Unveiling the Menace of Incidental Insidious Chronic Kidney Disease due to Posterior Urethral Valve - A Case Report

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Key Clinical Message:

Posterior Urethral Valve should be considered as a possible diagnosis for chronic kidney disease in any child, even if their age at presentation is not typical, and even if the initial diagnostic assessment was normal.

Keywords:

Posterior Urethral valve, Chronic Kidney Disease, Pediatrics, Nephrology, Urology.

Introduction:

Posterior Urethral Valve (PUV), the etiology of which is not fully understood, is considered the most prevalent congenital cause of bladder outlet obstruction in male children, with an incidence of 1 in 3800.[1–3]

PUV can lead to enuresis, urosepsis, chronic kidney disease, and even death. It is very rare for it to present as macroscopic hematuria.[4,5]

In 40% of cases, PUV is associated with other malformations including aneuploidy, cardiac anomalies, and gastrointestinal anomalies.[1] Diagnosis can be made prenatally through sonography.[3,5] However, some cases are detected postnatally during investigations for other causes.[5]The median age at which patients present is 5 months; presentations at older ages are considered unusual.[4]

The primary management focuses on protecting the kidneys through transurethral or suprapubic catheterization for drainage. After that, the main treatment is primary valve ablation through transurethral incision. If there is difficulty in removing the valve, vesicostomy, ureterocutaneostomy or nephrostomy can be performed. Complications include irreversible hypoplastic and dysplastic kidneys as well as lifelong bladder dysfunction and chronic kidney disease.[3]

It is rare to have follow-up cases as the majority of patients believe that valve ablation is a complete solution.[4]

What makes this reported case unique is the age at which the patient presented and the main complaint of seizures. During evaluation, an insidious progressive chronic renal failure was observed.

Case History and Examination:

A 5-year-old boy from consanguineous parents presented to the emergency room of Children's Hospital in Damascus with a chief complaint of a seizure episode. According to the parents, the child had two episodes of limb shaking that lasted for minutes and were consistent with Tonic-Clonic seizures. These episodes were considered benign febrile seizures as they occurred during periods of hyperthermia following pharyngitis and flu. The last episode lasted for 2 minutes and the patient lost consciousness afterward. The patient was admitted to the emergency room for evaluation.

Upon arrival, the patient had woken up and vital signs were within normal ranges for his age. Medical history revealed a slight delay in motor and language development and a history of surgically treated undescended testes at the age of 1 year.

Physical examination showed ascites and tenderness in the right iliac region. Additionally, enlarged lymph nodes were observed in the neck, axilla, and groin.

Differential diagnosis, investigations and treatment:

Basic laboratory tests showed abnormal renal function with urea levels at 60 g/dl and Creatinine levels at 2 g/dl. Full laboratory findings are available in (Table 1)

Abdominal ultrasound confirmed ascites and revealed second-degree hydronephrosis in the left kidney and first-degree hydronephrosis in the right kidney. The diameter of ureter was increased bilaterally with bilateral loss of corticomedullary differentiation. The bladder on ultrasound appeared full and enlarged, filling most of the abdomen. Its walls showed thickness and multiple diverticula along with a classified polyp. These findings are consistent with neurogenic bladder and lower urinary tract obstruction. MRI of the spine was performed to determine the cause but no abnormalities were observed. MCUG confirmed bilateral hydronephrosis with two large diverticula within the bladder wall (Unfortunately unavailable). Renogram revealed a glomerular filtration rate (GFR) of 40 ml/min for right kidney and 24 ml/min for left kidney which is consistent with insidiously progressive chronic renal failure. Initial management focused on protecting the kidneys from further damage by performing ureterocutaneostomy as it was believed that patient had neurogenic bladder.

After the surgery, urodynamics studies were conducted to rule out any neurological bladder issues. However, these studies did not find any evidence of a neurological bladder. As a result, the patient was referred for a urethroscopy procedure.

During the urethroscopy, it was discovered that the patient had a posterior urethral valve. The valve was treated by ablation during the procedure. Subsequently, another urethroscopy was performed to assess the condition of the urethra (video 1).

Outcome and follow-up:

Based on the findings and treatment provided, it was recommended that retransplantation of the ureters onto the bladder should be carried out after three months. During the period of follow up there was not any complications reported and no urinary infections were observed.

Discussion:

Posterior urethral valves (PUV) are the most common form of lower urinary tract obstructions (LUTO) found in male pediatric patients. The embryonic abnormality responsible for PUV formation remains unknown. Some authors suggest that it may result from the abnormal fusion of the mesonephric duct into the urethra, while others suggest it is due to the persistence of the cloacal membrane. The overall incidence of LUTO is approximately 3.3 per 10,000 pregnancies and 2.3 per 10,000 live births. The overall prevalence is reported to be 3.34 per 10,000 births, ranging from 2.95 to 3.72.[3,6]

About 35% of cases are diagnosed in utero, and these cases are suspected when there is bilateral hydronephrosis, megacystis, and oligohydramnion or anhydramnion. On the other hand, 42% of cases are diagnosed during childhood, and the presenting symptoms include signs of urinary obstruction and infection.[3] Our patient presented with loss of consciousness after a seizure; laboratory studies revealed high levels of creatinine and urea consistent with chronic kidney disease (CKD). Further assessment showed that the patient had PUV.

The predominant presentation that persists in almost all patients with PUV is voiding dysfunction (identified by poor stream, straining, and dribbling). Additional features may include anemia, bladder distension palpable above the pubic region, ascites, fever, ballotable kidneys, and inability to thrive.[4] Many complications can occur due to the obstruction induced by PUV and it varies depending on the degree of obstruction and the patient's age. The obstruction can range from minimal to severe resulting in bilateral severe hydroureteronephrosis because of the high pressure above the valve placement, chronic kidney disease (CKD) which may progress to end-stage kidney disease (ESKD), diverticular formation in the bladder, and even bladder rupture in severe cases.[7,8] Even though PUV manifests with voiding dysfunction and CKD is considered a complication, our patient presented with no signs or symptoms of voiding problems, and CKD was detected during evaluation.

Micturating cysto-urethrogram (MCUG) is the most useful technique for diagnosis. Ultrasonography is considered an assistive diagnostic method especially for detecting complications such as diverticular formation in the bladder and hydroureteronephrosis.[7,8] In this case, MCUG was not able to detect the valve which was then ablated during urethroscopy.

As we have seen in this case report, PUV can lead to an insidious progressive chronic kidney disease even after years of birth. As we know, PUV is not uncommon; therefore, practitioners should consider it as a differential diagnosis for CKD in any child even if their age at presentation is not typical. Moreover, heigh suspicion is needed if diagnostic assessment investigations show normal findings as in this case.

List of Abbreviations:

PUV: Posterior Urethral Valve

MCUG: Micturating cysto-urethrogram

CKD: Chronic kidney disease

ESKD: End-stage kidney disease

LUTO: lower urinary tract obstructions

GFR: Glomerular filtration rate

Declarations:

Ethics Approval:

Ethical approval was taken from Faculty of Medicine at Damascus University.

Informed Consent:

Written informed consent was obtained from the patient's parents for their daughter's anonymized information to be published in this article.

Availability of Data and Materials:

The datasets used during the current study are available from the corresponding author on reasonable request

Competing Interests:

The authors have declared that no competing interests exist.

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All authors participated in writing and reviewing the whole paper

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Data curation, Methodology, Writing – original draft, Writing – review & editing

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Table 1: laboratory findings when the patient was presented to the emergency room.

Video 1: the second urethroscopy which was performed after removing the valve in the first one.

Hosted file

`table 1.docx` available at https://authorea.com/users/771398/articles/853818-unveiling-the-menace-of-incidental-insidious-chronic-kidney-disease-due-to-posterior-urethral-valve-a-case-report