Spontaneous rupture in a renal and pyelic fusion with a single system associated with obstructive megaureter secondary to ureterovesical junction stricture


**Abstract**

Congenital anomalies of the urinary tract are diverse and they include the total absence of the kidney, its location, orientation, or aberrant shape, as well as collecting system and blood irrigation alterations.

We present the case of a 2-month old male infant with symptoms of progressive abdominal distension and fever secondary to spontaneous kidney rupture. A renal and pyelic fusion with a single system associated with ureterovesical junction stricture was identified. Division was carried out by means of nephrostomy, after which Politano ureteral reimplantation and Hendren ureteral refashioning were performed. We also present a literature review.

In 1938 Wilmer first classified anomalies presenting with renal fusion, and then in 1957 McDonald and McClellan improved and broadened that classification to include crossed ectopia with fusion, crossed ectopia without fusion, solitary crossed ectopia, and bilaterally crossed ectopia. Anomalies with fusion can be 1) unilateral fused kidney with

**Resumen**

Las anomalías congénitas del tracto urinario son diversas, estas involucran la ausencia total del riñón, la ubicación, orientación o forma aberrante de éste, así como alteraciones del sistema colector y de la irrigación sanguínea.

Presentamos el caso de un masculino de dos meses de edad, quien debutó con un cuadro de distensión abdominal progresiva y fiebre, secundario a una ruptura renal espontánea. Se evidenció una fusión renal y piélica, con un sistema único asociado a estenosis de la unión ureterovesical (UV). Se realizó derivación mediante nefrostomía, posteriormente reimplante ureteral tipo Politano y remodelación ureteral tipo Hendren.

Wilmer en 1938 clasificó las anomalías con fusión renal por primera vez. Posteriormente, McDonald y McClellan en 1957, mejoraron y ampliaron esa clasificación e incluyeron la ectopia cruzada con fusión, la ectopia cruzada sin fusión, la ectopia cruzada solitaria y la ectopia cruzada bilateral. Las anomalías con fusión pueden ser: 1) riñón fusionado unilateral con ectopia inferior, 2) riñón sigmoide o con forma de S, 3) riñón en bulbo o
INTRODUCTION

Congenital anomalies of the upper urinary tract include a great variety of abnormalities, ranging from complete absence of the kidney, to its location, orientation, and shape. Congenital obstructive nephropathy (CON) is the most common cause of chronic nephropathy in children. It is among the three principal causes of end-stage renal disease (ESRD) and is classified together with dysplasia, aplasia, and other abnormalities of the urinary system, such as a heterogeneous entity called congenital abnormalities of the kidney and urinary tract (CAKUT). Relatively common, CAKUT affects more than 2% of pregnancies and is responsible for 51% of chronic renal disease in North America. Among the diagnostic varieties of CAKUT, obstructive disease has the greatest risk for developing ESRD.1

CASE PRESENTATION

The patient is a 2-month-old male with no important past medical history. Illness began with progressive abdominal bloating 3 weeks prior to admittance, associated with fever and progressive reduction in urinary volume until reaching acute renal failure and then anuria. Physical examination revealed great abdominal distension and plain radiograph of the abdomen showed a generalized radio-opaque image (Figure 1).

Upon patient admittance, laboratory studies reported serum creatinine 6.22 mg/dL, urea 141 mg/dL, blood urea nitrogen (BUN) 66 mg/dL, leukocytes 26.2, and hemoglobin 7.44 g/dL. Abdominal ultrasound and computed axial tomography scan identified renal fusion with renal cavity ectasia and loss of renal parenchymal continuity, along with a large quantity of free perirenal fluid, suggestive of urinoma (Figure 2).

As a result of the tomography findings, cystoscopy was carried out that showed a single left ureteral meatus. Catheterization for carrying out retrograde pyelography was not possible and so surgical exploration by lumbotomy was performed; the urinoma was drained and the nephrostomy was placed. Anterograde pyelography revealed renal and pyelic fusion with a single ureter associated with obstructive megaureter secondary to ureterovesical junction stricture (Figure 3). In a second surgical stage, Hendren ureteral refashioning with Politano-Leadbetter reimplantation was performed. The patient progressed satisfactorily and passage of contrast material to the bladder was corroborated by control anterograde pyelography (Figure 4). The nephrostomy catheter was removed definitively and at present the patient’s renal function is conserved.

DISCUSSION

Ureteropelvic junction (UPJ) stricture and ureterovesical junction (UVJ) stricture are upper urinary tract congenital obstructions, along with ureterocele and other abnormalities of ureteral structure and location. Obstruction of the UPJ presents in one out of every 1,000-2,000 births and is bilateral in 20-25% of cases. It is predominant in males, with a 3:1 male/female ratio. Intrinsic obstruction of the UPJ arises in a proximal segment of the adynamic ureter. This segment of the dysfunctional ureter often
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Renal fusion abnormalities are divided into crossed-fused renal ectopia (CFE) and horseshoe kidney. The frequency of CFE is 1 in 7500 autopsies and is less frequent than horseshoe kidney, which has a reported frequency of 1 in every 400. More than half of the patients with CFE have other urogenital, gastrointestinal, cardiovascular, skeletal, or chromosomal abnormalities.²

Crossed ectopia is referred to when a kidney is located on the side opposite that of the insertion of its ureter in the bladder. Ninety percent of crossed ectopic kidneys are fused with the kidney of the same side and thus represent the majority of fusion defects. Wilmer (1938) first classified renal fusion abnormalities and then McDonald and McClellan (1957) improved upon and broadened that classification to include crossed ectopia with fusion, crossed ectopia without fusion, solitary crossed ectopia, and bilaterally crossed ectopia (Figure 5). Abnormalities with fusion can be 1) unilateral fused kidney with inferior ectopia, 2) sigmoid or S-shaped kidney, 3) lump or “cake” kidney, 4) L-shaped or tandem kidney 5) disc or shield kidney, and 6) unilateral fused kidney with superior ectopia³ (Figure 6). Despite the fact that in the present case the conformation of both kidneys could be considered to correspond to L-shaped kidney, we believe it does not fall into any of the abovementioned classifications due to the fact that there is a single excretory system associated with obstructive megaureter secondary to UVJ stricture. Only 5 cases of renal and pyelic fusion with a single ureter have been reported in the literature. Up to now, the present case is the only reported case associated with UVJ stricture and spontaneous kidney rupture in a pediatric patient.

In the past, diagnosis was made through excretory urography, but today ultrasound and tomography have

Figure 1. Plain radiograph of the abdomen with radio-opacity of the entire abdominal region.

Figure 2. A) Renal ultrasound showing cavity dilation. B) CAT axial view showing loss of the renal parenchymal continuity and free perirenal fluid. C) CAT coronal view showing renal fusion with a large quantity of free fluid.
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identified a greater number of asymptomatic cases. Cystoscopy and retrograde pyelography are useful for evaluating the collecting system and draining pattern. Computed axial tomography and magnetic resonance provide high quality images through which the majority of renal fusion abnormalities can be classified. With the advent of prenatal ultrasound, it is possible to suspect the presence of a congenital urinary tract abnormality and attempt to avoid the development of chronic nephropathy by carrying out opportune management of the different pathologies.

**CONCLUSIONS**

Spontaneous urinoma is rare in the pediatric population, given that most cases occur in adults and are associated with trauma. However, when such a case is detected in the pediatric patient, it is necessary to suspect obstructive congenital pathologies (strictures of the ureteropelvic junction, the ureterovesical junction, the urethral valves, among others) that can cause severe renal cavity
Spontaneous rupture in a renal and pyelic fusion with a single system associated with obstructive megaureter secondary to ureterovesical junction stricture. The association with renal fusion abnormalities is uncommon, but the possibility of their relation to these obstructive pathologies should always be considered. The present case clearly illustrates the sequence of clinical events that are a repercussion of the association with multiple congenital urinary tract abnormalities.5–10

REFERENCES