Cent. Eur. J. Med. • 8(3) • 2013 • 343-345
DOI: 10.2478/s11536-012-0083-z

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Central European Journal of **Medicine**

Unrecognized Anterior Urethral Valves as a Cause of Renal Failure

Case Report

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Received 21 January 2012; Accepted 29 June 2012

Abstract: Congenital obstruction of the male urethra is usually caused by posterior urethral valves. Anterior urethral valves (AUV) represent a rare anomaly with a wide spectrum of presentation varying from mild voiding difficulties to end-stage renal disease. Prompt diagnosis and appropriate treatment is essential to prevent renal impairment. We report the case of a 13 month-old boy who presented with deterioration of kidney function caused by unrecognized AUV disorder. Temporary cutaneous vesicostomy was necessary to protect the upper urinary tract from further damage and to stabilize renal function. Even though a voiding cystourethrogram (VCUG) demonstrated obstruction of distal urethra, AUV were initially overlooked but finally diagnosed on additional VCUG followed by urethroscopy.

Keywords: *Anterior urethral valves • Renal failure • Urethral obstruction • Urethra*

1. Introduction

Anterior urethral valves (AUV) represent a fairly uncommon cause of bladder outlet obstruction in the pediatric population [1]. The clinical presentation of AUV is highly variable, and it depends on age of the patient and the degree of obstruction. Examination of the infravesical obstruction is usually focused on posterior portion of urethra because posterior urethral valves (PUV) are a much more frequent congenital anomaly compared with AUV. Unless suspected, AUV can be easily overlooked during cystourethrographic imaging or endoscopic examination. Nevertheless, this rare entity can cause significant urethral obstruction with a severe impact on the urinary tract and subsequent irreversible renal impairment. Therefore, proper and rapid diagnosis and treatment of AUV is essential for preservation of kidney function.

2. Case report

A 13-month-old boy was referred to our institution as a patient with impaired kidney function resulting from vesicoureteral reflux (VUR) that had been unsuccessfully treated by antibiotic prophylaxis. The diagnosis of VUR was made by voiding cystourethrography (VCUG) in a community pediatric center. The patient had a history of three previous urinary tract infections, a weak voiding stream but normal appearance of external genitalia. Two out of the three urinary tract infections occurred under continuous antibiotic treatment that lasted for six months.

After admission to our department, initial laboratory findings confirmed a markedly elevated serum creatinine level indicating deterioration of kidney function, followed by severe electrolyte disturbance. Urine analysis revealed a urinary tract infection. Ultrasonography showed ureterohydronephrosis on the left side, enlargement of left kidney and hyperechogenicity of the renal parenchyma in both kidneys. The bladder wall was moderately thickened and a post-voiding residual volume of 40 mL was noted.

There was no proven difficulty with urethral catheterization, which was undertaken as an emergency treatment. Because there was no sign of recovery of kidney function, a temporary cutaneous vesicostomy was created as a salvage treatment.

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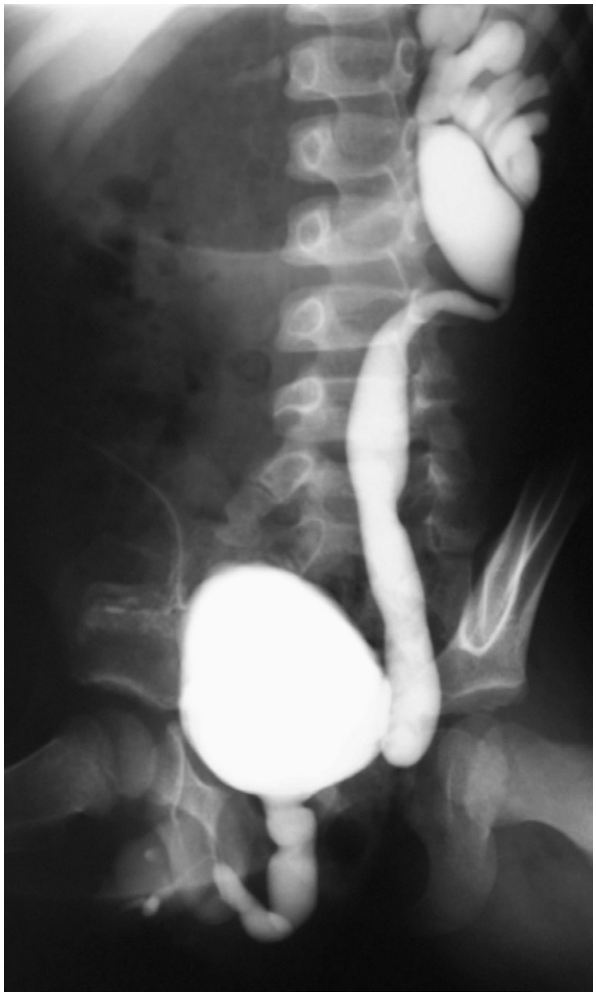
Ten days after the vesicostomy, serum creatinine and electrolytes normalized and a voiding cystourethrogram was performed. VCUG demonstrated left (grade IV) vesicoureteral reflux and dilated posterior urethra with an obstruction at the distal bulbar urethra (Figure 1). Kidney function was subsequently evaluated using renal scintigraphy using dimercaptosuccinic acid, which showed that the right and left kidneys contributed 83% and 17%, respectively.

During cystourethroscopy, cusp-like fibrotic valves at the bulbar part of the urethra were revealed, as well as moderate dilatation of the entire urethra proximal to the valves. There was no urethral diverticulum, urethral stricture or posterior urethral valve shown by the urethroscopy. Hyaluronic acid/dextranomer gel was injected in the left vesicoureteral junction, and simultaneously, a transurethral endoscopic fulguration of the anterior urethral valves was carried out. The patient had an uneventful recovery and became symptom-free on follow-up. The vesicostomy was closed three months later and a significant improvement in patient's voiding stream was noted. A follow-up VCUG demonstrated normal appearance of the urethra and complete resolution of vesicoureteral reflux (Figure 2). Antibiotic prophylaxis was discontinued after the normal VCUG was confirmed. Clinical status, blood tests and urine samples did not reveal any urinary tract infection. Serum creatinine levels remained within the normal range. In fourth year of age when the patient was able to cooperate, uroflowmetry was performed; it revealed a normal voiding pattern.

Figure 1. Voiding cystourethrogram: dilated proximal part of urethra and abrupt change of urethral caliber at the distal part of urethra with vesicoureteral reflux on the left side

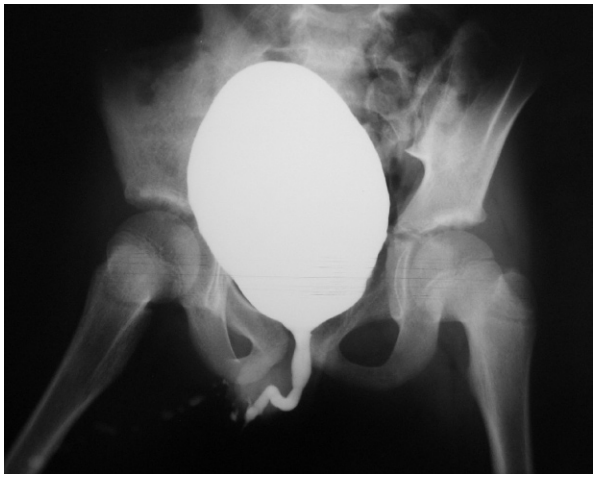


3. Discussion

Urethral obstruction is one of the few life-threatening conditions found in babies. In contrast to the higher prevalence of posterior urethral valves, anterior urethral valves (AUV) remain a fairly rare cause of urinary obstruction. AUV are nine times less common than posterior urethral valves [2]. Other common causes include urethral polyps, bladder and urethral diverticulum, meatal stenosis, ureterocele and bladder stones [1].

Morphologically, AUV appear as diaphanous to thick bands of tissue on the ventral aspect of the urethra, assuming an iris-like, semilunar or cusp-like configuration [3]. AUV can be located anywhere in the anterior urethra. Approximately 40% of AUV are located in the bulbous urethra, 30% at the penoscrotal junction and 30% in the penile urethra [4]. AUV are often present in the form of a urethral diverticulum with a distal wall acting as an obstructive valve, but a saccular diverticulum could be a consequence of valvular obstruction of the urethra [5]. In this case, our patient had isolated AUV without noticeable diverticulum formation that could be the reason for missing proper diagnosis.

Figure 2. Follow-up voiding cystourethrogram: normal appearance of the urethra, no vesicoureteral reflux is present



Firlit et al classified obstructive changes as consequences of AUV depending on the degree of urethral dilatation, the presence of a diverticulum and the grade of upper tract dilatation. Type I is represented by AUV and proximal urethral dilatation, type II includes urethral diverticula formation, in type III there is a combination of valve, diverticulum, proximal urethral distention and vesical enlargement but not massive ureterectasis, and type IV is represented by severe changes of upper urinary tract [6]. Undoubtedly, severity of urinary tract impairment correlates with degree and duration of obstruction.

Clinical manifestations may vary from mild voiding difficulties to hydroureteronephrosis and severe renal impairment. If a diverticulum is associated, ballooning of the penile urethra may be visible during voiding and dribbling of the urine can occur. Recurrent urinary tract infections, particularly if combined with straining to void and a weak voiding stream should evoke a high suspicion of urethral valves. In neonates, AUV, as well as other infravesical obstructions, may present as failure to thrive, biochemical disturbances, hydronephrosis, urinary ascites, urosepsis and end-stage renal disease.

VCUG and retrograde urethrography are very helpful imaging methods for defining the anatomy of male urethra [7]. Typically, on VCUG the urethra is dilated proximal to the obstructive valve and narrow distal to it. Demonstration of the entire anterior urethra during the voiding phase of VCUG is crucial; otherwise AUV may easily be missed. A concomitant diverticulum usually facilitates diagnosis; however, our patient had no diverticulum formation but rather uniform dilatation of entire proximal urethra. Vesicoureteral reflux can be found in up to one-third of patients with AUV [5].

Sonography can be used for diagnosis of AUV associated with a urethral diverticulum [8]. Finally, cystourethroscopy represents the diagnostic tool for visualizing and confirming the true cause of urethral obstruction.

Surgical correction is essential for management of AUV. Transurethral fulguration or resection are effective treatments in most cases, but sometimes result in severe urethral stricture, often with incomplete valve resection in very young patients. In cases where an associated diverticulum is present, open repair with diverticulectomy and urethroplasty is reasonable treatment.

A temporary cutaneous vesicostomy could be necessary in patients with severe renal impairment. Our patient underwent vesicostomy as a salvage treatment because clinical and laboratory findings indicated severe refractory deterioration of kidney function as a result of the unrecognized AUV.

AUV are rare, but may cause severe urethral obstruction in a silent and very rapid manner leading to end-stage renal disease. We would like to emphasize necessity for evaluation of the anterior urethra in any male child with suspected urinary tract obstruction. If AUV are detected, prompt and appropriate treatment is essential for salvation of kidney function.

Acknowledgments

Supported by the Ministry of Science, Republic of Serbia, Project No. 175048.

References

[1] Casale A., Posterior Urethral Valves and Other Urethral Anomalies, In: Wein A., Kavoussi L., Novick A. et al. (Eds.) Campbell-Walsh Urology, 9th ed. WB Saunders, 2007

[2] Williams D.I., Retik A.B., Congenital valves and diverticula of the anterior urethra, Br. J. Urol. 1969, 41, 228-234

[3] Arena S., Romeo C., Borruto F.A., et al., Anterior urethral valves in children: an uncommon multipathogenic cause of obstructive uropathy, Pediatr. Surg. Int., 2009, 25(7), 613-616

[4] Narasimhan K.L., Choudhary S.K., Kaur B., Anterior urethral valves, Indian. Pediatr., 2005, 42, 708–710

[5] Zia-ul-Miraj M., Anterior urethral valves: a rare cause of infravesical obstruction in children, J. Pediatr. Surg., 2000, 35, 556–558

[6] Firlit R.S., Firlit C.F., King L.R., Obstructing anterior urethral valves in children, J. Urol., 1978, 119, 819–821

[7] Levin T.L., Han B., Little B.P., Congenital anomalies of the male urethra, Pediatr. Radiol. 2007, 37(9), 851-862

[8] Goyal M., Sharma R., Gupta D.K., et al., Congenital anterior urethral diverticulum: sonographic diagnosis, J. Clin. Ultrasound., 1996, 24, 543–544