Case Report

Anterior Urethral Valve and Diverticulum in a Neonate with Febrile Urinary Tract Infection

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Anterior urethral valve (AUV) was first described by Watts in 1906 as a cause of urethral obstruction. Since then, a few cases have been reported in which AUV led to variable urinary tract symptoms [1]. If left untreated, AUV can also result in end-stage renal disease [2]. As a result, AUV should be immediately evaluated and managed.

We present a case of a neonate with an AUV with diverticulum that was successfully treated by open surgery.

CASE REPORT

A 15-day-old boy was referred to our urologic department for evaluation of a febrile urinary tract infection that developed 2 days prior to presentation. He was born at 38 weeks of gestation via spontaneous vaginal delivery and weighed 3,200 g. The pregnancy and routine prenatal ultrasound (US) had been normal. His body temperature was 38°C. He presented with urinary dribbling. The physical examination demonstrated a soft and palpable mass at the penoscrotal junction that collapsed completely on manual pressure and emptied its contained urine through the urethra. Urinalysis showed pyuria with a positive nitrite result and significant culture of Staphylococcus aureus (>10⁵ CFU/ml). The complete blood count revealed leukocytosis. Routine investigations included an unremarkable blood urea nitrogen, serum creatinine, and electrolyte panel. Abdominal US examination displayed a thick-walled bladder (7 mm) with normal kidneys. Transpenile US revealed...
an AUV with diverticulum (Fig. 1). A voiding cystourethrogram (VCUG) showed an anterior urethra diverticulum with dilation of the proximal urethra but no vesicoureteral reflux (Fig. 2). We tried to operate by means of endoscopy, but it was not possible owing to the small caliber of the urethra. Open diverticulectomy and urethroplasty was performed with complete excision of the obstructing AUV, and a 5 Fr feeding tube was subsequently placed (Fig. 3). After the catheter was removed postoperatively at 7 days, a normal urinary stream was visualized. Three months after the operation, a retrograde urethrogram showed no urethral abnormalities (Fig. 2).

**DISCUSSION**

AUV is a congenital mucosal fold located distally to the membranous urethra. The sites of an AUV can be anywhere distal to the membranous urethra and are reported in the following frequency: bulbar urethra (40%), penoscrotal junction (30%), pendulous urethra (30%), and occasionally in the fossa navicularis [3].

Even though the exact etiology is unclear, various theories have been proposed as possible mechanisms since the first report of AUV in 1906 by Watts. AUV may represent an abortive attempt for urethral duplication in the first 12 to 14 weeks of intrauterine life. Another possible theory is that AUV results from the sequestration of an epithelial...
nests after closure of the urethral folds. Incomplete focal development of the corpus spongiosum with bulging of the urethral mucosa due to ineffective support, defective fusion of the ventral folds over the urethral groove, and cystic dilatation of the urethral glands are suggested as plausible mechanisms for the development of AUV and ruptured cystic dilatation of Cowper's duct cyst [4]. In our case, the microscopic view of the resected urethral diverticulum showed complete development of the corpus spongiosum (Fig. 4).

AUV and diverticulum are rare congenital urethral anomalies that can lead to penile swelling, urethral obstruction, urinary retention, incontinence, nocturnal enuresis, bladder rupture, and end-stage renal disease [5]. Depending on the severity of the anatomical obstruction, it may present soon after birth or later in childhood [6].

The severity of an AUV can be classified into 4 classes. The simplest form is seen in type I, which consists of a demonstrable AUV associated with proximal urethral distortion. Type II obstruction is associated with the presence of a definitive urethral diverticulum. Type III obstruction is a combination of valve, diverticulum, proximal urethral distention, and vesical enlargement without massive ureterectasis. Type IV obstruction represents severe changes of the upper tract [4]. The case presented here was indicative of a type II obstruction.

VCUG remains the most important imaging technique for the evaluation of urethral abnormalities and confers the additional advantages of demonstrating associated diverticulum, megacystis, vesicoureteral reflux, or other associated anomalies in the proximal urinary tract. Typically, the urethra appears dilated proximal to the valve and narrow distal to it. If no abnormality is detected on VCUG, a retrograde urethrogram should be performed. However, it should be noted that the valves are likely to be missed on a retrograde urethrogram, because they may remain open with retrograde flow. Cystourethroscopy may show cusp-like valves in the anterior urethra, although it is less accurate because retrograde flow induces the valve to lie flat against the urethral wall.

The early diagnosis of AUV appears advantageous in neonates if they have more severe urinary obstruction, as indicated by greater degrees of hydronephrosis, renal dysfunction, and the presence of urinary ascites at birth in the most severe cases. Thus, the routine use of prenatal ultrasonography will probably alter the mode of presentation and clinical outcomes [7,8].

Most recently, the majority of authors agree that transurethral valve ablation is the treatment of choice for AUV, because the new small diameter pediatric urethroscope can be used for valve ablation in neonates [9]. This minimally invasive transurethral approach has become feasible through improvements made in endoscopic instruments since the 1990s. In general, open reconstruction is recommended for large diverticula with inadequate spongiosum or in the case of a poorly emptying urinary tract, especially in association with urinary tract infection [10]. Because our patient presented with a febrile urinary tract infection owing to his poorly emptying urinary tract and a large diverticulum, a surgical approach with 1-stage open reconstruction was pursued.

Congenital anterior urethral obstruction in children has a generally good prognosis but may occasionally result in poor renal outcomes, such as renal insufficiency, renal failure, or patient death. We performed a successful open reconstruction of the valve with no postoperative complications such as extravasation of urine, stricture formation, urethrocystostomy, or other complications. AUV is a rare congenital anomaly that is often difficult to diagnose. AUV is an uncommon cause of lower urinary tract obstruction and renal failure. All patients with poor stream and recurrent urinary tract infection should be carefully evaluated and immediately treated.

CONFLICTS OF INTEREST
The authors have nothing to disclose.

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