Antenatally Diagnosed Posterior Urethral Valve: A Case Report

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Abstract

Posterior urethral valve (PUV) are the most common congenital obstructive lesion of the urethra and a common cause of obstructive uropathy in infancy. Clinical presentation depends on the severity of the obstruction. In case of severe obstruction, the diagnosis is usually made antenatally. Here, we present a case of antenatally diagnosed PUV of a fetus of a lady in her 9th month of pregnancy with mild lower abdominal pain for several hours. On ultrasound (US) examination, we found 36 ½ ± 2 weeks of pregnancy with mild to moderate oligohydramnios.. Fetal urinary bladder was over distended; both the kidneys were grossly hydronephrotic and PUV like echo lucent area was seen at the prostatic region (Key hole sign). Emergency caesarian section (CS) was done and US of the baby showed typical US finding of PUV. The prognosis of antenatal diagnosis of PUV in early pregnancy is poor. But in this case due to the late onset of symptoms and as immediate necessary steps were taken, the baby was totally cured. This case was reported to aware about importance of antenatal anomaly scan and to share our experience.

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Introduction

Posterior urethral valves (PUVs), also referred as congenital obstructing posterior urethral membranes (COPUM), are the most common congenital obstructive lesion of the urethra and a common cause of obstructive uropathy in the male fetus in the antenatal and immediate postnatal period[1, 2]. The estimated incidence of this condition was thought to be 1 in 10,000 deliveries[2]. However more recent studies have suggested that the incidence could be higher at about 1 in 2500-5000[3] and this appears to be due to the widespread use of antenatal ultrasonography. Treatment of PUVs remains a clinical challenge, requiring long-term management from early infancy into adulthood in order to avoid progressive bladder dysfunction and deterioration of both upper and lower urinary tracts.

Case Report

A 22 year old lady in a non consanguineous marriage, presented in her 9th month of pregnancy to out patient department (OPD) with mild lower abdominal pain from previous night. She was a primigravida and by her dates the lady was in her 38 ± 2 wks of gestation. She did not give any history in the first trimester bleeding per vagina, febrile illness and took some drug like folic acid, iron and zinc combination and calcium. She did multiple ultrasounds at 9 ± 1 wks, 24 ± 1 wks and 33 ± 2 wks. All reports were normal and corresponded to the period of amenorrhoea. All her routine antenatal investigations were normal.

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On examination, her general physical condition was within normal, except mild lower abdominal pain. Obstetrical examination revealed that the uterine fundal height corresponds to 36 wks gestation. A check ultrasound was adviced and the findings showed a single live fetus of gestational age ~ 36 ½ ± 2 wks. There was mild to moderate oligohydramnios. The fetal urinary bladder was over distended, both the fetal kidneys were grossly hydronephrotic and PUV like echo lucent area was seen at the prostatic region (typical key hole sign).

The couple was called for the counseling and as the lady was near last stage of term pregnancy, the consulting obstetrician advised admission for CS and to consult with pediatric surgeon about fetal condition. Emergency CS was done next day and the baby was apparently healthy. Only abdomen was mildly distended and dribbling of urine was present.

Parents consulted pediatric surgeon who advised US examination of whole abdomen of the baby. US examination was done in our department and findings were moderately hydronephrotic changes of both kidneys with bilateral dilated ureters. Urinary bladder was well filled, wall was thick (> 5 mm) and mildly irregular. And there was a small deepening of wall at proximal urethra (key hole sign).

The pediatric surgeon seeing the US report diagnosed the case as PUV with bilateral gross hydroureter and hydronephrosis. He advised urine for routine microscopic examination (RME), culture and sensitivity (CS), serum creatinine, electrolyte, Hb % and retro grade urethrogram (RGU) with micturating cystourethrogram (MCU) and referred to the urology department of Bangabandhu Sheikh Mujib Medical University (BSMMU). At BSMMU pediatric OPD the same advice was given. All of the pathological reports were within normal limit. It was stated in the RGU and MCU report:-

Saccular dilatation of the posterior urethra was seen with abrupt narrowing in the distal part. However anterior urethra appeared to be normal without any abnormal dilatation or narrowing.

Wall of the urinary bladder was thick and irregular.

Significant amount of post voidal residue was present.

Impression: posterior urethral valve with cystitis.

Then the consulting pediatric surgeon planned to do valve fulguration or cutaneous vesicostomy.

At the age of 13 days baby's valve fulguration operation was successfully done. After operation catheterization was done for 10 days.

After that regular US and serum creatinine were done in our hospital, Serum creatinine was always normal and on US it showed that cortical thickness was increasing, cortico-medullary differentiation was improving and urinary bladder wall thickness was becoming normal day by day. There was no abnormality in micturition of the baby.

Images:

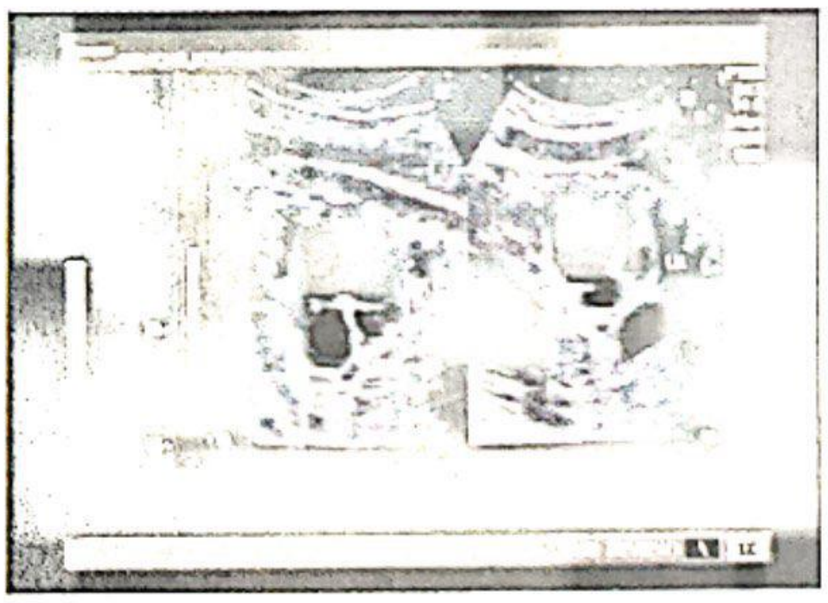

Figure 1: Fetal hydronephrosis

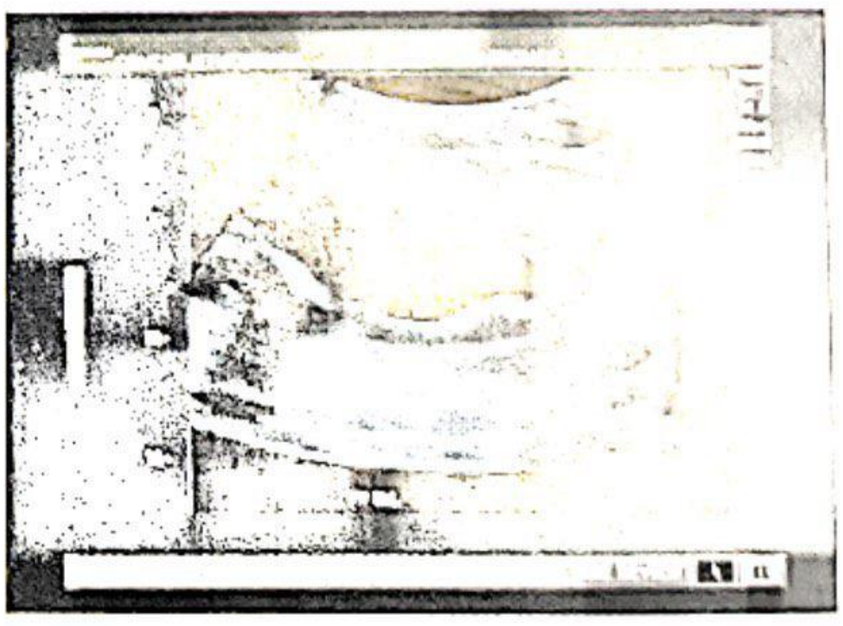

Figure 2: a

Figure 2: b
Figure 2 :(a & b) Key Hole Sign

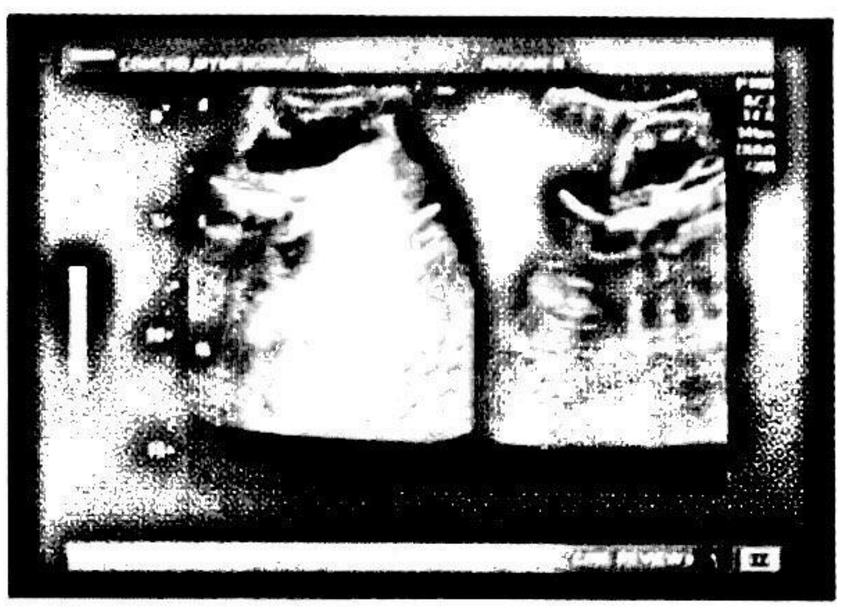

Figure 3: Bilateral hydronephrosis in new born.

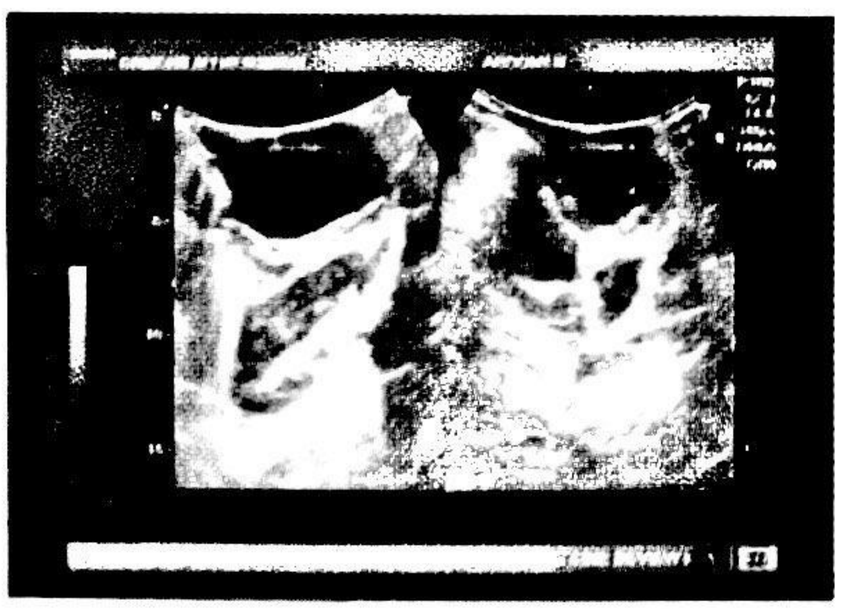

Figure: Thick wall of urinary bladder in new born.

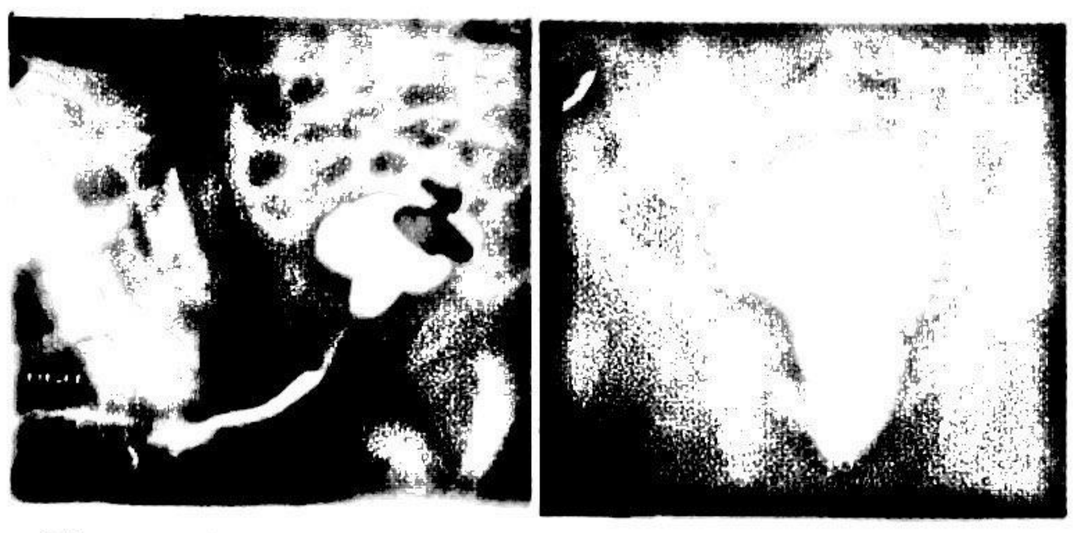

Figure 4 and 5 : RGU and MCU of new born.

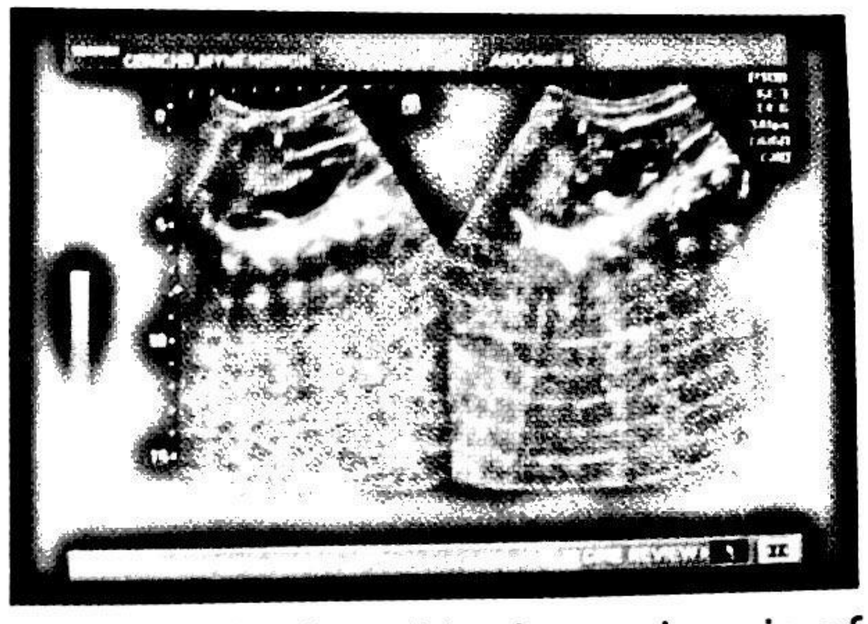

Figure 6: Reduced hydronephrosis of the baby after operation.

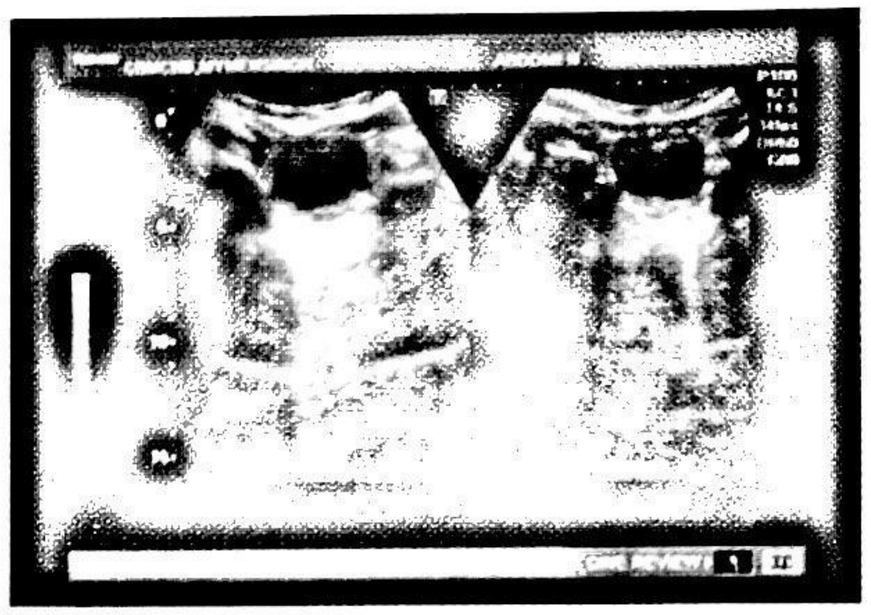

Figure 7: Reduced urinary bladder thickness of the baby after operation

Discussion

PUV are the most common congenital obstructive lesion of the urethra and a common cause of obstructive uropathy in infancy.

Clinical presentation depends on the severity of obstruction. In severe obstruction the diagnosis is usually made antenatally. The fetus will be small for gestational age and US examination will demonstrate oligohydramnios and associated abnormalities[4]. In less severe cases the diagnosis is often not apparent until early infancy. Urinary tract infections are common in this group[4].

PUV results from the formation of a thick valve like membrane from tissue of Wolffian duct origin (failure of regression of mesonephric duct) [5] that course obliquely from the verumontenum to the most distal portion of the prostatic urethra. This is thought to occur in early gestation (~5-7 wks)[6]. The valve is actually a diaphragm with a central pinhole, however as it is more rigid along its line of fusion it gradually distends and becomes distended[2-4, 7, 8].

The vast majority of cases are sporadic, although rare examples of PUV occurring in families have been reported[5]. PUV is sometimes related to other congenital and chromosomal abnormalities including-, e.g. Down syndrome, bowel atresia and cranio spinal defects[5].

On antenatal US the appearance is that of marked distention and hypertrophy of the bladder with or without hydronephrosis and hydroureter. And severity is depending on the oligohydramnios and renal dysplasia[7]. Unfortunately such findings are generally not seen before 26 wks of gestation and as such are not frequently identified on routine morphology and anomaly screening carried out around 18 wks gestation[8]. Assessing echogenicity of the kidneys are an indication of poor function[5,8]. A key hole sign may be seen on US due to the distention of both the bladder and urethra immediately proximal to valve.

Micturating cystourethrogram (MCU) is the best imaging technique for the diagnosis of PUVs. The diagnosis is best made during the micturition phase in a lateral or oblique view, such that the PU can be imaged adequately[7, 9]. Findings include-

* Dilatation and elongation of the PU (equivalent of the USG key hole sign).
* Linear radiolucent band corresponding to the valve (only occasionally seen).
* Vesico-ureteric reflux – seen in 50% of patients[7].
* Bladder trabeculation /diverticula.

Antenatal treatment is possible, consisting of vesico amniotic shunting (allowing urine to exit the bladder via the shunt, by passing the obstructed urethra). Essentially this procedure consists of a supra pubic catheter performed under US guidance. The efficacy of this procedure is controversial, as often despite this significant renal and pulmonary morbidity exists[8].

Postnatal, definitive treatment is simple and involves transurethral ablation of the offending valve[4].

The overall prognosis is most affected by the degree and duration of obstruction. Severe cases with obstructive cystic renal dysplasia, oligohydramnios and pulmonary hypoplasia are often incompatible with life.

In the correct age group and with clear dilatation of the posterior urethra there is usually little differential other than urethral atresia, which is far less common[8]. When only the bladder is clearly abnormal, thickened wall and trabeculated, other conditions need to be considered[4]– neurogenic bladder, prune-belly syndrome.

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