

A new born with anterior urethral diverticulum and posterior urethral valve

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Introduction

The presentation of posterior urethral valve (PUV) and anterior urethral diverticulum (AUD) or valve (AUV) is rare and few cases reported in current literature. Both congenital anomalies lead to bladder outlet obstruction. Therefore, early surgical interventions to prevent further renal damage and renal failure are needed. The association of the PUV and AUD fits the theory of a wide spectrum of mesenchymal defects including megalourethra.

Case presentation

A newborn baby was transferred from the peripheral hospital for further management of posterior urethral valve and bulge at the penoscrotal junction. Antenatal history revealed that anomaly scan identified bilateral hydronephrosis and hydroureter with suspected PUV. During the neonatal examination, a bulging was noted at the site of the penoscrotal junction with a poor stream of urine and palpable bladder with ballotable kidneys. Consequently, blood examinations revealed high serum creatinine 280-300mg/dl while other blood reports were normal.

Furthermore, the ultrasound scan showed bilateral hydronephrosis and hydroureter with suspected PUV and due to rising serum creatinine with reduced urine output suprapubic cystostomy was performed on 6 days of age. However, reduced urine output was noted from the suprapubic catheter and as a result, bilateral ureterostomy was performed at 14 days of age to relieve the obstructive uropathy. Moreover, antegrade cystourethrogram was arranged after ureterostomy creation. It revealed the AUD at the bulbar urethra, posterior urethral valve and vesicoureteric reflux on both sides (Figure. 1).

Subsequently, the serum creatinine level was reduced to normal and at the same time, there was pus discharge through

the urethra from the infected AUD. As a result, urethrostomy was created over the AUD at day 26 of age (Figure. 1). Thereafter, there was no evidence of urinary tract infection and serum creatinine level was normal (48mg/dl) up until the age of one when cystoscopic guided posterior urethral valve ablation (Figure. 2) and ureterostomy closure was done. Excision of the AUD and repair of the urethra will be done later.

Discussion

Lower urinary tract obstruction in children can occur due to AUV or diverticulum or posterior urethral valve. It can be presented as an isolated entity or together with a proximal diverticulum; illustrating the spectrum of the disease [1].

AUD may occur without AUV as seen in our case. PUV is the main cause of bladder outlet obstruction in children. The association of PUV with anterior urethral valve (AUV) is uncommon. In this case, the posterior urethral valve and anterior urethral diverticulum were diagnosed/presented at birth.

Various proposed hypotheses have been described which can range from “an abortive attempt at urethral duplication, failure of alignment between the proximal and distal urethra, imbalanced tissue growth in the developing urethra leading to a remnant of excess tissue acting as a valve and congenital cystic dilatation of periurethral glands leading to a flap-like valve (1)”. AUVs can occur at the level of the bulbar urethra (40%), pendulous urethra (30%) and penoscrotal junction (30%) [1]. Our patient showed no evidence that AUV is the cause of diverticulum. PUV occurs at the level of the verumontanum (membranous urethra).

The observed clinical manifestation is dependent on the age of the patient and the degree of obstruction, which increases the variability in the manifestation. It may present with bilateral hydronephrosis and hydroureter. Additionally, voiding cystourethrogram (VCUG) can also show a dilated urethra with abrupt calibre change or ending in a smooth bulge [2].

VCUG serves as the diagnostic modality in the diagnosis of AUV. It can reveal a urethral calibre change such as the dilated or elongated posterior urethra, dilatation of the anterior

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Figure 1 a. Antegrade CUG shows posterior urethral valve and anterior urethral diverticulum and B/L VUR

Figure 1 b. Bilateral ureterostomy and urethroscopy over anterior diverticulum

urethra, and urethral diverticulum. Apart from these it shows thickened trabeculated bladder, a hypertrophied bladder neck and vesicoureteric reflux (VUR) [2]. In our case, the patient had bladder outlet obstruction and bilateral reflux with renal impairment. Therefore, the patient had suprapubic cystostomy and bilateral ureterostomy. Cystourethroscopy is beneficial in confirming the diagnosis which would precede the treatment that comprises of the destruction of the AUV and PUV by electrocautery or by a resecting hook [3].

In our case, cystourethrogram and cystourethroscopy led to diagnosis and resection of the posterior urethral valve and urethroscopy was done for anterior urethral diverticulum which needs excision later. To prevent further renal damage, infection and vesicoureteral reflux [4], it is very important to diagnose this rare condition earlier in order to efficiently manage it.

Conclusion

Our baby presented with a bulge at the penoscrotal junction and bladder outlet obstruction at birth with renal impairment. Suprapubic cystostomy was performed to relieve obstruction and antegrade cystourethrogram had confirmed AUD and posterior urethral valve with bilateral reflux. These rare presentations were managed simultaneously. Therefore, VCUG is the diagnostic modality in bladder outlet obstruction in newborn.



Figure 2. Cystoscopic views of posterior urethral valve and anterior urethral diverticulum

All authors disclose no conflict of interest. The study was conducted in accordance with the ethical standards of the relevant institutional or national ethics committee and the Helsinki Declaration of 1975, as revised in 2000.

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Learning Points:

- There were few reported cases in the literature with posterior urethral valve and congenital anterior urethral valve but there are no reported cases of AUD and PUV in literature.
- Even though it is rare, the possibility of the posterior urethral valve in association with anterior urethral diverticulum should be considered in differential diagnosis whenever a neonate presents with bladder outlet of obstruction.
- In this case, the emergency and subsequent long term management of both pathologies, while safeguarding renal function, is a challenge.