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A Case of Type 1 RTA in an Adult Associated With Sjogrens Syndrome Presenting As Periodic Paralysis Mimic

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Introduction

RTA type 1 is a defect in the distal acidification in kidneys resulting from impaired H^+ secretion in the distal tubule, it may be genetic or acquired. Patients with distal RTA have low ammonia secretion rates. We present a case of acquired distal RTA secondary to sjogrens syndrome who had multiple episodes of pure motor quadriparesis with positive such history in her sibling (sister) and was a close mimic of hypokalemic periodic paralysis.

Case history

A 35 year old female presented with generalized weakness of body, inability to walk, inability to extend the head from past two days. Patient had significant past history in the form of two such similar episodes six years back. The cranial nerves including fundus examination was normal, deep tendon reflexes were absent, power grade 2 in all limbs with hypotonia, plantars bilaterally flexor and rest of the examination was unremarkable. Investigations revealed Normal CBC, Normal Kidney Function Test, Liver Function Tests, normal ultrasonography of abdomen with severe hypokalemia (K^+ 1.2 mmEq/L) and normal serum sodium and hyperchloremia. Arterial blood gases showed metabolic acidosis (pH 7.27). ECG showed features of hypokalemia. Chest radiograph was normal. Thyroid function tests and serum cortisol was normal. Urine examination was normal however urine pH was > 5.5 inspite of acidemia and Type 1 RTA was suspected. 24 hour urinary potassium excretion was increased with positive urinary anion gap which substantiated the diagnosis of type 1 RTA. Secondary causes of type 1 RTA were looked into and patient proved to have sjogrens syndrome with positive SSA- Ro and SSB-La. Her sibling also had positive sjogrens profile. Patient was managed on the lines of type 1 RTA and patient responded well.

Discussion

Clinically, Type 1 RTA is characterized by failure to thrive, polyuria, and nephrocalcinosis,[1] however in our patient no such features were present. Biochemically, it is characterizedby hypokalemic, hyperchloremic metabolic acidosis as seen in our patient . The basic defect in type 1 RTA lies in a failure of the distal tubular sodium hydrogen ion exchange or chloride-bicarbonate exchanger.[2,3] resulting in sodium being wasted by the kidneys in association with bicarbonate wasting. The

ensuing contraction of the extracellular volume[4] gives rise to hyperaldosteronism and consequent potassium wasting. The resulting hypokalemia is manifested symptomatically as muscle weakness and lack of renal concentrating ability. Osteomalacia in the adult and rickets in the child may be seen. Finally, the hyperchloremic metabolic acidosis gives rise to hypercalciuria and hypocitraturia,[5] leading to nephrocalcinosis and nephrolithiasis as a complication of unrecognized and untreated RTA ultrasonography of abdomen didn't show nephrocalinosis in our patient. Chronic metabolic acidosis obligates the skeletal buffering of the extra hydrogen ion. In addition, acidosis stimulates mitochondrial citrate oxidation, resulting in a reduction in the renal excretion of citrate, which is a physiologic calcium chelater that acts to increase calcium solubility. Thus, the hypercalciuria coupled with hypocitraturia make supersaturation probable and nephrocalcinosis likely. Nephrocalcinosis is encountered more frequently in type 1 RTA and uncommonly in type 2 RTA.The retention of hydrogen ion in type 1 RTA obligates the hypercalciuria and increases the risk of nephrocalcinosis. In contrast, type 2 RTA is more difficult to treat because of the frequent and massive doses of citrate needed to counteract the massive bicarbonate wasting.

Type 1 RTA (also known as classic RTA) is characterized by hyperchloremic metabolic acidosis and hypokalemia as seen in our case.The urinary pH is always in excess of 5.5 with net acid excretion of less than 70 mEq/min per 1.73m2. When metabolic acidosis is corrected by base therapy, less than 3% of the filtered load of bicarbonate is excreted in the final urine output. When patients respond by conserving potassium, and urinary potassium excretion falls below 40 mEq/d. However, in patients who have distal RTA, the degree of potassium wasting continues to be a reflection of the hyperaldosteronism associated with the volume contraction in type1, distal RTA.

The treatment of type 1 and type 2 RTA is relatively simple, requiring the use of sodium bicarbonate or the slightly more palatable compound Shohl solution (or Bicitra), which contains citric acid and sodium citrate, providing 1 mEq/mL of alkali. Polycitra solutions contain potassium citrate to provide 2 mEq/mL of alkali and 2 mEq/mL of potassium designed to correct both the acidosis and hypokalemia.. Nonetheless, the large spectrum of variability likely reflects medical noncompliance. The effects of therapy should be monitored every month for the first 6 months. Close monitoring of serum bicarbonate concentration, urinary calcium/creatinine ratio, and linear growth allow better adjustments of medication to maintain serum bicarbonate to more than 22 mEq/L (22 mmol/L) and to reverse hypercalciuria.

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