Distal Renal Tubular Acidosis with Grade 4 Vesicoureteral Reflux in a Child with Single Kidney

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Abstract

Introduction
Renal tubular acidosis (RTA) is a non-uremic defects of urinary acidification. It is characterized by a normal anion gap hyperchloremic metabolic acidosis; plasma potassium may be normal, low or high depending on the type of RTA. These syndromes differ from uremic acidosis which is associated with a high anion gap, decreased glomerular filtration with enhanced proton secretion by the remaining nephrons.

Case Report
We presented a 2 year-old male child with features of acute kidney injury with growth retardation. On evaluation the child was diagnosed to have distal renal tubular acidosis with grade 4 vesicoureteral reflux with right sided single kidney.

Conclusion
The child had congenital malformation of renal system which was not evaluated previously and remains untreated for long duration which leads to growth retardation and presented in a serious condition in our case.

Key Words: Child, Distal RTA, Single kidney, Vesicoureteral reflux (VUR).

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Introduction

In 1946, Albright et al. described dRTA as a distinct entity (1). The clinical syndrome described, consists of hypokalemia, hyperchloremic metabolic acidosis, inability to lower urine pH below 5.5 in the face of systemic acidosis, nephrocalcinosis and nephrolithiasis. Additional features included osteoporosis/osteomalacia, autoimmune primary hypothyroidism and stunted growth. The syndrome was designated “distal renal tubular acidosis,” since the establishment of a large pH gradient between urine and blood which is a function of the distal nephron. The physician should be familiar with the clinical presentation, and the correct management of the illness, in order to prevent nephrocalcinosis, rickets/osteomalacia or growth retardation. Renal agenesis is relatively common congenital anomaly, usually an isolated sporadic abnormality (2). But sometimes it is associated with complications like vesicoureteric reflux leading renal tubular acidosis, growth retardation and hypertension. Single kidney with vesicoureteric reflux with or without other anomalies like posterior urethral valve, ureterocele etc. is collectively called as Congenital abnormalities of kidney and urinary tract (CAKUT) (3).

Case Report

A 2 year-old male child, weighing 7 kg, born out of non consanguineous marriage, by normal vaginal delivery, presented with fever for 5 days, polyuria, polydipsia, anasarca for 2 days and respiratory distress for one day.

On initial evaluation the patient had high fever, pulse-148/min, sinus rhythm with normal volume, respiratory rate-66/min, blood pressure 112/54 mm of Hg (above 95th percentile) with severe pallor. He had severe malnutrition with height and weight both below 3rd percentile on Centers for Disease Control and Prevention (CDC) growth chart.

Systemic examination revealed bilateral coarse crepitations in chest and firm, non tender liver with span 10 cm without any bruit with ascites in abdomen. Urine output was 16 ml/kg/hr. Investigations revealed blood glucose was 75 mg/dl. Arterial blood gas analysis revealed pH -7.08, PO2 -186 mm of Hg, PCO2 -29 mm of Hg, HCO3 -8.5 mmol/l, Na+ 140 mmol/l, K+ 7.9 mmol/l, Ca++ 0.7 mmol/l and Chloride -101 meq/l which was suggestive of partially compensated raised anion gap metabolic acidosis with hyperkalemia with hypocalcemia. Hb -6.1 gm/dl, TLC -24500, N62 L25, Platelet 1.41 lakh/cm², ESR-120 mm in 1st hr, Urea-110 mg/dl, Creatinine-1.8 mg/dl, Serum albumin -3.0 mg/dl, Liver enzymes were normal. Routine urine examination showed pH-7 with few pus cells with mild albuminuria. Chest X-ray revealed right sided pneumonia. We planned for urgent peritoneal dialysis to control refractory acidosis and anasarca. Strict input output chart was maintained and antibiotics were given after adjusting dose for renal clearance. Intravenous bicarbonate was given 2 meq/kg over 2 hrs. Tab Amlodipine with tab Prazosin was given to control hypertension. After 4 days of bedside peritoneal dialysis general condition of patient improved (Figure.1), swelling subsided, polydipsia and polyuria decreased. Urine output was 8 ml/kg/hr, Urea -77 mg/dl, Creatinine-1.3 mg/dl. Arterial blood gas analysis revealed normal anion gap metabolic acidosis. Further investigations revealed serum calcium and phosphate were 6mg/dl and 3mg/dl respectively with normal urinary phosphate excretion and urinary calcium creatinine ratio > 0.27. Serum osmolality was 303 mOsm/kg of water, urine osmolality was 264 mOsm/kg of water (all
these were random values). Thyroid profile was normal. Urine examination shows pH -8, protein-trace, glucose/ketone bodies-nil, RBC-nil, Pus cells-few, Urine culture shows no growth. X-ray of bilateral wrist joint revealed rachitic changes (Figure.2) and USG abdomen revealed single hypertrophied kidney on right side with dilated pelvicalyceal system (Figure.3). Micturating cystourethrogram revealed grade 4 vesicoureteral reflux (Figure.4). We could not able to do radionuclide scan and urodynamic studies as the patient party could not afford. We continued oral bicarbonate solution and gave calcium with vitamin D supplements. Thereafter we referred the child to a pediatric surgeon and discharged her on antibiotic prophylaxis, antihypertensives and bicarbonate solution with calcium and vitD supplementation and appropriate diet chart.

**Fig.1:** Child after peritoneal dialysis

**Fig.2:** Bilateral wrist joint revealed rachitic changes

**Fig.3:** Single hypertrophied right kidney with dilated pelvicalyceal system

**Fig.4:** Grade 4 vesicoureteric reflux

**Discussion**

The signs and symptoms of kidney and urinary tract diseases are subtle and misleading in infants and young aged children. Vesicoureteric reflux (VUR) can be an isolated finding and called primary reflux, or associated with urological abnormalities such as posterior urethral valves or ureterocele and referred to as secondary reflux (4). VUR can also occur as part of multiorgan malformation syndromes (5). The finding of collections of abnormalities of kidney and ureteric development has lead to the term congenital abnormalities of the kidney and ureteric tract (CAKUT) (3). In CAKUT, VUR is the most common abnormality among other disorders (6, 7) such as duplex systems, obstruction, dysplasia, and single kidneys (8). This suggests the developmental processes that have gone
away in these syndromes have many phenotypic effects. Long standing VUR can lead to diffuse tubulopathy with chronic kidney disease (9).

In our case the 2 year-old child was grossly malnourished with acute renal failure superimposed on a chronic kidney disease which was associated with distal renal tubular acidosis and grade 4 vesicoureteric reflux with single right sided kidney. The child had a past history of recurrent vomiting and diarrhoea for long duration with failure to thrive but was not properly investigated and she developed serious complications. We managed the child with peritoneal dialysis and supportive treatment by which the child improved and was referred to a surgeon for further management.

Conclusion

Long standing untreated vesicoureteral reflux with renal tubular acidosis can leads to growth retardation and subsequently serious outcome. Grades of reflux, type of renal tubular acidosis and severity of renal scarring is important for prognosis. A single evaluation of reflux is of slight value for predicting future functional tubular impairment, and the duration of reflux and other associated factors may be more important. We should always keep an eye of suspicion in young children for renal diseases who presents with recurrent vomiting, malnutrition and growth failure and go for a thorough investigation to prevent future complications.

Conflicts of interest: None.

Author contribution

All authors contributed together in patient evaluation, doing investigations, data compiling and cross checking references. All authors read and approved the final manuscript.

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