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Laurence-Moon-Biedl syndrome: Scintigraphic appearance of kidneys

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We report a 7-year-old child with Laurence-Moon-Biedl syndrome, an autosomal recessive syndrome, with impaired renal function detected by means of technetium-99m diethylenetriaminepentaacetic acid (Tc-99m DTPA), technetium-99m dimercaptosuccinic acid (Tc-99m DMSA) scintigraphy, and ultrasonography. The altered renal morphology and decreased renal functions are documented.

Key words: Laurence-Moon-Biedl syndrome, renal function, Tc-99m DTPA, Tc-99m DMSA

INTRODUCTION

LAURENCE-MOON-BIEDL SYNDROME (LMBS) is an autosomal recessive syndrome characterized by mental retardation, hypogonodism, short stature, obesity, retinitis pigmentosa, polydactily, skull deformities, gastrointestinal and renal abnormalities.[1] Recently it has been recognized that renal abnormalities occur as commonly as the other cardinal features.[1] We report a patient with Laurence-Moon-Biedl Syndrome who had an abnormal renal morphology assessed by Tc-99m DMSA scintigraphy and impaired renal function evaluated with Tc-99m DTPA scintigraphy.

CASE REPORT

A 7-year-old girl with a known history of LMBS had been under examination for her recurrent urinary tract infection. She underwent radiological and scintigraphic examinations for a possible underlying urinary system abnormality. Ultrasound examination showed minimally atrophic kidneys with marked lobulation. The echogenity of the kidneys and the ureters was within normal limits. Intravenous pyelography (IVP) showed mild calyceal blunting and asymmetrical parenchymal loss.

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Tc-99m DTPA scintigraphy was used to determine the glomerular filtration rate (GFR), perfusion index (PI) and differential renal function (DRF). Following i.v. administration of 185 MBq of Tc-99m DTPA, serial images were recorded with a gamma camera (Toshiba 501) equipped with a low energy all purpose collimator (Fig. 1). Both kidneys showed decreased and delayed perfusion and impaired glomerular function bilaterally. Her GFR, calculated according to Gates' method,[2] was 41.1 m*l*/min (20 m*l*/min for the left and 21.4 m*l*/min for the right kidney). Perfusion indices were calculated as 286 for the left kidney and 500 for the right, using time activity curves and the formulae;[3]

PI: (Integral of the aortic curve to T_{max}/Integral of the renal curve to aortic T_{max}) × (pixel size of renal ROI/pixel size of aortic ROI) × 100.

The kidneys showed slightly reduced concentrating function. Although the excreting function of the right kidney was minimally delayed, both kidneys excreted a sufficient amount of radioactivity in 20 minutes (Fig. 2). DRF was calculated as 48.8 for the left and 51.2 for the right kidney, using the ratio of radioactivity measured between 80 and 140 seconds in one kidney to the total radioactivity in both kidneys over the same period.

Tc-99m DMSA scintigraphy was performed 6 hours after i.v. administration of 110 MBq Tc-99m DMSA. The distribution of the radioactivity was inhomogeneous, indicating that the renal morphology was disturbed bilaterally due to a combination of parenchymal scar formation and dilated pelvicalyceal structures (Fig. 3). The

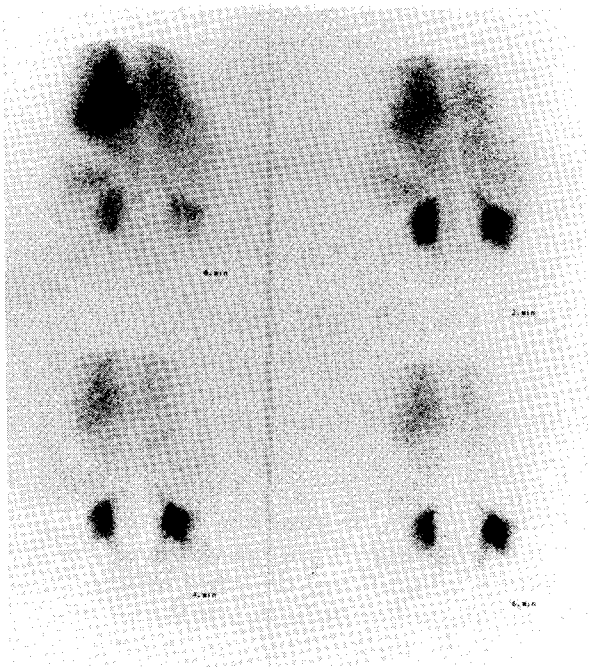

Fig. 1 Tc-99m DTPA study of a 7-year-old child with Laurence-Moon-Biedl syndrome. Bilateral hydronephrosis, that is more severe on the right side, show decreased radioactive tracer accumulation and delayed excretory function of both kidneys. The background activity is noticed as high uptake.

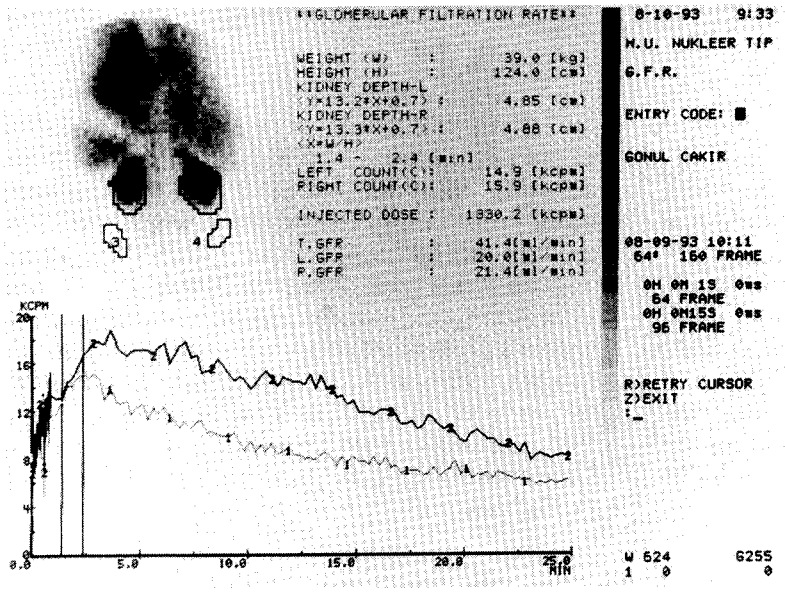

Fig. 2 The computerized analysis of the kidneys. Using Gates method and the time activity curves (curves 1 and 2, for the left and the right kidney, respectively) the total glomerular filtration rate was calculated as 41.4 m*l*/min.

uptake of the radiopharmaceutical agent was decreased in both kidneys (8% and 9% of the injected dose for the left and the right kidney, respectively).

DISCUSSION

Renal involvement may occur in the majority of patients (70–90%) with Laurence-Moon-Biedl syndrome and is an important factor in predicting mortality. Renal findings are now considered as a cardinal feature of this syndrome with variation in kidney size, hydronephrosis, parenchymal damage, renal hypoplasia, vesicoureteral reflux, mesangial proliferation, sclerosis, intestinal fibrosis, focal sclerosing glomerulonephritis and tubulointerstitial disease.[1,4–7] Microscopic changes may be important findings in the genesis of functional disturbances, and a wide variety of histopathologic changes in cortical and medullary cyst have been described.[4–7] Hypertension may accompany in 50% of these patients.

Ultrasonographic appearances of the kidneys in the neonatal period may be indistinguishable from autosomal recessive polycystic kidneys.[4] The IVP findings of this syndrome have been well documented with a reduction in renal size, asymmetrical parenchymal loss and calyceal clubbing, and the cystic changes have been said to attribute to vesicoureteral reflux.[5]

In our patient, impaired glomerular filtration and renal scarring were documented with DTPA and DMSA

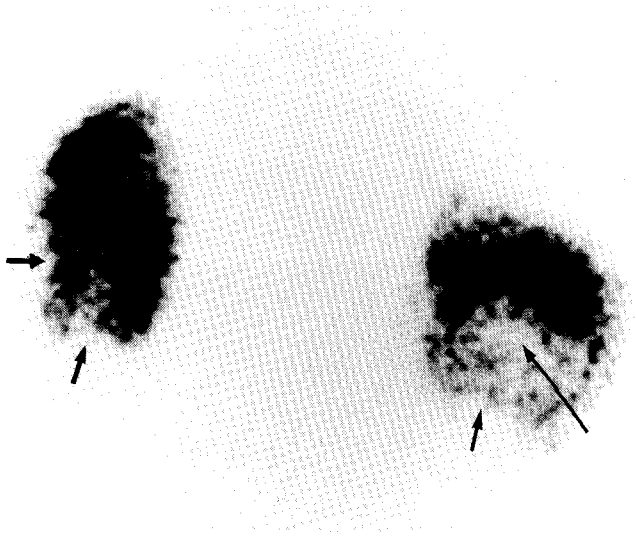

Fig. 3 Tc-99m DMSA study showing amorph appearance of the kidneys; irregular uptake of the tracer due to the combination of parenchymal scar formation in the left kidney (short arrow). The right kidney is in an amorph shape, the irregular and decreased uptake of the tracer was suggestive of dilated pelvicalceal structures (long arrow) and parenchymal scar (short arrow).

scintigraphy. Renal scarring may be due to vesicoureteral reflux and recurrent urinary infection, but DTPA renography is not a sensitive test to document the evidence of reflux. DTPA renography and/or DMSA cortical scan would be a useful tool to document and monitor the global and differential renal function in such patients non-invasively. Since this syndrome most likely results in chronic renal failure, early diagnosis and follow-up of renal dysfunction should be essential.

To the best of our knowledge, this is the first report which describes the scintigraphic appearance of the kidney involvement in Laurence-Moon-Biedl syndrome. This syndrome should be on the list of differential diagnoses of diseases responsible for renal morphologic changes and impaired renal function.

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