

CONGENITAL MEGALOURETHRA ASSOCIATED WITH URETHRAL DUPLICATION: A CASE REPORT

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Abstract

A 28 years old male with well developed secondary sexual characteristics was admitted in BSMMU with a grossly swollen, malformed phallus and passage of urine mostly through an opening in the perineum. After birth, his parents noticed that when the baby micturates the phallus is swollen and few drops of urine comes out through the external urethral meatus but most of the urine comes out through an opening in the perineum. No treatment was taken by his parents for this complaint. At puberty his secondary sexual characteristics developed normally. He felt sexual urge towards females but his penis was not erected. At the peak of his sexual excitement, orgasm occurs with release of whitish seminal fluid which comes mostly through perineal opening. His general physical examination was unremarkable & examination of the genitalia revealed well developed scrotum. Both of the testes were normal in size, shape and consistency but the penile shaft was large and flabby, and the ventral aspect of the penis appeared as a sac with mild pseudo phimosis. Careful palpation suggested the absence of the corpus spongiosum & corpora cavernosa. Both these findings were subsequently confirmed by a duplex colour doppler study. The penis distended ventrally when he tried to micturate and the urinary stream was narrow. About 75% of urine flow evacuates through perineal opening which was located about 2 cm from anal opening in the midline ventrally. A voiding cystourethrogram revealed that whole of the anterior urethra was grossly dilated with narrowing at the bulbar part & external urethral meatus. There was another passage (about 5 cm) from the scrotal margin up to proximal posterior urethra. Cystogram showed a bladder diverticulum at left lateral wall. An urethroscopic examination revealed that just proximal to the stenosed external urethral meatus the urethra is widely dilated which seemed like a transparent fusiform sac with irrigating fluid. Whole of the anterior urethra was very thin walled with transmitting lights through it. Proximal part of the urethra near the external sphincter was grossly narrowed. Urethroscopic was also introduced through the abnormal urethral opening into the perineum which was located in the midline raphe of the perineal region about 2 cm from the anal verge. This urethra joined with the penile urethra distal to the external sphincter. Prostatic part of the common urethral channel was normal containing verumontanum and a normal bladder neck.

Key words: *Corpora cavernosa, Urethral duplication, Megalourethra.*

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Introduction:

Urethral duplication and megalourethra both are very rare anomalies and their concomitant presence is extremely rare, with only a few published cases worldwide in the literature[1]. Here we present a case report of concomitant

presence of megalourethra and urethral duplication that was severely stenotic in its bulbar part and meatus, with the ventral urethra grossly dilated.

Case report:

A 28 years old male with well developed secondary sexual characteristics was presented with a grossly

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swollen, malformed phallus and passage of urine mostly through an opening in the perineum since birth. He was the product of a spontaneous vaginal delivery. Since birth his mother noticed that the baby has an abnormally flaccid phallus. When he micturates the fallus is enlarged and few drops of urine comes out through the external urethral meatus but most of the urine comes out through an opening in the perineum. His parents could not avail any treatment for the baby due to financial constraint and he was brought up as such. At puberty, his secondary sexual characteristics developed normally. He felt sexual urge towards females but his penis was not erected. At the peak of his sexual excitement, orgasm occurs with release of whitish seminal fluid which comes mostly through perineal opening. His general physical examination was unremarkable. Examination of the external genitalia showed an abnormality of the penile shaft. The penile shaft was large and flabby, and the ventral aspect of the penis appeared as a sac with mild pseudophimosis. The external urethral meatus was narrowed (Fig. 1). Careful palpation suggested the absence of the corpus spongiosum. Corpora cavernosa were also not palpable. Both these findings were subsequently confirmed by a duplex colour doppler



Fig.-1: Abnormally flaccid phallus.

study of the penis. The penis distended ventrally when filled with urine, and the urinary stream was narrow. About 75% of urine flow evacuates through perineal opening which was located about 2 cm from anal opening in the midline ventrally. A voiding urethrogram (Fig. 2) revealed that whole of the anterior urethra was grossly dilated with narrowing at the bulbar part. Posterior urethra was thin in caliber. There was another passage of contrast (about 5 cm) from the scrotal margin up to proximal posterior urethra. Cystogram showed a bladder diverticulum at left lateral wall. External urethral meatus was stenosed. The serum creatinine and blood urea nitrogen levels were normal.

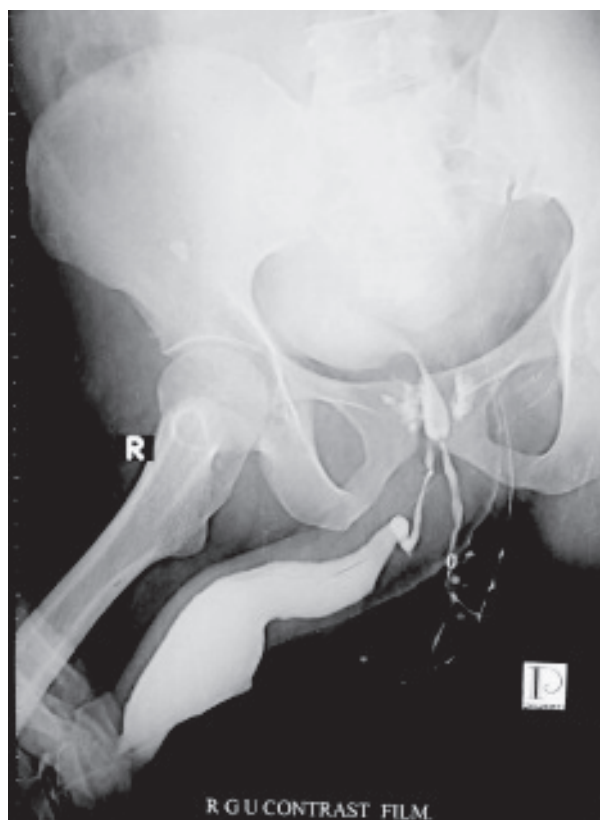


Fig.-2: Voiding urethrogram revealed that whole of the anterior urethra was grossly dilated with narrowing at the bulbar part & another passage of contrast (about 5 cm) from the scrotal margin up to proximal posterior urethra.

An urethroscopic examination (Fig. 3) revealed that just proximal to the stenosed external urethral meatus the urethra is widely dilated; urethra seemed like a transparent fusiform sac with irrigating fluid. Whole of the anterior urethra was very thin walled with transmitting lights through it. Proximal part of the urethra near the

external sphincter was grossly narrowed through which cystoscope could not be negotiated. Urethrocystoscope was then introduced through the abnormal urethral opening into the perineum. This urethral opening was located in the midline raphe of the perineal region about 2 cm from the anal verge. Urethral lining seemed like well developed urothelium. About 5 cm proximal to this opening, this urethra joined with the penile urethra distal to the external sphincter. Prostatic part of the common urethral channel was normal containing verumontanum and a normal bladder neck. Both the ureteric orifices were normally placed in the trigone. Bladder mucosa was normal except a small diverticulum arising from the left lateral wall of the urinary bladder.



Fig.-3: Urethrocystoscopy showed whole of the anterior urethra was very thin walled with transmitting lights through it.

Discussion

Urethral duplication, a rare congenital anomaly, with < 200 cases reported in the literature till date[2]. Most of the reported cases are that of incomplete duplications. Complete duplication is rare only 20 cases have been reported until now[3]. This anomaly is most common in males and very few cases have been reported in females.

The most common is the Y-subtype of duplication⁴. The ventral or perineal urethra in this duplication is usually the most functional and contains the sphincter mechanism, as well as the verumontanum. The dorsal (orthotopic) urethra is typically poorly developed. Y-type duplications are often associated with other severe congenital anomalies such as imperforate anus, cloacal exstrophy, prune belly syndrome, handfoot- genital syndrome, and so forth[4]. Most patients are asymptomatic except for a double stream, though some of them can present with urinary tract infections, incontinence and bladder outflow tract obstruction. A child may be brought, by the mother, just because of the dorsal opening. Genitourinary anomalies and lumbar spine anomalies of the colon have been reported to be associated with this condition. Patients can present with urinary tract obstruction, which in severe cases can progress to chronic renal insufficiency.

The term megalourethra was originally used by Nesbitt in 1955[6], and refers to a rare congenital disorder characterized by primary, non obstructive, diffuse dilatation of the penile urethra. The condition is very rare with only around 100 cases reported in the literature. The urethral dilatation is not secondary to distal obstruction but to poor embryogenic development of the corpus spongiosum. The corpus cavernosum also may be involved depending on the severity of the condition. Megalourethra is usually associated with other congenital anomalies, not only in the urogenital system but in other organ systems, especially in the severe forms. Two varieties of congenital megalourethra have been described—scaphoid and fusiform megalourethra. These can be distinguished according to the severity. In the scaphoid type, there is absence of the corpus spongiosum, so that no support is available for an otherwise normally- formed urethra which results in ventral sacculation of the penile urethra. In the fusiform type (floppy penis), there is either absence or incomplete development of the erectile bodies, so that the urethra may balloon dorsally as well[1].

Various intermediate forms have also been reported; thus, the distinction between these 2 types, although commonly accepted, has not been clearly defined. Therefore, megalourethra should be considered as a spectrum of disease[7]. According to this classification, our patient was diagnosed as having a fusiform megalourethra, based on the physical and radiological findings. Megalourethra is also associated with urinary tract or other congenital anomalies, such are prune belly

syndrome, cloacal exstrophy, renal dysplasia/hypoplasia, hydronephrosis, polycystic kidney, vesicoureteral reflux, ureteral and urethral duplication, cryptorchidism, intestinal malrotation, anorectal anomalies, VATER [vertebral (defects), (imperforate) anus, tracheoesophageal (fistula), radial and renal (dysplasia)] and VACTERL (vertebral, anal, cardiac, tracheal, esophageal, renal, limb) association, and so forth. Urethral stenosis can also be observed⁸.

Conclusion:

Our case of complete urethral duplication associated with megalourethra is an extremely rare and complex case. Only 4 similar cases have been published in the past[1]. Almost every such case is exceptional, and the operative technique must be tailored to each individual case.

Conflict of Interest : None declared

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