CASE REPORT

Missed Diagnosis of Anterior Urethral Valve labelled as Neurogenic Bladder

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ABSTRACT

Anterior urethral valve (AUV) is a long known but rare anomaly, which is occasionally encountered in boys with obstructive uropathy. We present a case of Anterior urethral valve which was misdiagnosed as neurogenic bladder and was planned for Augmentation cystoplasty with catheterizable stoma elsewhere. This article highlights the frequently missed diagnosis of Anterior urethral valve by most practitioners because of its rarity. JMS 2015; 18(2):150-152

Key words; Anterior urethra, valve, neurogenic bladder

INTRODUCTION

Anterior urethral obstruction is rare compared with posterior urethral valves. Although these obstructive structures may be called valves, they often occur in the form of a diverticulum of the urethra with one wall acting as in the form as obstructive" valve." A ratio of the incidence of anterior urethral valves relative to that of PUV is 1:8; PUV occurs in 1 in 8000 to 1 in 25,000 live male births¹.

As classically described, these take the form of either a fenestrated diaphragmatic membrane or a mucosal cusp arising from the ventral wall of the urethra. This fold enlarges during micturition and is flattened against the urethral roof, becoming a potentially serious obstacle. The effect on the proximal urethra is variable and appears to be separate from bladder and upper urinary tract repercussions.²

Case History

8 year old male child with H/o inability to achieve normal voiding since birth, dribbling day and night. Passed urine

very rarely on his own. H/o recurrent febrile urinary tract infection (UTI) since last 1 year. Pattern of voiding could not be assessed. He gave history of fall with fracture neck of right femur, operated on 2010. No history of constipation. On Examination growth, activity and intelligence were normal. No signs of nervous system involvement. He was circumcised. His baseline investigations were normal with creatinine 1.0 mg per decilitre. X ray KUB region was normal. Ultrasonography abdomen revealed bilateral hydronephrosis, left more than right side with small volume trabeculated bladder (30cc). DMSA scan showed Left kidney 27 percent (%) split function with moderate to severe impairment of function and diffuse cortical scarring. Right kidney showed 73% split function with mild impairment of function. MRI spine was normal. VCUG (vesicocystourethrogram) was showing grossly trabeculated bladder with grade 4 vesico ureteric reflux (VUR) on left side with dilated posterior urethra and abrupt change in caliber of urethra at peno-scrotal junction (Fig-1). Urodynamic study showed peak flow rate of 6ml/minute with low capacity, low compliance and high pressure unstable urinary bladder with no demonstrable DSD (detrusor sphincter dysynergia). Cystoscopy was done twice elsewhere which showed severely trabeculated

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urinary bladder with wide open left UO (ureteric orifice), normal right UO, and normal anterior and posterior urethra.

After reviewing previous records especially VCUG, and in view of no evidence of neurogenic component we decided to subject him to cystoscopy again which revealed an anterior urethral valve extending from 3 to 9° clock position in the penile urethra (Fig.2), with grossly trabeculated bladder and patulous left UO. Valve was incised at 6° clock position by Sachse's knife and 12 french two way foley catheter indwelled.

Post operatively Catheter was removed after 4 days. He voided with good flow and was dry for over one hour on the same day with a satisfactory uroflowmetry (Fig.3). He is still on our follow up since last 1 month and is voiding at regular intervals of 2-3 hours and is on prophylactic antibiotics.

DISCUSSION

Anterior urethral valves are rare causes of obstructive uropathy in boys. They are an important cause of urinary tract obstruction in children and can be easily missed by an unaware clinician^{3,4,5}. They occur 8 to 10 times less frequently than PUV. Anterior urethral valves are congenital in nature and can be located anywhere in the anterior urethra viz., approximately 40% in the bulbar urethra, 30% at the peno scrotal junction and 30% in the penile urethra. Some of them exist without an associated diverticulum⁶. The clinical presentation of anterior urethral valves is similar to that of PUV. The spectrum ranges from mild urethral dilatation to bilateral hydronephrosis with renal insufficiency. Anterior urethral valves can cause histological changes in the bladder similar to PUV and produce valve bladder like picture. The presentation in the developed countries is usually as an antenatally diagnosed hydronephrosis because of routine antenatal screening. However, in our country the presentation is usually seen in older children with obstructive urinary symptoms, and/or urinary tract infections. Symptoms are principally dependent on age at time of presentation and degree of obstruction.⁷ These patients commonly have dribbling of urination, difficulty in voiding, incontinence, hesitancy or urinary retention, poor urinary stream, and recurrent urinary tract infections.⁸ Older children may also have enuresis, postvoid dribbling, or failure to thrive. An early, precise diagnosis and awareness of the AUV in boys with obstructive symptoms can reduce incidence of advanced uropathies.⁸

It is recommended that a VCUG be done for diagnosis, since the valves are likely to be missed on retrograde urethrography, as they remain open with retrograde flow. The whole of the anterior urethra must be included in the field of exposure while doing VCUG in order to avoid missing the site of obstruction. The valve itself may be visualized as a linear defect along the ventral wall but also may only be noted as an abrupt change in the caliber of the urethra. Vesicoureteral reflux has been noted in approximately one-third of patients, and upper tract dilation noted in one-half. In our case, the patient was being managed as a case of neurogenic bladder with normal MRI lumbo sacral spine. A high index of suspicion is essential to diagnose AUV at an early stage. This is particularly so when more common PUV is not seen during cystoscopy.

Endoscopically, the valve appears as a filmy, ventrally located cusp or flap of tissue. It occasionally has been described as an iris-like membrane. One must examine the urethra carefully at the distal end of any urethral diverticulum because retrograde flow of irrigant may flatten the valve mechanism against the urethral wall. Suprapubic pressure with the bladder full and irrigation ports on the scope open may demonstrate the valve mechanism more readily⁹. Furthermore, elevation of the valve with an endoscopic loop is invaluable in the identification of the lesion. Treatment is by endoscopic incision. Transurethral incision of the valve is effective for most patients, but one must be careful to aggressively incise to avoid any distal obstructing lip. If the urethra is of sufficient caliber and support, transurethral fulguration is recommended. Open urethroplasty is useful in patients with a large urethral diverticulum, urethral duplication and thin urethra.

CONCLUSION

In children with infravesical obstruction, A high index of suspicion is essential to diagnose AUV at an early stage, particularly when PUV is absent and radiographic evidence of neurogenic bladder is present in a neurologically normal child.

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Fig.1: VCUG showing grade 4 left side VUR with Pine tree urinary bladder typical of neurogenic bladder.

Fig.2: Cystoscopy showing Anterior urethral valve in penile urethra extending from 3 to 9 °clock position ventrally.

Fig.3: Post operative uroflowmetry showing peak flow 11.8ml/sec, voiding volume 94ml and voiding time 20sec.