A rare congenital fusion anomaly of the urinary tract: Pancake kidney

Ahmet Midhat Elmacı[1], Muhammet İrfan Dönmez[2]

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ABSTRACT

Pancake kidney is a rare subtype of cross fused renal ectopia. Fusion of both upper and lower poles of the kidney in pelvis results in a disc or cake shaped kidney appearence. In this report, we are presenting two cases (a 3-month-old male and a 3-year-old girl) with different presentations and their follow-up results. Usually, renal fusion anomalies do not pose a risk for deterioration of renal function. With this fact kept in mind, cases should be managed individually. Additionally, magnetic resonance imaging should be the preferred modality in which further evaluation is required since it can give both functional and anatomical detail with no radiation exposure.

Keywords: Crossed fused renal ectopia; magnetic resonance imaging; pancake kidney; pediatric urology.

[1]Department of Pediatric Nephrology, Konya Dr. Ali Kemal Belviranlı Women's Maternity and Children's Hospital, Konya, Turkey
[2]Department of Pediatric Urology, Konya Dr. Ali Kemal Belviranlı Women's Maternity and Children's Hospital, Konya, Turkey

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Corresponding Author: Muhammet İrfan Dönmez
E-mail: m_irfan83@yahoo.com

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Introduction

Crossed fused renal ectopia is a rare congenital anomaly caused by abnormal growth of ureteric bud and separation failure of metanephric tissues. Although its true incidence is unknown, autopsy series has shown an estimated incidence of 1 in 2000 with male predominance. [1] Pancake kidney is an even rare subtype of crossed fused renal ectopia. Fusion of both upper and lower poles of the kidney in pelvis results in a disc or cake shaped appearance. In this report, we are presenting two cases with different presentations and their follow-up.

Case presentations

Case 1

A 3-month-old male infant was admitted to our clinic with suspected solitary kidney in prenatal counselling. There was no history of oligohydramniosis and birth weight was within normal range (3500 g). Upon admittance, he was 5800 g (25-50%) in weight and 62 cm (50-75%) in height. His physical examination was unremarkable. Additionally, urine analysis, serum blood urea nitrogen, sodium, potassium, chloride and creatinine levels were within normal range. Ultrasonography of the urinary tract revealed empty renal fossa bilaterally while there was a pelvic kidney with uncertain boundaries. Subsequently, a Technetium-99m Dimercaptosuccinic acid (DMSA) scintigraphy was ordered and a pancake kidney was discovered on the left side of the pelvis (Figure 1). Both kidneys had normal uptake with no signs of parenchymal problems. Magnetic resonance urography (MRU) did not reveal any evidence of hydroureteronephrosis (Figure 2). There were no urinary tract infections or increase in urinary tract dilatation during 1-year follow-up.

Case 2

A 3-year-old girl was referred to our clinic after she was evaluated for vomiting and her kidneys was not found in their normal positions. Her prenatal history was unremarkable. There was no history of urinary tract infections. However, her elder sister has been diagnosed with pelvic ectopic left kidney. Physical examination was normal and her growth were all within normal age percentiles. Urinalysis, serum blood urea nitrogen, sodium, potassium, chloride and creatinine levels were within normal range. Ultrasonography showed that kidneys were located

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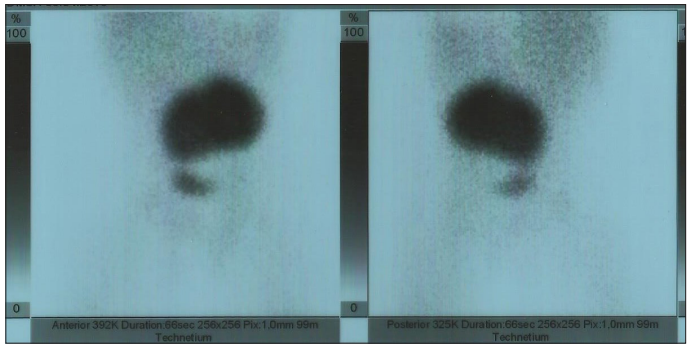

Figure 1. Dimercaptosuccinic acid scan of the first patient

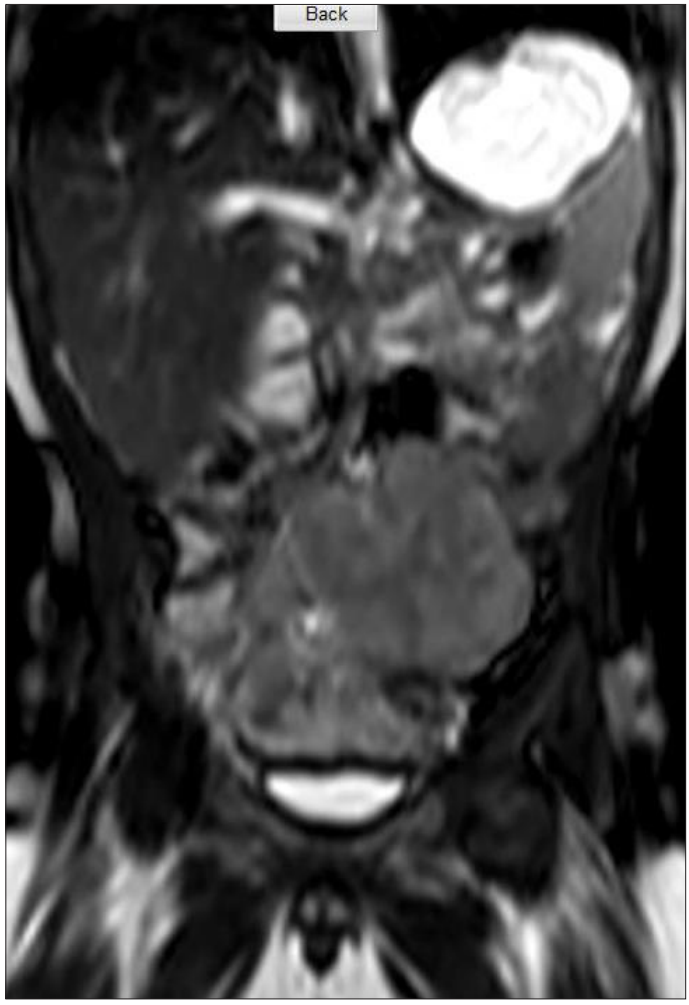

Figure 2. Magnetic resonance urography image - pancake kidney of the first patient

in pelvis with connection to each other. Finally, DMSA scintigraphy revealed two kidneys fused at midline of the pelvis. Uptakes of the both kidneys were normal (Figure 3). Patient was free of urinary tract infections during the 9 months of follow-up period.

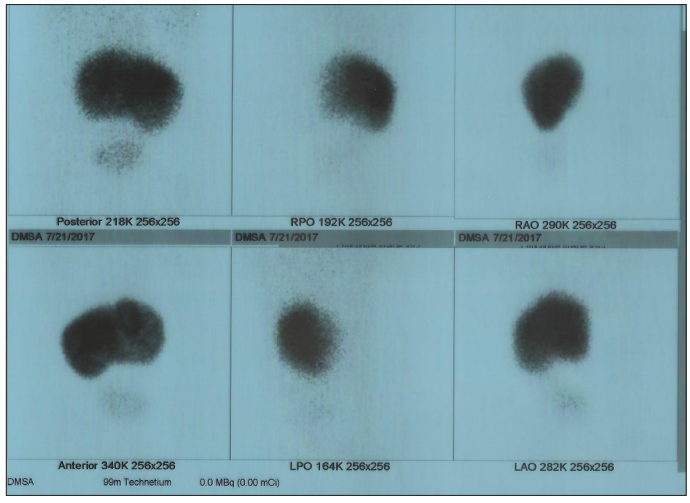

Figure 3. Dimercaptosuccinic acid scan of the second patient

Informed consents were obtained from the parents of both patients prior to writing this case report.

Discussion

Most ectopic kidneys are incidentally found.[2] There is also a rise in the detection of urinary tract anomalies in prenatal period due to increased use of antenatal ultrasonography.[3] Pancake kidney is a rare subtype of crossed fused renal ectopia. Usually, each kidney has its own collecting system and ureters. However, there are reports of single ureter draining two systems.[4] It commonly occurs on the right side, left kidney being the crossing component, however our first case was presented in the left hemipelvis.[4]

Mainly, patients with renal ectopia are asymptomatic but urinary tract infections, ureteropelvic junction obstruction, vesicoureteric reflux and renal stones may be associated with it. In our first case, an anomaly was detected prenatally. The patient without kidneys in bilateral renal fossa was counselled due to increased the risk of renal ectopia. Postnatal evaluation revealed normal renal function and DMSA scan demonstrated normal functioning kidneys.

Ultrasonography followed by renal scintigraphy is the most commonly used imaging modality to diagnose ectopic kidneys.[4,5] In adults, multiphasic abdominal computed tomography is used for the evaluation of those anomalies in terms of anatomical details.[6] On the contrary, use of computed tomography is limited in children due to harmful ionizing radiation. Alternatively, magnetic resonance urography is the preferred option.[5] In our first case, MRU was used to identify vascular and ureteric anatomy. However, on the second patient no further imaging methods were required possibly due to normal ultrasonographic

findings. Also, even the individual has no history of renal problems, vascular imaging prior to any surgeries for complete renal fusion anomalies may be beneficial.

Other urogenital or vertebral anomalies may accompany renal fusion anomalies. Ureteropelvic junction obstruction, vesicoureteral reflux, single ureter and ureterocele have been reported as associated abnormalities.[4,8] Those anomalies should be treated accordingly. Also, non-obstructing renal stones has been reported in a middle aged male with crossed fused renal ectopia.[9] Even more, multicystic dysplastic kidneys have been historically reported in fusion anomalies.[8] Therefore scintigraphic evaluation plays an important role in determining the diagnosis.

Usually, renal fusion anomalies do not pose a risk for deterioration of renal function. With this fact kept in mind, cases should be managed individually. To conclude, pancake kidney is a rare subtype of a rare congenital anomaly with a low risk of developing problems. However, magnetic resonance imaging should be the preferred modality in children when further evaluation is required.

Informed Consent: Informed consents were obtained from the parents of the both patients prior to writing this case report.

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