Case Report

Poor Tc-99m dimercaptosuccinic acid uptake, re-evaluation with Tc-99m MAG3 scintigraphy in Lowe syndrome

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ABSTRACT

Tc-99m dimercaptosuccinic acid (DMSA) is filtered through the glomeruli and reabsorbed by the proximal tubules as low molecular weight proteins. In Lowe syndrome this mechanism is impaired and so poor DMSA uptake is seen. Poor DMSA uptake was shown in very few studies, but none mentioned normal Tc-99m MAG3 uptake. In this case, the patient had poor DMSA uptake, normal MAG3 uptake and a neurogenic bladder in anterior to the left kidney that attenuates left kidney.

Keywords: Lowe syndrome, Tc-99m dimercaptosuccinic acid, Tc-99m MAG3

INTRODUCTION

Lowe syndrome (oculocerebrorenal syndrome) was first described by Lowe *et al.*, in 1952. It is characterized by X-linked inheritance, congenital cataracts, mental retardation, and renal Fanconi syndrome.[1] Lowe's syndrome is a very rare disease, with estimated prevalence in the general population of approximately 1 in 500,000.[2] Under physiological conditions, low molecular weight proteins are freely filtered from the glomeruli and then reabsorbed by proximal tubular epithelial cells via receptor-mediated endocytosis.[3,4] Tc-99m dimercaptosuccinic acid (DMSA) scintigraphy is a widely used imaging method for detecting renal cortical function. In this imaging technic, DMSA is filtered in the glomeruli and then reabsorbed by proximal renal tubular epithelial cells via megalin- and cubilin-mediated endocytosis.[4] In Lowe syndrome, this mechanism is impaired and renal tubular proteinuri is seen. The patern of Tc-99m DMSA is shown in very few studies, but up to date none showed Tc-99m MAG3 patern in Lowe syndrome, In this case report, the patient had also neurogenic bladder, which makes the state interesting.

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CASE REPORT

A 13-year-old boy who had Lowe syndrome that was known since newborn, admitted to our clinic from another hospital to take a DMSA kidney scan. His serum electrolytes were, urea: 33 mg/dL (normal 8.4-38.4), creatinine: 0.91 mg/dL (normal 0.84-1.25), Na: 139 mmol/L (normal 135-150), K: 4.3 mmol/L (normal 3.5-5), Cl:109 mmol/L (normal 96-110). In his venous gas analyzes, pH: 7.35 (normal 7.31-7.41), HCO_3 content: 20.7 mmol/L (normal 24-28), pCO_2:38.4 mmHg (normal 41-51). In DMSA scan [Figure 1], there was a huge high-grade radioactivity accumulation in anterior to the left kidney and extends through the pelvis. The left kidney can be hardly seen in the posterior images because of the radioactivity accumulation that belongs to the urinary bladder. Both kidneys accumulated the activity low grade. In left posterior oblique images, the left kidney is seen when the attenuation goes away. We took an ultrasonography (USG) image and confirmed the huge activity accumulation was the urinary bladder. In his CT, the anatomical details were better. Furthermore, dilated ureter may be seen like deviated neurogenic bladder. However, there was no ureter abnormality in USG or CT. We took Tc-99m MAG3 scintigraphy in 10 days. The concentration functions of the radioactivity of both kidneys were normal, as expected and seen in patients with renal tubular acidosis and renal tubular dysfunction in the literature. However, it was shown in Lowe syndrome for the first time as we know.

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DISCUSSION

In this case report, the patient with Lowe syndrome had poor Tc-99m DMSA and huge and more activity accumulation that belongs to the urinary bladder in anterior to the left kidney [Figure 1]. In USG [Figure 2] and abdominal CT [Figure 3], urinary bladder can be seen in anterior to the left kidney and extends through the pelvis. The kidneys had normal Tc-99m MAG3 uptake with his dynamic renal scintigraphy [Figures 4a and b].

Yee, *et al.*,[5] studied on rats and acid base imbalance is associated with DMSA uptake. Kim, *et al.*,[6] showed poor renal uptake of Tc-99m DMSA and Tc-99m methylene diphosphonate in a patient with Fanconi syndrome and poor DMSA uptake is associated with impairement of tubular reabsorption of Tc-99m DMSA. Green and Davies's[7] study showed poor DMSA uptake in renal tubular acidosis. Quinn, *et al.*,[8] showed poor Tc-99m DMSA uptake and near normal Tc-99m DTPA uptake in a patient with tubulointerstitial renal disease. Caglar, *et al.*,[9] show the same patern with Tc-99m DMSA and Tc-99m MAG3 scintigraphy in renal tubular acidosis.

Poor activity accumulation of kidneys in Lowe syndrome was defined by Kim, *et al.*,[10] in a case report for the first time. A clinical study was prepared with five patients who had Lowe syndrome. They showed poor DMSA uptake by kidneys and it was attached to the defect in megalin- and cubulin-mediated endocytosis in proximal tubules.[4] Both of these two studies did not mention normal Tc-99m MAG3 uptake.

Up to date, as we know no neurogenic urinary bladder is defined in a patient with Lowe syndrome. In this case, the boy also had a neurogenic bladder and it is deviated to the left side. As mentioned in the literature, DMSA accumulation is more in the urinary bladder than kidneys. In this case, during

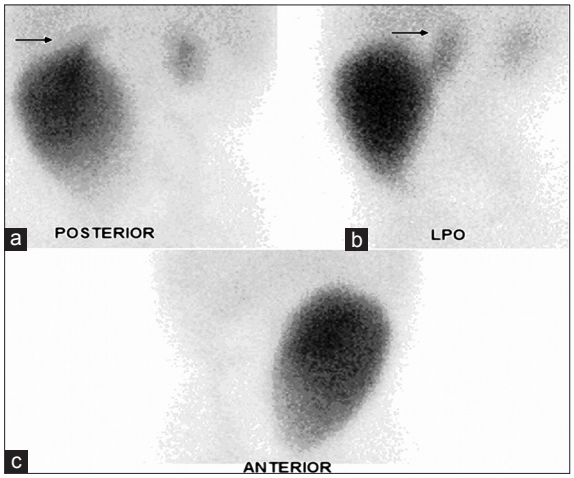

Figure 1: Poor Tc-99m dimercaptosuccinic acid (DMSA) uptake of kidneys. Left kidney (arrow) cannot be seen clearly in posterior image due to attenuation of urinary bladder (a), left kidney is seen clearly in left posterior oblique image (b), huge and high activity accumulation in urinary bladder in anterior image (c)

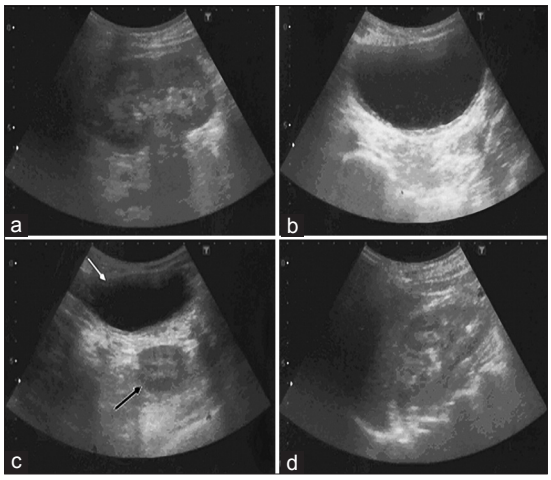

Figure 2: (a) Left kidney, (b) urinary bladder, and urinary bladder (white arrow) is seen in anterior of the (c) left kidney (black arrow) and (d) right kidney

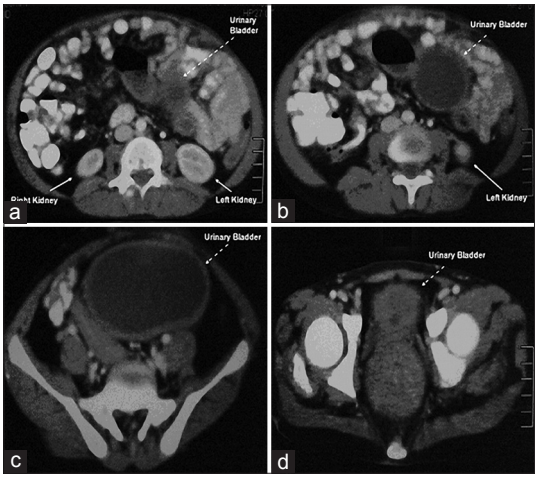

Figure 3: (a) Upper part of the urinary bladder is seen in anterior of the left kidney and (b) lower part of the urinary bladder is seen in the pelvis

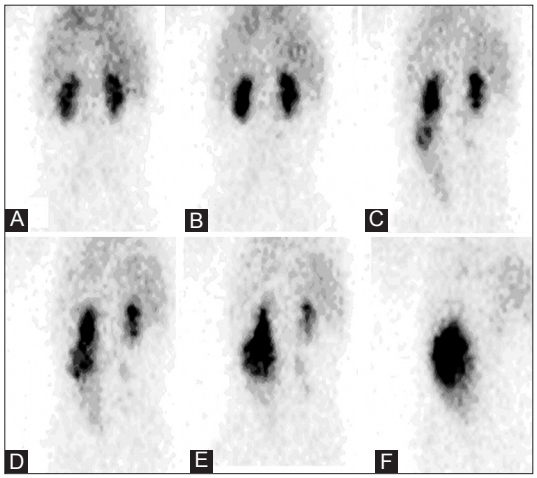

Figure 4a: Both kidneys have normal Tc-99m MAG3 uptake when the urinary bladder fills with radioactivity, it attenuates the lower part of the left kidney (A, B: extraction phase, C-F: excreation phase)

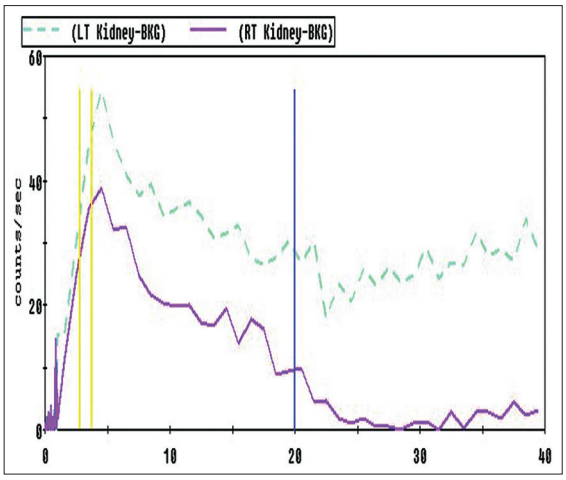

Figure 4b: Renogram curve

MAG3 scintigraphy, urinary bladder fills with radioactivity after the kidneys started to excrete the radioactivity. When the urinary bladder fills, it started to attenuate the lower part of the left kidney and extended through the pelvis. In Lowe syndrome, the kidneys' concentration function is actually normal, if there is no another kidney abnormality, despite DMSA accumulation being poor. Therefore, DMSA scintigraphy should not be used to evaluate the concentration function, instead, MAG-3 scintigraphy examination should be used.

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