Case Report

Posterior urethral valve with unilateral vesicoureteral reflux and patent urachus: A rare combination of urinary tract anomaliess

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Abstract

Posterior urethral valve (PUV) is a common cause of lower urinary tract obstruction in male infants with an incidence of 1:5000-8000. PUV continues to be a significant cause of morbidity and ongoing renal damage in infants and children. It can coexist with vesicoureteral reflux (VUR) in about 50% of cases and also with patent urachus in about one-third of cases. It is a case of a 22-day-old full-term male child who presented with poor urinary stream and progressive abdominal distension of 5-day duration as well as leakage of clear fluid from umbilicus of 1-day duration. Abdominopelvic ultrasonography showed bilateral hydronephrosis. Micturating cystourethrogram also showed features of bladder outlet obstruction and PUV. In addition, a grade V left VUR and a fistulous tract between the dome of the urinary bladder and the umbilicus, which was consistent with a patent urachus was demonstrated. In conclusion, this case demonstrates a rare combination of congenital urinary tract anomalies involving PUV, left VUR and patent urachus.

Key Words: Posterior urethra valve, urachus, vesicoureteral reflux

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INTRODUCTION

Posterior urethral valve (PUV) continues to be a significant cause of morbidity and end-stage renal failure in childhood.[1-4] Vesicoureteral reflux (VUR) is commonly associated with PUV[5,6] and is present in 50% of cases.[7,8] Patent urachus frequently coexists with congenital lower urinary tract obstruction and it is associated with PUVs in about one-third of cases.[9,10]

The increased incidence of VUR in cases of urachal anomalies (UA) had also been reported,[11] but the combination of these three entities has not been reported to the best of our knowledge.

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CASE REPORT

This is a case of a 22-day-old full-term male child who presented at the Emergency Pediatric Unit of the University of Ilorin Teaching Hospital with a history of poor urinary stream and progressive abdominal distension of 5-day duration as well as leakage of clear fluid from umbilicus of 1-day duration.

He was delivered per vagina at about 38-week gestational age. The pregnancy and delivery were uneventful.

Physical examination revealed a grossly distended abdomen with dilated superficial veins. The umbilicus was swollen, hyperemic and discharging clear fluid.

Electrolyte, urea and creatinine (E/U/Cr) result showed; Na = 136 mmol/L, K = 3.7 mmol/L, U = 18.1 mmol/L,

Cr = 124 mmol/L. Full blood count and urine microscopy, culture and sensitivity (m/c/s) are within the normal limits.

An impression of bladder outlet obstruction presumably due to PUV with infected urachal cyst was made.

Abdominopelvic ultrasonography showed bilateral hydronephroses [Figure 1]. The urinary bladder showed multiple trabeculations, sacculations and diverticula. The posterior urethra was also dilated giving a "keyhole" appearance.

Micturating cystourethrogram (MCUG) also showed thickened bladder wall, trabeculations, sacculations and diverticular formation. There was dilated crescentic filling defect at the distal end of a dilated posterior urethra consistent with PUV. Left VUR up to the calyces was also demonstrated (grade V) [Figure 2]. A contrast filled beaded fistulous tract was also seen between the dome of the urinary bladder and the umbilicus [Figure 3].

A diagnosis of PUV complicated by grade V left VUR with urachal fistula was made.

Cystoscopy revealed dilated posterior urethra and two valve-like folds arising from the caudal end of verumontanum and ascending along the urethra to meet at 12 O' clock. Thick-walled bladder with extensive trabeculations, sacculations and diverticula as well as incompetent right vesico-ureteral orifice was also noted.

Valve ablation using Mohan's valvotome was carried out. The post-operative urine stream was good and the E/U/Cr improved significantly.

Post-operative E/U/Cr showed; Na = 135 mmol/L, K = 4.5 mmol/L, U = 2.3 mmol/L, Cr = 44 mmol/L.

The patient was discharged 10th day after surgery.

DISCUSSION

A PUV is an abnormal congenital obstructing membrane that is located within the posterior male urethra. The valve is believed to result from abnormal embryologic development of the fetal posterior urethra.[2]

PUV is by far the most common congenital obstructive lesion of the urethra, occurring only in phenotypic boys.[3,4] It continues to be a significant cause of morbidity and end-stage renal failure in childhood.[1,2] The mechanical obstruction that occurs with PUVs, increases voiding pressure and may alter normal development of the fetal bladder and kidneys.[2]

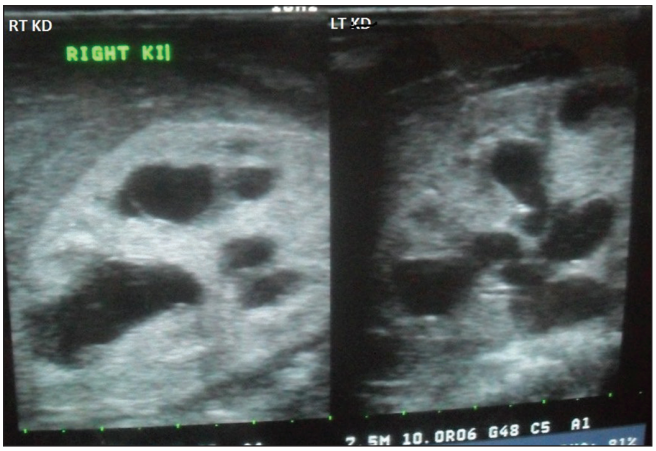

Figure 1: Sonogram of both kidneys showing bilateral hydronephroses worse on the left

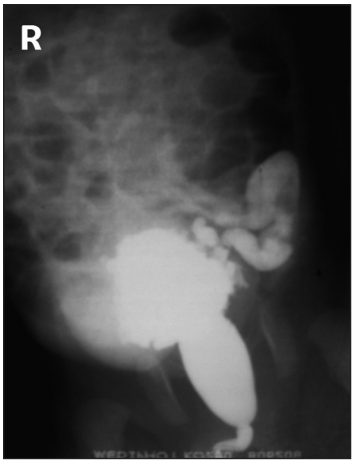

Figure 2: Micturating cystourethrogram (anteroposterior view) showing dilated posterior urethra and urinary bladder with trabeculations, sacculations and diverticular formation. Dilated and tortuous left ureter consistent with vesicoureteral reflux is also demonstrated

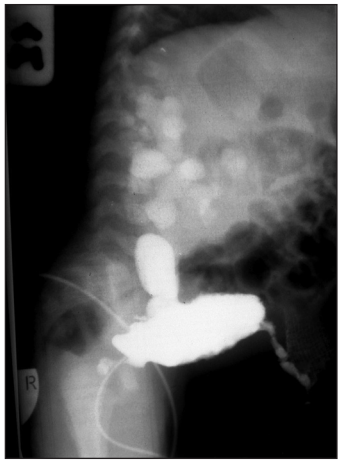

Figure 3: Micturating cystourethrogram (lateral view) showing grade V vesicoureteral reflux and a contrast filled tract between the urinary bladder and the umbilicus consistent with patent urachus

Signs and symptoms may range from mild obstructive symptoms of voiding dysfunction, to severe obstruction with resultant renal failure and pulmonary hypoplasia.[2]

MCUG is the best imaging technique for the diagnosis of PUVs.[4] Radiologic findings include dilatation and elongation of the posterior urethra and occasionally, a linear radiolucent band corresponding to the valve.[6] Bladder wall hypertrophy, trabeculation, sacculation, diverticula and VUR may be seen as demonstrated in this case.

VUR is the abnormal flow of urine from the bladder into the upper urinary tract.[1] It is commonly associated with PUV as seen in this case.[5,8] In the majority of cases, it occurs as a result of a primary maturation abnormality of the vesicoureteral junction or a short distal ureteric submucosal tunnel in the bladder that alters the function of the valve mechanism.[6,10]

VUR is present in about 50% of patients with PUVs[2,7] and is more often unilateral than bilateral[2] as reported in this case.

Unilateral reflux, may occur in up to 35% of boys with PUV and has been linked with protected renal function.[3,4] This might be the reason for the preservation of renal function in this patient. This patient had severe left unilateral VUR, characterized by gross dilatation and tortuosity of the right ureter; gross dilatation of the renal pelvis and calyces regarded as a grade V based on the International Reflux Committee Study.

The urachus develops from the superior portion of the urogenital sinus and connects the dome of the bladder to the allantoic duct during fetal life. The urachus is located behind the abdominal wall and anterior to the peritoneum in the space of Retzius. Before birth, the urachus is obliterated and becomes a vestigial structure known as the medial umbilical ligament. In the absence of complete obliteration, the urachus persists as either a patent urachus, urachal cyst, urachal sinus or urachal diverticulum.[9]

Patent urachus represents the failure of the entire course of the urachus to close, resulting in an open channel between the bladder and the umbilicus.[10]

A patent urachus frequently coexists with congenital lower urinary tract obstruction such as prune belly syndrome (PBS) and PUVs, but it is commonly associated with PBS.[3]

Fox *et al.* reported that VUR was demonstrated in 14 of the 22 children (64%) with UA and the rates were similar among the various types of UA.[7]

A patent urachus is usually diagnosed in the neonate when urine is noted leaking from the umbilicus.[9] This anomaly is demonstrated by sinography or MCUG in the lateral projection as seen in this case. A contrast filled tubular structure is seen between the dome of the bladder and the umbilicus.

Radiologic investigation, especially MCUG has proved to be an important source of clinical information in the urinary tract disorders.[9] MCUG as demonstrated in this case is the best imaging technique for the diagnosis of PUVs with VUR and patent urachus.

The presence of vesicoureteric reflux on one hand and patent urachus on the other hand in patients with PUV had been extensively reported.[2,3,9,10]

The increased incidence of VUR in cases of UA had also been reported,[11] but the rare combination of these three entities to the best of our knowledge has not been reported.

Reflux nephropathy is a common cause of renal failure; therefore, it is important that this condition be detected as early as possible to allow prompt prophylactic antibiotic treatment and hopefully reduce the risk of scarring and reflux nephropathy.[4]

In the absence of renal isotope scan, it is difficult to conclude that the refluxing kidney has retained renal function; however, this is most likely considering the study of Donnelly *et al.*[12] in which all eight patients with unilateral reflux studied had normal renal function on long-term follow-up. This also gives credence to the assertion that unilateral VUR in PUV is associated with protected renal function.[4]

In addition, the presence of a patent urachus with continuous dribbling of urine through the umbilicus and subsequent lowering of the intravesical pressure might also partly contribute to the preservation of renal function in this patient.

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