Scintigraphic and Radiologic Findings of Pancake Kidney in a Patient with Fanconi Aplastic Anemia

Adem Maman¹, Fatih Fırat², İdris Kaya³, Arif Kürşat Ayan⁴

¹Tokat State Hospital, Department of Nuclear Medicine, Turkey
²Tokat State Hospital, Department of Urology, Turkey
³Buhara Hospital, Department of Radiology, Turkey
⁴Atatürk University, Department of Nuclear Medicine, Turkey

Abstract
In this case, we have presented that a patient has fanconi aplastic anemia with pancakes kidney in scintigraphy and ultrasonography. The patient is 10 years old and a girl who fanconi aplastic anemia had been diagnosed since three years. In physical examination her general status is good. There was not left hand thumb and she had double the distal phalanx in his right hand thumb in her inspection. We observed 2/6 systolic murmur in cardiovascular system examination. Other systems were natural. Abdominal ultrasonography was observed that both the kidney were ectopic location and fused view in the left lower quadrant. Similarly in Tc-99m DTPA and DMSA renal scintigraphy, both kidneys were fused and in the left hemipelvis. The right kidney function were significantly lower by comparison with the left kidney functions. Radiological imaging is necessary in patients with Fanconi aplastic anemia without present clinical symptoms. The renal ultrasonography is important for determining pancakes. In addition, static and dynamic renal scintigraphy plays an important role in revealing the functional status of the kidneys.

Key words: Fanconi aplastic anemia, pancakes kidney, scintigraphy

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Introduction
Fanconi aplastic anemia is a rare genetic syndrome characterized with various congenital anomalies and bone marrow failure. Low birth weight, café au lait spots, finger abnormalities and typical facial appearance is present at birth. Approximately 30% of the patients with this disease also have urinary system anomalies (1). Various types of congenital renal abnormalities such as renal agenesis, ectopy or horseshoe kidney can be encountered. Pancake kidney is a rare renal anomaly characterized by lobular shaped fused kidneys via upper and lower renal poles without presence of septum in-between (2). In this report we aim to present ultrasound and scintigraphy findings in a case of pancake kidney anomaly in a patient with Fanconi aplastic anemia.

Case Report
Ten year old female patient with a history of Fanconi aplastic anemia was admitted to pediatric outpatient clinic with abdominal pain. Systemic examination revealed left thumb aplasia and duplication of distal phalanx of the right index finger. A grade 2/6 cardiac murmur was detected in cardiovascular examination. Family history was insignificant. Blood test results were normal except anemia. In abdominal ultrasound examination, kidneys were fused and located in the left lower quadrant (Figure 1). Tc-99m DMSA cortical renal scintigraphy demonstrated two kidneys in left lower quadrant with fusion from medial aspects (Figure 2). Tc-99m DTPA scintigraphy showed similar imaging findings with normal functioning left kidney and reduced right renal function (Figure 3).

Discussion
Fanconi aplastic anemia is an autosomal recessive disorder and can be seen with various congenital anomalies.
anomalies. It is reported that in patients without significant malformations, diagnosis might be delayed until the obvious hematopoietic changes occur (3). Rest of the patients usually present with growth retardation, multisystem anomalies, genital and urinary anomalies. We detected left thumb aplasia and duplication of distal phalanx of the right index finger in our patient and diagnosis was established with following test and examinations. Renal structural abnormalities such as renal agenesis, ectopy and horseshoe kidney can be encountered relatively more frequent in patients however, co-occurrence of pancake kidney which is a pelvic fusion anomaly is very rare. Pancake kidney is first described in 1926 by Looney and Dodd (4). It is seen more frequently in males and can be diagnosed in patients with various ages. It is characterized by presence of a lobulated renal mass with fused poles without a septum in between kidneys (2). Each renal collecting tubules are placed anteriorly with a short ureter which open the bladder normally. Incidence of renal ectopy in autopsy series is about 1/4000 whereas 85% of these are fused kidneys. The most common type of renal ectopy is horseshoe kidney and its incidence is 1/700 in autopsy series (2). However incidence of pancake kidney is remain unknown (4).

Reports present in literature regarding the co-occurrence of pancreatic agenesis, uterine anomalies and caudal regression anomalies with pancake kidney (5, 6). Renal ectopy, cross ectopy and associated urogenital malformations (e.g. hypospadias and vesico-ureteral reflux) were reported in patients with Fanconi aplastic anemia (7), however, co-occurrence of pancake kidney was not reported to our knowledge.

Diagnosis of pancake kidney is usually established by coincidence. Intravenous pyelography was usually the method of diagnosis in the past. However, in recent years, diagnosis has been established by ultrasonography, computed tomography and scintigraphy. In our patients ultrasonography revealed the presence of fused kidneys and diagnosis was validated by renal cortical Tc-99m DMSA and Tc-99m DTPA scintographies. Scintigraphic evaluation also revealed that left kidney was functioning normally whereas reduced function was detected in right kidney.

References

4. Looney WW, Dodd DL. An Ectopic (pelvic) Completely

Figure 2. Renal cortical Tc-99m DMSA scintigraphy image shows two kidneys fused from medial aspects in lower abdomen.

Figure 3. Tc-99m DTPA scintigraphy image shows two fused kidneys in lower abdomen. Left kidney is normal functioning whereas reduced function is seen in right kidney.
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