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Concomitant anterior urethral valve, distal urethral diverticulum and posterior urethral valve with five-year follow up; A case report and literature review

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ABSTRACT

Concomitant anterior urethral valve and diverticulum (AUVD) and posterior urethral valve (PUV) is an extremely uncommon congenital anomaly that causes infra-vesical obstruction. We present our experience with one case of concomitant AUVD and PUV as well as the related literature review. Early diagnosis and successful management of these anomalies can improve renal function and prevents recurrent urinary tract infections and subsequent renal failure.

1. Introduction

The anterior urethral valve and diverticulum (AVUD) and posterior urethral valve (PUV) are extremely rare congenital anomalies that present with various urinary symptoms. The presentation of urethral anomalies is dependent on the age of the patient and severity of the obstruction.[1,2] High-quality oblique view voiding cystourethrography (VCUG) is the mainstay of diagnostic imaging for obstructive urethral pathologies in children.[3] Here we present a case of concomitant AUVD and PUV with degenerative changes of the right kidney along with a five-year follow-up and a review of the literature.

2. Case presentation

A 3-year-old male patient with uncomplicated birth history and record of recurrent urinary tract infections (UTIs) was referred to our center by a pediatric nephrologist for evaluation of anatomical urinary tract abnormalities. The patient was admitted to the hospital on his seventh day of life due to urinary retention. Per-urethral catheterization was failed so vesicostomy was placed. The ante-grade VCUG demonstrated dilated urethra, suggestive of AUV. Endoscopic fulguration of AUV was done at 6-month-old and vesicostomy was closed one month after the AUV repairing surgery. Kidney-ureter-bladder ultra-sonographies were recommended as post-operative follow-up examination. The decrease of the size and the thickness of the parenchyma of the right kidney and the increase of corticomedullary echo were noticed after 4 months of follow-up. In addition, right hydronephrosis and dilated right ureter with an increased anterior-posterior (AP) diameter of the right pelvis were noted in the follow-up ultra-sonographies. The left kidney appeared normal in ultra-sonographic examinations. The patient was referred to our center due to deterioration of his general condition, hydronephrosis and recurrent UTIs. The initial VCUG demonstrated AUVD and PUV as shown in Fig. 1A.

For further evaluation, dimercaptosuccinic acid (DMSA) and diethylenetriamine pentaacetic acid (DTPA) scans were performed. According to the DMSA-scan, the right kidney was small in size with moderate to severe diffusely decreased parenchymal function. The differential renal function of the right kidney and left kidney were 12% and 88%, respectively. Based on DTPA-scan, the left kidney had acceptable perfusion, function and prominent pyelocaliceal system without significant mechanical obstruction; whereas, the right kidney was small in size with severely decreased perfusion and function with dilated pyelocaliceal system and ureter, raising the possibility of ureterovesical junction obstruction (UVJO). Eventually; after gaining consent form, the patient underwent endoscopic fulguration of the posterior urethral valve (at 5-, 7-, 12-o'clock positions), anterior urethral valve and diverticulum (at 6-o'clock position); also a bladder neck incision (BNI) (at 6-o'clock position) was done due to bladder neck hypertrophy.

The patient was discharged with no complications the day after the

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Abbreviations

AUVD	anterior urethral valve and diverticulum
PUV	posterior urethral valve
AUV	anterior urethral valve
AUD	anterior urethral diverticulum
VCUG	voiding cystourethrography
UTIs	urinary tract infections
AP	anterior-posterior
DMSA	dimercaptosuccinic acid
DTPA	diethylenetriamine pentaacetic acid
UVJO	ureterovesical junction obstruction
MRU	magnetic resonance urography

atrophied right kidney with severe cortical loss, in accordance with previous results, normal-sized left kidney with normal cortical thickness; bladder diverticulum in the anterior portion with 24*15mm diameter; and angulation at the distal part of the right ureter at the UVJ (Fig. 2A, B & C). The above-mentioned findings required no further intervention.

3. Discussion

Among several theories explaining the embryologic basis of developing concurrent anterior and posterior urethral anomalies, the theory of delay in mesenchymal tissue migration and abnormal absorption of the Wolffian duct could clearly explain the mentioned concurrency. The incidence of AUVD is 10- to 30-times lower than PUV. The concomitant occurrence of anterior and posterior urethral valves is extremely rare.[2] To the best of our knowledge, this is the sixth case of concomitant AUD

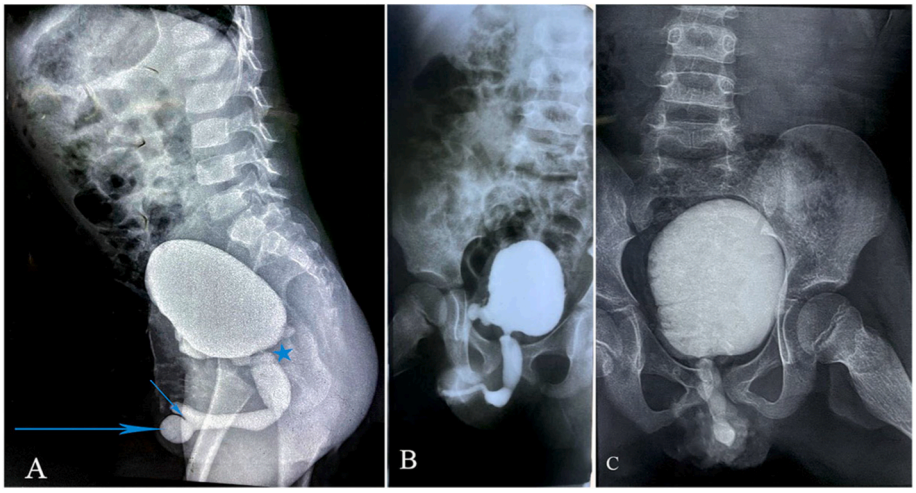

Fig. 1. Voiding phase VCUG: A. Pre-operative voiding phase oblique-view VCUG which was performed at 3-year-old revealing AUVD and PUV, B. voiding phase of VCUG in the first year of follow-up revealing response to the endoscopic fulguration with no signs of AUVD and PUV, C. voiding phase AP view VCUG in the fifth-year follow-up revealed no signs of AUVD and PUV. The asterisk shows PUV, the short arrow shows AUV and the long arrow shows AUD.

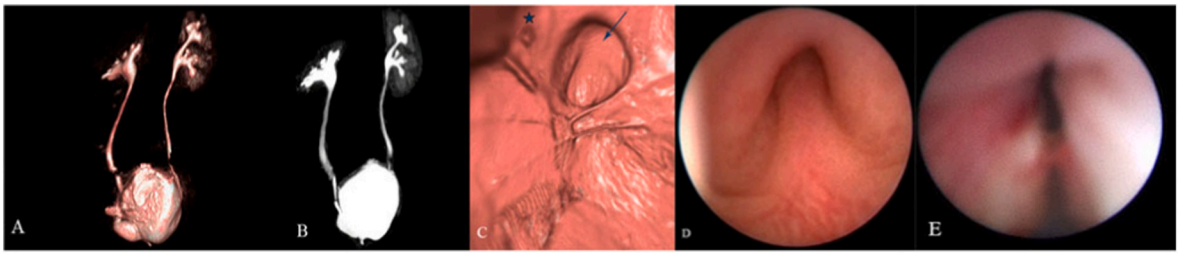

Fig. 2. MRU and virtual cystoscopy: A. MRU view of the urinary tract in the fifth year follow-up demonstrating atrophied right kidney, normal-sized left kidney and angulation at the distal part of the right ureter at the UVJ. B. MRU view of the urinary tract in the fifth year follow-up demonstrating atrophied right kidney with severe cortical loss, normal-sized left kidney with normal cortical thickness and angulation at the distal part of the right ureter at the UVJ, C. Right lateral view of virtual cystoscopy of the bladder in the fifth year follow-up revealing bladder diverticulum in the anterior portion with 24*15mm diameter. The arrow shows the bladder diverticulum in the right lateral aspect of the bladder; the asterisk shows the right ureter orifice. Cystourethroscopic view of urethral anomalies at the time of surgery: D. Cystourethroscopic view of type II PUVs. E. Cystourethroscopic view of hypertrophied bladder neck indicating modified BNI intervention.

operation. The urethral catheter was removed one week later, after voiding per urethra was noted with good caliber. Follow-up was every 3 months in outpatient's clinic for one year and then annually on regular basis except during the COVID pandemic. VCUG was included in the first and fifth year follow-up sessions as demonstrated Fig. 1B and C.

On the last follow-up visit, a magnetic resonance urography (MRU) and 3-dimensional virtual cystoscopy were performed for further evaluation of right kidney function and bladder. The MRU depicted

and PUV and the second case of concomitant AUV, AUD, and PUV as reported in Table 1. This case is reported as another experience in the successful management of a rare congenital urethral anomaly in the literature in the hope of consensus on the management of concomitant AUVD and PUV.[1]

The kidney-ureter-bladder ultrasonography, VCUG along DTPA or DMSA scan should be included in the initial evaluation to determine the renal function.[1,2] Precise cystourethroscopy would be a valuable

Table 1
Literature review of the concurrent anterior urethral valve/diverticulum (AUVD) & posterior urethral valve (PUV).

Literature review of the concurrent anterior urethral diverticulum (AUD) & posterior urethral valve (PUV)							
Source DOI	age	presentation	Ultra-sonography	VUR	Renal scan	management	comment
10.1016/S0022-5347(17)52936-5	0 d	Urinary retention, penoscrotal mass	Bilateral hydronephrosis	–	Dilation of upper tracts	Marsupialization of AUD, Endoscopic fulguration of PUV	High creatinine resolved by surgery
10.1055/s-2005-872916	15 d	Straining during voiding, dribbling, fluctuating penoscrotal swelling	Bilateral hydronephrosis, dilated bladder	Left: grade 5 reflux	Left kidney: no function	Electrocautery ablation of PUV, vesicostomy, primary repair of AUD surgery, left nephrectomy	High creatinine resolved by surgery
10.1016/j.jpedsurg.2012.09.055	2 d	Antenatal hydronephrosis, Weak stream	Bilateral hydronephrosis, severely dysplastic kidney, penile cyst, thick wall small bladder	–	–	Endoscopic resection of the dorsal wall of the diverticulum, resection of PUV, further excision of entire AUD including fistulous tract	High creatinine resolved by surgery
10.1016/j.eucr.2020.101447	18 mo	Weak stream, straining during micturition, febrile UTI, swelling at the ventral penis	bilateral severe hydronephrosis, thick wall bladder	Left: high-grade reflux	–	Endoscopic PUV valve ablation, vesicostomy, open excision of anterior urethral diverticulum involving membranous and bulbar urethra and urethroplasty	High creatinine
10.14534/j-pucr.2021267550	1 d	swelling of ventral penile skin,	Bilateral hydronephrosis predominantly in left kidney, thickened bladder wall, dilation of the proximal urethra	Left: grade 5 reflux	Left kidney: no function	Endoscopic PUV valve ablation, open diverticulum excision & urethroplasty	Not mentioned
Literature review of the concurrent anterior urethral valve (AUV), anterior urethral diverticulum (AUD) & posterior urethral valve (PUV)							
10.1016/j.jpurol.2020.11.002	1 y	Antenatal hydronephrosis, urinary dribbling, recurrent UTI, penile swelling	Bilateral hydronephrosis, thickened wall bladder	–	–	Anterior urethral diverticulectomy, TUR of AUV	PUV fulguration surgery was performed at 1-month-age

VUR, vesicoureteral reflux; AUD, anterior urethral diverticulum; PUV, posterior urethral valve; UTI, urinary tract infection; TUR, transurethral resection.

confirming evaluation in urethral anomalies. Urethral anomalies could be managed through endoscopic *trans*-urethral or open surgical intervention.[2] A gradual urinary obstructive process following bladder dyskinesia presented in patients with PUVs. In younger patients, chronic increase in detrusor pressure may lead to myogenic decompensation. The gradual deterioration of bladder function and larger post-void residues might be secondary to bladder neck obstruction. In the cases of PUV and hypertrophied bladder neck simultaneous BNI and valve ablation can prevent future myogenic failure.[4] Although, in this case previously performed vesicostomy and repeated per-urethral catheterization subsided typical radiologic manifestations of PUV on VCUG; there was evidence of PUVs in cystourethroscopy (Fig. 2 D). In addition, an increase in bladder wall thickness in follow-up ultra-sonographies and hypertrophied bladder neck in cystoscopy (Fig. 2 E) led the surgeon to perform modified BNI to prevent future myogenic failure.

In this case, urethral anomalies especially AUD were all successfully managed using endoscopic fulguration as illustrated by 1-year and 5-year follow-up VCUGs (Fig. 1B and C) which suggests not all AUDs require open surgical intervention. The physician must carefully follow the patient, as it has been reported several times that one anomaly could obscure the symptoms of another.[2] The authors also suggest that a meticulously reconstructed 3-dimensional MRU can strikingly demonstrate the abnormal changes in bladder structure.

4. Conclusions

The concomitant AUVD and PUV can result in urinary retention. The early surgical repair can prevent recurrent febrile UTIs and subsequent renal insufficiency. A precise cystourethroscopy and high-quality oblique view VCUG are the cornerstones of diagnosing urethral anomalies. In the matter of urethral anomaly, urologists must bear in mind the complete evaluation of the urinary tract.

Informed consent

Written consent was obtained by the authors from the legal guardian of the patient prior to submission.

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Declaration of competing interest

The authors declare no conflict of interest.

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