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Distal Renal Tubular Acidosis Presenting As Hypokalemic Periodic Paralysis

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Abstract

We report a 4 year old child with periodic paralysis secondary to distal renal tubular acidosis. Although, hypokalemia with muscle weakness is associated with distal renal tubular acidosis, hypokalemic periodic paralysis without typical features, is very uncommon. The patient responded well both, clinically and biochemically, to alkali and potassium supplements.

Key words: Distal renal tubular acidosis, Hypokalemic periodic paralysis

INTRODUCTION

Renal tubular acidosis (RTA) is a disorder of renal acidification resulting in hyperchloremic metabolic acidosis and inappropriately high urine pH. Type 1 (distal) RTA can cause hypokalemia but initial presentation of type 1 RTA with hypokalemic quadriparesis is uncommon. We report a case of hypokalemic periodic paralysis in previously unsuspected distal renal tubular acidosis.

CASE REPORT

A 4 year old boy presented with lower limb weakness that progressed to involve upper limb over a period of 12 hours. The patient had no prodromal symptoms and no history of drug consumption or vaccination in recent days. Patient did not have any seizures, weakness of cranial nerve or respiratory muscle and any sphincteric disturbances. Patient had similar history 1 year back, for which he was treated as Guillan Barre Syndrome and received Intravenous Immunoglobulin for 2 days. Birth history was normal with birth weight 2.6 kg. Siblings were normal.

On general examination, he was growth retarded (weight - 11.5 kg, 3rd percentile; length - 92 cm, 5th percentile). His vital parameters were normal. Central nervous system examination showed decreased power (2/5 and 3/5 in both lower limb and upper limb respectively), with normal tone, normal deep tendon reflex, flexor plantar response, normal sensory and cranial nerve examination. Meningeal signs were absent. Other systemic examinations were unremarkable.

Patient was investigated for cause of quadriparesis. Routine investigations such as hemogram, blood urea (26 mg/dl), and serum creatinine (0.5 mg/dl) were normal. Serum electrolytes showed severe hypokalemia with normal sodium levels (K+ 2.2 meq/l, Na+ 142 meq/l); however, serum Ca++ 10.1 meq/l, and Phosphorus 5.2mg/dl were normal with elevated Alkaline phosphatase 1307 IU/L. Nerve conduction velocity was normal with normal CPK (92 IU/L) values. We investigated for the cause of hypokalemia. The main reasons for hypokalemia excessive losses (renal or gastrointestinal loss) or redistribution within body compartments were considered. Arterial blood gas analysis showed normal anion gap metabolic acidosis (pH 7.178, HCO3 9.8 meq/l, anion gap 12). Urine output was adequate (4 ml/kg/h). Urinary examinations showed (pH 7, anion gap 125, Na+ 143 meq/l, K+ 49 meq/l, Cl- 67 meq/l, Ca++ 13.4 meq/l). Urinary potassium excretion (210 meq/L; normal 40-80 meq/L) and 24 hour urinary calcium excretion (104 mg; normal <4 mg/kg/day) were high. Ammonium chloride test was not done. Other investigations such as X-ray wrist and Ultrasound of adrenals, renal and ureteric system were normal. ECG was showing hypokalemic changes as ST segment depression, T wave flattening and prominent U waves.

Thus, renal potassium losses lead to hypokalaemic paralysis in this patient. Laboratory finding such as absence of hypochloremia, and hyponatremia ruled out Bartter syndrome. Characteristic features of hyperaldosteronism like hypertension and polyuria were absent with normal adrenal gland in the Ultrasound abdomen. Normal thyroid function test ruled out thyrotoxicosis. Normal ultrasound KUB ruled out meullary sponge kidney/renal cyst. Evidence of gastrointestinal potassium loss was not observed and it should be associated with appropriately low urine potassium excretion, so ruled out. Therefore, diagnosis of distal renal tubular acidosis was made on the basis of laboratory findings of hyperchloremic metabolic acidosis with normal anion gap, hypokalemia, increased calcium excretion, urine pH > 5.5 and positive urinary anion gap. 24-hour urinary potassium level confirmed an abnormally excessive urinary potassium loss despite severe hypokalemia.

Initially, intravenous potassium was given to correct hypokalemia slowly over 24 hour, and intravenous sodium bicarbonate was given to correct metabolic acidosis over 48 hour. Then, potassium citrate supplement, polycitra (containing Na:K:Bicrbonate in ratio 1:1:2) was started and dose was titrated as per pH and serum bicarbonate level. With treatment, patient's weakness improved gradually and discharged on oral Polycitra solution. On follow up, growth improved (4 kg weight gain and 5 cm gain in height in 1 year) with near normal biochemical parameters (Na+ 145 meq/l, K+ 3.6 meq/l, pH 7.38, HCO3- 22 meq/l, 24 hour K+ excretion 114 mg, and 24 hour calcium excretion 63mg). After 1 year of regular follow up, patient is normal with no recurrence of paralysis and is doing well.

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DISCUSSION

Renal tubular acidosis is a defect of renal acidification, which results in hyperchloremic metabolic acidosis and inappropriately high urine pH. A group of transport defect secondary to reduced proximal tubular resorption of bicarbonate or distal secretion of proton or both.[1] In comparison to glomerular disease, RTA is less common. There is no significant impairment of glomerular filtration/tubule interstitial inflammation. Distal RTA (primary/ secondary) is due to one or more transporters involved in the acidification process including H+/K+ ATPase, HCO3-/Cl- anion exchange or component of aldosterone pathway. Because of impaired H+ ion excretion, urine pH cannot be reduced <5.5 despite severe metabolic acidosis. Owing to the lack of H+ to bind to in the tubular lumen, loss of bicarbonate distally results in increased Cl- absorption and increased K+ excretion leading to hyperchloraemia and hypokalemia respectively. Hypercalciuria is usually present. Chronic metabolic acidosis impairs citrate excretion and increases risk of calcium deposition in the kidney.[2-4] Mobilization of organic component to serve as buffer to chronic acidosis leads to bone disease.

Distal renal tubular acidosis may be sporadic or inherited due to intrinsic renal or urological causes, toxins, or related to systemic disease. In our case, we could not get a possible cause, so it was assumed to be sporadic or primary. Mutational study could not be done due to financial constraints and non-availability in our setup.

Clinically, most of the patients present with poor growth, polyuria and polydipsia. It may present with severe rachitic deformity and multiple fractures.[5] Hypokalemia may lead to muscle weakness and transient paralysis. Cases of RTA presenting with musculoskeletal pain.[6] and even rhabdomyolysis have been reported in adults. Nephrocalcinosis and renal calculi may occur. Sensorineural hearing loss has been reported in familial cases.[7] Chang et al reported 3 girls with secondary hypokalemic periodic paralysis with different types of RTA. One girl had primary distal RTA with nephrocalcinosis, second had primary Sjögren syndrome with distal RTA, and the third had isolated proximal RTA complicated with multiple organ abnormalities, unilateral carotid artery stenosis, respiratory failure, and altered consciousness.[9]

In our patient, recurrent paralysis associated with hypokalemia and metabolic acidosis was observed as the sole manifestation of distal RTA. Our case had no polyuria, polydipsia, bony changes or renal morphological changes. Hypokalemia leading to periodic paralysis may be due to very high 24-hour potassium excretion leading to potassium depletion, and absence of nephrocalcinosis may be due to moderate metabolic acidosis and lesser calcium excretion.

Treatment of children with distal RTA is aimed to improve growth parameters; however, final height remains compromised. First two years of therapy are crucial,[8] so early diagnosis and management is required. Treatment typically included alkali supplementation. The dose of bicarbonate used is usually 2-3 mEq/kg/day, which can be increased till normalization of pH is achieved. Hypokalemia should be treated along with acidosis. This patient required prolonged potassium and alkali supplementation. Vitamin D and calcium should also be given; however, they were not given in view of normal bone mineralization and coexisting hypercalciuria in our patient.

Table (1) Biochemical investigations

ABG [pH=7.178,HCO3=9.8 meq/l]
Anion gap =12
Na+=142 meq/l
K+=2.2 meq/l
Ca+2 =10.1 meq/l
PO4=5.2mg/dl
ALP=1307IU/L
B Urea=26 mg/dl
S Creatinine=0.5 mg/dl
TSH=5.09 IU
ECG Showing hypokalemic changes
Nerve conduction velocity- normal
CPK=92 IU/L
Urinary PH=7
Urinary potassium excretion=210 meq/L (normal 40-80 meq/L).
24 hour Urinary calcium excretion=104 mg (normal <4 mg/kg/day)
Urinary anion gap=125

CONCLUSION

Clinicians should search for a secondary cause of hypokalaemia in asymptomatic patients, who present with paralysis, particularly in presence of atypical metabolic features. Distal RTA should be kept in the differential diagnosis of hypokalemic periodic paralysis. A high index of suspicion and prompt management can reverse the symptoms which can be life threatening.

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