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Fused pelvic kidneys: "hourglass sign" on Tc-99m DTPA diuresis renography

Ozgur KARACALIOGLU, Seyfettin ILGAN, Nuri ARSLAN, Ozdes EMER and Mehmet OZGUVEN

Department of Nuclear Medicine, Gulhane Military Medical Academy and School of Medicine, Ankara, Turkey

A 13-year-old girl with fused pelvic kidneys detected on ultrasonographic examination was referred to our department for a diuresis renography to assess urine drainage dynamics and exclude the possibility of urinary tract obstruction. Renal scan demonstrated fused pelvic kidneys functioning normally without obstruction. The authors present this case to demonstrate the diuresis renography findings of the combined position, fusion and rotation anomalies of the kidneys that is a rare congenital anomaly of the urinary tract.

Key words: pelvic kidney, renal scintigraphy, Tc-99m DTPA

INTRODUCTION

DURING NORMAL EMBRYOLOGY DEVELOPMENT, the permanent kidneys are initially positioned in the pelvis opposite the sacral somites, with their pelves facing anteriorly. Through the combination of axial trunk lengthening, elongation of the ureter, intrinsic renal growth, and rotation, the metanephros ascends to a higher position. By the 8th week of gestational life, migration has been completed, and the kidney resides in the upper retroperitoneum opposite the second lumbar vertebra with the hilum facing medially.

Bilateral congenital renal ectopia of the case described here represents a developmental arrest during renal ascent so that the kidneys occupy a pelvic location having a shorter than normal ureter and probably an abnormal vascular supply. Actually, fused pelvic kidneys are an advanced form of horseshoe kidney, because large portions of the renal parenchyma fuse. The resulting kidney loses its horseshoe shape and becomes a flattened disc or lump kidney as in our case. The fusion of the renal parenchyma prevents normal medial rotation to a varying degree, and therefore malrotation is a usual accompaniment, with the pelvis typically directed anteriorly. Although, unilateral pelvic renal ectopia is not an uncommon finding and is usually discovered during routine urologic evaluation, bilateral pelvic kidneys with or without fusion are rare. Clinically these kidneys are usually asymptomatic; however, effective renal plasma flow is often diminished and drainage may be impeded. The incidence of stone formation and infection is increased.[1] Sometimes, conventional radiological imaging methods are not helpful in the diagnosis of urinary tract obstruction in these patients, because anomalies of renal position often are associated with bizarre and sometimes puzzling alterations in morphology and function due to the interdependence of the renal ascent, rotation, and vascular supply. Since it is based on the rate of washout of the radiopharmaceutical from the collecting system in the upper urinary tract, diuresis renography is the only study that evaluates renal function and urodynamics in a single test.[2] Furthermore, dysmorphic appearance does not affect the results of this objective study.

CASE REPORT

A 13-year-old girl with fused pelvic kidneys detected by ultrasound was referred to our department for a diuresis renography to evaluate the urine drainage dynamics and the possibility of obstruction. Pelvic examination revealed a pelvic mass. Physical examination was otherwise normal. The patient had no other complications attributable to the anomaly. There was no family history of any kidney problems.

A conventional diuresis renography with an IV injection of furosemide (1 mg/kg) 25 minutes after the IV administration of 5 mCi (185 MBq) of Tc-99m DTPA was

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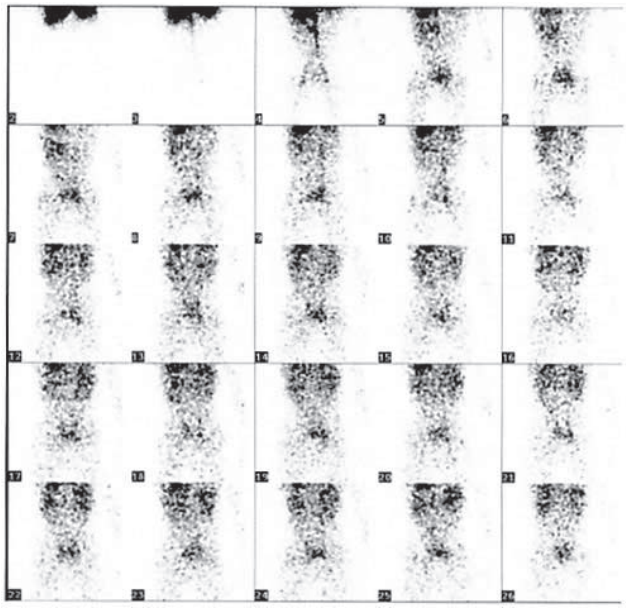
For reprint contact: Ozgur Karacalioglu, M.D., Gulhane Military Medical Academy and School of Medicine, Department of Nuclear Medicine, 06018-Etlik, Ankara, TURKEY.

E-mail: aokaracali@yahoo.com



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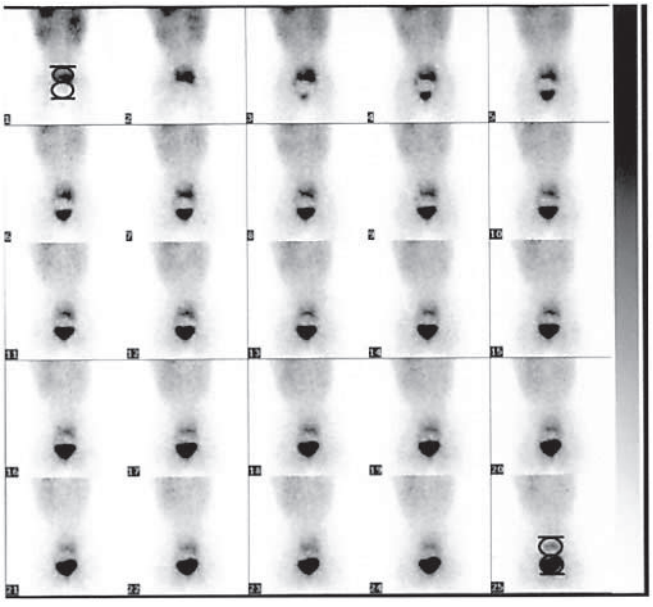

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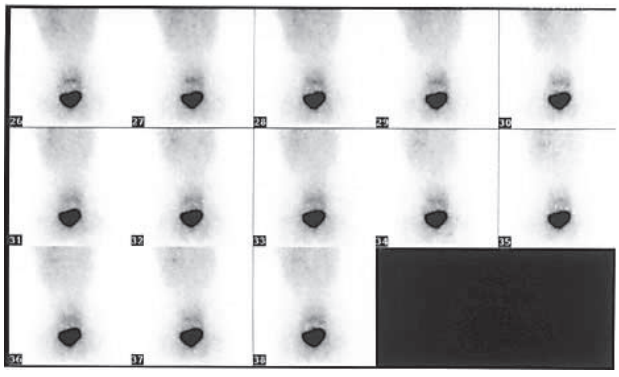
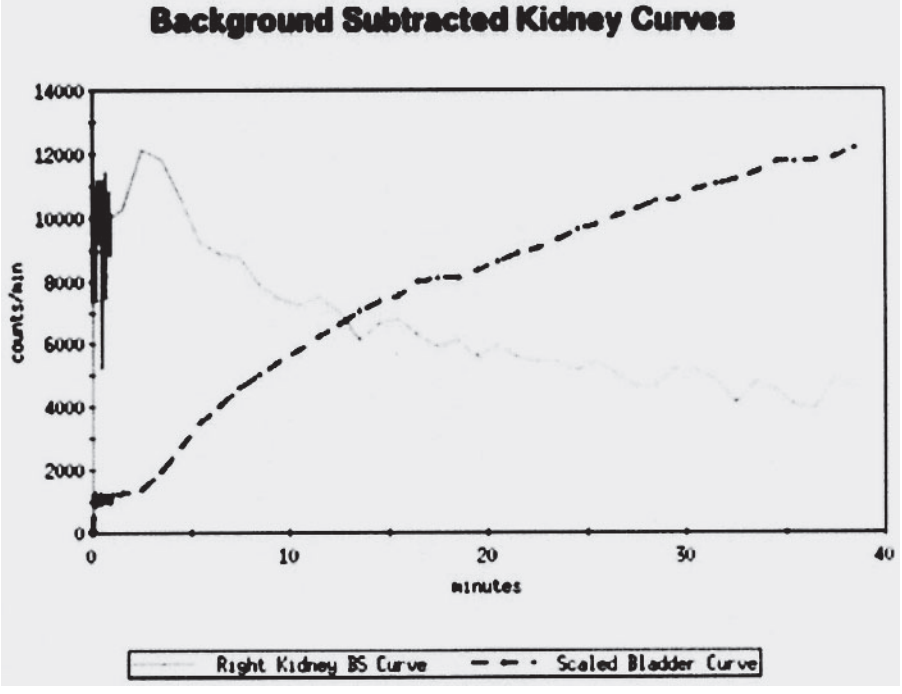


c

Fig. 1 Tc-99m DTPA diuresis renography images. Sequential 2 second images of the perfusion phase of the study revealed a mid-pelvic blush (a). When the bladder started to fill with urine and renal parenchymal concentration of the radiopharmaceutical started to decrease, an "hourglass" appearance was formed in sequential 1 min images of the concentration phase following the perfusion (b, drawing). There was no evidence of obstruction or even pelvicaliceal stasis (c). The renogram curve generated after cortical ROI was assigned around the fused moieties. Solitary renogram curve demonstrates the preserved function of both fused moieties (d).

d



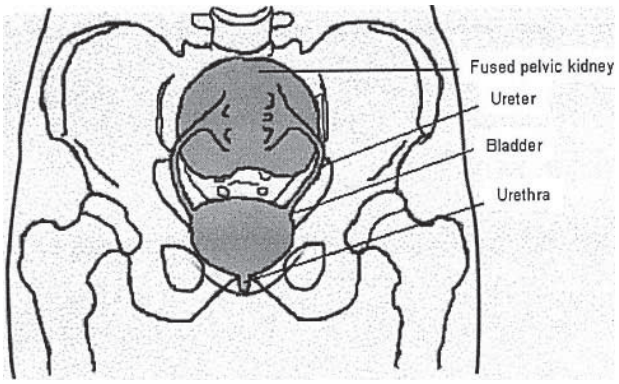


Fig. 2 Illustration of fused pelvic kidneys.

performed. The patient was instructed not to restrict her water intake before the test, and oral hydration of between 5 and 10 ml/kg body weight was given 1 hour before the tracer injection. The images were acquired from anterior projection while the patient was in the supine position. Rapid sequential digital flow images at 2 sec/frame for 1 min and functional images at 60 sec/frame for 38 min were recorded in a 64 × 64 matrix following the intravenous injection. Acquisition of images was performed with a large field of view gamma camera peaked with a 20% window at 140 keV, fitted with a low-energy, general-purpose collimator. During the perfusion phase a mid-pelvic blush was noticed (Fig. 1a). In the concentration phase an increasing activity was seen at the same location of the previously noticed pelvic blush with no activity at the normal renal anatomic sites (Fig. 1b). At the same time, the concentration phase of the dynamic renal perfusion study revealed pelvic kidneys fusing at the midline with both ureters retaining their normal insertion into the bladder after coursing laterally. An "hourglass" appearance was formed as the excreted renal activity started to fill the bladder (Fig. 1b, c). It was difficult to discriminate the fused moieties, and therefore the region of interest was drawn around the fused kidneys. Then, the renogram curve demonstrated the preserved function and also excluded obstruction (Fig. 1d). The position, fusion and rotation of this rare congenital anomaly of the urinary tract are illustrated in Figure 2.

DISCUSSION

Henot first described fused pelvic kidneys in 1830. Judd and Harrington made the first radiologic diagnosis of this condition in 1919.[3] Both the conditions of bilateral pelvic kidneys and fused pelvic kidneys appear to be very rare. Faulty ureteral bud development, abnormalities of renal vasculature limiting ascent, and teratogenic factors have been offered as explanations.[4,5] Cook and Stephens have proposed a theory that relates the position and fusion anomalies to abnormal variation in growth or flexion of the hind end of the developing embryo.[6] Another postulation is renal fusion resulting from the pressing together of the nephrogenic blastema by the umbilical arteries as the developing kidneys ascend out of the pelvis.[7] However, mechanical factors alone do not provide a comprehensive explanation when one considers that cardiovascular and skeletal abnormalities are known to be associated with renal ectopia and fusion.

Many patients with pelvic renal ectopia remain entirely asymptomatic. When symptoms are present, they typically relate to calculi, hydronephrosis, infection, or hematuria, because fusion anomalies and pelvic ectopia are prone to cause stasis of urine, pyelectasis, calicectasis, infection and stone formation. Besides, the anomalous position of the kidneys, pelvis, and ureter predisposes to poor drainage and may result in extensive hydronephrosis.[8] The ureters are also short in fused pelvic kidneys and have a tangential course, which increases the risk of ureteric obstruction, stone formation, and infection.[9] As a result, pelvic kidneys are more susceptible to calculus formation than normal ones and are also more frequently observed to be hydronephrotic, the cause of which may be true obstruction or nonobstructive dilatation due to vesicoureteral reflux, dysmorphism, malrotation, and so on.[10–12]

Malrotation is often accompanied by dysmorphism of the pelvis and calyces, especially in the presence of renal ectopy and fusion. In other words, the malrotated kidney and pelvis may look hydronephrotic or deformed or may have a bizarre shape that suggests extrinsic compression or the presence of tumor, but in most instances these appearances are illusory. Diagnostic difficulty may be further compounded by poor renal function. In this respect, sometimes special diagnostic studies may be needed to exclude obstruction and to establish a correct diagnosis. Because kidneys are located within the confines of the bony pelvis, they overly the pelvic bones and lumbosacral vertebrae, which makes their visualization on urography difficult even when renal functions are well preserved.

Although they can give correct information about the position and morphological characteristics of the kidneys, both the increased incidence of obstructive pathologies and the inherent dysmorphism in fused pelvic kidneys may cause problems in differential diagnosis by conventional rediological imaging modalities. The assessment of the patency of the urinary tract in these patients is important and diuresis renography remains the modality of choice for demonstrating the function of kidneys located in the pelvis, because diuretic renography is the only study that can evaluate renal function and urodynamics in a single test.[13] At the same time, the fused pelvic kidneys have an aberrant blood supply from the distal aorta, where atherosclerosis is common, leading to renal artery stenosis and hypertension.[9] Therefore, if hypertension is detected in these patients, captopril enhanced renal scintigraphy may be helpful in the diagnosis.

The prognosis ultimately depends on whether the

pelvic kidney is solitary and whether the contralateral kidney is normal, as reflected in the series by Down and et al. in which renal disease developed in 40% of patients with a solitary ectopic kidney (representing approximately 10% of patients with pelvic kidney).[14] Prognosis also depends on the severity of associated anomalies, which are particularly common with renal ectopy, and may coexist in up to 85% of patients.[15] The high incidence of associated abnormalities requires thorough investigation, particularly in young children found to have renal ectopy.

According to our knowledge, this report is the first reported case of fused pelvic kidneys with Tc-99m DTPA diuresis renography findings, although it has been reported with other conventional imaging modalities.[8,16–18]

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