Rare Case of Atrophic Ectopic Kidney With Giant Hydronephrosis in a 7-Year-Old Girl

Ming-Yu Hsieh, Min-Sho Ku, Teng-Fu Tsao, Shan-Ming Chen, Yu-Hua Chao, Jeng-Dau Tsai, Ko-Huang Lue, and Ji-Nan Sheu

Ectopic kidney is a rare condition. Giant hydronephrosis is also an uncommon lesion in children and is usually due to ureteropelvic junction obstruction. We report a case of 7-year-old girl presenting with abdominal fullness. The imaging characteristics of magnetic resonance urography of the lesions are reported. The findings from the radiologic investigations were suggestive of a dysplastic ectopic pelvic kidney with giant hydronephrosis. Subsequent surgery confirmed the diagnosis. An ectopic kidney with giant hydronehrosis is an extremely rare condition in children and can present as an asymptomatic abdominal mass. A careful survey for other structural anomalies is mandatory in such cases. UROLOGY 81: 655–658, 2013. © 2013 Elsevier Inc.

An ectopic kidney is a rare condition, with a low clinical incidence of 1 in 5000 children.[1,2] Giant hydronephrosis is also an uncommon lesion that is usually due to ureteropelvic junction obstruction.[3,4] The term has been used to describe a renal pelvis with massive dilation containing >1 L of fluid in adults or the equivalent of 1 day's urine output in children or, alternatively, 2%-4% of the body weight in fluid.[3,4] We report a rare case of ectopic pelvic kidney with a giant hydronephrosis presenting as a huge asymptomatic abdominal mass in a 7-year-old girl. This is the first reported case in a child in which magnetic resonance urography (MRU) was used as a diagnostic modality.[5-8]

CASE REPORT

A 7-year-old girl was brought to the out-patient department because of abdominal fullness that had been present for several months. A large abdominal mass across the midline was palpable, and she was subsequently admitted for additional assessment. Her personal and urologic histories were unremarkable.

On physical examination, the patient's vital signs were normal. Her abdomen was markedly distended, and a large, nontender, palpable mass was noted on the left side, extending across the midline. Her external genitalia were normal. Her urinalysis findings were normal, and the serum urea nitrogen and creatinine was 12.1 mg/dL and 0.3 mg/dL, respectively. Abdominal ultrasonography revealed a large cystic mass localized in the left abdomen and extending across the midline and the right kidney in the normal position with mild hydronephrosis. The left renal fossa and pelvis contained a large cystic structure but no discernible kidney.

For additional investigation, abdominal magnetic resonance imaging with a 1.5-T commercial scanner (Signa Horizon Echospeed, General Electric Medical Systems, Milwaukee, WI) was performed. Morphologic evaluation of the urinary tract using MRU and T_1- and T_2-weighted magnetic resonance imaging demonstrated a very large, lobulated cystic lesion in the abdomen, probably arising from the retroperitoneum (Fig. 1A). The right kidney showed mild dilation of the pelvis but not of the calices (Fig. 1A), and the left renal fossa was filled with a cystic lesion instead of any recognizable renal tissue. The right ureter and bladder were normal, but the left ureter was not visible (Fig. 1B). Instead, a small ovoid structure in the left part of the pelvis was noted, suggesting a small, atrophic kidney (Fig. 1B). Technetium-99m dimercaptosuccinic acid (DMSA) scan revealed an area of increased DMSA uptake in the left pelvis adjacent to the bladder, which was suspected to be a dysplastic ectopic pelvic kidney (Fig. 1C). On the basis of these findings, the diagnosis of a small ectopic pelvic kidney associated with giant hydronephrosis was made. The voiding cystourethrography findings were normal.

A temporary percutaneous nephrostomy tube was placed to assist with drainage of the kidney and allow assessment of renal function before proceeding with surgery. Approximately 2000 mL of clear yellow urine was subsequently drained; <40 mL of urine output was drained during a 4-week period of nephrostomy. The urinalysis findings revealed isosthenuria, proteinuria, hematuria, and waxy casts.

Cystoscopy was performed at surgery and revealed a normal bladder and the right ureteral orifice at the

Ming-Yu Hsieh and Min-Sho Ku contributed equally to this work.

Financial Disclosure: *The authors declare that they have no relevant financial interests.*

From the Departments of Pediatric Surgery, Pediatrics, and Medical Imaging, Chung Shan Medical University Hospital, Taichung, Taiwan; and Institute of Medicine, and Department of Pediatrics, School of Medicine, Chung Shan Medical University, Taichung, Taiwan

Reprint requests: Ji-Nan Sheu, M.D., Department of Pediatrics, Chung Shan Medical University Hospital, No. 110, Section 1, Jianguo North Road, Taichung 402, Taiwan. E-mail: cshy098@csh.org.tw

Submitted: October 28, 2012, accepted (with revisions): November 26, 2012

0090-4295/12/$36.00

http://dx.doi.org/10.1016/j.urology.2012.11.036

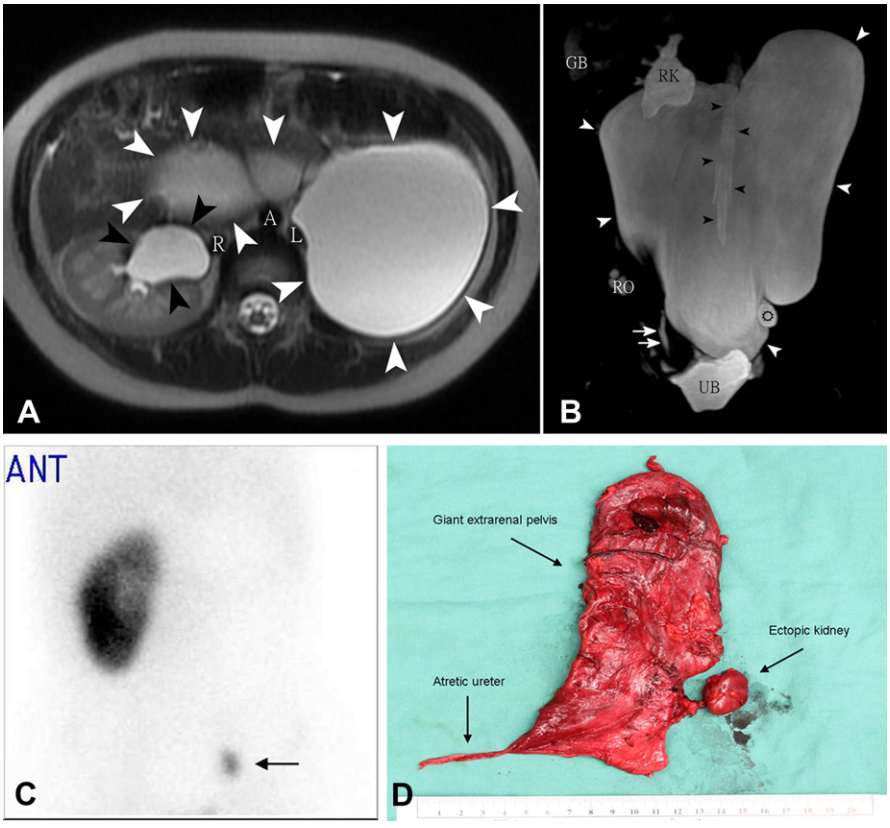

Figure 1. (A) Axial T_2-weighted magnetic resonance imaging of small ectopic kidney and giant extrarenal pelvis using half-Fourier acquisition single-shot turbo spin-echo sequence (repetition time 3231 ms, excitation time 91 ms) through right renal hilum showing lobulated cystic lesion (white arrowheads) in left abdomen and extending across midline, with dilated renal pelvis (black arrowheads) and normal calices of right kidney but nonvisible left kidney. A, aorta; L, left inferior vena cava; R, right inferior vena cava. **(B)** Coronal maximum-intensity projection imaging of heavily T_2-weighted MRU scan (repetition time 5000 ms, excitation time 700 ms) showing huge cystic lesion in abdomen and pelvis (white arrowheads) and other fluid-containing structures: gallbladder (GB), right kidney (RK), cerebrospinal fluid (black arrowheads), right ovary (RO), right ureter (white arrows), and urinary bladder (UB). Small ovoid structure (clear star) on left part of pelvis. **(C)** DMSA renal scan revealing area of increased uptake in left pelvis adjacent to bladder, suggesting presence of small, dysplastic ectopic kidney. **(D)** Grossly, specimen showed excised giant extrarenal pelvis due to ureteropelvic junction obstruction in dysplastic ectopic kidney with short calcified atretic ureter. (Color version available online.)

normal position but an absent left ureteral orifice. Surgery was subsequently performed through a left lower quadrant incision of the abdomen. Additional exploration revealed a giant extrarenal pelvis (22 × 10 cm) connected to the ectopic kidney and a short calcified atretic ureter (5 × 0.3 cm; Fig. 1D). The blind-ended atretic ureter did not drain into the bladder, consistent with the cystoscopy findings. The kidney was small and atrophic (2.5 × 1.6 cm). Therefore, we elected to perform nephrectomy rather than pyeloplasty. The patient's postoperative course was uneventful.

COMMENT

Failure of the embryonic kidney to reach the usual position results in an ectopic kidney. Factors that can prevent the orderly movement of the kidneys include ureteral bud maldevelopment, defective metanephric tissue, genetic abnormalities, maternal illness, teratogenic causes, or an anomalous vasculature acting as a barrier to ascent.[5,9] The chief characteristics of an ectopic kidney, which is often small and tends to lie obliquely, are an abnormally short ureter and an aberrant blood supply. Most pelvic kidneys are clinically asymptomatic and are incidental findings during physical or radiologic investigations in the hospital for other medical complaints. Ectopic kidneys are more susceptible to disease, including hydronephrosis, obstruction, and lithiasis, than normally positioned kidneys.[1,6] Although the overall incidence of hydronephrosis was 0.2% in a general screening study,[2] a greater incidence (33%-56%) was reported in children with an ectopic kidney.[1,10]

Ultrasonography is typically the first imaging modality used to assess for urinary tract abnormalities. However, ultrasound diagnosis of a solitary kidney can miss the presence of a small, atrophic renal unit. In the present case, abdominal and pelvic ultrasonography revealed an empty left renal fossa and did not show the very small,