Urethral duplication with congenital megacystis and obstructive megaureter – A rare association

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INTRODUCTION

Urethral duplication (UD) in a female is a rare congenital anomaly. Although UD is commonly associated with other congenital anomalies of the urinary tract, its association with congenital megacystis with obstructive megaureter has not yet been reported. We present the case of a 9 year old girl child with complete sagittal duplication of the urethra associated with congenital megacystis and left obstructive megaureter.

CASE REPORT

A girl aged 9 years was investigated for continuous dribbling of urine since with recurrent urinary tract infection (UTI) and occasional pain in the left lumbar region. She also had a normal urge for urination with good stream. Physical examination revealed two urethral openings, one below the other in the sagittal plane, below the clitoris.[Figure 1a,1b and 3b (Diagramatic representation)]. Her renal function tests (RFTs) were normal. Ultrasound scan of the abdomen showed gross hydrourerteronephrosis of the left kidney extending up to the lower ureter and the urinary bladder had a very large capacity with irregular lobulated contours. Intravenous pyelogram (IVP) showed a large capacity bladder with left gross hydrourerteronephrosis and severely tortuous left ureter [Figure 2a] suggestive of left obstructive megaureter. Micturating cystourethrogram also revealed a very large capacity bladder [Figure 2b]. The magnetic resonance imaging of the spine was normal. Cystoscopy showed a double urethra, one above the other in the sagittal plane. The posterior urethra was in the orthotopic position and was opening into the large capacity bladder [red arrow in Figure 1a]. The anterior urethra was present just posterior to the clitoris [anterior/accessory urethra – black arrow in Figure 1a]. The internal opening of the accessory (anterior) urethra was located about 2 cm cranial to the bladder neck on the anterior wall of the urinary bladder (Figure 1c). Green arrow in Figure 1a shows the vaginal opening. Line diagram showing the position of urethra and vagina are given in Figure 3. Excision of the accessory anterior urethra with reduction cystoplasty and left ureteric tailoring with reimplantation was performed in a single setting. The excision of the accessory urethra(Figure 3a) was performed with the child in the lithotomy position through the abdominoperineal route. Since the excised urethra was

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posterior to the clitoris, the nerve supply to the clitoris was not disrupted. The orthotopic urethra and the bladder neck were also safeguarded during the dissection. The resected accessory urethra had transitional cell lining with thin, poorly muscular wall on the histopathology. The girl was followed up with 3 monthly urine microscopy and culture, RFT, ultrasound scan of the abdomen at 6 months, and IVP at the end of 1 year of surgery. Since these were normal, she was followed up with only RFT and ultrasound scan of the abdomen annually for the past 5 years and is doing well.

**DISCUSSION**

UD is a rare congenital anomaly of the urinary tract, the etiology of which is not fully understood. The proposed theories include (i) a persisting cloacal membrane with urogenital tubercle mesoderm displacing posteriorly causing division of the urethral plate and (ii) inflammation or vascular injuries during the development of urethra causing obstruction, urethral plate division, and duplication.[2]

The UD in a female is classified according to the plane (frontal or sagittal) and different anatomical variations: (1) double urethra with double bladder, (2) double urethra with single bladder, (3) accessory urethra posterior to the normal urethra, (4) double proximal urethra with single distal urethra, and (5) single proximal urethra with duplicated distal urethra.[3] Based on this classification, our patient falls in category 2. Majority of the cases reported in the literature had UD in the sagittal plane, as in our patient. Most of the UD in the females were in a subclitoral or supraclitoral position, and were blind ending or stenotic. The duplicated urethra in our case was in the subclitoral position, and was patent, with urinary leakage as this accessory (anterior) urethra lacked a sphincter mechanism. The bladder was of very large capacity without outlet obstruction (congenital megacystis) and due to its large size, it had an irregular contour. However, the cystoscopy showed normal mucosal lining without trabeculations. Continuous leakage of the urine, absence of sphincter mechanism, and obstructive megaureter would have contributed to the recurrent UTI. This association has not been previously reported in the literature so far. The treatment of UD varies depending on the anatomical type and associated urinary tract pathologies. In our case, the girl was treated with excision of the anomalous urethra along with reduction cystoplasty and left ureteric reimplantation after tailoring of the left ureter.
CONCLUSION

Female UD is a rare congenital anomaly leading to continuous urinary incontinence. Excision of the duplicated or accessory urethra along with other necessary procedures gives good results. This is the first-ever report of UD associated with congenital megacystis and obstructive megaureter.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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