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Unfused renal ectopia: a rare form of congenital renal anomaly

Gül Nihal NURSAL and Gülgün BÜYÜKDERELİ

Department of Nuclear Medicine, Çukurova University Faculty of Medicine, Adana, Turkey

Unfused crossed renal ectopia observed 1 in 75,000 autopsies is a rare congenital anomaly. Typically one kidney is located in the proximity of the other kidney, and the ureter of the anatomically anomalous kidney crosses the midline to insert to the bladder in its normal anatomic position. Although renal function is usually not affected, the condition is generally accompanied by other congenital anomalies. In this case report, static and dynamic scintigraphic images of two patients with unfused crossed renal ectopia are presented. Besides properties of imaging modalities, clinical features are discussed in light of the available literature.

Key words: unfused crossed renal ectopia, renal scintigraphy, congenital anomaly

INTRODUCTION

ANATOMIC VARIANTS of the kidneys are often noted during renal scintigraphy and other imaging studies. These conditions may have a variable effect on overall renal function and can cause confusion, both clinically and on initial imaging.[1]

Crossed renal ectopia (CRE), which was first described by Pannorlus in 1964, is a rare congenital anomaly consisting of transposition of a kidney to the side opposite its normal position. The associated ureter crosses the midline to insert in its normal position in the bladder.[2,3]

In this report we present two patients with unfused crossed renal ectopia. Renal scintigraphic imaging methods (static and dynamic studies) and their advantages over each other were discussed in the context of the available literature.

CASES

Case 1: A 4-year-old boy who had hydrocephalus and meningomyelocele was treated in the neonatal period. He had additional medical problems namely neurogenic bladder, severe thoracolumbal scoliosis and pes equinovarus deformity at right foot.

He was being assessed for recurrent urinary tract infections and was referred to us for dynamic and static renal scintigraphy. After the injection of 74 MBq (2 mCi) technetium-99m ethylenedicysteine (Tc-99m EC), dynamic images were obtained with the patient in the supine position, posteriorly. Immediate and delayed images showed normally localized left kidney and a horizontally placed ectopic right kidney situated posterior to the left renal pelvis. Left ureter was shown to be inserting into the bladder normally, but right ureter inserted into the superior wall of the bladder. Both ureters were visualized on the 7-minute image (Fig. 1a). Although a minimal radiopharmaceutic stasis was demonstrated in both kidneys, this activity disappeared after diuretic administration.

Following injection of 185 MBq (5 mCi) Tc-99m dimercaptosuccinic acid (Tc-99m DMSA), planar imaging was performed 3 hours later to identify the nature of the abnormality. The study was performed with the patient in the supine position. Images of 400 kcounts per view were obtained in the posterior, anterior, left posterior oblique and right posterior oblique positions in a matrix of 256 × 256. It was shown that the malrotated right kidney was located posterior to the left kidney (Fig. 1b). No cortical scarring was seen on the right kidney, and a relative hypoactive area was observed on the lateral aspect of the left kidney.

Ultrasonography was done and two separate kidneys

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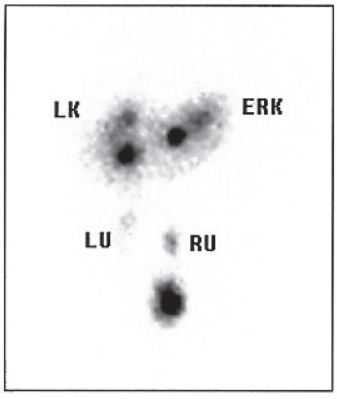
For reprint contact: Gül Nihal Nursal, M.D., Çukurova Universitesi, Nukleer Tıp Anabilim Dali, 01330, Adana, TURKEY.

E-mail: gnnursal@yahoo.com



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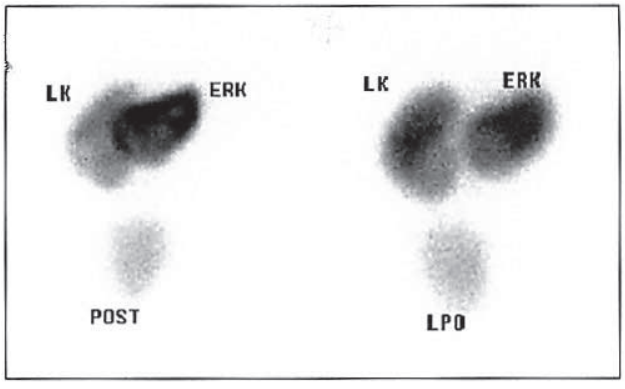

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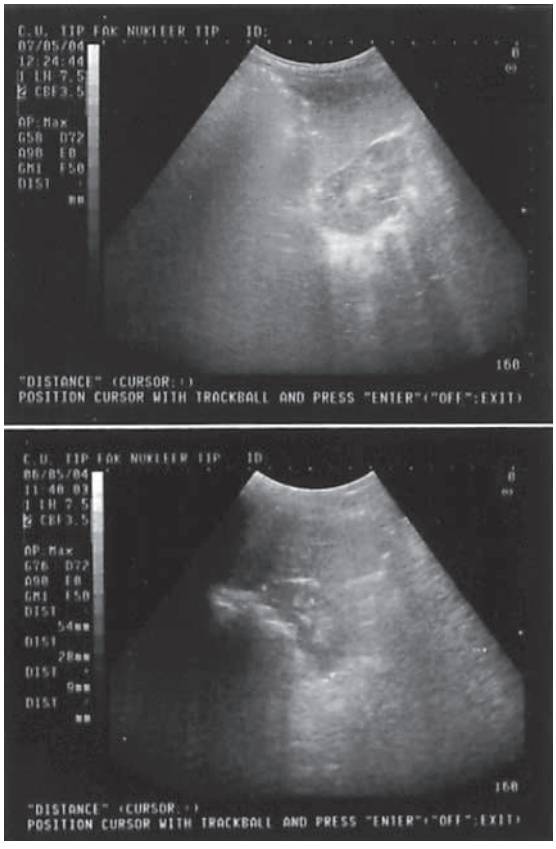

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Fig. 1 a: Tc-99m EC renogram obtained from posterior view. Dynamic images demonstrate normally localized left kidney and ectopic right kidney, which is placed posterior to the left renal pelvis. Both ureters are clearly visible on 7-minute image (LK: left kidney, ERK: ectopic right kidney, LU: left ureter, RU: right ureter). b: Tc-99m DMSA scintigraphy shows normally situated left kidney and crossed ectopic right kidney on the left renal fossa (POST: posterior, LPO: left posterior oblique, LK: left kidney, ERK: ectopic right kidney). c: Renal ultrasound images of the left kidney (*upper*) and the ectopic right kidney (*lower*).

with normal collecting systems were demonstrated on the left side (Fig. 1c).

Case 2: A 10-year-old boy with a known history of crossed renal ectopia was referred to us for evaluation of renal functions. The condition has been diagnosed by ultrasonography in the newborn period when he was treated for anal atresia.

Tc-99m diethylenetriaminepentaacetic acid (Tc-99m DTPA) dynamic and Tc-99m DMSA static imaging was performed in the same manner as for patients 1. Tc-99m DTPA imaging showed the presence of crossed renal ectopia with well functioning kidneys and two ureters on the 8-minute image. Left ureter was crossing midline before inserting to the bladder in its normal location. The right ureter was normally placed (Fig. 2a).

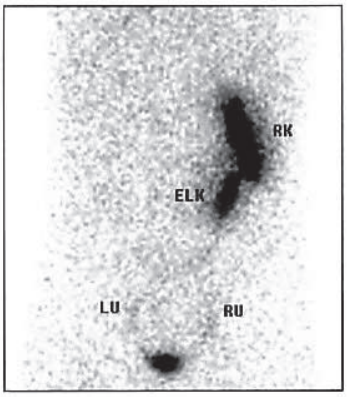
Tc-99m DMSA images showed that there was no kidney on the left and ectopic left kidney was located nearby to the anterior lower pole of the right kidney (Fig. 2b). Radiopharmaceutical uptake of the two kidneys was normal.

Ultrasonography showed two unfused kidneys with normal collecting systems on the right renal fossa (Fig. 2c).

DISCUSSION

Embryologic development of CRE has not been clearly determined but many theories have been offered to explain this congenital anomaly. It is deduced that mechanical factors are of primary importance in ectopia without fusion.[3] Being more frequent in males (M/F = 1.4/1), crossed renal ectopia is two to three times more common on the right than on the left. Furthermore unfused forms were noted to be usually on the right. The condition is generally diagnosed in the third decade.[4]

The overall autopsy incidence for crossed renal ectopia is reported as 1:2000 to 1:7000.[5] The incidence of unfused crossed renal ectopia however has been reported to be 1 in



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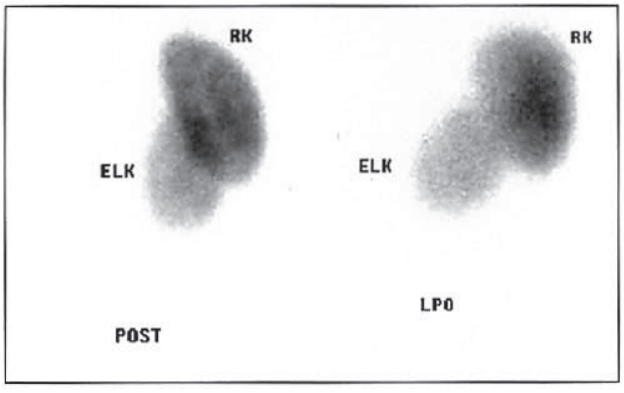

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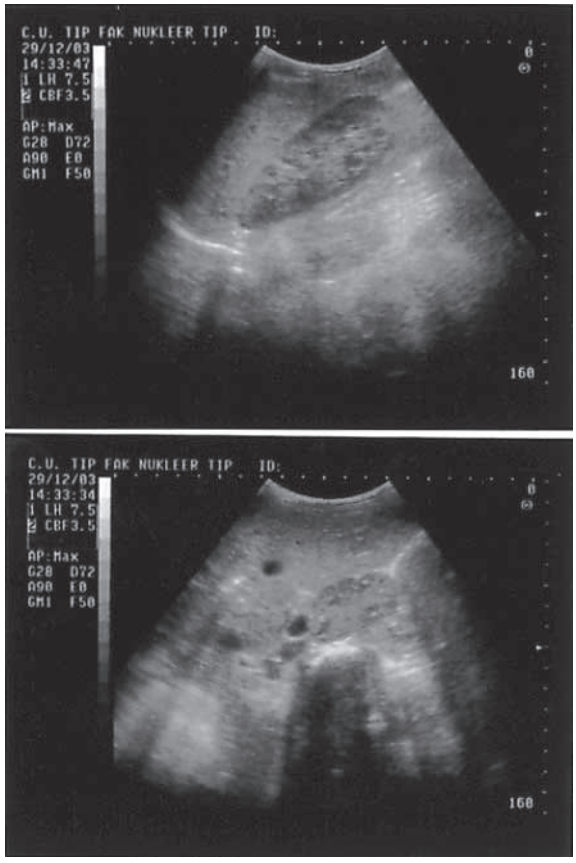

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Fig. 2 a: Tc-99m DTPA renogram. Dynamic images at 8 minutes show normally functioning right kidney and ectopic left kidney on the right renal fossa. Left ureter crosses the midline and both ureters insert into bladder at their respective normal location (RK: right kidney, ELK: ectopic left kidney, LU: left ureter, RU: right ureter). b: Tc-99m DMSA scintigraphy demonstrating right kidney in normal anatomic position and crossed ectopic left kidney (POST: posterior, LPO: left posterior oblique, RK: right kidney, ELK: ectopic left kidney). c: Renal ultrasound images of the right kidney (*upper*) and the ectopic left kidney (*lower*).

75,000 autopsies, an incidence ten times lower than that of fused crossed renal ectopia.[3]

Crossed renal ectopia can be anatomically classified into four groups; 1) CRE with fusion (the majority of cases: 90%), 2) CRE without fusion (uncommon), 3) solitary CRE (very rare) and 4) unfused bilaterally CRE (also very rare).[6] In the first instances, the ectopic kidney is usually located inferiorly to the orthotopic kidney. Malrotation of the crossed ectopic kidney is the rule. Although the kidneys are reported to be usually vertically oriented, in our first case the ectopic kidney was in a perpendicular position relative to the other kidney.[4]

Crossed renal ectopia initially can be difficult to diagnose since the common symptoms such as abdominal or flank pain, palpable mass, hematuria, urinary tract infections, and dysuria are nonspecific. Therefore, incidentally diagnosed patients constitute 20 to 30% of the cases.[5] Urinary tract diseases such as vesicoureteral reflux, urinary infections, ureteroceles, renal calculi, and renovascular hypertension can co-exist with ectopic kidneys, which are likely to be complicated by ureteropelvic junction obstruction because of their frequent abnormal shape, melrotation and aberrant vasculature.[4] Additional urinary tract findings include megaureter, hypospadias, cryptorchism, urethral valves, and cystic dysplasia.

Other congenital anomalies may accompany CRE such as unilateral agenesis of fallopian tubes and ovaries, skeletal abnormalities (radial clubhand, hemivertebrae, spina bifida, scoliosis, and congenital hip dislocation), gastrointestinal abnormalities (imperforate anus and esophageal atresia with tracheoesophageal fistula), and cardiopulmonary anomalies.[4] Concistent with the literature, both of our cases had additional congenital anomalies besides CRE such as scoliosis, pes equinovarus deformity and anal atresia.

Computerized tomography (CT) scan, ultrasound, renal scan and arteriography in selected cases, have all been used to better clarify CRE.[3] There are certain pitfalls to be

avoided when using ultrasound, particularly in cases in which an empty renal fossa may be filled with a loop of bowel, simulating a kidney.[1] Although CT is a superior tool to US for topographical anatomic details, ectopic kidney may be difficult to identify in scans where intravenous radio-contrast injections were not used.[1] The clinical complaints did not warrant a CT scan in our cases.

Tc-99m DMSA is taken up specifically in the tubular cells of the renal cortex and facilitates assessment of function and identification of aberrantly located kidneys.[4,7] Placing the patient in multiple positions during scintigraphy has been used to differentiate between fused and unfused forms.[3] However, confirmation of nonfusion may be truly achieved only in surgery as fibrous bands may bridge two kidneys that may not be detectable with imaging studies.[3] Besides DMSA, dynamic renal scintigraphy is also needed for diagnosis as it shows the ureter anatomy in detail. Therefore normal insertion of the ureter after crossing the midline may be visualized.[7] Renal scintigraphy is therefore even advantageous to the above-mentioned methods as it is capable of clearly demonstrating the location of functioning kidneys as well as providing information about perfusion and level of function.[1] In the instance where one of the kidneys is diseased, it will be of benefit beforehand to know that there are two functioning separate units, even if there are fibrous bands or nonfunctioning bridging tissue between these units anatomically. Therefore the necessary intervention could be specifically targeted with confidence. Otherwise, the clinician might mistakenly assume that he/she is dealing with a single unit which could delay or change the clinical strategy.

In conclusion, CRE is usually diagnosed when other disease states are being investigated. It rarely causes significant clinical problems. Treatment is only indicated for the complications of the anomaly rather than for the anomaly itself.[3]

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