Case Report IJCR (2019) 4:67



International Journal of Case Reports (ISSN:2572-8776)



Late Presentation of Posterior Urethral Valve in a 24-Year-old Man

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INTRODUCTION

Posterior urethral valve (PUV) is the most common cause of lower urinary tract obstruction in male neonates, with an incidence of one case per 8,000 to 25,000 live births1. The diagnosis is usually made prenatally or at birth, when male newborns are evaluated for prenatal hydronephrosis, or during early childhood, but rarely during adolescence or adulthood. Late presentation of PUV is rare and it has been estimated to account for 10% of cases2. Presentation in adulthood may occur due to the presence of a mild form of the disease devoid of the life threatening complications seen in children with the severe form3.

A rare case of PUV which manifested in adulthood with lower urinary symptoms is presented to create awareness of the likelihood of the disease outside the paediatric age group.

How to cite this article:

OO Moemenam, NR Njeze and OD Ray-Offor. Late Presentation of Posterior Urethral Valve in a 24-Year-old Man. International Journal of Case Reports, 2019 4:67

Conflict of interest: None

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CASE REPORT

O. J. is a 24-year-old student who presented to the Urology Clinic of Federal Medical Centre, Owerri, with history of increased frequency in urination of 10 weeks duration. The symptom was said to have started gradually and progressively worsened over time. There was good streaming of urine, even though he felt the quantity increased because he spent more time passing out urine. There was associated dribbling, urge incontinence, hesitancy and feeling of incomplete emptying of bladder. There was also early morning swelling of the feet up to the ankle, noticed mainly in the right, and relieved when he walked around for a while. No associated facial puffiness, fever, pain on micturition, lumbar pain or urethral discharge. For these symptoms, he had been treated with antibiotics which were prescribed at a medical laboratory, but without any improvement. He then decided to come to our hospital for expert management.

Past medical history revealed that he was diagnosed and treated for syphilis about 3 years ago at a peripheral hospital. He also had dysuria about 7 months prior to presentation, which he treated using herbal medications. Family and social history was not significant.

Physical examination revealed a young man, not pale, anicteric, afebrile, acyanosed, mildly dehydrated with no pedal oedema.

Abdominal examination revealed a mass in the suprapubic region which was 20 weeks in size, well defined, rounded and firm. Chest and cardiovascular system examinations were not contributory. Urinalysis showed pus cells (+), while urine culture did not yield any growth of organisms. Serum electrolytes, urea and creatinine values obtained were deranged: urea 131mg/dl (normal range = 15 - 40), creatinine $2.6 \text{mg/dl} (0.5 - 1.5), \text{ K}^+ 6.7 \text{mmol/l} (3.5 - 5) \text{ and}$ 111mmol/l (96 – Cl 108). Abdominal ultrasound (US) scan revealed moderate bilateral hydronephrosis. The urinary bladder was markedly dilated with thickened wall. A

diverticulum was noted on the right side of the urinary bladder. Provisional diagnosis of urinary secondary bladder outlet obstruction? urethral He stricture was made. was catheterized and sent for micturatingcystourethrogram (MCUG) and retrograde urethrocystogram (RUCG).

The RUCG showed the anterior urethra that is normal in calibre and outline (Fig.1). The MCUG showed a grossly dilated posterior urethra. The bladder wall was thickened and had trabeculations with a contrast filled outpouching on the right (diverticulum) (Fig.2). There was no vesicoureteric reflux. He was then booked for urethrocystoscopy and resection of posterior urethral valve.

During the urethrocystoscopy, a posterior urethral valve was noted just after the bladder neck and internal sphincter. The posterior urethral valve was avulsed as the beak of the urethrocystoscope was advanced into the urinary bladder. He was placed on antibiotics and catheterized in order to maintain continuous bladder drainage for one week post op.

He is currently being jointly managed with the nephrology unit for chronic renal failure. On follow up, he has remained well with resolution of his symptoms as well as improvement in his deranged electrolytes, urea and creatinine.

DISCUSSION

Posterior urethral valve (PUV) is a congenital malformation found in males that characterized by the presence of abnormal obstructing membranes in the posterior urethra. The exact embryology of the condition is not completely understood but it is believed to arise mainly as a result of an anomalous insertion of the mesonephric duct into the primitive fetal cloaca4. This leads to an obstruction in the flow of urine through the urethra with the classic presentation of poor stream at birth. It is the common cause of bladder outlet most obstruction in male children and can be complicated by urinary tract infection (UTI),

structural changes in the urinary tract and impaired renal function⁵.In cases with mild obstruction, presentation may be delayed until later in life.

Young et al² classified PUV into three types.

 Type 1 - mucosal folds extend distally from each side of the verumontanum

- towards the membranous urethra where they fuse anteriorly.
- Type 2 -mucosal folds extend cephalad from the verumontanum to the bladder neck.
- Type 3 represents a diaphragm or ringlike membrane with a central aperture just distal to the verumontanum⁶.



Fig.1. RUCG showing the normal anterior urethra (white arrow). The posterior urethra is mildly dilated.

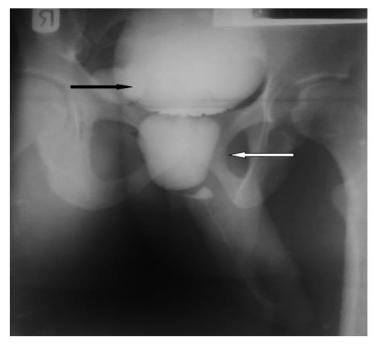


Fig.2. MCUG showing the markedly dilated posterior urethra (white arrow) and the urinary bladder diverticulum (black arrow). The urinary bladder is also thick-walled and trabeculated.

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The incidence of PUV in Nigeria is not known. However PUV together with other congenital obstructive uropathy accounted for 1.4–6.4% of all renal diseases in most of the centres in Nigeria^{6, 7}. No racial, ethnic or geographical predilection is noted.

The usual presentation of PUV is prenatal or at birth. The symptoms and signs of late presentation of PUV depend mainly on the degree of obstruction and the presence of complications. The most common presentations include poor or weak urine stream, dribbling at voiding, urinary frequency, feeling of incomplete bladder emptying urinary and incontinence, as in the index case. Patient may present with repeated urinary tract infection and chronic renal failure, which are common clinical features in adolescents and adults. Other rare situations that could be related to late presentation of PUV may include retained ejaculation and infertility. Agbuguiet al³ reported an unusual case of primary male infertility in an adult with PUV in Benin Nigeria.

Imaging is frequently employed the confirmation of diagnosis of PUV. Ultrasonography may show hydronephrosis, with or without cortical thinning. Hydroureters and thick-walled bladder with trabeculation and diverticula may also be seen. In younger patients, a dilated posterior urethra with a hypertrophic bladder neck may be noted. This feature was not demonstrated in the index patient, on ultrasound. Echogenic lines that are the actual valve leaflets might be seen. The combination of the dilated, thick-walled bladder dilated posterior urethra has been described as having a keyhole appearance. MCUG is considered the diagnostic study of choice for the evaluation of posterior urethral valves8. The diagnosis is best made during the micturition phase in a lateral or oblique view, such that the posterior urethra can be imaged adequately. Findings may include dilatation and elongation of the posterior urethra, vesicoureteric reflux (VUR), bladder trabeculation or diverticular. Linear radiolucent

corresponding to valve band the may occasionally be seen. In the present patient, ultrasonography showed the hydronephrosis, while the MCUG was able to the dilated posterior clearly demonstrate urethra. Intravenous urography (IVU) is not routinely used in the diagnosis of PUV, but may show hydroureteronephrosis when present. Delayed images may show bladder pathology, but the lower urinary tract is better visualized with MCUG. Computed tomography (CT) and magnetic resonance imaging (MRI) are rarely necessary because they cannot reliably depict valves, although they may reveal the sequelae of bladder outlet obstruction.

Generally, of PUV treatment involves transurethral ablation of valve. the Electrosurgical fulguration with a Bugbee electrode through aurethrocystoscope is the commonest treatment option for management of PUV9. The valves may also be incised with a cold knife. In this patient, the valve was avulsed using the beak of the urethrocystoscope which gave a satisfactory result. Late presentation of PUV could be a cause of chronic renal failure whether or not it has been treated. Parkhouseet al¹⁰ reported on long-term follow-up among treated postpubertal patients and showed that 26% of them had chronic or end-stage renal failure. The index patient is currently being managed for chronic renal failure

SUMMARY

A rare case of posterior urethral valve in a 24-year-old man has been presented. The varying clinical presentation, diagnostic modalities and treatment options have also been discussed. This case highlights the need to remain alert to the possibility of late diagnosis of PUV.

Conflict of interest: None

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