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Unusual Cause of Acute Renal Failure in Infancy

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Acute renal failure due to intratubular obstruction is uncommon in infants. Two infants presenting with acute renal failure associated with acute gastroenteritis were found to have bilateral global nephrocalcinosis secondary to oxalosis.

Key words: *Nephrocalcinosis, Renal biopsy, Oxalosis.*

Acute renal failure (ARF) is frequent in newborns and infants. Clinical conditions causing hypovolemia, hypoxemia and hypotension in newborns and infants may lead

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to renal insufficiency. Few leading causes are perinatal anoxia, ischemia and sepsis. ARF accounts for 8-24% of all NICU admissions. Intrinsic renal failure accounts for 11% of all admissions of which intra-renal obstruction is rare(1,2). We report two infants with intra-tubular obstruction presenting as ARF.

Case Reports

Case 1

A 3-month-old girl, first living child born to third degree consanguineous parents was referred for management of ARF with oliguria, which was noticed during evaluation of acute gastroenteritis. The patient's antenatal, perinatal, birth and developmental history was unremarkable except for repeated episodes of vomiting. On examination the infant was dehydrated, tachypneic, acidotic with a short systolic murmur over the left precordium medial to the mid-clavicular line with normal blood pressure. There was no renal or bladder mass.

Investigations showed blood urea level of 333 mg/dL, creatinine 9.1 mg/dL, bicarbonate 12 mEq/L and hemoglobin 6 g/dL. Ultrasonogram of abdomen showed both kidneys of 5.1 cm size with uniformly hyperechoic renal parenchyma and global nephrocalcinosis. Echocardiography showed a small ventricular septal defect. The patient was started on intravenous fluids with sodium bicarbonate and peritoneal dialysis. Ultrasound guided renal biopsy, done five days after admission, showed 16 glomeruli with some degree of immaturity, shrinkage of the tuft and corona of prominent visceral epithelial cells. The cellularity and basement membrane thickness appeared normal. Striking features were seen in the tubules, many of which were filled with crystalline deposits, obstructing the lumina completely. In some areas the deposits were seen within the interstitium extending from the tubular wall. Patchy infiltrates of lymphocytes were seen in the interstitium. Blood vessels appeared mildly thickened and the biopsy picture was suggestive of oxalosis (*Fig. 1*). 24-hr urine oxalate excretion was 14 mg for the 5 kg child (normal <2 mg/kg/day). The patient was started on treatment with oral pyridoxine 100 mg daily and discharged on request with serum creatinine level of 4.5 mg/dL.

Case 2

A 3-month-old girl born to non-consanguineous parents was referred for acute gastroenteritis with dehydration, hurried breathing and oliguria. Blood pressure was normal. Evaluation showed a serum creatinine

Fig. 1. Kidney biopsy showing oxlate crystals in the tubules.

of 7.9 mg/dL, urea 162 mg/dL and bicarbonate 8 mEq/L. Ultrasonogram showed global nephrocalcinosis. She was treated with intravenous fluids and peritoneal dialysis. An ultrasound guided renal biopsy showed 17-18 glomeruli with mild increase in mesangial cellularity and increased mesangial matrix. Many tubules showed destruction of the lining epithelium and contained crystalline material, consistent with oxalate crystals, which was birefringent under polarized light. The interstitium showed patchy aggregates of lymphocytes and mononuclear cells. The patient was treated with oral pyridoxine 100 mg daily and discharged on request with a serum creatinine level of 3.2 mg/dL.

Discussion

Oxalosis is characterized by elevated levels of oxalic acid. Oxalosis can be primary or secondary. Primary oxalosis is of two types, type I and type II. The childhood and young adult form of oxalosis present with history, signs and symptoms of urolithiaisis whereas infantile type of primary oxalosis present with renal failure. Hence sibling with oxalosis, neonate with ARF and an infant with echogenic kidney should be taken as clues for early diagnosis(3,4). It is imperative to screen a sibling of a child with oxalosis antenatally, with the help of amniocentesis, chorionic villus sampling and fetal liver biopsy(5). Liver biopsy tissue showing deficiency of the enzyme alanine glyoxylate aminiotransferase is the diagnostic investigation. Preemptive liver transplantation has a better therapeutic strategy than combined liver and kidney transplantation after the onset of renal failure, to improve the outlook of these patients(6).

Difficulties in the diagnosis of oxalosis in infancy are many. The normal range of urinary and plasma oxalate, glycolate, glyoxylate and glycerate in infants are not known. Serum oxalate level tends to be higher in children with oxalosis and renal failure when compared to children with same status of renal failure without oxalosis. Diagnosis of oxaluria may be complicated by the presence of renal failure as urine oxalate excretion may decrease and even fall within the normal range when renal failure becomes more advanced.

Oxalosis should be considered in the differential diagnosis of intrinsic renal failure in infancy especially when on clinical evaluation there are no signs or symptoms other than those, which can be explained by renal failure. An ultrasonogram must be done in every child with renal failure(7). Documentation of cortical/global nephrocalcinosis in an infant warrants further careful evaluation. Thus ARF when associated with cortical nephrocalcinosis is a strong pointer for oxalosis especially with an insignificant past history. Exact diagnosis is important since it has consequence concerning genetic counseling and treatment. Hence in the absence of a liver biopsy, which is confirmatory, an ultrasonogram will suggest and renal biopsy will confirm oxalosis. Dialysis and transplantation are hard to justify in infantile primary oxalosis at present.

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Idiopathic Granulomatous Hepatitis

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A 12-year-old male child reported with history of fever for last seven years. Hepatosplenomegaly, hepatic and bone marrow granulomas were the main features. Idiopathic Granulomatous Hepatitis (IGH), a rare syndrome amenable to immunosuppressive therapy was diagnosed.

Key words: *Granulomatous Hepatitis*

Idiopathic Granulomatous Hepatitis (IGH) is a rare cause of pyrexia of uncertain origin (PUO). Generally reported to affect middle aged adults, IGH is uncommon in children. We report a child with IGH who became symptomatic at the age of 5 years and was diagnosed and treated successfully with immunosuppressive drugs at 12 years of age.

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Case report

This male child first presented at the age of 5 years with high grade fever and hepatosplenomegaly. Investigations for PUO were inconclusive. The child responded to an empirical course of antimalarials. During the subsequent seven years, the child was repeatedly hospitalized with recurrent episodes of high grade fever almost every year. Each febrile episode lasted for 2 to 12 weeks and was associated with massive hepatosplenomegaly. Defervescence, either spontaneous or induced by empirical therapy was associated each time with regression of liver and spleen sizes. Additional clinical features and investigations done over the period of illness are outlined in *Table I*. Bone marrow and liver showed presence of non-caseating granulomas with epitheloid cells. These were diffusely scattered in the bone marrow, while in the liver, they were mainly