Case Report

In Utero Treatment of Obstructive Ureterocele

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Abstract

Lower urinary tract obstruction consists of a heterogeneous group of conditions in which the normal urethral egress of urine from the fetal bladder is impaired. The most frequent diagnoses are posterior urethral valves, urethral atresia, and less common obstructive ureterocele. We report a case of a fetus with prenatal diagnosis of obstructive ureterocele who presented progressive bilateral hydronephrosis. A fetal cystoscopy with laser ablation was performed.

Keywords: Fetal cystoscopy, lower urinary tract obstruction, obstructive ureterocele

Introduction

Lower urinary tract obstruction (LUTO) is an entity that comprises different diseases, often causing bladder neck obstruction.[1] It occurs in approximately one out of 5000 newborns, with up to 90% being males.[2]

LUTO can be due to different causes. The most frequent is the presence of posterior urethral valves, which account for about 70%–80% of cases. The second most frequent cause corresponds to urethral atresia, especially prevalent in cases diagnosed early in gestation (11–14 weeks) and in females. The third obstructive pathology to consider is the obstructive ureterocele.[2] Other causes of LUTO include prune belly syndrome, persistent cloaca, caudal regression, and megacystis-microcolon-intestinal hypoperistalsis syndrome, which although were less common, must be considered. In about 20% of cases, there are other structural or chromosomal anomalies associated.[3]

We describe a case of in utero diagnosis and treatment of an obstructive ureterocele by fetal cystoscopy with laser ablation.

Case-Report

Thirty-nine-year-old G2P1 with no prior medical history. The previous pregnancy, 5 years ago, was normal. The father has left renal duplication.

Received: 16-01-2022 **Revised:** 14-02-2022 **Accepted:** 15-03-2022 **Available Online:** 05-07-2022

Access this article online

Quick Response Code:

Website: https://journals.lww.com/jmut

DOI: 10.4103/jmu.jmu_8_22

The first trimester combined screening revealed a low risk for aneuploidy. At the second trimester ultrasound, at 21 weeks and 6 days, an intravesical ureterocele [Figure 1] associated with right renal duplication and hydronephrosis (right renal pelvis of 7 mm) was diagnosed. The left kidney was sonographically normal. A complete anatomical evaluation, including fetal echocardiography, was performed which allowed exclusion of associated anomalies. In addition, an amniocentesis showed a normal 46, XX karyotype.

Follow-up ultrasounds showed bladder outlet obstruction from the ureterocele, an increasing of megacystis and parietal bladder thickness, a progressive calyceal and ureteric dilatation with bilateral hydronephrosis, and a decreasing of amniotic fluid volume [Figure 2]. Consequently, at 26 weeks and 4 days, the pregnant woman was referred to a fetal therapy center.

At the admission, these ultrasonographic findings were confirmed and a fetal surgery to decompress the urinary system was proposed. Fetal cystoscopy showed an ureterocele extending from the right side of the bladder to the urethra. Therefore, ureterocele's laser ablation was performed and proceeded without complications. Postsurgical recovery was uneventful with no maternal complications. Twenty-four h after the procedure, an ultrasound evaluation showed normal

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How to cite this article: Solinho M, Saraiva S, Lourenço C, Brito C. In utero treatment of obstructive ureterocele. J Med Ultrasound 2023;31:154-6.

bladder size with thickened hypertrophic walls and bilateral ureterohydronephrosis. No signs of ureterocele were identified and amniotic fluid index was normal.

Follow-up revealed a normal bladder and improvement in bilateral hydronephrosis, although right hydronephrosis had persisted until delivery [Figure 3]. Amniotic fluid index remained normal throughout the remainder of the pregnancy. At 36 weeks and 6 days, the pregnant woman went into spontaneous labor. A 2800 g female was delivered by emergent cesarean section due to nonreassuring fetal status, with an Apgar score of 9/10.

The newborn was admitted to the neonatal intensive care unit, as she had an iatrogenic abdominal eventration and for renal function surveillance [Figure 4]. On 1st day of life, the abdominal eventration was corrected. After 5 days, she was discharged and referred to pediatric urology outpatient care clinic under antibiotic prophylaxis.

Through the postnatal evaluation, the mother refused radionuclide renal scans several times. However, several routine renal/bladder ultrasounds were performed and at 18 months old, the child showed a right kidney of smaller dimensions, with no hydronephrosis and a right ureterocele was identified; the left kidney was normal. From now on, the child was lost follow-up.

Discussion

Despite being one of the less common causes of LUTO, the obstructive ureterocele represents the most frequent form of bladder outlet obstruction in female.[4] Therefore, we must be aware of this diagnosis.

The prenatal diagnosis of LUTO is classically based on ultrasound identification of several signs: a dilated/thick-walled bladder, bilateral hydronephrosis, dilated ureters, a visible posterior urethra (also known as the *keyhole sign*) and reduced amniotic fluid; however, these signs are not always present together.[3] In our case report, we identified several of these findings.

Regarding postnatal renal function, it is difficult to predict it from prenatal appearance, however, studies have recently identified predictive features.[3] Measurement of amniotic fluid volume and the appearance of the renal cortex at the time of diagnosis of LUTO show promising predictive accuracy for postnatal renal function.[1] It has been proven that the presence of oligo/anhydramnios early in pregnancy almost invariably implies the development of pulmonary hypoplasia, lung failure, kidney failure, and death.[2,5]

Prenatal assessment of fetal renal function may be also performed to select the fetuses who will benefit from in utero therapy and to predict the postnatal outcomes. Recent studies show that fetal urinalysis between 13 and 23 weeks can distinguish fetuses that would benefit from intrauterine fetal therapy.[6] In addition, some studies demonstrated that β2-microglobulin values are predictive of glomerular function.[7] Recently, staging systems have been established to select patients for in utero intervention. Ruano *et al*. proposed

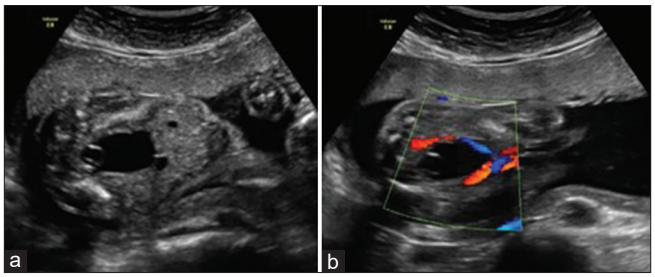

Figure 1: (a) Fetal intravesical ureterocele. (b) Fetal intravesical ureterocele

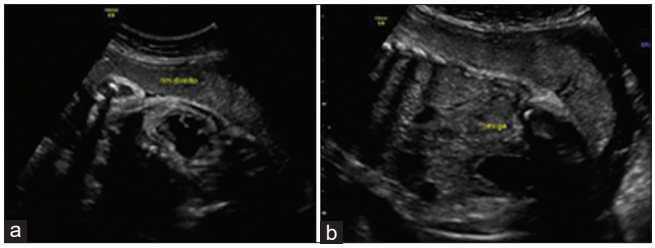

Figure 3: Right kidney with hydronephrosis (a) and normal-sized bladder (b)

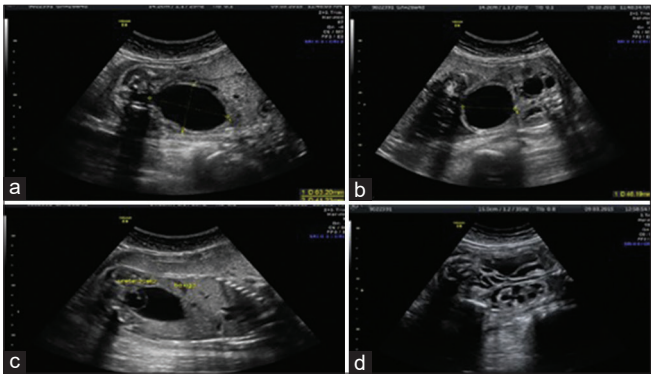

Figure 2: (a and b) Progressive megacystis. (c) Ureterocele inside the bladder. (d) Bilateral hydronephrosis and megaureter

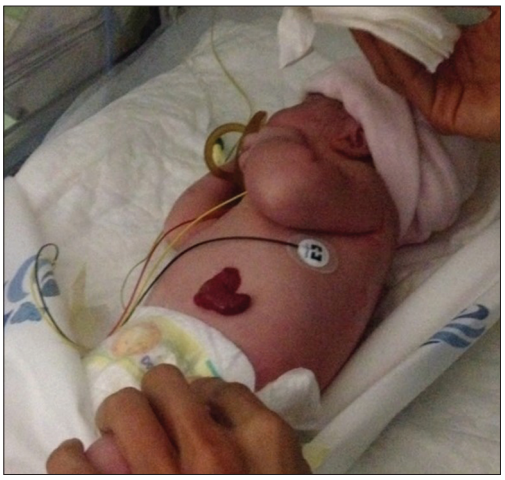

Figure 4: Iatrogenic abdominal wall eventration

a standardized prenatal evaluation based on four stages of obstruction,[8] whereas Fontanella *et al*. proposed an alternative system based on amniotic fluid and bladder volume correlating them to gestational age at diagnosis.[9] The intervention window is also important to select patients for intervention, as a late one may not be beneficial to the patient's renal or pulmonary outcome.[3]

Despite the efforts to define the optimal candidates for treatment, the prognosis remains reserved due to the fetal affectation's variability of the different nosological entities and the current therapeutic limitations.[3] After appropriate patient selection and thorough counseling, the choice of technique depends on local expertise.[3] The vesicoamniotic shunting has been used for the past 30 years with poor-moderate results. Recently, fetal cystoscopy has been introduced both for the evaluation and for the possible treatment of fetuses with obstructive uropathy.[10] We referred the patient to a specialized center that opted for this technique. The role that fetal cystoscopy will have in the management of this pathology remains to be determined. In our case report, since renal MAG3 scan in postnatal period was not performed, we cannot take conclusions about the remaining renal function nor assess the relative contribution of each kidney to overall renal function. In addition, the follow-up after the 18 months old was lost.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest statement

There are no conflicts of interest.

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