

Pancake Kidney, Rare Kidney Fusion Anomaly in Pediatrics

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Abstract Pancake or doughnut kidney is a very rare anatomical abnormality. It is subgroup of fusion anomaly. Most of the time, diagnosed incidentally because affected patients are asymptomatic. Here we report Pancake kidney in Toddler boy with proteinuria and developmental delay.

Keywords: pancake kidney, renal fusion anomaly, proteinuria

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1. Introduction

Pancake or doughnut kidney is a very rare developmental anomaly of kidneys. It is created when extensive medial fusion of both kidneys happened and due to this complete fusion they seem like discoid or shield shape mass. There are usually two separate ureters, entered normally into bladder. There have been some case reports and in majority of them it was diagnosed incidentally. The incidence of pancake kidney is not exactly determined. Here we presented 13months boy with proteinuria and pancake kidney.

2. Case Reports

13months old boy was brought to clinic with complaint of proteinuria, which was detected in laboratory assessments of poor weight gain. Patient is the first child of consanguineous parents. His mother had complete prenatal follow up, she had normal vaginal delivery. The child was born with good Apgar score and he had no history of admission. On physical examination his head circumference: 46cm (more than 25 percentile), body weight: 8.100 kg (less than 5 percentile) Height: 73cm (10percentile), Blood pressure: 85/65 mmHg pulse rate: 102. All other findings were normal, no edema and no organomegaly, except in his developmental assessment which showed he couldn't sit without help and just made monosyllabic babble. Laboratory exams revealed: WBC: 10700 neut:35%, Hb:10.7 MCV: 78, PLT:223000, BUN:24 ,Cr:0.4 and in urine analysis SG:1005, Prot:2+ Blood: negative with no RBC Casts. Morning Spot Urine random protein to creatinine ratio: 1.1, Albumin: 3.3, C3:101, C4:70. Sonography of kidneys and ureters revealed fused and small kidneys, which was

seemed, liked crossed ectopic kidneys. DMSA scan was performed. In DMSA scan two fused kidneys, attached medially with acceptable good cortical function in right and irregular border in lower pole of left kidney, has been seen which was remarkable of pancake kidney and pyelonephritis. (Figure 1) Due to Association between vesicoureteral reflux and ectopic kidneys, voiding cysto- urethrogram (VCUG) was performed that no vesicoureteral reflux was detected. Neurological imaging (brain MRI) was normal and developmental delay had been followed up.

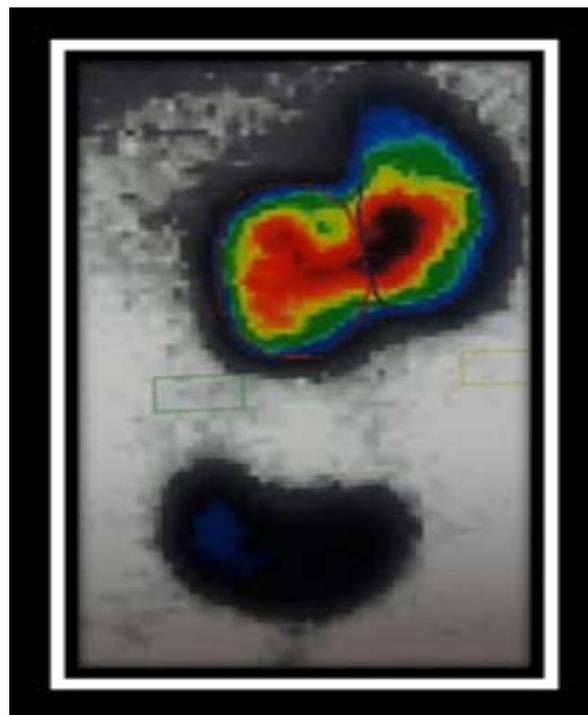


Figure 1. DMSA scan posterior view

3. Discussion

Renal fusion anomaly was first described by Wilmer in 1938. He proposed, these anomalies, were created as a result of pressure from abnormally placed umbilical arteries that prevents cephalad migration of the renal unit. [1] Among fusion anomalies, the most common form is horseshoe kidney with the incidence of 0/25 % population. McDonald and McClellan in 1957 categorized the fusion anomalies and modified into different types, depending on the fusion type. There are two kinds of renal ectopia: simple renal ectopia and crossed renal ectopia (i.e., crossed ectopia with and without fusion, solitary and or bilateral crossed ectopia). [2]

Pancake kidney is a very rare fusion anomaly which was first described by Looney and Dodd.

Both kidneys are completely fused to each other at medial border and seem like a mass. Pancake kidneys also ectopic and placed in pelvic cavity. [3] In most cases it has been recognized incidentally because most patients are asymptomatic, but some presentation like urinary tract infection, urolithiasis due to obstructions and stasis has been reported. [4] There have been some reports between pancake kidney and structural anomalies like fallot tetralogy, caudal regression and sacral agenesis. [5] Pancake kidneys are usually situated anterior to the bifurcation of the abdominal aorta and can be demonstrated on ultrasound, CT, IVU, MRI and scintigraphy. [6] As mentioned, pancake kidney is structural anomaly and proteinuria which is a result of glomerular pathology, demonstrating parenchymal involvements. Recently, in India a case of glomerulonephritis in pancake kidney has been reported. [7] Here we report a toddler age patient with pancake kidney and non-nephrotic range proteinuria that did not reported in literatures .parenchymal involvement was seen in DMSA scan and may be it is due to division parenchymal abnormality or pyelonephritis. Further investigation must be done for pancake kidney to clarify

its anatomical and functional abnormality. They need long term follow up to assess their kidney function and complications.

4. Conclusion

Pancake kidney is a very rare renal fused anomaly. Most affected patients are asymptomatic and it is diagnosed incidentally. Unfortunately there is no clinical guideline for management of this disease. According to our research, this is first report of Pancake kidney and developmental delay in toddler age group and need more investigation in future.

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