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Unusual presentation of female bladder outlet obstruction- female hypospadias with urethral stenosis

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ABSTRACT

Hypospadias is a rare birth deformity characterised by shortening of urethra with dorsal ectopia of the urethral meatus. The occurrence of hypospadias in female patients is extremely rare. We present a young female complaining of recurrent urinary tract infection and voiding difficulty caused by hypospadias.

Introduction

Ectopic dorsal location of the urethral opening (female hypospadias) is very rare condition causing chronic recurrent cystitis in women because of a retrograde delivery of urogenital infection from the introitus and vagina to the short wide urethra and urinary bladder. Many patients present with voiding difficultly. Female hypospadias can be surgically corrected by transposition of ectopic urethral opening from vagina to the perineum in a sub clitoral location.

Case report

A 14 year girl complained of slow urinary flow and recurrent urinary tract infections (UTI) for the last 6 years. She had very poor flow on uroflowmetry suggestive of urethral stenosis (Qmax 1 ml/sec, voided volume 400 ml). She had achieved her menarche about a year previously. She had history of successful abdominal surgery for sacrococcygeal teratoma at age of 17 days. On clinical examination clitoris and labia majora were normally placed. Vaginal orifice was normally placed. Perineal sensation was normal. The urethral orifice was not located. Abdominal and pelvic examination were normal. Neurological examination of the lower limbs and perineum were normal. CT scan of the abdomen showed bilateral hydroureteronephrosis and MRI examination of the spine were normal. There were no abdominal masses nor spinal anomalies. Uterus was normal.

On examination under anaesthesia the urethral meatus was located with great difficulty on the anterior vaginal wall 2–3 cm proximal to the hymeneal margin buried in rugose vaginal epithelium (Fig. 1). This was sequentially dilated with Teflon and metal dilators to 20 F (Fig. 2). She subsequently voided well with good flow rates but after a few months flow was diminished needing a second urethral dilatation. After meatal dilatation patient greatly improve symptomatically, upper tract dilatation resolve and uroflowmetry show normal flow pattern. Patient didn't have any clinical evidence of neuropathy so that urodynamic evaluation not performed. It was urethral maldevelopment with meatal stenosis, patient was school going adolescent, we have recommended surgical correction to prevent future problems in form of recurrent urinary tract infection when patient become sexually active. In addition we also anticipate problem of recurrent urethral meatal stenosis as location of urethral orifice is abnormal, urethral dilatation is difficult in that situation.

Discussion

In the female hypospadias is a rare entity. The urethral meatus opens on the anterior vaginal wall anywhere between the introitus and the fornix. This may be associated with other anomalies like 46XX Disorders of sexual development (DSD), non-neurogenic neurogenic bladder and urethral duplication. We report a case of idiopathic female hypospadias in a fourteen year old with the urethral opening on the anterior vaginal wall 2–3 cm proximal to the hymeneal ring. She presented with bladder outlet obstruction (BOO) and was treated by urethral dilatation. She will need urethral reconstruction. Urethral folds arising from urogenital sinus mesenchyme fuse medially to form the mammalian urethra.

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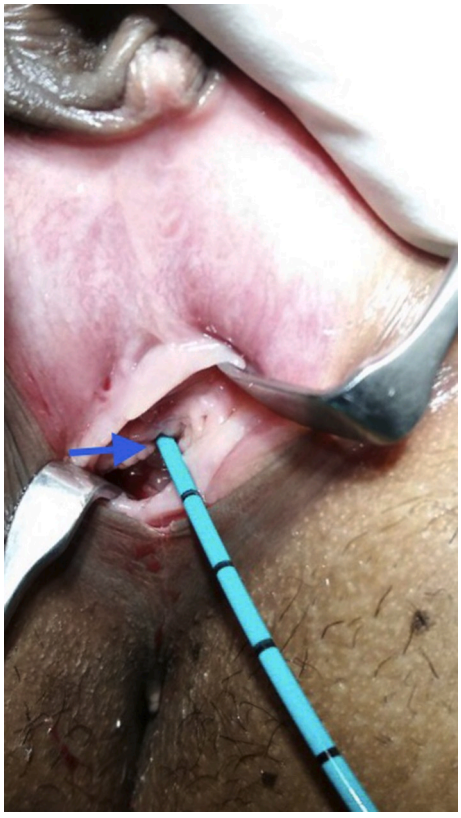

Fig. 1. Showing ureteric catheter placed through abnormally located urethral opening in Anterior vaginal wall.

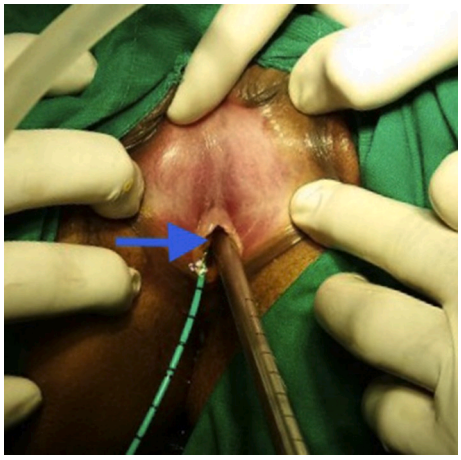

Fig. 2. Showing cystoscope & ureteric catheter in urethral opening through Anterior vaginal wall.

Failure of both urethral-fold fusion and distal urethral migration result in female hypospadias. Embryologically this could represent an arrest during a late stage of urogenital sinus development.[1] Classification of anatomic variants of female hypospadias includes low vaginal ectopia of the external urethral opening; high vaginal ectopia of the external opening of the urethra; vesicovaginal fusion of the neck of the urinary bladder with vagina accompanied with enuresis, urogenital sinus in females (ectopia of the external urethral opening in the urogenital sinus); any of the above variants of female hypospadias in combination with false or true hermaphroditism.[2,3] Late presentation is common because of difficulty in clinical diagnosis. Recurrent cystitis occurs due to exposure of the urinary tract to pathogenic vaginal microflora. Voiding dysfunction may result from stenosis of the meatus.[4] All the variants of female hypospadias may be surgically corrected by transposition of the external opening of the urethra from the vagina to the perineum under the clitoris.[5]

Learning points

Isolated female hypospadias is rare abnormality, usually it is associated with other urogenital abnormalities.

It usually comes to notice when catheterisation try for another reasons.

It may present in different form like recurrence UTI, BOO with or without obstructive uropathy, incontinence.

Neonatal examination in female patient should include finding of three openings in perineum to miss this entity and avoid future complications.

Genital reconstruction using various flap procedures is treatment of choice.

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Declaration of competing interest

None.

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