Stony kidneys in a child: Clue to a rare diagnosis

A. Mukund, P. Gupta, S. Gamanagatti
Department of Radiology, All India Institute of Medical Sciences, New Delhi, India

A five-year-old boy presented to the Emergency Department with altered sensorium. He had nausea, with occasional vomiting and anorexia for the past one week. His parents denied any history of fever, diarrhea or a significant past medical history or significant family history. The child was unconscious, afebrile, with a pulse rate of 68/minute, blood pressure of 140/80 mmHg and respiratory rate of 17/minute. The systemic examination was unremarkable.

Urinalysis showed calcium oxalate crystals in abundance and blood investigations revealed elevated urea and creatinine levels. A twenty-four hour urinary oxalate was found to be elevated.

Abdominal radiographs showed bilateral diffusely dense kidneys [Figure 1]. Ultrasonography revealed normal-sized kidneys with a smooth outline and uniformly raised cortical echogenicity. No calculus or hydronephrosis was present. Non-contrast computed tomography (NCCT) showed diffusely increased density of both the kidneys with calculi in the gall bladder [Figure 2]. Based on the findings of urinalysis and imaging characteristics, a diagnosis of primary hyperoxaluria was suspected and later confirmed on liver biopsy.

Primary hyperoxaluria is a group of autosomal recessive disorders caused by deficient metabolism of glyoxalate to glycine, leading to an increased production of oxalate that gets deposited in various organs. At least three different types of hyperoxalurias are known.[1] Type-1 results from deficiency of the peroxisomal enzyme alanine-glyoxalate transaminase. Type 2 is caused by the deficiency of cytosolic enzymes glyoxalate reductase and D-glycerate dehydrogenase. Type 3 results from a primary increase in the absorption of oxalate from the gastrointestinal tract.

Classic presentation is nephrocalcinosis with nephrolithiasis, which leads to renal failure in early adulthood. Once the kidneys fail, oxalate accumulates in the body and gets deposited at various sites. Preferred

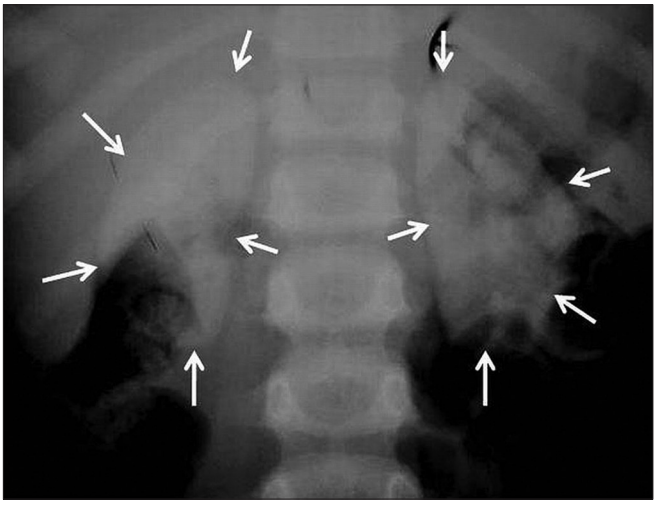

Figure 1: X-ray KUB shows diffusely dense renal shadows on both sides (outlined by white arrows)

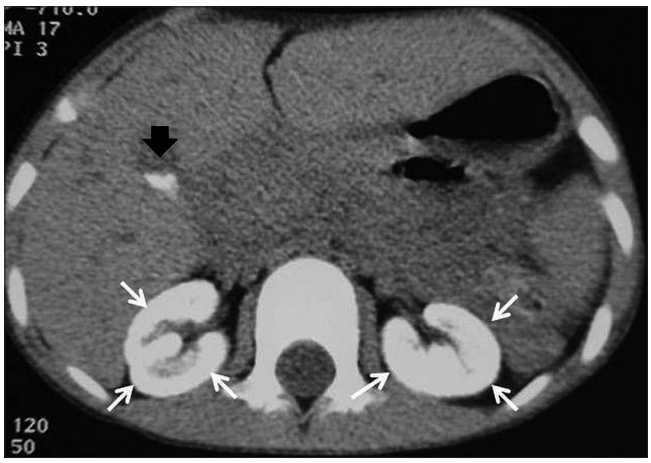

Figure 2: Axial non-contrast CT showing increased attenuation of the kidneys (white arrows) with a calculus in the neck of the gall bladder (black arrow)

Address for correspondence:
Dr. Shivanand Gamanagatti, Department of Radiology, All India Institute of Medical Sciences, New Delhi - 110 029, India. E-mail: shiv223@rediff.com

Access this article online	
Quick Response Code:	Website: www.indianjnephrol.org
	DOI: 10.4103/0971-4065.83749

sites for deposition are bones, bone marrow, blood vessels, cartilage, and the male genitourinary system.

Radiological investigations provide an important clue to the diagnosis. Abdominal radiographs may show increased density in both the kidneys, with or without renal calculi. CT shows markedly increased attenuation of kidneys or calcification/calculi within the kidneys. The liver and heart also show increased attenuation.[2] These imaging features are practically pathognomonic of primary hyperoxaluria. In the present case, the abdominal radiograph shows diffusely dense kidneys on both the sides and the CT images show diffuse calcification of the renal parenchyma predominantly involving the renal cortex, suggestive of cortical nephrocalcinosis, however, no renal calculus was seen. Diffuse, homogeneous, and predominant involvement of the cortex points to the probable diagnosis of primary hyperoxaluria. Other common causes of cortical nephrocalcinosis include acute cortical necrosis, chronic glomerulonephritis, and the Alport syndrome.[3] However, in these conditions the calcification seen is thin and rim-like, having a 'tramline' appearance or there may be a spotty appearance due to calcification of the necrotic glomeruli.

Diagnosis of primary hyperoxaluria should be considered when a child presents with recurrent nephrolithiasis and nephrocalcinosis or early onset renal failure with raised oxalates. Diagnosis is confirmed by genetic analysis, liver biopsy, and analysis for the activity of glyoxylate aminotransferase and glyoxalate reductase.

Combined liver–kidney transplantation is the treatment of choice for patients with progressive renal failure.[4] Medical therapy is aimed at increasing excretion and decreasing the deposition of calcium oxalate.

References

1. Tintillier M, Pochet JM, Blackburn D, Delgrange E, Donckier JE. Hyperoxaluria: An underestimated cause of rapidly progressive renal failure. Acta Clin Belg 2001;56:360-3.
2. Akhon O, Ozmen MN, Coskun M, Ozen S, Akata D, Saatci U. Systemic oxalosis: pathological renal findings and specific extrarenal findings on US and CT. Pediatr Radiol 1995;25:15-6.
3. Diallo O, Janssens F, Hall M, Avni EF. Type 1 primary hyperoxaluria in pediatric patients: renal sonographic patterns. AJR Am J Roentgenol 2004;183:1767-70.
4. Jeyarajah DR, Mcbride M, Klintman GB, Gonway TA. Combined liver-kidney transplantation: What are the indications? Transplantation 1997;8:1091-6.

How to cite this article: Mukund A, Gupta P, Gamanagatti S. Stony kidneys in a child: Clue to a rare diagnosis. Indian J Nephrol 2011;21:295-6.

Source of Support: Nil, **Conflict of Interest:** None declared.