Incomplete left and complete right bilateral double collecting system with renal hypoplasia of right superior pole and right ureterocele

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ABSTRACT

The case of a seventeen-month-old female patient sent to the urology service of the Centro Médico Nacional del Noreste in Monterrey, Nuevo León, Mexico, for right hydronephrosis is presented. There was important medical history of right hydronephrosis detected by ultrasound at 27 weeks of gestation that persisted at 31 weeks seen in repeat ultrasound. A third ultrasound image was taken at 14 days of extrauterine life, showing right hydronephrosis and ipsilateral ureter dilatation with left ureterocele. At 6 months of age another ultrasound study revealed the same findings.

Prenatal hydronephrosis is the most commonly detected prenatal genitourinary abnormality. Differential diagnosis can range from a self-limited condition with no clinical significance (physiological hydronephrosis with normal postnatal ultrasound) to conditions that require surgery or that cause renal destruction.

Key words: Hydronephrosis, ureterocele, double collecting system, Mexico.

RESUMEN

Se presenta el caso de una paciente de un año y cinco meses de edad, enviada al servicio de urología del Centro Médico Nacional del Noreste en Monterrey, Nuevo León por hidronefrosis derecha. Los antecedentes de importancia con los que cuenta son: hidronefrosis derecha detectada a las 27 semanas de gestación por ultrasonografía. Persistieron estos datos a las 31 semanas de gestación, y aún a los 14 días de vida extrauterina: hidronefrosis derecha y dilatación del uréter ipsilateral con ureterocele izquierdo. A los seis meses de edad, se mantiene con los hallazgos mencionados.

La hidronefrosis prenatal es la anormalidad prenatal genitourinaria más comúnmente detectada. El diagnóstico diferencial puede establecerse contra una gama de situaciones que van de una condición autolimitada, sin significancia clínica (hidronefrosis fisiológica con una ultrasonografía postnatal normal) a situaciones que requieren intervención quirúrgica o que provoca destrucción renal.

Palabras clave: Hidronefrosis, ureterocele, doble sistema colector, México.
INTRODUCTION

Urinary tract development arises from the intermediate mesoderm. The pronephros is a non-functional tissue that is formed at 3-5 weeks of gestation (WOG). The mesonephros is then formed and produces a small quantity of urine at 5-11 WOG. After mesonephros regression the ureteral bud appears during the 4th WOG and induces metanephros formation that will form the definitive kidney. The collecting systems, calyces, renal pelvis, and ureter arise from the ureteral bud of mesonephric origin, while the nephrons and tubules arise from the metanephric blastema. 1-4

The first sign of tubular renal function appears in the metanephric kidney between 9 and 12 WOG. At the 14th WOG the loop of Henle becomes functional and tubular reabsorption occurs. In the human fetus, new nephrons are formed after the 36th WOG. Nephrogenesis is complete at the time of birth in full-term infants but nephron formation continues after birth in pre-term infants. 4

Hydronephrosis diagnosis and treatment in newborns and preadolescents is still controversial among urologists, radiologists, pediatricians, and nephrologists. In the last 20 years numerous cases of fetal hydronephrosis have been diagnosed thanks to obstetric ultrasound (approximately 1.4-4.5% of all imaging studies).1

CASE PRESENTATION

The case of a seventeen-month-old female patient referred to the urology service of the Centro Médico Nacional del Norte for hydronephrosis is presented. Past medical history included right hydronephrosis that had been detected at 27 weeks of gestation with prenatal control ultrasound, identifying right pyelocalycectasis. Repeat ultrasound at 31 weeks of gestation showed that condition persisted. Another repeat ultrasound at 14 days of extraterine life revealed right hydronephrosis and right ureteral dilatation with left ureterocele. At 6 months of age these findings persisted. Physical examination did not provide any relevant data. Kidney scintigrafm reported retention in the pyelocalyceal systems, dilated primarily on the left side and marked urine passage to the bladder. Important dilatation of left ureter was striking. Total glomerular filtration rate was 118 mL/min. Contribution of right kidney was 38% and left 62% (Figure 1).

Other studies:

Kidney ultrasound that showed bilateral pyelic dilatation.

Micturition cystogram with filling defect in middle bladder with no reflux data (Figures 2 and 3).

Excretory urography: Delay in collection and elimination of contrast medium in right kidney unit with slight ureteropyelocalycectasis and incomplete double collecting system; the left with megaureter in left inferior collecting system with large filling defect at bladder level (Figures 4 and 5).
Abdominopelvic computed tomography in diffuse cortical nephrographic phase with reduction of contrast medium collection in upper pole of right kidney and image of double collecting system in left kidney (Figure 6).

Urethrocystoscopy revealed right ureterocele, left horseshoe meatus in B position draining clear urine. Left double collecting system union 2 cm from meatus was observed with 7F pediatric cystoscope. Through surgical intervention complete right double collecting system was found with renal dysplasia of the upper pole and ureterocele with integral inferior system. Upper pole partial nephrectomy and superior ureterectomy, ureterocele detachment, and double-J catheter placement in inferior system were carried out.

Histopathological report stated: ureter with chronic follicular ureteritis, renal pelvic remnant that had scant consistent renal parenchyma and renal cystic dysplasia.

DISCUSSION

Ureteropelvic duplication is the most frequent upper urinary tract abnormality. It affects 0.8% of the population and is seen in 1 out of every 125 autopsy studies. Its incidence is twice as high in females, affecting both the left and right sides equally, and unilateral cases are six times more frequent than bilateral cases. There is a genetic predisposition and its incidence is up to 8 times higher in parents and siblings of the affected patient. 

Embriologically it is caused by complete or incomplete duplication of the ureteral yolk or metanephric diverticulum at the 8th WOG. There are two ureters at the inferior level in ureteral duplication. The opening of the ureteral orifices in the bladder follows a pattern - the Weigert-Myer law - in which the ureter that drains the upper pole is always medial and caudal in relation to the ureteral orifice of the lower pole. Therefore, the ureter of the lower pole that is more lateral and cranial will have poorer bladder wall muscle support and in turn a shorter submucosal tunnel, facilitating the possibility of presenting with vesicoureteral reflux. Fehrenbaker (1972) described the presence of reflux in more than two-thirds of infants that presented with urinary infection and that had complete double pyeloureteral system. Double collecting systems can include a multicystic dysplastic portion, generally in the upper pole and rarely in the lower pole.

It is more likely for the ureter that drains the upper pole in a duplicated system to have an abnormality at the bladder level. It can open in a more caudal position, constituting an ectopic ureter, or it can be in the form of a cystic dilatation of the ureteral distal extreme in its submucosal or intramural portion, forming a ureterocele.

Prenatal ultrasound that is usually ordered at 16-20 WOG has revealed many cases of uterine hydronephrosis. Prenatal hydronephrosis is the most commonly detected prenatal genitourinary abnormality. Differential diagnosis can be from a self-limited clinically
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Figure 5. Excretory urography showing incomplete left double collecting system with megaureter in left inferior system, bilateral ureteropyelocaliectasis and a large bladder filling defect.

Figure 6. Abdominopelvic computed tomography in diffuse cortical nephrographic phase with reduction of contrast medium collection in right kidney upper pole and image of double collecting system in left kidney.

Insensitive condition (physiological hydronephrosis with normal postnatal ultrasound) to conditions requiring surgery or that cause renal destruction.²

Important factors to be taken into consideration in hydronephrosis evaluation include well-being of the fetus, gestational age, unilaterality or bilaterality of the pathology, and liquid amniotic volume. Differential diagnoses in hydronephrosis are:²

- Ureteropelvic junction obstruction
- Vesicoureteral reflux
- Primary megaureter with no reflux
- Ureterocele
- Ureterovesical junction obstruction
- Ectopic ureter
- Posterior ureteral valves
- Multicystic renal dysplasia
- Recessive autosomal renal polycystosis
- Exstrophy
- Prune Belly syndrome

Hydronephrosis presents in 0.25% of all pregnancies. The American Society for Maternal-Fetal Medicine has formulated a scale to evaluate dilatations based on uterine ultrasound findings:

- Anteroposterior diameter of renal pelvis (APDP) from 3-5 mm is considered to be slight
- APDP from 6 - 9 mm = moderate
- APDP above 10 mm (in the 30th WOG) or 15 mm (in the 33rd WOG) is considered to be serious dilatation called obstructive dilatation

The last two findings require postnatal isotope renogram for the purpose of determining optimum treatment. Studies show spontaneous recovery in 50% of mild dilatations, in 15% of moderate dilatations, and 0% of serious dilatations.¹

Liquid amniotic total volume ranging from 500-2000 mL is an important factor in fetal hydronephrosis. In early gestation the source of amniotic liquid is a maternal plasma transudate. Around the 16th WOG the majority of amniotic liquid is fetal urine. The volume constantly increases up to the end of the second trimester when it then stabilizes and slowly decreases before the end of gestation. Oligohydramnios refers to a reduced quantity of amniotic liquid that will then have an effect on the development of the lungs.²
Prenatal hydronephrosis can have obstructive and non-obstructive causes. When there is oligohydramnios and an increase in renal echogenicity these factors are highly predictive of obstructive etiology with 100% sensitivity and 93% specificity. This has implications in prenatal and postnatal management since the fetus presenting with obstructive etiology can be a candidate for prenatal surgery.

Fetal abnormalities of the urinary tract are a common finding that occur in 0.2 – 1.5% of all pregnancies. Hydronephrosis is the most common abnormality found in prenatal ultrasound studies; it is not the diagnosis but it serves as a sign leading to its cause. Infants presenting with prenatal hydronephrosis have an approximately 12 times higher risk of having pyelonephritis requiring hospitalization during their first year of life and it is more highly associated with females. 3

The type of surgical treatment to be used should be based on different parameters: diagnosis, age of patient, general health status, associated pathologies, overall kidney function and that of the affected segment.

### CONCLUSIONS

Surgical repair techniques at both the upper level (ureteropelvic or ureteroureteral anastomosis) and lower level (ureterovesical neoinplant with or without ureteral modeling and ureterectomy plus ureteral reimplant) have a high success rates. Even when complications present due to failure in the technique used, anatomical or functional injury, or pathology recurrence, they are acceptable based on the principle of maximum kidney function preservation possible.

However, when there is no function or it is minimal, maintaining the affected renal segment is not justified, and even less so if the repair attempt could compromise the viable healthy or functional segment.

Heminephrectomy is the surgical procedure of choice when the affected pole is not functioning. 3

### BIBLIOGRAPHY