract. MRI showed accessory tract originating from the dorsum of the proximal penile shaft, passing below the symphysis pubis and ending blindly anterior to the urinary bladder with normal bilateral upper tracts (figure 3C, D).

DIFFERENTIAL DIAGNOSIS

A diagnosis of incomplete dorsal urethral duplication with severe dorsal penile curvature was made.

TREATMENT

During surgical exploration through a circumcoronal incision, a 5-cm-long sinus tract and proximal fibrous cord were excised which was extending beyond the pubis and ended blindly behind the pubic symphysis anterior to the urinary bladder (figure 4A, B). Intraoperatively, Gittes and McLaughlin's artificial erection test was performed to look for the degree of curvature (after applying a tourniquet at the base of the penis, heparinised saline (1000 units per 100 cc saline) was injected through a scalp vein needle into the corporal body until full erection was achieved).⁵ At the point of the maximum curvature, two pairs of

Figure 4 (A, B) A 5-cm-long sinus tract followed by a fibrous cord extending beyond the pubis was excised.

Figure 3 (A) Retrograde urethrogram showed a blind-ended epispadiac urethra (arrow) with the normal orthotopic anterior urethra. (B) Voiding cystourethrography revealed no communication between the urinary bladder and the epispadiac urethra. (C, D) MRI showing two accessory tracts (two white arrows) from the dorsum of the proximal penile shaft, passing below the symphysis pubis and ending blindly.

ventral corporal plication were performed, and visible correction of the curvature was noted.

OUTCOME AND FOLLOW-UP

Histologically, the epispadiac urethral tract was lined by stratified squamous epithelium distally with urothelium lining and few smooth muscle bundles proximally confirming the diagnosis of urethral duplication. Postoperative period was uneventful. At 6 months of follow-up, he had a minimal residual curvature (figure 2B) and comfortable sexual intercourse.

DISCUSSION

Urethral duplication is a rare congenital anomaly having multiple described anatomical variants. Duplications in the sagittal plane can be dorsal or ventral, complete or partial, and may be associated with other congenital urogenital anomalies.^{1–3} Duplications occurring in the coronal plane are the rarest and are usually associated with bladder duplication. Based on radiological findings, Effman has classified urethral duplication into (figure 5): type I (incomplete and blind ending) divided into distal, not communicating with urethra (type IA) and proximal, arising from urethra but ending blindly in periurethral tissue (type IB); type II (complete and patent duplication) divided on the basis of number of meatus into type IIA having two meatus, either arising independently from the bladder (type IIA1) or second tract arising from first tract (type IIA2) and type IIB having two urethral channels arising either from bladder or posterior urethra but terminating as a single meatus. There is also a subset of type IIA2 in which the accessory urethral channel opens in the region of perineum known as Y-duplication (type IIA2-Y). Type III is having coronal or caudal duplication.³

Figure 5 Diagrammatic presentation of the Effman's classification of urethral duplication (shaded part is the accessory tract).

Prepubic sinus is a variant in which a tract originating in the skin overlying the pubic symphysis or the base of the penis extending toward the bladder without any communication with it.⁶ Multiple theories have been proposed to explain the embryological aetiology for congenital prepubic sinus, among which the theory suggesting that it is a variant of urethral duplication predominates.⁷ In our case, patient's anomaly was similar to the type IA variant of Effman's classification suggesting it to be a variant of epispadiac urethral duplication. Intraoperatively, dorsal chordee has been observed in patients with epispadiac urethral duplication, but an adult patient presenting with dorsal penile curvature is rarely observed. Similar to the hypospadias, in which the ventral chordee impairs the development of corpora and surrounding structures in the ventral aspect, the fibrous cord located dorsally at the root of the penis in our case may limit the development of the corporal bodies dorsally and thus may have lead to asymmetrical development.⁸ During childhood, the penile growth is not rapid enough to cause a definitive differential growth rates between the dorsal and ventral sides of the corporal bodies, and thus penile deviation is not much appreciated in this age group. However, in our case, due to minimally symptomatic nature of the disease and also due to low medical awareness, the patient presented

to us late in the adulthood when his erected penis used to lay parallel to the abdominal wall. As the duplicated urethral tract ended blindly as a fibrous tract and the distal opening was located at the dorsum of the proximal penile shaft, dorsal hooding of the prepuce was not present but dorsal curvature used to accentuate during tumescence due to the hinge effect of the atretic urethral tract.

Although the primary indication for surgery in a child with incomplete urethral duplication without any voiding symptoms remains cosmesis, parents should be counselled that if it is left untreated and the accessory tract is not excised, it may lead to the development of dorsal penile curvature later. We report this rare case to share our experience which might be helpful in managing such uncommon presentations of these congenital deformities.

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Learning points

- Evaluation of urethral duplication should aim to delineate anatomy, ascertain the functional urethra and identify coexisting anomalies.
- Even if the urethral duplication is incomplete, blind ending and asymptomatic, the hypoplastic urethral tract must be excised as long-standing cases may develop penile curvature.
- Surgical treatment for urethral duplication must be individualised according to the anatomical and radiological findings.
- Both the attending physician and parents should be made aware of such congenital deformities and its long-standing complications.

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