Distal Renal Tubular Acidosis Presenting as Recurrent Paralytic Crises A Case Report

Rupali Malik[1], Nabadwip Pathak[2]

[1] Senior Resident, Department of Internal Medicine, VMMC and Safdarjang Hospital, New Delhi-110029, India

[2] Student (MD Medicine), Department of Internal Medicine, VMMC and Safdarjang Hospital, New Delhi-110029, India

Abstract

Renal tubular acidosis (RTA) is a syndrome resulting from tubular dysfunction leading to deranged acid-base balance. One of the subtypes, distal RTA manifests as renal parenchymal stone disease with nephrocalcinosis, evidence of vitamin D deficiency and growth faltering in children with ultimate short stature in adulthood. The physician should be familiar with the condition, in order to expedite correct management and ameliorate the complications. We hereby report a case of distal renal tubular acidosis (dRTA), who presented with recurrent hypokalemic paralysis and osteopenia but, unusually, without any nephrocalcinosis.

Keywords: Distal renal tubular acidosis, paralytic crises, acid-base balance

(Rec.Date: Mar 31, 2015 Accept Date: Apr 09, 2015)

Corresponding Author*:* Rupali Malik, Senior Resident, Department of Internal Medicine, VMMC and Safdarjang Hospital, New Delhi-110029, India
E-mail: drvickyster@gmail.com

Introduction

Renal tubular acidosis (RTA) type I or distal RTA is a rare disorder consisting of a defect in renal regulation of acid-base balance [1,2]. Though generally asymptomatic, it can present with significant morbidity and is usually manifested as chronic, normal anion gap metabolic acidosis in combination with persistent hypokalemia and a persistently high urine pH. Life-threatening hypokalemia with muscular paralysis (ie, serum potassium level < 2.0 mmol/L) can occur in patients with untreated distal RTA. We describe the case of a patient with this disorder, who presented with recurrent acute hypokalaemic paralysis.

Case

A 38-year-old male presented to our hospital with complaint of acute onset weakness in all four limbs for past 2 days with similar history of four similar short-lasting episodes in last 2 years. All the episodes were acute in onset, with symmetric weakness of all four limbs without a preceeding history of diarrhoea, dysarthria, diplopia, respiratory difficulty, fever, joint pain, skin rash, photosensitivity, dry eyes, dry mouth, parotid swelling, jaundice, drug intake. or any other illness. Also, there was no sensory, bladder, or bowel involvement during these episodes though definite history of increased urination (5-6 litres per day) was evident. He was found to have hypokalemia (documented) on all the occasions and complete recovery occurred in a span of 36-48 hours on receiving intravenous and oral potassium supplementation. There was no family history of similar or significant illness.

At presentation, he had an average built with otherwise normal general physical examination. Neurological examination revealed normal higher mental status, a power of 3/5 in all the 4 limbs with generalised hypotonia and absent superficial and deep reflexes.

He was admitted and evaluated for the cause of the illness. His laboratory evaluation showed severe hypokalemia with supportive findings of electrocardiogram with normal other electrolytes, renal and liver function tests [Table 1]. Intravenous therapy followed by oral potassium supplementation brought serum potassium to normal values. Arterial blood gas analysis showed normal anion gap metabolic acidosis.

On further evaluation, an initial diagnosis of underlying distal RTA was considered in view of an alkaline urinary pH (> 5.5) in the setting of metabolic acidosis. Urine output was > 5 litres

per day, with urinary pH of 6.0 and specific gravity of 1.015. Urinary electrolytes showed urinary potassium of 25 meq/L along with gross urinary potassium wasting as 24-hrs urinary potassium was 189 mEq/L (normal range 40-80 mEq/L). Urinary calcium excretion was high as 24-hrs urinary calcium excretion was 310 mg in 24 hrs (normal 100-300 mg/day). Urinary anion gap was positive (49 mEq/L) indicating decreased ammonium chloride secretion. Urine examination did not show any glucose, proteins or pus cells while 24 hour urinary protein excretion were normal. Ammonium chloride test was carried out by giving 0.1 mg/kg ammonium chloride orally after obtaining blood pH, serum bicarbonate and urine pH. Six hours monitoring of these parameters revealed lack of acidification of urine inspite of increasing acidosis of blood from 7.29 to 7.17 suggesting distal RTA. Altogether all the tests were consistent with features of distal RTA. There was no evidence of any proximal tubular dysfunction.

He was evaluated for etiology of distal RTA, which revealed negative rheumatoid factor (RF) and antinuclear antibody (ANA). Thyroid function tests and Parathyroid hormone levels were normal. X-ray of long bones showed osteopenia. Ultrasonography of abdomen was normal with normal kidney size without any evidence of calcifications.

Patient was managed with potassium and alkali supplementation to which he responded well with resolution of symptoms and normalising dyselectrolytemia. He was later discharged on long term oral (potassium and bicarbonate) supplements and since then, did not develop any further episodes of weakness for last 1 year.

Discussion

Renal tubular acidosis (RTA) is a constellation of renal tubular defects characterized by a normal anion gap hyperchloremic metabolic acidosis; plasma potassium may be normal, low or high-depending on the type of RTA.

In Distal RTA (Type 1), the collecting duct of the distal nephron fail to secrete acid into the urine leading to systemic acidemia and classical clinical features including normal anion gap hyperchloremic metabolic acidosis; hypokalemia; nephrocalcinosis; nephrolithiasis and hypercalciuria [1]. RTA type I is either inherited or acquired. The more commoner, acquired

form, is caused by entities like drugs, autoimmune diseases, or by infection including sjögren syndrome, lupus, hepatitis, amphotericin B, toluene toxicity, and chronic pyelonephritis [2-4]

The clinical manifestations of RTA type I depend upon the disease type, severity and whether it is acquired or inherited. Hypokalemia that is universal in these cases results in muscle soreness, flaccid paralysis, and electrical cardiac disturbances. The most common cause of death is hypokalemia-induced cardiac dysrhythmia.

The mechanisms underlying hypokalemia in distal renal tubular acidosis have not been fully elucidated. The hypokalemia is particularly prevalent in the acquired forms. Hypokalemia probably results from increased kaliuresis due to renal tubular leakage, decreased proximal tubular reabsorption in the face of acidosis and hypocapnia, and aldosterone stimulation [5]

It is important to differentiate from other causes of hypokalemia like familial periodic paralysis (FPP), thyrotoxicosis, hyperaldosteronism and gastrointestinal loss; because the treatment is different and bicarbonate therapy can deteriorate familial hypokalemic periodic paralysis; on the other hand it is the mainstay of therapy in RTA. In FPP oral potassium and acetazolamide are useful in preventing the attacks but acetazolamide is contraindicated in RTA as it produces acidosis.

Bone is critically involved in buffering during the chronic stages of metabolic acidosis [6, 7]. Chronic metabolic acidosis may trigger the release of alkali and calcium from the bone and eventually lead to reduction of bone mass and osteoporosis. Hypercalciuria, hyperphosphaturia, hypocitraturia and a high urinary pH are the main events that predispose RTA patients to develop renal stones [8]. Urinary citrate is an inhibitor of the crystal aggregation and precipitation. When citrate excretion is reduced, more calcium is chelated, thus aggravating urolithiasis and nephrocalcinosis [9]. But in our patient we did not find evidence of nephrolitiasis on x ray and ultrasound abdomen

Conclusion

Recurrent hypokalemic paralysis with apparently progressive symptoms should be evaluated for an underlying disorder such as dRTA that could be potentially treatable. Also it is important to differentiate it from other causes of hypokalemia for appropriate management.

References

1. Batlle D, Moorthi KM, Schlueter W, Kurtzman N. Distal renal tubular acidosis and the potassium enigma. Semin Nephrol. 2006;26(6):471-8.
2. Rodríguez Soriano J. Renal tubular acidosis: the clinical entity. J Am Soc Nephrol. 2002;13(8):2160-70.
3. McCurdy DK, Frederic M, Elkinton JR. Renal tubular acidosis due to amphotericin B. N Engl J Med. 1968;278(3):124-30.
4. Carlisle EJ, Donnelly SM, Vasuvattakul S, Kamel KS, Tobe S, Halperin ML. Glue-sniffing and distal renal tubular acidosis: sticking to the facts. J Am Soc Nephrol. 1991;1(8):1019-27.
5. Blake-Palmer KG, Karet FE. Cellular physiology of the renal H+ATPase. Curr Opin Nephrol Hypertens. 2009;18(5):433-8.
6. Barzel US. The effect of excessive acid feeding on bone. Calcif Tissue Res. 1969;4(2):94-100.
7. Green J, Kleeman CR. Role of bone in regulation of systemic acid-base balance. Kidney Int. 1991;39(1):9-26.
8. Rothstein M, Obialo C, Hruska KA. Renal tubular acidosis. Endocrinol Metab Clin North Am. 1990;19(4):869-87.
9. Simpson DP. Citrate excretion: a window on renal metabolism. Am J Physiol. 1983;244(3):F223-34.