Posterior urethral valve: typical and atypical mode of presentation

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Abstract

Posterior urethral valve is an obstructive developmental anomaly of the urethra of male newborns. It is the most common cause of bladder outlet obstruction in male newborns. We hereby present two cases of posterior urethral valve because of the usual and unusual times of presentation in a six month old boy and a 38 year old man respectively.

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Key Words:

Introduction

Posterior urethral valve is a congenital anomaly of the male urethra in which thick folds of mucous membrane develop within the posterior urethra. The incidence is 1:5,000 8,000 boys and it is the most common cause of urinary tract obstruction in the neonates and infants with end stage renal damage occurring in untreated cases[1]. However, presentation in adulthood has been described[1]. It is almost exclusively seen in males[1-4].

The common and uncommon time of presentation is highlighted in these two case studies of a 6 month old boy and a 38 year old man with posterior urethral valves.

CASE I

Master O.O. a 6 month old male presented to the children emergency room of University of Benin Teaching Hospital with a history of recurrent fever since birth associated with poor appetite, poor weight gain and occasional vomiting. On further questioning the mother admitted that baby dribbles urine, a symptom she attributed to poor feeding. She had been attending a nearby private clinic with no improvement. Baby had even been subjected to various laboratory investigations where it was discovered he had urinary tract infection. He was treated with appropriate antibiotics with no improvement and subsequently referred to University of Benin teaching Hospital. The patient is the 5th sibling born to a 52 year old security man and a 40 year old housewife. His mother had no antenatal care or obstetric scan and he was born at home by spontaneous vaginal delivery. He cried immediately after birth but his birth weight was not known though his mother admits he was very small. He was only fed on breast milk and water.

On examination he was pale, febrile and asthenic. There was no jaundice or dehydration. His anterior fontanelle was still open and not under pressure. His chest was clear. Abdominal examination revealed a distended abdomen with a palpable pelvic mass. His penis appeared normal with no epi or hypospadias. A provisional diagnosis of failure to thrive secondary to bladder outlet obstruction was made. Various laboratory and radiological investigations were ordered. He was immediately catheterized suprapubically.

His hematocrit was 25% WBCC

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11,600/ml, leucocytes 68%, others 32%. Urine microscopy, culture and sensitivity yielded growth of coliforms sensitive to rocephine+++ and he was immediately placed on appropriate medications. His chest radiograph was normal while abdominal ultrasound showed bilateral moderate hydronephrosis and hydroureters. His urinary bladder was distended with mural thickening and a dilated posterior urethra was also seen. A micturatingcystourethrogram showed a severely trabeculated urinary bladder with left sided diverticulum and a characteristic "spinning top" appearance of the posterior urethra (Fig.1a). Vesicourethral reflux was not demonstrated. The rest of the urethra showed poor urine stream. The post void film showed residual urine within the diverticulum (Fig.1b). Based on these findings a diagnosis of posterior urethral valve was made. He has since had corrective surgery and discharged home. He has been visiting the paediatric outpatient department and is showing signs of weight gain.

Figure1a: Voiding cystourethrogram . The urinary bladder is distended with sacculations, trabeculations and a left sided diverticulum (white arrows). The posterior urethra is dilated and funnel shaped tapering into a narrow stream anterior urethra (black

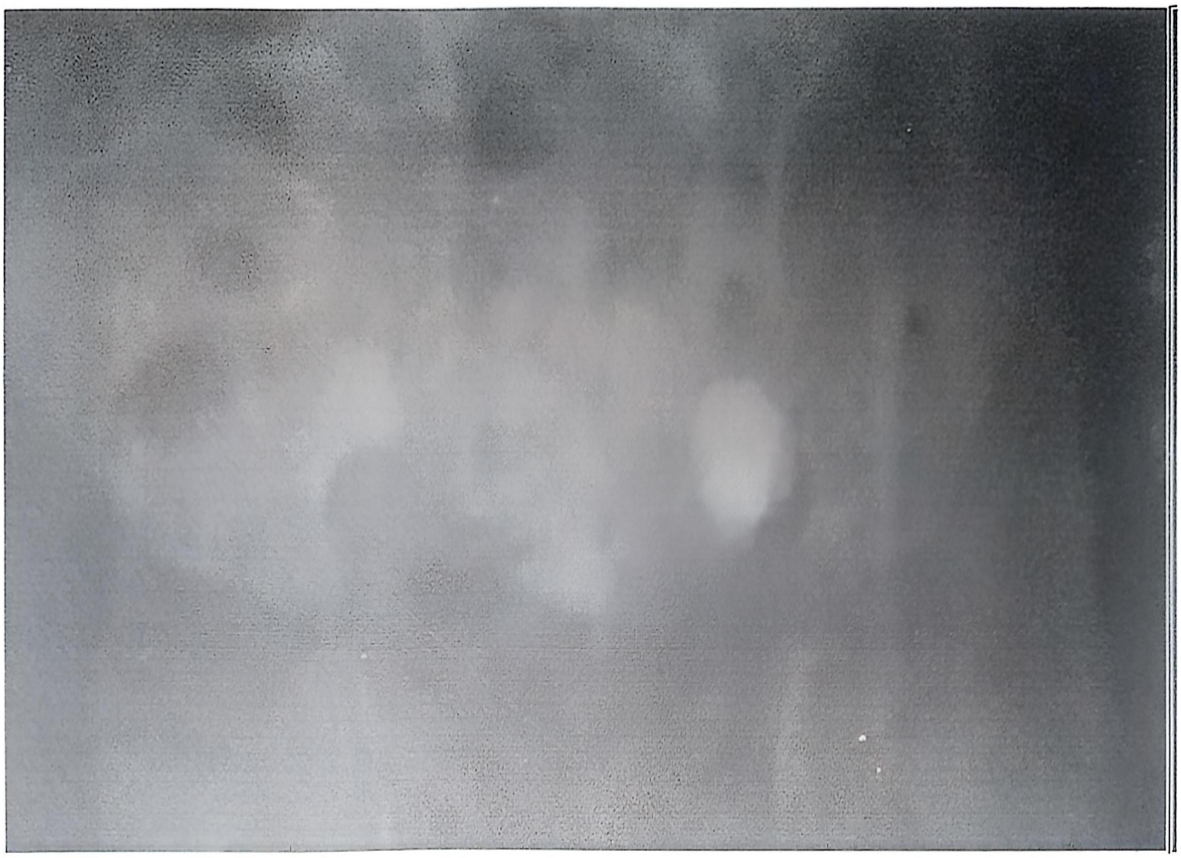

Figure1b: Post void film. Urine is retained within the diverticulum (white arrows). There is no significant urine within the true bladder lumen (black arrows)

CASE 2

Mr O.R. is a 38 year old motor mechanic who was referred to the urology unit of University of Benin Teaching Hospital with a provisional diagnosis of urethral stricture. As long as he could remember he had always had poor urine stream associated with straining. There was no history of fever or dysuria and he had never had trauma to the groin or any form of instrumentation to his penis. There was also no history of urethral discharge.

He is single and has no children. He is the first sibling in a monogamous setting with 6 others. His father is a retired clerk while his mother is a petty trader. He presently lives alone.

On examination, he was healthy looking, not pale, afebrile and anicteric. His chest and abdominal examination revealed no abnormality. A rectal examination done did not reveal an enlarged prostate gland. His laboratory investigations and chest radiograph were unremarkable. Abdominal ultrasound scan revealed mildly enlarged hydronephrotic kidneys without signs of renal calculi. The urinary bladder showed thickening of its walls with a dilated posterior urethra. The prostate gland appeared normal in size and echotexture. There were no bladder calculi.

A provisional diagnosis of urethral stricture was made and he was referred to radiology for retrograde urethrography. This revealed a normal anterior urethra. A micturatingcystourethrogram demonstrated a normal capacity urinary bladder with some sacculations and a funnel shaped posterior urethra (Fig 2).

Bladder outlet obstruction secondary to posterior urethral valves was made and cystourethroscopy was advised. This confirmed posterior urethral valves. He has since had corrective surgery and he is presently attending the outpatient department with no more symptoms.

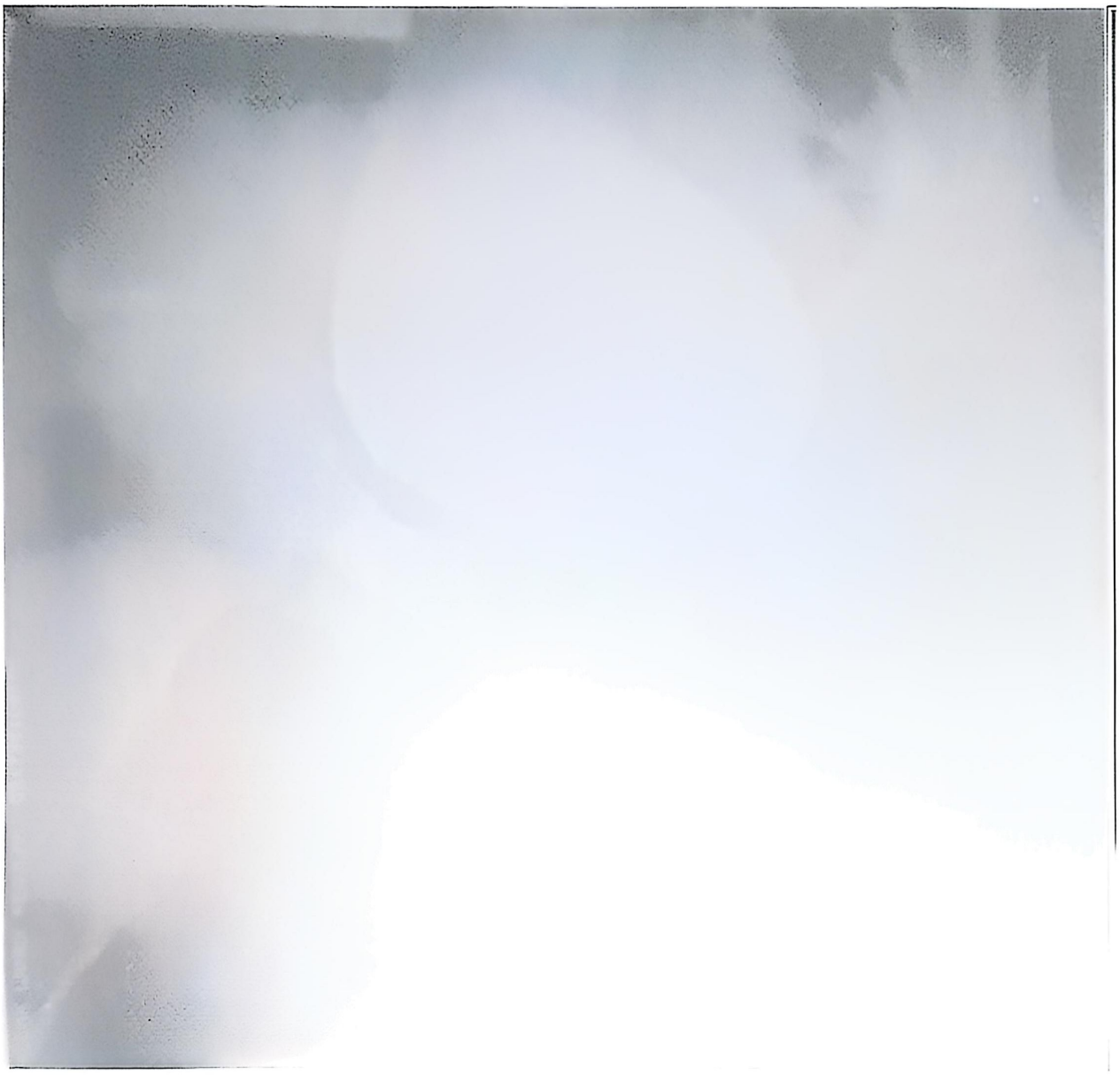

Figure2: Voiding cystourethrogram. The urinary bladder was adequately distended with sacculations of the fundal margin (white arrow). The posterior urethral is dilated and funnel shaped tapering into a narrow stream anterior urethra (black arrows)

DISCUSSION

Posterior urethral valve is categorized into 3 depending on the position of the mucosal folds[1,2].

1. **Type I:** This is the most common type in which the mucosal folds extend anteroinferiorly from the lower aspect of the verumontanum. These folds are believed to be vestiges of the wolffian duct.
2. **Type II:** This is a rare form in which the mucosal folds extend anterosuperiorly from the verumontanum towards the bladder neck.
3. **Type III:** In this type, a diaphragm-like membrane is located below the verumontanum. It is believed to be caused by abnormal canalization of the urogenital diaphragm.

Anterior urethral valves have been described but this is very rare[4,5]. It produces symptoms of varied intensity with most severe ones leading to urinary tract infection and renal failure while the milder ones may not produce any obstruction. Unlike the posterior urethral valves this anomaly has milder and reversible sequelae in majority of cases. A high level of suspicion as well as the awareness of the existence of this condition is essential while diagnosis is based on micturatingcystourethrographic studies.

Differentiation of posterior urethral valves into types is only possible at urethroscopy or autopsy. The patients presented were found to have type 1. Time of presentation varies but more than 65% present before 2 years of age, 23% between 2 to 16 years while a small 8% are diagnosed inutero[1,3]. Presentation in adulthood accounts for only 3%[1]. Maranya[6] reported two adult cases of posterior urethral valves, one aged 19 years and the other 21 years. They both presented with symptoms of bladder outflow obstruction. Urinary tract ultrasound and micturatingcystourethrograms were the radiological methods of confirmation and symptoms resolved completely following surgery. The patient in the second case presented in adulthood while the first case presented in infancy.

Symptoms vary from recurrent urinary tract infection to symptoms of bladder outlet obstruction which include urinary hesitancy, straining or dribbling. Case 2 presented with poor urine stream and straining and was misdiagnosed to have urethral stricture most likely because of his age. The possibility of posterior urethral valve was not considered. The only other lesion which presents this way in an adult is prostatic gland hypertrophy/hyperplasia which this patient did not have. Others present with failure to thrive and/or a palpable abdominal mass which could represent hydronephrotic kidneys, a distended urinary bladder or urinary ascites[14]. The first case presented had similar features. Enuresis is also a common presentation[4].

Posterior urethral valve causes obstruction to urinary flow with consequent development of obstructive uropathy. The urinary bladder dilates and its wall undergoes hypertrophy with formation of trabeculations, sacculations and diverticuli. Subsequently there will be vesicoureteral reflux, progressive bilateral hydronephrosis and hydroureters. In some instances, there may be partial or complete rupture of the dilated bladder, pelvis or calyces leading to urinary ascites and urinomas.

Children with posterior urethral valves suffer from recurrent urinary tract infection worsened by stasis which was the case with the first patient and may eventually develop calculi in the urinary tract. The age at presentation will depend on the severity of obstruction.

Radiological diagnosis of posterior urethral valves is by conducting a micturating (voiding) cystourethrogram. However, voiding cystourethrosonography(VCUS) with echo contrast has now been advocated for the diagnosis of posterior urethral valve and vesicoureteral reflux from any cause. Posterior urethral valve is a common cause of vesicoureteral reflux. It was originally developed to avoid X-ray exposure during detection of vesicoureteral reflux.

Bosio*et al*[7] carried out a study in 100 males using voiding cystourethrosonography to determine the efficacy of the procedure in adequately visualizing the male urethra and to differentiate the normal from the obstructed urethra. Of the 100 boys, 8 were correctly diagnosed with posterior urethral valves as confirmed by subsequent voiding cystourethrography. None of those with normal ultrasound imaging showed clinical signs suggestive of urethral obstruction and remained asymptomatic 12 to 54 months later. Xhepa*et al*[8] compared the diagnostic efficacy of voiding cystourethrosonography in 22 patients and obtained a diagnostic concordance of 66.6%. Overall cystourethrosonography showed a sensitivity

superior to voiding cystourethrography. The only major limiting factor of voiding cystourethrosonography include the operator training and experience as well as cost of the contrast media. None of the cases presented in this report had voiding cystourethrosonography. Urethral sonography using a perineal approach has also been used by some authors in evaluating for anomalies in infants. Schoellnast*et al*[9] reviewed 88 patients with a mean age of 64 days who had undergone both voiding cystourethrography and urethral sonography. Sonography correctly showed absence of anomalies in 73 patients. However, it facilitated the correct diagnosis of 3 patients with posterior urethral valves, one patient each with urethral diverticulum, ectopic ureteric insertion into the urethra, urogenital sinus and urethrovaginal fistula. They therefore concluded that urethral sonography is a valuable tool for diagnosis of urethral anomalies. However, positive or equivocal sonographic findings should be an indication for voiding cystourethrography. Inutero diagnosis can be made by serial prenatal ultrasound scans which, shows a persistently dilated urinary bladder, posterior urethra with progressive bilateral hydronephrosis and hydroureters. There may be associated oligohydramnios, urinary ascites, urinoma and hypoplastic lungs depending on the severity of obstruction. Antenatal/prenatal diagnosis of urinary tract anomalies allow for immediate prophylactic treatment of urinary infections and decrease the risk of severe complications like pyelonephritis developing. It also allows for proper planning of postnatal management options[10].

Recent advances in ultrasound technology with improvement in antenatal care has made it possible to recognised this anomaly inutero even though prenatal diagnosis carries a poorer prognosis because of the development of irreversible renal damage, severe oligohydramnios and potter's syndrome. Both cases presented did not have the benefit of prenatal obstetric scan.

Abdominopelvic ultrasound scan and intravenous urography are used to demonstrate upper urinary tract abnormalities with the later providing a clue to renal function. Abdominopelvic ultrasound is a non invasive modality of demonstrating the effects of posterior urethral valve on the urinary system. However, the presence of dilated urinary system is not conclusive of posterior urethral valves[1,3,11], but when this is seen in a male it should be considered as a likely cause. The bladder will be seen to be dilated with thickening of its wall and diverticuli may also be seen. The posterior urethra will also be seen to be dilated. There may be debris mixed with urine within the bladder lumen suggesting stasis and infection. Urine leakage into the perivesical area or intraperitoneally will be demonstrated ultrasonographically[3]. Both patients presented demonstrated a distended urinary bladder with thickened walls ultrasonographically. In addition there was dilatation of the posterior urethra.

Micturatingcystourethrogram is the investigation of choice in demonstrating posterior urethral valves[1,2,3,4,7]. This will demonstrate the dilated and hypertrophied urinary bladder with/without diverticuli. The posterior urethra will show a fusiform enlongation and dilation of its proximal end persisting throughout voiding and leading into a diminished calibre anterior urethra. A transverse or curvilinear filling defect representing the posterior urethral valve may be seen in the posterior urethra. There will be poor urinary stream distal to the site of the valves. Vesicoureteral reflux may be seen in up to 50% of cases and a large post void residual urine remains within the bladder or diverticulum[3].

The two cases presented showed fusiform dilatation of their posterior urethra giving the 'spinning top' configuration with poor urine stream within the anterior urethra. The first case in addition had bladder trabeculations, sacculations and a left sided diverticulum which did not empty on the post

void film. However, none of the cases showed vesicoureteral reflux.

Treatment of posterior urethral valves is by surgical ablation or primary avulsion using Fogarty balloon catheter. Prognosis will depend on the amount of renal damage done prior to definitive treatment.

The diagnosis of posterior urethral valve is often overlooked in an adult who presents with poor urinary stream, hesitancy and symptoms of prostatism as was the case in the second patient. This is because urethral strictures and prostatic hypertrophy is a commoner cause of these symptoms in the adult population. Therefore, it is essential for every male undergoing retrograde urethrogram to have a voiding cystourethrogram at the same time to demonstrate the posterior urethra, otherwise dilatation of the posterior urethra due to valves may be overlooked. This was the case with the second patient presented.

Conclusion

Two cases of posterior urethral valves in extremes of age are presented. The need for prenatal ultrasound diagnosis and voiding cystourethrogramis discussed. The appearances on the various imaging modalities are also highlighted.

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