Case Report

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Radiology Section

Prenatal Diagnosis of Posterior Urethral Valve Presenting with the Keyhole Sign: A Case Report

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ABSTRACT

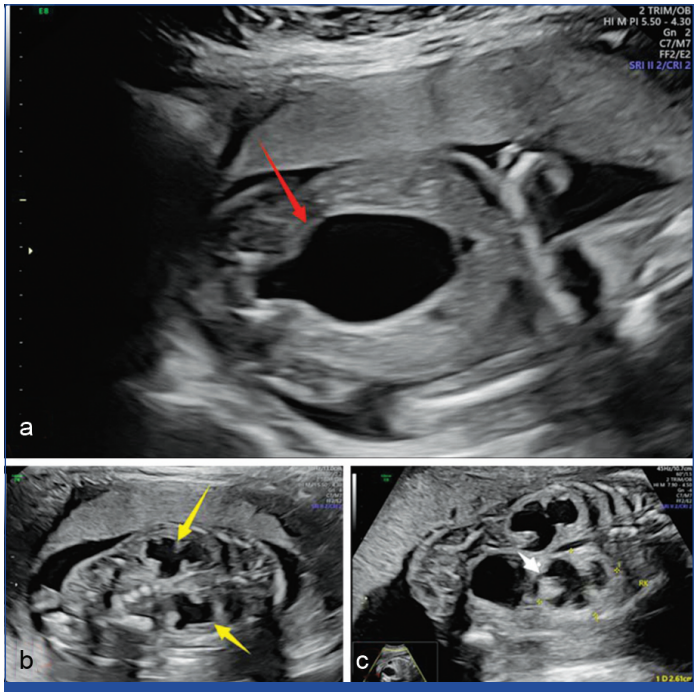
The most common cause of Lower Urinary Tract Obstruction (LUTO) in the male foetus is the Posterior Urethral Valve (PUV). It can cause severe complications such as renal dysfunction and dysplasia of the lung. Early prenatal diagnosis is vital for assessing prognosis, providing parental counselling, and planning additional care. This is a case report of a 22-year-old primigravida in whom characteristics of foetal bladder outlet obstruction were identified on a routine scan of the second trimester. Ultrasonography showed a significantly distended foetal bladder at gestational week 21, indicative of megacystis. Moreover, the classical keyhole sign was also seen due to dilatation of the proximal urethra. Despite these anomalies, amniotic fluid was within the normal range, suggesting that renal function was not yet severely impaired. The patient decided to have the pregnancy terminated. Foetal autopsy revealed the existence of megacystis and bilateral hydroureter with hydronephrosis, which were in line with the prenatal imaging findings. This case highlights the importance of identifying sonographic signs, in particular the keyhole appearance and related urinary tract dilatation. Early identification of the above findings is crucial in appropriate clinical decision-making.

Keywords: Anomaly scan, Bladder outlet obstruction, Counselling, Foetal lower urinary tract obstruction, Hydronephrosis

CASE REPORT

A 22-year-old primigravida with a history of non-consanguineous marriage and known hypothyroidism on levothyroxine, who had conceived after ovulation induction. She had no history of diabetes, hypertension, infections, or teratogenic exposure during pregnancy. Target ultrasound performed at around 21 gestational weeks showed a singleton foetus with a normal cranium, spine, thorax, and limbs. Nevertheless, the urinary system was severely affected. The foetal bladder was significantly distended, and the craniocaudal diameter was about 37 mm. The proximal urethra was dilated, giving the typical keyhole appearance suggestive of bladder outlet obstruction. Both kidneys were swollen with renal pelvicalyceal dilation. The right renal pelvis was found to be about 7.9 mm, and the left was about 7 mm in antero-posterior diameter, and there was corresponding central calyceal dilatation. Bilateral ureters were also dilated [Table/Fig-1]. Amniotic fluid volume was normal despite these anomalies. Results were indicative of foetal bladder outlet obstruction and a high likelihood of a PUV, causing bilateral urinary tract dilatation {Urinary Tract Dilatation (UTD) grade 2-3} [1]. Multidisciplinary counselling was provided to the patient by the obstetrician, paediatric surgeons, and foetal medicine specialists. They were informed that the renal function was preserved with normal liquor volume at the time of the scan. However, there was a probability that the condition may progress in later gestation and present with renal dysplasia and oligohydramnios. In such a case, the possibility of requiring a vesico-amniotic shunt in later gestation was explained to the couple. In case the couple opted to continue the pregnancy, serial follow-up scans were recommended to assess renal function and amniotic fluid volume.

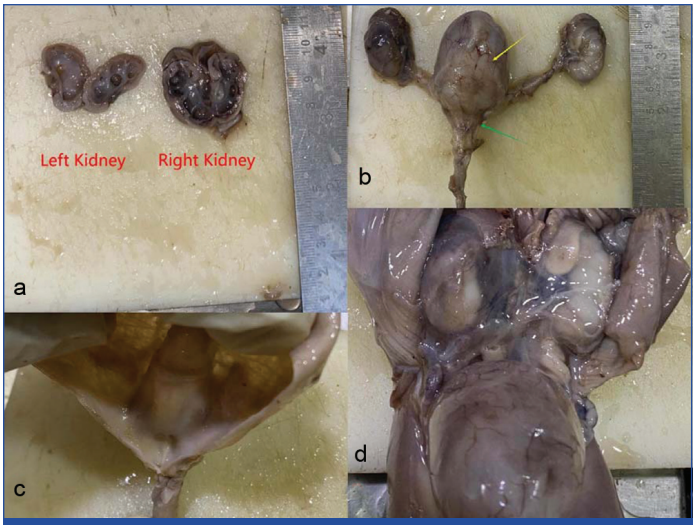
The patient chose to terminate the pregnancy. Oral mifepristone followed by vaginal misoprostol was used for medical termination. The pregnancy was terminated without any complications, and the foetus was expelled. The patient was haemodynamically stable. The foetus was male, with an estimated age of 21 weeks and a weight of 500 grams. The abdomen was distended. Thorough examination revealed a significantly enlarged urinary bladder of about 4×3.5×3 cm. Bilateral ureters were dilated. Both kidneys exhibited cystic enlargement of the pelvicalyceal systems, consistent with hydronephrosis. The microscopic observation revealed primitive glomeruli, tubular structures, and dilated collecting systems. Other organs, such as the thymus, spleen, heart, and adrenal glands, were not remarkable. Placental inspection showed that the umbilical cord had three vessels and had no signs of inflammation. The final autopsy diagnosis was the PUV and megacystis with bilateral hydroureter and hydronephrosis [Table/Fig-2].



[Table/Fig-1]: Ultrasound images using standard curvilinear probe (3.5-5 MHz) showing: a) Classic keyhole sign- dilated bladder and proximal urethra (red arrow); b) Bilateral dilated pelvicalyceal system (yellow arrows); c) Dilated right ureter (white arrow).

DISCUSSION

The most prevalent congenital pathophysiologic cause of LUTO in male infants is a PUV, which is caused by abnormal membranous



[Table/Fig-2]: a) Cut sections of bilateral kidneys showing cystic dilatation of pelvicalyceal systems; b) Gross specimen showing overdistended bladder (yellow arrow) and dilated proximal urethra (green arrow); c) A longitudinal fold was seen in the posterior urethra; d) Gross specimen of kidneys and bladder in the abdomen and pelvis after incision.

folds along the posterior urethra and results in urinary outflow obstruction and progressive dilation of the bladder and upper urinary tract. It contributes to a high percentage of the cases of obstructive uropathy that cause paediatric renal impairment [2,3].

With routine obstetric ultrasonography, prenatal diagnosis has become possible. The common sonographic findings are an overdistended bladder, a dilated proximal urethra (also known as the keyhole sign), bilateral hydronephrosis, and hydroureter (found in severe cases and accompanied by oligohydramnios and renal dysplasia) [4,5]. With an incidence of 1:5000 to 1:25 000 of live births, PUV may lead to renal failure in 25% to 43% of affected patients [6].

In a case of PUV, the pathologic obstruction is at the site of the posterior urethra and may result in progressive increase of intravesical pressure, which in turn causes dilation of the bladder, ureters and renal collecting systems [3].

The amniotic fluid volume status is also an important prognostic factor in foetal obstructive uropathy. Foetuses diagnosed antenatally with LUTO who demonstrate preserved amniotic fluid volume at mid-gestation generally have a favourable perinatal outcome, with good survival rates and minimal requirement for prolonged respiratory support. Nevertheless, despite this relatively reassuring early course, these children remain at a significant risk for long-term renal sequelae, including chronic kidney disease, progression to end-stage renal disease, and the potential need for renal replacement therapy [7].

According to Pellegrino C et al., despite early diagnosis and treatment, a considerable number of the affected children had a dysfunctional bladder with different stages of chronic kidney disease [2].

Joudar I et al., have reported foetal ascites secondary to bladder rupture caused by a PUV [8]. Gerede E et al., have presented the case, which confirms that this type of congenital malformation is associated with risks of renal impairment until failure, which is also the basis of the parents' decision to terminate the pregnancy [9].

Treatment of foetal PUV remains challenging, and in most cases, management decisions are individualised. Foetal interventions like vesico-amniotic shunting have also been proposed to decompress the urinary tract to improve perinatal outcomes, but the efficacy of foetal therapy is questionable, and the procedures may result in complications [10].

Differentials for foetal bladder enlargement include urethral atresia, which is characterised by complete urethral obstruction with distal underdevelopment, typically presenting as a markedly enlarged, irregular bladder with oligohydramnios [11]. Demonstration of a normal distal urethra on pathology excluded this entity in our case. Prune belly syndrome consists of deficient abdominal musculature, undescended testes, and urinary tract abnormalities, often accompanied by multisystem anomalies and reduced amniotic fluid on prenatal imaging [12]. In our case, the abdominal wall musculature was normal, thus excluding this diagnosis. Megacystis-Microcolon-Intestinal Hypoperistalsis Syndrome (MMIHS) presents with a persistently enlarged non-obstructed bladder, normal or increased amniotic fluid, and associated bowel dilatation, more frequently seen in female foetuses [13]. In our case, a PUV was suggested by bladder outlet obstruction, with dilatation of the posterior urethra and progressive upper tract involvement.

CONCLUSION(S)

The PUV is a major cause of foetal bladder outlet obstruction that may have long-term renal outcomes. This case has underscored the critical importance of prenatal ultrasonography in early diagnosis, particularly for detecting typical findings such as the keyhole sign, megacystis, and bilateral urinary tract dilatation.

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