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CASE REPORT

Renal pelvis rupture in a kidney with ureteropelvic junction obstruction and extrarenal calyces

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Abstract The extrarenal calyx (ERC) is a rare congenital anomaly, associated with various other abnormalities of the urogenital system. We report a unique case of ERC in a solitary functioning kidney with a massively dilated pelvis that developed spontaneous rupture. A dismembered pyeloplasty was performed at the time of rupture. The patient did well post-operatively with a stable creatinine and stable SFU grade 2 hydronephrosis.

Introduction

First reported in 1925 by Eisendrath, extrarenal calyces (ERC) have since been described in less than 30 case reports [1]. In 1961, Malament characterized the anomaly as one in which the major calyces and renal pelvis lie outside the renal parenchyma [2]. In most case reports, ERC has been described in association with other renal anomalies including bifid kidney [1], horseshoe kidney [3], ectopic kidney [4], and ureteropelvic junction obstruction (UPJO) [5,6]. We report a case of spontaneous pelvis rupture of a massively hydronephrotic solitary functioning kidney with UPJO and ERC. To our best knowledge, this is the first such report in the literature.

Case

The female described herein first came to urologic attention at 32-weeks gestational-age when prenatal ultrasound (US) demonstrated cystic structures in both kidneys. She was born at 38 weeks by normal spontaneous vaginal delivery, with a weight of 3.3 kg and normal physical exam. Post-natal US showed a multicystic dysplastic kidney (MCDK) on the

Abbreviations: ERC, extrarenal calyx; MCDK, multicystic dysplastic kidney; UPJO, ureteropelvic junction obstruction.

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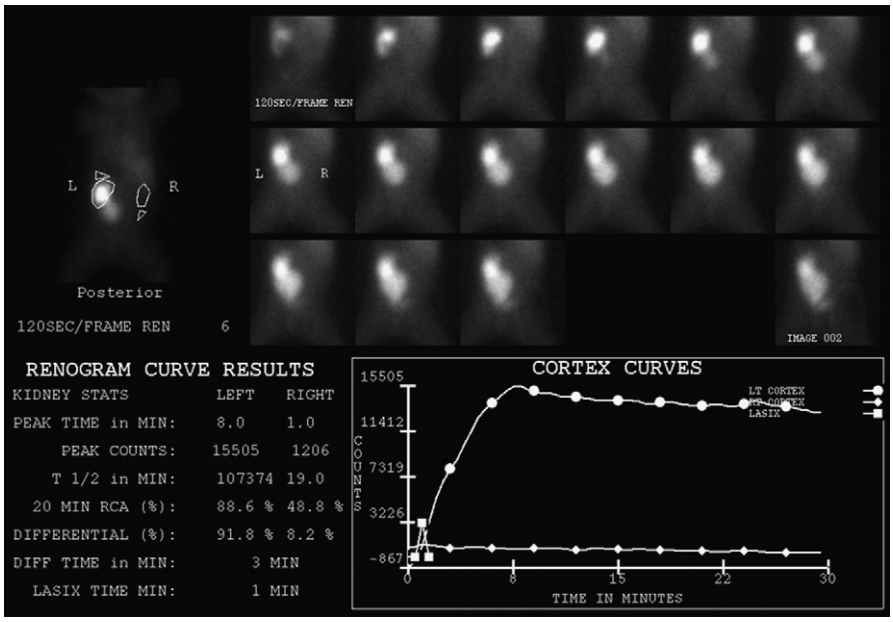

Figure 1 MAG-3 scan (Lasix at 1 min) at 7 weeks of age showing no function on the right (MCDK) and left hydronephrosis with excretion into a large pelvis.

right and left hydroureteronephrosis down to the bladder, with severe pelviectasis but relatively preserved parenchyma. Voiding cystourethrogram was normal with no reflux. She was placed on prophylactic antibiotics and demonstrated normal feeding, weight gain, and urine output over the next several weeks. Her creatinine level came down appropriately to 0.4 mg/dl during this time. At one month of age she presented to the emergency department with abdominal distension. Her abdominal exam prompted a CT scan, which demonstrated findings consistent with the postnatal US, as described above. MAG-3 Lasix renal scan performed at 7 weeks of age showed no function in the right kidney, and cortical excretion of material into a severely dilated pelvis and ureter on the left (Fig. 1). She did clinically well over the next four months, although she did have a single urinary tract infection that responded promptly to antibiotics. During this period she had further imaging studies, including MRI and repeat US, which were consistent with prior findings with stable dilation of the left renal pelvis and ureter and continued preservation of parenchyma. The presumptive diagnosis at that time was UPJ or UVJ obstruction. At 6 months of age surgical options were discussed with the family. Because the renal scan had been equivocal and the level of obstruction was still uncertain, a decision was made to perform cystoscopy with retrograde pyelogram and possible pyeloplasty or reimplant depending

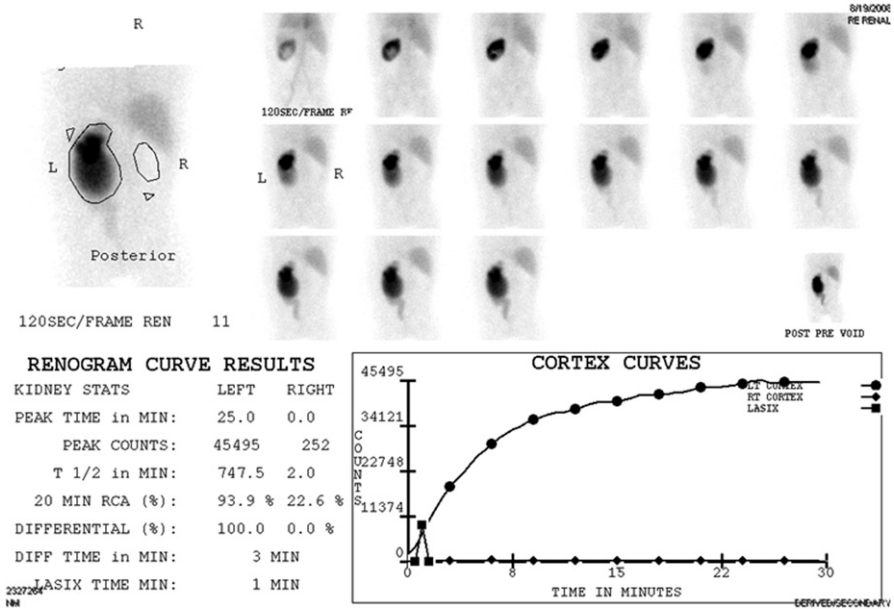

Figure 2 MAG-3 scan at 18 months showing prompt cortical excretion into a large extrarenal pelvis on the left.