Tc-99m DMSA Scintigraphy in the Diagnosis of Renal Anomalies: A Turner Syndrome Case

Renal Anomali Teşhisinde Tc-99m DMSA Sintigrafisi: Bir Turner Sendromu Olgusu

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ABSTRACT Turner syndrome (45, X0) is a chromosomal abnormality characterized by short stature, ovarian failure, streak gonad, hypertension, autoimmune thyroid disease, heart and renal anomalies. Here we report a case of Turner syndrome associated with crossed renal ectopia with fusion anomaly. A 8 year-old girl who had growth retardation and frequent urinary tract infection was investigated by Tc-99m DMSA scintigraphy. DMSA scan imaging showed crossed renal ectopia with fusion anomaly. The current case showed us once again that static renal Tc-99m DMSA scintigraphy imaging is an important diagnostic tool. It can not only determine renal cortical function but also can determine the congenital anomalies of the kidney including fusion anomalies.

Key Words: Technetium Tc 99m dimercaptosuccinic acid; Turner syndrome

ÖZET Turner sendromu (45, X0) boy kısalığı, ovarian yetmezlik, çizgi (band tarzında) gonad, hipertansiyon, otoimmün tiroid hastalığı, kalp ve renal anomalilerle karakterize bir kromozom anomalisidir. Bu yazıda füzyon anomalili kros renal ektopinin eşlik ettiği bir Turner sendrom vakası sunulmaktadır. Gelişme geriliği olan ve sık idrar yolu enfeksiyonu geçiren 8 yaşında bir kız çocuğuna Tc-99m DMSA sintigrafisi uygulandı. DMSA sintigrafisinde füzyon anomalili kros renal ektopi saptandı. Bu vaka bize statik renal DMSA sintigrafisinin önemli bir teşhis aracı olduğunu bir kez daha göstermiş oldu. DMSA sintigrafisi ile sadece renal kortikal fonksiyon değerlendirmesi değil, aynı zamanda füzyon anomalilerini de içeren renal anomali teşhisi yapılabilmektedir.

Anahtar Kelimeler: Teknesyum Tc 99m dimerkaptosüksinik asid; Turner sendromu

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Turner syndrome (TS) is a chromosomal abnormality that results from the complete or partial loss of the second sex chromosome. Initially described in 1938 by Henri Turner, it affects one in 2500-3000 live-born females.[1-3] TS patients phenotype is variable and the wide spectrum of clinical features includes: short stature, ovarian failure, streak gonad, webbed neck, lymphedema heart and renal anomalies, hypertension, and autoimmune thyroid disease.[4,5] Renal malformations have been found in 30-40% of TS cases. Some renal abnormalities that can be seen in Turner syndrome are: malrotation, horseshoe kidney, ectopic kidney, renal agenesis and renal cysts.[6] Therefore, it is possible to suggest the investigation of renal anomalies in any case of TS with or without such a clinical finding. Especially searching of renal anomalies is very important in the treatment of urinary tract infections.

Renal cortical scintigraphy by technetium-99m dimercaptosuccinic acid (Tc-99m DMSA) is a highly sensitive method for the detection of cortical abnormalities related to urinary tract infection.[7,8] It also gives crucial information the relative renal function and the morphology of the kidney. Common indications of DMSA scintigraphies are; detection of focal renal parenchymal abnormalities, detection of renal sequelae, detection of acute pyelonephritis, and detection of associated abnormalities such as abnormal duplex kidney, small kidney, dysplastic tissue, horseshoe kidney, ectopic kidney.[9] Compared to ultrasound and intravenous urography, the sensitivity of DMSA scintigraphy is high, both in acute and chronic pyelonephritis. Especially in pediatric nuclear medicine, DMSA scintigraphy is a widely used imaging technique for the detection of renal cortical lesions and renal abnormalities.[10]

CASE REPORT

A 8 year-old girl, who had growth retardation and frequent urinary tract infection, was referred to Onsekiz Mart University Faculty of Medicine Department of Nuclear Medicine and Medical Genetics. On physical examination she had short stature and webbed neck. The height and the weight of the patient were, 94.5 cm and 14.8 kg respectively. In order to investigate the etiology of growth retardation, chromosomal analysis was done in cytogenetics laboratory of medical genetics. Chromosomal analysis from pheripheral blood sample was performed and the karyotype was centrally reviewed. Heparinised blood sample was obtained for cytogenetic diagnosis and lymphocytes were processed using PHA stimulated short-term (72-hour) culture. GTG banding chromosomes were evaluated by fully automated karyotype analysis system (Leica DM2500/Germany). In a total of 40 metaphase cells were analyzed for the current case. The karyotype analysis was revealed 45,X and case was diagnosed as classical Turner syndrome due to X chromosome monosomy in her all methaphases that evaluated (Figure 1).

Clinical and laboratory investigations revealed urinary tract infection. The ultrasonographic examination performed at another institution showed

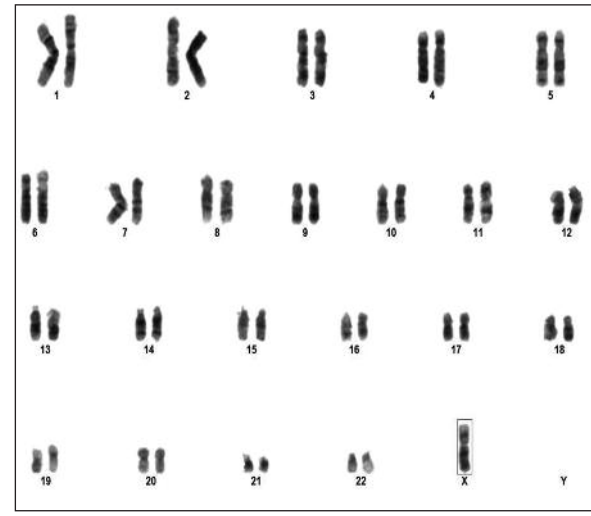

FIGURE 1: The chromosomal analysis of a 8 years old female. GTG banding karyotype showing the 45.XO cell population (Turner Syndrome).

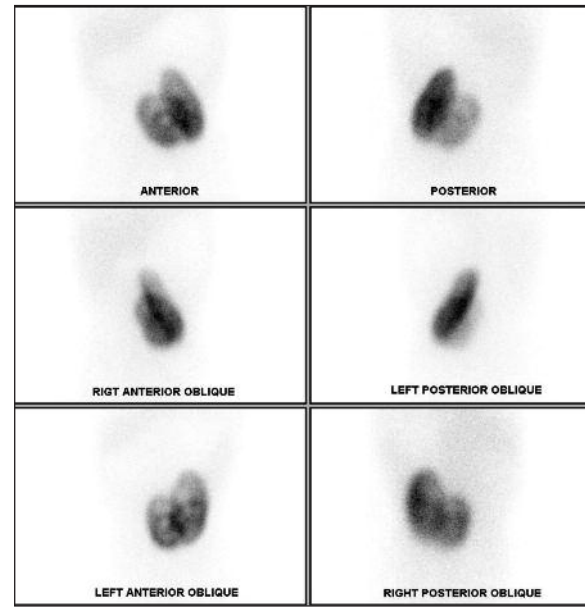

FIGURE 2: Scintigraphic images showed a right crossed fused renal ectopia. The planar images of Tc-99m DMSA scintigraphy in anterior, posterior, right anterior oblique, left posterior oblique, left anterior oblique and right posterior oblique positions.

that the right kidney was not at the normal anatomical location, probably fused to the left kidney. Tc-99m DMSA scintigraphy was performed to investigate the renal parenchymal scar and abnormality. Written informed consent was obtained from her parents before scintigraphy. Scintigraphy was performed with a dual head gamma camera (General Electric Infinia). Anterior, posterior, left anterior oblique, right posterior oblique, left posterior

oblique and right anterior oblique planar images were taken precisely 2 hours after injection of an age-adjusted dose (74 megabecquerel MBq) of 99mTc-DMSA (Figure 2). After planar study, SPECT (Single-photon emission computed tomography) projection was carried with 360 degree rotation. The SPECT images were reconstructed with OSEM/MLEM, filter Butterworth filter (critical frequency=0.5cm, power value=10) and the transverse, sagittal and coronal slices were obtained (Figure 3).

The planar and SPECT DMSA scintigraphy showed a crossed renal ectopia with fusion anomaly (Figure 2,3). SPECT is a three dimensional imaging tool which has been used to investigate space-occupying lesions and anatomical abnormalities. SPECT imaging has additional benefit for determining and correlating the findings of planar imaging. In current case SPECT imaging more clearly showed that the fusion area of kidneys (Figure 3). Left kidney was in the normal position whereas the right kidney was located and fused inferiorly to the lower pole of left kidney (Figure 2, 3). The parenchymal distribution of DMSA showed a minimal heterogeneity probably due to the dilation of pelvicalyceal system but a photopenic area compatible with renal scar was not identified. Otherwise right kidney dimension was smaller than the left kidney. It was difficult to calculate differential renal function because of fusion but it was calculated to be 60% on left, 40% on right kidney.

DISCUSSION

Turner syndrome is an abnormality in the X chromosome that affects development in females. The

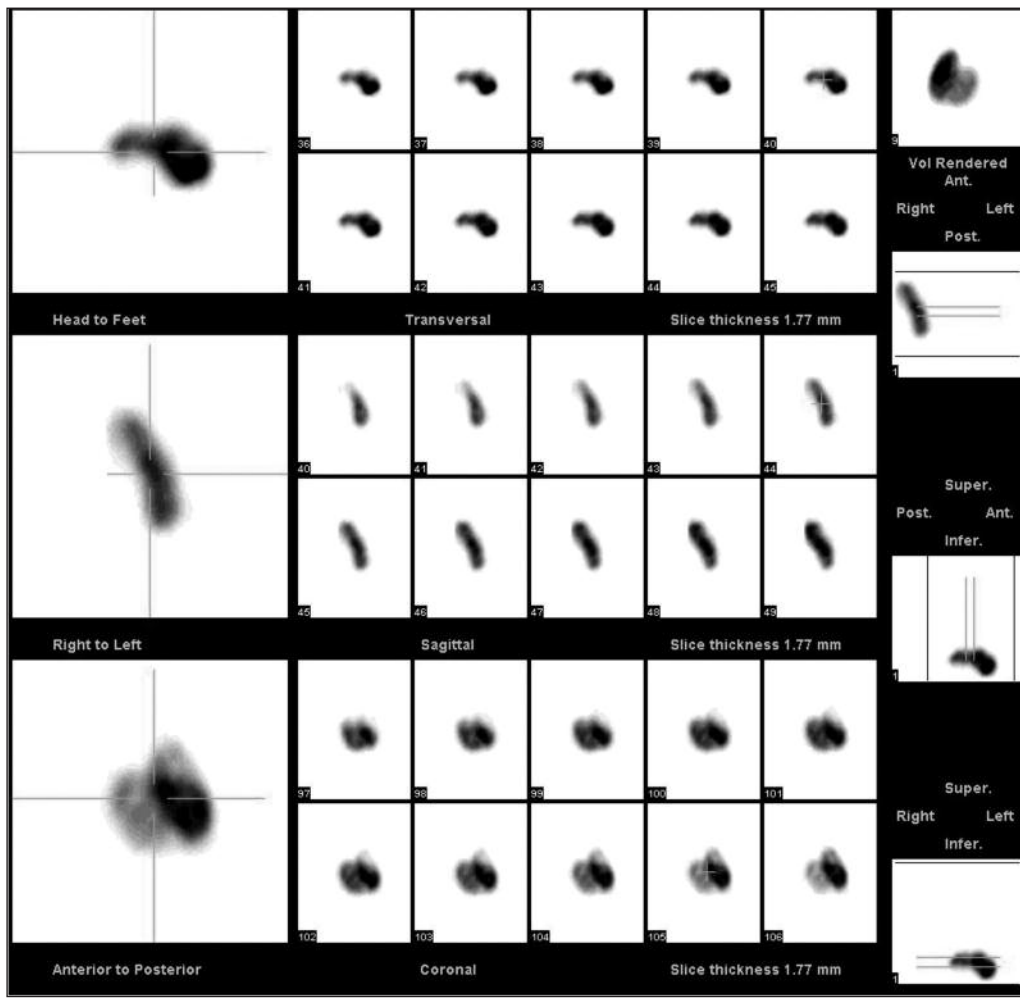

FIGURE 3: The transverse (head to feet), sagittal (right to left) and coronal (anterior to posterior) slices of Tc-99m DMSA SPECT. SPECT imaging more clearly showed that the fusion area of kidneys.

features vary widely and are associated with short stature, ovarian failure, streak gonad, heart and renal anomalies, hypertension, and autoimmune thyroid disease. Renal abnormalities are commonly observed in TS.[11] Aguigha et al. reported that the most frequent kidney anomalies were malrotations, horseshoe kidneys and double kidneys.[12] Diagnosis of renal abnormalities such as malrotation, horseshoe kidney, ectopic kidney, renal agenesis and renal cysts are very important for accurate treatment of renal disease. Most structural anomalies may be initially asymptomatic but still there may be a higher risk of hypertension, urinary tract infection and hydronephrosis. For this reason strict clinical follow-up was recommended to the current case who has been diagnosed as crossed renal ectopia with fusion anomaly. As a result, it may be suggested that the use of DMSA may be helpful in patients with congenital renal anomalies.[13] This allows diagnosis of clinically significant abnormalities and it also detects collecting system malformations, horse-shoe kidneys and malrotation or other positional abnormalities.

In clinical cases like Turner syndrome which is associated with frequent renal malformations, it is suggested to investigate the existence of renal malformation by DMSA scintigraphy. Therefore, it is possible to investigate the damages of kidney parenchyma caused by frequent urinary system infections at the same time with the diagnosis of any renal abnormality by the same investigation tool. In DMSA scintigraphies the radiation dose that the patient is exposed to is very low. The effective Tc-99m DMSA dose is estimated to be approximately 1mSv examination, regardless of the age of the child and using the dose schedule put forward by the European Association of Nuclear Medicine Paediatric Commitee.[9] However the effective dose intravenous urography is estimated to be approximately 1-1.8mSv.[14] The effective dose of DMSA is generally less than the intravenous urography.[9] Although ultrasonography dose not utilize radiation and is a noninvasive method it is less sensitive than DMSA scintigraphy to detect renal cortical scars. Studies comparing ultrasonography with renal cortical sintigraphy indicated that ultrasonography cannot be substituted for DMSA scan in the detecting of renal scar.[15]

As a result, because of its high sensitivity DMSA scintigraphy today is one of the safest tools that can be used on patients at any age, for the investigation of both renal parenchymal damages and for the diagnosis of renal abnormalities.

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