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CASE REPORT

Multiple and bilateral renoureteral malformations. Case report and literature review

Malformaciones renoureterales múltiples y bilaterales. Reporte de caso y revisión de la literatura

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ABSTRACT

Congenital malformations constitute anomalies of structure and/or function of prenatal origin, which are often asymptomatic or cause few symptoms, but currently have a high incidence. With the aim of showing the findings detected in the imaging studies carried out, the case of a young adult with repeated mild urinary sepsis is presented, who was diagnosed with multiple and bilateral renoureteral malformations in the infertility consultation. The majority of congenital renoureteral malformations imply serious biopsychosocial problems for the patient, with the consequent impact on the psychological sphere and the social life of family members when diagnosed in the prenatal stage, so they inevitably constitute a health problem, since they contribute to fetal and infant mortality or increased morbidity in any age group. For this reason, early diagnosis is essential to guarantee appropriate behavior and quality of life.
Keywords: Congenital Malformations; Renoureteral Malformations; Early Diagnostic; Ultrasound; Descending Urogram; Double Excretory System; Ureterocele.

RESUMEN

Las malformaciones congénitas constituyen anomalías de estructura y/o función de origen prenatal, que muchas veces son asintomáticas o provocan escasos síntomas, pero exhiben alta incidencia en la actualidad. Con el objetivo de mostrar los hallazgos detectados en los estudios imagenológicos realizados, se presenta el caso de una adulta joven con sepsis urinarias leves a repetición, a quien en consulta de infertilidad se le diagnosticaron malformaciones renoureterales múltiples y bilaterales. La mayoría de las malformaciones congénitas renoureterales implican serios problemas biopsicosociales para el paciente, con la consecuente afectación de la esfera psíquica y la vida social de los familiares cuando se diagnostican en etapa prenatal, por lo que inevitablemente constituyen un problema de salud, pues contribuyen a la mortalidad fetal e infantil o al aumento de la morbilidad en cualquier grupo etario. Por esta razón se hace imprescindible el

diagnóstico precoz para garantizar la conducta adecuada y la calidad de vida.

Palabras clave: Malformaciones Congénitas; Malformaciones Renoureterales; Diagnóstico Precoz; Ultrasonido; Urograma Descendente; Doble Sistema Excretor; Ureterocele.

INTRODUCTION

The first references to the kidney and its pathology date back to ancient Egypt (1500 BC), but it was Hippocrates (460–370 BC) who first recognized and described various subtle macroscopic changes in urine that reflected specific diseases in different organs, including the kidney.[1] Kidney formation begins in week 3 and nephrogenesis continues until week 36, therefore the kidneys and urinary tract are susceptible to the influence of environmental risk factors that disrupt development throughout pregnancy.[2] In line with the above, congenital anomalies *of the kidney and urinary tract* (CAKUT) are a series of malformations that can occur in the kidney, the collecting system, the bladder, or the urethra. The malformations may have underlying causes, such as specific genetic factors, epigenetic factors, and environmental influences on fetal development. According to Díaz García et al., congenital malformations occupy an important place among human diseases, mainly because of their functional, psychological, and social impact on the life of each individual. They also suggest that CAKUTs constitute a heterogeneous group of pathologies with a wide range of symptoms and signs, ranging from asymptomatic cases to cases incompatible with life. According to Mejía Mesa et al.[3], congenital renal malformations are very common, with the most common anomaly being dilation of the urinary tract. They assert that CAKUTs are a significant cause of morbidity and mortality in newborns and are also related to other non-renal congenital disorders in approximately 30 % of cases. They have even been identified as the leading cause of chronic renal failure in children, especially those with severe malformations.

Data from the World Health Organization (WHO) confirm that congenital anomalies are the fourth leading cause of neonatal death and the seventh leading cause of death in children under 5 years of age worldwide. The prevalence of major malformations ranges from 2 % to 4 %, depending on the age of the population evaluated and whether the diagnosis was made pre- or postnatally. Isolated minor malformations are much more common and their prevalence is even more variable, with figures ranging from 14 % to 35,8 %. Fetuses with three or more minor malformations are at increased risk of having a genetic syndrome or a major malformation.[5] In the Americas, birth defects are among the leading causes of death and, although in lower-income countries they account for less than 5 % of infant mortality causes in relative terms, in higher-income countries this group of causes accounts for 30 % of deaths recorded before the age of one. They are also one of the leading causes of disability in children.[6] Congenital anomalies of the kidney and urinary tract account for 20-30 % of anomalies detected by prenatal ultrasound. ([4] The first report of a prenatal ultrasound was published in 1958, and the first diagnosis of a renal anomaly by prenatal ultrasound was in 1970.[7] In this regard, imaging plays a decisive role, as it offers multiple modalities that facilitate early and accurate diagnosis in the prenatal or immediate postnatal stage, which favors adequate follow-up and, therefore, a better prognosis for the patient.

The above constitutes the motivation for this case presentation, which aims to show the images of the findings detected in the imaging studies performed on an apparently asymptomatic adult with multiple bilateral renoureteral malformations.

CASE REPORT

A 28-year-old female patient, obstetric history $E_4P_0A_4$ (spontaneous), with a family history of hypertension in her living mother, which is controlled with diet and treatment, personal history of “good health,” asymptomatic, in good general condition, began to be studied in an infertility consultation because “despite having a stable partner and not using contraception, she has not been able to conceive.” After a thorough interview, the patient complained of “repeated mild urinary tract infections” for which she did not seek treatment or follow-up with a doctor. The physical examination was negative. Several tests were ordered to confirm the diagnosis and determine the course of action.

Laboratory tests were performed, including Hb, erythrocyte sedimentation rate, cyturia, vaginal exudate, and blood chemistry, which were normal. Chest X-ray was unremarkable. An abdominal ultrasound showed no abnormalities in the liver, gallbladder, pancreas, aorta, or spleen. The kidneys were normal in size, echogenicity, and position, appearing to have an uncomplicated double excretory system (no pyelocaliceal dilatation) and bilateral double ureters (both upper thirds visible). The bladder was of good capacity, with thin walls and no focal lesions. Intravesical dilatation of both ureters was confirmed, more pronounced on the left, which increased with the Valsalva maneuver due to vesicoureteral reflux extending to the distal segments, and was also accompanied by slight dilatation of the excretory system. Uterus of normal size, homogeneous texture, with IUD in the endometrial cavity. Adnexa without abnormalities. No ascites.

DISCUSSION

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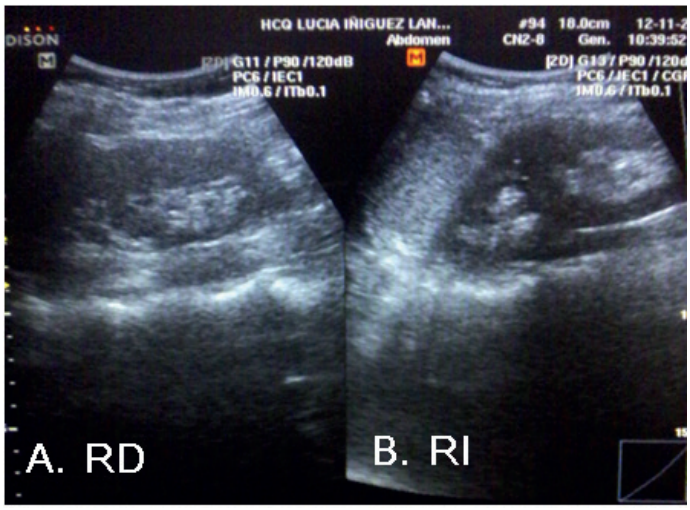

Figure 1. Renal ultrasound: kidneys of normal size and echogenicity, appearing to have an uncomplicated double excretory system

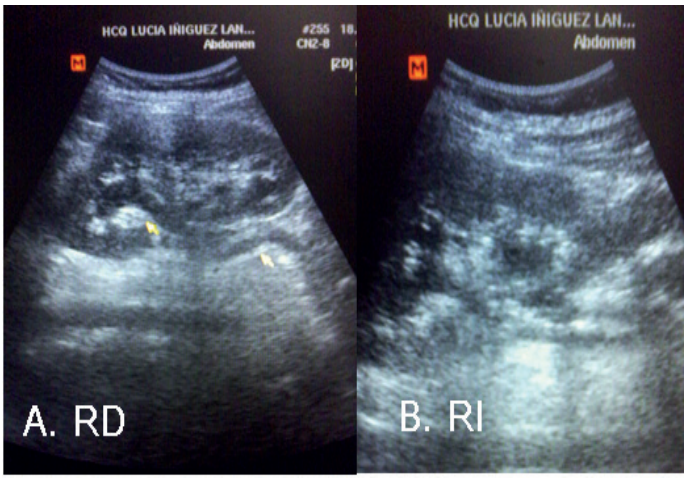

Figure 2. Ultrasound with Valsalva maneuver: bilateral double excretory system associated with double ureter, both dilated

Based on the findings described, a descending urography was indicated to confirm the presence of renoureteral malformations and assess renal function. The following images were observed in the study.

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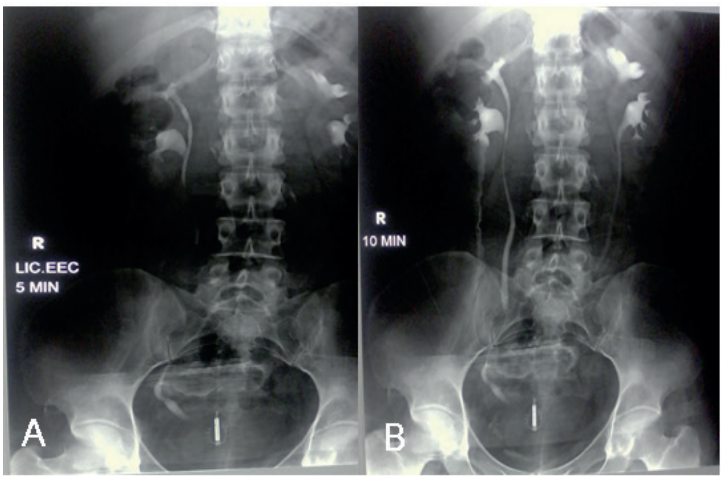

Figure 3. DESCENDING UROGRAM: in the 5- and 10-minute views (A and B), there is good contrast concentration and elimination by both kidneys. The presence of a bilateral double excretory system with double ureter is confirmed. IUD in pelvic excavation. Rachischisis. Scoliotic attitude of the lumbar spine with left convexity

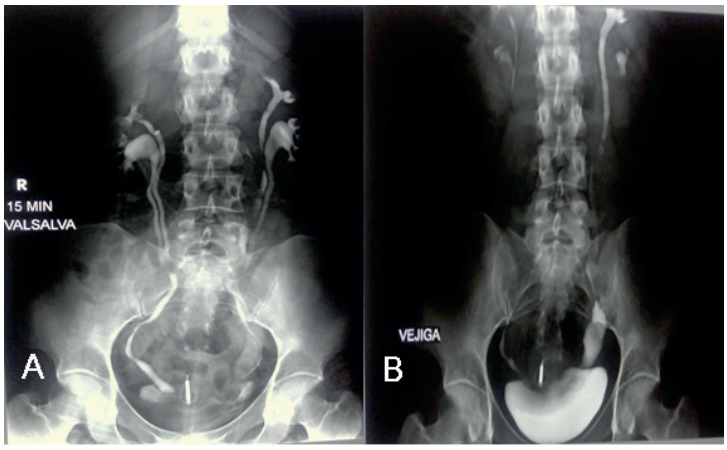

Figure 4. DESCENDING UROGRAM: A: view at 15 minutes with Valsalva maneuver, where both incomplete double ureters and dilation of their distal end are more clearly visible, predominantly on the left. B: bladder insufficiently full, however, this cystographic view confirms the markedly dilated distal third of the left ureter

The case is concluded as a bilateral double excretory system with bilateral incomplete double ureter associated with double ureterocele.

DISCUSSION

Congenital anomalies are defined as structural or functional defects in the body that are present at birth and are therefore of prenatal origin, including genetic, environmental, or unknown defects, even if the defect is not apparent in the newborn and manifests itself later. They can be major or minor; major anomalies significantly affect health and generally require medical or surgical treatment, while minor anomalies are defined as alterations in the phenotype with no functional or aesthetic consequences. The pathogenesis of the different entities encompassed by CAKUT is complex, in line with the complicated mechanism of formation and embryological development of the urinary system. CAKUT is more common in males than in females, and there are cases of familial aggregation. They may be part of multiorgan syndromes.[9] In this regard, the scientific literature[10] also describes hereditary syndromes with renal involvement, some of which are Coloboma-Renal Syndrome, also known as papillo-renal syndrome, Kallman Syndrome, and Fraser Syndrome. On the other hand, the possible etiologies of these defects are characterized by genetic factors, consanguinity increases the prevalence of rare genetic congenital anomalies and almost doubles the risk of neonatal and infant death, intellectual disability, and other congenital anomalies in marriages between first cousins. Maternal infections, such as syphilis or rubella, are a major cause of congenital anomalies in low- and middle-income countries. It should be noted that the mother's nutritional status, iodine and folate deficiencies, overweight, and diseases such as diabetes mellitus are related to some congenital anomalies. Environmental factors, such as maternal exposure to certain pesticides and other chemicals, as well as alcohol, tobacco, psychoactive drugs, and radiation during pregnancy, can increase the risk of congenital anomalies in the fetus or newborn. Likewise,

risk factors for congenital malformations include: advanced maternal age (over 35), low weight and height for gestational age, fetal growth restriction, family history of congenital malformations, physical factors, maternal exposure to agrochemicals, and acute maternal illness in the first trimester of pregnancy.[8]

Therefore, the evaluation of a patient with CAKUT should include, in addition to studies directly related to their nephrourological structural alteration, a specific search for extrarenal manifestations and a thorough family history to help identify the underlying molecular cause.[9] CAKUT is classified into several groups: renal parenchymal abnormalities, upper and lower urinary tract abnormalities, and renal position abnormalities. Renal parenchymal abnormalities include renal agenesis, renal hypoplasia, and renal dysplasia. Renal agenesis is defined as the absence of a kidney and ureter. Hypoplasia is defined as a decrease in the number of nephrons. Renal dysplasia is defined as the presence of dysplastic elements in the renal parenchyma, such as interstitial fibrosis or the presence of non-renal tissue. The most severe form of renal dysplasia is multicystic renal dysplasia. Anomalies of the upper and lower urinary tract include double excretory system, stenosis of the ureteropelvic junction, stenotic megaureter, vesicoureteral reflux, ectopic ureter, ureterocele, bladder exstrophy, and posterior urethral valves. The main abnormalities in kidney position include ectopic kidney, malrotated kidney, and horseshoe kidney.[10] A "double" kidney is one that has two separate pyelocaliceal systems and therefore has an upper and lower hemisystem. The ureters may join together. If this occurs at the ureteropelvic junction, it is called a bifid pelvis; if they join more distally but before reaching the bladder, it is called a bifid ureter. And if they do not join and drain their respective poles or separate hemisystems into or outside the urinary tract, it is called a double excretory system. The double excretory system is the most common congenital anomaly of the urinary tract, estimated to occur in 1 in 125 live births. It is identified more frequently in females at a ratio of 2:1, and the unilateral form is six times more common than the bilateral form.[11] The double ureter is divided into complete and incomplete.[12] The identification of a double ureter may be an incidental diagnosis. The three complications most commonly associated with complete ureteral duplication are vesicoureteral reflux, ectopic ureterocele, and ectopic ureteral insertion. Vesicoureteral reflux is the most common cause of acquired kidney disease in patients with ureteral duplication, with a prevalence of 22 % for incomplete ureteral duplication and 69 % for complete duplication. Typically, reflux occurs in the lower pole ureter through a ureteral orifice with lateral ectopia and a short submucosal tunnel. The upper ureteral meatus is located more caudally and medially (Weigert-Meyer's law), has a longer submucosal tunnel, and therefore, reflux is less in this orifice. Reflux to the upper pole may occur if the ureteral orifice is located in the bladder neck or urethra, since at this level, outside the trigone, there is no submucosal tunnel to prevent retrograde reflux. The lower ureter is usually refluxing and the upper ureter is obstructed by a ureterocele.[13]

Ureterocele is a congenital malformation characterized by cystic dilatation of the intravesical segment of the distal ureter. It is classified as orthotopic or simple when it has a normal anatomical position and ectopic when it is located elsewhere. It has an estimated incidence of 1/4000 children (female-to-male ratio, 4-6:1). Ultrasound: intravesical thin-walled anechoic cyst. Color Doppler ureteral flow helps rule out complete obstruction of the ureterocele. Because ureterocele is dynamic in nature and can change shape and size, ultrasound offers greater diagnostic accuracy thanks to the possibility of obtaining continuous images.[14] There are different imaging modalities that facilitate the diagnosis and follow-up of these conditions. The most commonly used in our setting are M-mode and D-mode (Doppler) ultrasound and radiographic techniques such as descending urography or computed axial tomography. Prenatal ultrasound (from week 20) has enabled early diagnosis and prognostic assessment. Other imaging tests include voiding cystography (if vesicoureteral reflux or VUR is suspected) and nuclear medicine tests: renal scintigraphy with dimercaptosuccinic acid (DMSA) Tc 99m (gold standard for the diagnosis of kidney damage) and MAG3 Tc 99m diuretic renogram (if obstruction is suspected). Kidney survival varies considerably depending on the cause and the presence of risk factors for progression of kidney damage.[15]

That is why, although some congenital anomalies of the urinary tract are only incidental findings with no clinical relevance, others are a risk factor for the development of complications such as hypertension, cardiovascular disease, or end-stage renal disease. Early diagnosis remains a goal for radiologists, as does knowledge of their clinical features and repercussions, especially in children.

CONCLUSIONS

Congenital malformations of the renoureteral tract represent an important public health problem due to their clinical and biopsychosocial impact. When identified in the prenatal stage, they not only condition a variable prognosis in terms of fetal and infant mortality, but also generate a significant emotional burden for the family, affecting its psychological and social dynamics. These anomalies, which can manifest with varying degrees of severity throughout life, increase morbidity in childhood and later stages if not addressed in a timely manner. Therefore, early prenatal diagnosis, based on appropriate imaging studies and a multidisciplinary evaluation, is essential to establish timely medical or surgical management that optimizes survival and preserves the patient's quality of life.

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CONSENT

The patient's consent was obtained for the performance of this work.

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CONFLICT OF INTEREST

The authors declare that there is no conflict of interest.

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