An unusual presentation of anterior urethral valve in a child with diabetes mellitus

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ABSTRACT

Anterior urethral valve (AUV) is identified to be a common source of congenital obstructive lesion of the anterior urethra. Up to 80% of children with AUVs develop bladder dysfunction, bladder instability, hyperreflexia, diminished compliance and capacity. We report a case of an unusual presentation of a child with AUV and diabetes mellitus.

Key words: *Anterior urethral valve, Nocturnal enuresis, Pretreatment azotemia, Urinary tract obstruction*

Anterior urethral valve (AUV) is known to be a common cause of congenital obstructive lesion of the anterior urethra [1]. This condition is often found in association with a large anterior urethral diverticulum, and the valve itself results in obstructing the urinary flow [2]. The embryology of AUV is not clear and may represent a faulty union between urethral mucosa and the epithelium of the fossa navicularis. A rupture of dilated bulbourethral glands has also been suggested as an etiology [3]. The valves may be located at the bulbar urethra, the penoscrotal junction, or the penile urethra [4].

Children with AUV present clinically at different ages depending on the severity of the obstructive process. Symptoms may consist of poor urinary flow, post-void dribbling and mild incontinence, significant bulging of the distal penis, palpable bladder with obstruction or even renal insufficiency, and urinary tract infections [5]. Diagnosis is usually made after a careful examination of the external genitalia, and compression of the distal shaft may result in expressing of urine as seen in a diverticulum. A voiding cystourethrogram (VCUG) is required to confirm the diagnosis and may demonstrate a dilated anterior urethra with proximal signs of chronic obstruction, including bladder diverticula and massive vesicoureteral reflux (VUR). Up to 80% of children with AUVs develop bladder dysfunction, bladder instability, hyperreflexia, diminished compliance, and capacity [6]. Pretreatment azotemia, VUR, and urinary tract infection together increased the risk of poor renal outcome in these children [7]. Children with milder and more subtle presentation usually have preserved renal function, and the outcome is better than that in posterior urethral valves, with 78% of patients having a normal renal function after treatment [7]. We report a case of an unusual presentation of a child with an AUV.

CASE REPORT

A 13-year-old male child presented to the pediatric urological services with a chief complaint of secondary nocturnal enuresis for 3 months. The child was diagnosed to have juvenile diabetes mellitus at the age of 5 years, and since then, he was on injectable insulin. The child had other symptoms of poor flow, post-void dribbling, and incomplete emptying of the bladder. The child was conscious, normal-built with normal intellectual abilities. Clinical examination did not reveal any abnormal findings before voiding, during voiding, and after voiding. His systemic including neurological examination was also normal.

On investigation, serum creatinine was found to be 1.67 mg% and blood hemoglobin was 10.8 g%. Ultrasonography revealed bilateral hydronephroureterosis with significant post-void residue. Uroflowmetry showed a maximum urinary flow rate of 3.8 ml/s (Fig. 1), voided volume of 68 ml, and post-void residue of 276 ml. Magnetic resonance imaging showed bilateral moderate hydronephroureterosis, tortuous ureters, and distended and elongated bladder with multiple diverticula (Fig. 2). Cystometry revealed a poorly complaint bladder with an obstructive flow (Fig. 3).

The child was taken up for the cystoscopy to rule out mechanical obstruction to the flow of urine. Cystoscopy revealed an epithelial bulge (Fig. 4) in the proximal bulbar urethra arising from the floor (ventral) and becomes prominent, whenever pressure was applied on the lower abdomen. No obvious opening could be made out. The bulge was punctured using a hook electrode and cutting current. The puncture was extended to open the bulge, releasing turbid contents. An 8 Fr catheter was introduced for 48 h. The

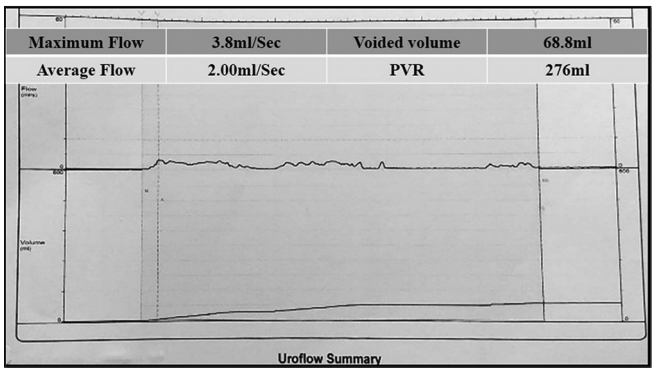

Figure 1: Uroflowmetry showing a maximum urinary flow rate of 3.8 ml/s

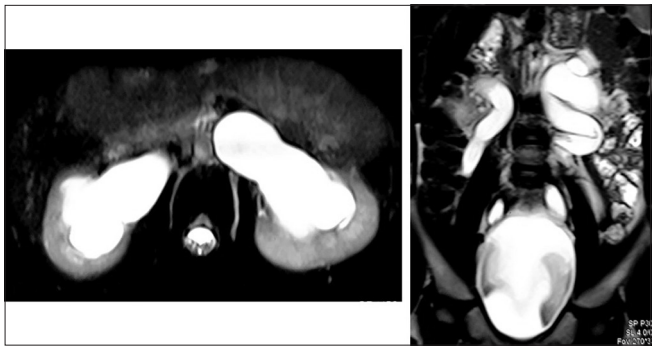

Figure 2: Magnetic resonance (MR) imaging showing bilateral moderate hydronephroureterosis, tortuous ureters, and distended and elongated bladder with multiple diverticuli

turbid contents were sent for culture and revealed the growth of *Escherichia coli* which was sensitive to most antibiotics. The child received antibiotics for 15 days.

Postoperatively (1 week later), the serum creatinine came down to 1.3 mg% and the child voided with good flow following the removal of the catheter. Currently, the child is on regular follow-up for the past 6 months. The child is voiding well and did not have any episodes of urinary tract infection.

DISCUSSION

AUV is a rare but well-known congenital anomaly [2]. It can occur as an isolated entity or in association with a proximal diverticulum, probably representing a spectrum of the disease. AUV and diverticula can cause severe obstruction, whose repercussions on the proximal urinary system can be severe and important. The clinical examination in these children often detects a highly distended bladder and lumbar mass. It is very important to observe during the act of micturition, more particularly, so as to observe diverticula or deformation of the penis or the appearance of a penoscrotal swelling, with post-void dribbling. Pressure on this swelling enables the emptying of its urinary contents.

The child in our report presented with lower urinary tract symptoms with nocturnal enuresis of recent origin being the most bothersome symptom. Moreover, the child was a known case of juvenile diabetes mellitus which made us think in terms

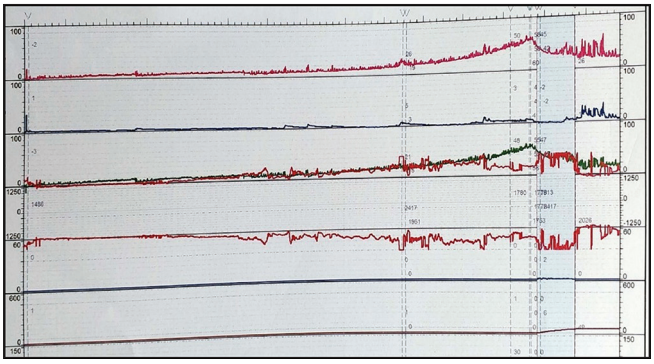

Figure 3: Cystometry revealed poorly complaint bladder with obstructive flow

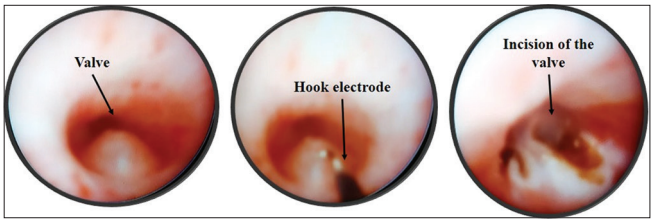

Figure 4: Cystoscopy revealed an epithelial bulge

of neurogenic diabetic vesicopathy or dysfunctional voiding syndrome. Children with juvenile diabetes can present with similar symptoms. This overlay of symptoms can confuse the diagnosis. It is better to evaluate these children in detail including urodynamic studies and visual imaging with cystoscopy. This child had no penile/penoscrotal swelling or abnormality. It was the findings on cystometry that made us look further for an obstructive cause. The appearance of a mucosal swelling in the proximal bulbar urethra clinched the diagnosis in this child.

VCUG is the diagnostic investigation of choice in the diagnosis of an AUV. It would reveal a dilated or elongated posterior urethra, a dilatation of the anterior urethra, a thickened trabeculated bladder, a hypertrophied bladder neck, VUR, and urethral diverticula. The urethra appears dilated proximal to the valve and narrowed distal to it on VCUG. A valve may be demonstrated on retrograde urethrogram as a linear filling defect along the ventral wall, or it may show a dilated urethra ending in a smooth bulge or an abrupt change in the caliber of the dilated urethra on VCUG [8]. VCUG may also reveal an associated anomaly in addition to demonstrating the valve. Urethroscopy usually helps in confirming the diagnosis. The treatment includes the destruction of the valve by electrocautery or by a resecting hook [9,10].

CONCLUSIONS

Congenital AUV is an uncommon but important cause of intravesical lower urinary tract obstruction. Clinical presentation depends on the severity of obstruction. Occasionally, they may present with unusual symptom of nocturnal enuresis and may not have the telltale sign of abnormal penile swelling.

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