



Case Report

Distal Renal Tubular Acidosis Associated with Mixed Connective Tissue Disease and Hypothyroidism

Razib Kumar Saha¹, Md. Azizul Haque², Mohammad Hasan Tarik³

Abstract

Renal tubular acidosis (RTA) is caused by defect in renal tubular acid transport. Sjogren's syndrome, rheumatoid arthritis, systemic lupus erythematosus and autoimmune hepatitis are the most common autoimmune causes of distal RTA. We are reporting a case of distal renal tubular acidosis associated with mixed connective tissue disease and hypothyroidism presenting as recurrent hypokalemic paralysis.

Keywords: Distal renal tubular acidosis, recurrent hypokalaemic paralysis, mixed connective tissue disease

TAJ 2017; 30: No-2: 76-78

Introduction:

Renal tubular acidosis (RTA) is characterized by hyperchloraemic acidosis with a normal anion gap and no evidence of gastrointestinal disturbances. The urine pH is inappropriately high (> 5.5) in the presence of systemic acidosis. RTA can be caused by a defect in one of three processes: impaired bicarbonate reabsorption in proximal tubule (proximal RTA); impaired acid secretion in the late distal tubule or cortical collecting duct (classical distal RTA); impaired sodium reabsorption in the late distal tubule or cortical collecting duct, which is associated with reduced secretion of both potassium and hydrogen ions (hyperkalaemic distal RTA).¹ Distal renal tubular acidosis (dRTA) may be inherited or acquired by drugs like carbonic anhydrase inhibitors and heavy metal toxicity like cadmium, lead, mercury, hyperglobulinaemic state or underlying autoimmune disorders like systemic lupus erythematosus, Sjogren's syndrome¹. Proximal RTA is frequently associated with urinary wasting

of amino acids, phosphate, glucose as well as bicarbonate and potassium. In the classical dRTA acid accumulation is relentless and progressive, resulting in mobilization of calcium from bone and consequent osteomalacia with hypercalciuria, renal stone formation and nephrocalcinosis. Potassium is also lost in dRTA¹. Here, we report a case of recurrent hypokalaemic paralysis due to dRTA associated with mixed connective tissue disease.

Case report

A 30-year-old female had previous history of multiple small and large joints pain and swelling which was more marked in morning and associated with morning stiffness and fever. She gave history of tightening and thickening of skin of both hands, feet and face which was associated with pain sensation and colour change of both hands and feet following cold exposure for last 10 years. She also had history of episodic both lower limb weaknesses several times for same duration.

¹ Assistant Registrar, Department of Medicine, Rajshahi Medical College Hospital.

² Associate Professor, Department of Medicine, Rajshahi Medical College.

³ Associate Professor, Department of Medicine, Rajshahi Medical College.

She gave no history of muscle pain, headache, seizure, generalized body swelling, chest pain, respiratory distress, dysphagia, abdominal pain. She took potassium supplement, thyroxin and occasional NSAID. She had history of intrauterine death of one fetus and another baby was died just after normal delivery and one normal alive child. On examination patient was mild anemic, pinched nose, small mouth, swelling of both hands and feet, sclerodactyly, Raynaud's phenomenon, muscle power of lower limb was 3/5, deep tendon reflexes were normal, plantar flexor but there was no lymphadenopathy, hepatosplenomegaly and thyroid gland was not enlarged.

Laboratory tests at admission revealed hypokalaemia (serum potassium 1.5mmol/L) and normal anion gap metabolic acidosis (bicarbonate 19mmol/L, blood anion gap 13.5mmol/L). Hypokalaemia was due to renal potassium losses (urine potassium 210 mmol/day). The metabolic acidosis was characterized by inappropriately high urine pH of 6.7, suggesting renal tubular acidosis (RTA). She was treated with potassium supplement and her condition improved. There was no hypouricemia, hypophosphatemia, no proteinuria or glycosuria and absence of paraprotein and urinary Bence-Jones protein suggested that this presentation is not due to proximal RTA. Hereditary dRTA is also excluded as there is no positive family history and onset is late. ANA was strongly positive, RF was negative, anti U1-snRNP and anti Ro/SS-A antibody were positive. Serum TSH was >100 uIU/ml and Serum T4 < 0.405 ug/dl. Serum calcium, magnesium levels were normal. There was no hypercalciuria. Ultrasonogram of whole abdomen, chest X-ray and X-ray KUB region were normal. Our patient fulfilled Kasukawa and Alarcon-Segovia criteria of MCTD.

Patient was treated with tablet sodium bicarbonate and tablet thyroxine during discharge. She was doing well during follow up after 2 months and her potassium and bicarbonate levels were normal.

Discussion

Renal tubular acidosis (RTA) should be suspected when there is a hyperchloraemic metabolic acidosis with normal anion gap and the urine pH is inappropriately high (> 5.5).¹ Many different conditions have been associated with distal renal tubular acidosis. Distal renal tubular acidosis (dRTA) may be inherited or acquired by drugs and toxins, hyperglobulinaemic state or underlying autoimmune disorders like systemic lupus erythematosus, Sjogren's syndrome.¹ The mechanisms underlying hypokalaemia have not been fully elucidated.

RTA is characterized by selective deficiency in H⁺ secretion in alpha intercalated cells in the collecting tubule. Despite acidosis urinary pH cannot be acidified and is above 5.5, which retards the binding of H⁺ to phosphate and inhibits titratable acid excretion. Furthermore, urinary excretion of ammonium chloride is decreased, and the urinary anion gap is positive. Enhanced K⁺ excretion occurs probably because there is less competition from H⁺ in the distal nephron transport system. Furthermore, hyperaldosterinism occurs in response to renal salt wasting, which will increase potassium excretion.²

It is difficult to tell the cause of distal renal tubular acidosis. Inherited causes may be excluded as there is no positive family history and presentation is late onset.

The concurrence of renal tubular acidosis and autoimmune thyroid disease has been reported before. Although the mechanism remains unclear, distal renal tubular acidosis has been well documented in the presence of a variety of autoimmune disorders including thyroid disease, Sjogren's syndrome and systemic lupus erythematosus. Symptoms of renal tubular acidosis usually resolve rapidly with substitution of thyroid hormone.³ But in our patient symptoms did not resolve after normalization of hormone level which implies of its non association. Our patient had mixed connective tissue disease which may be the underlying cause or association of dRTA.

Conclusion

Distal renal tubular acidosis has been known to be associated with connective tissue diseases like Sjogren's syndrome, rheumatoid arthritis and systemic lupus erythematosus. Though rarely reported, mixed connective tissue disease as an association of distal RTA should also be borne in mind.

References:

1. Brian R. Walker, Nicki R. Colledge, Stuart H. Ralston, Ian D. Penman. Davidson's principles and practice of Medicine. 22nd edition. Elsevier; 2014. 445-446
2. Maxine A. Papadakis, Stephen J. McPhee, Michel W. Rabow. CURRENT Medical Diagnosis and Treatment. 57th edition. Lange; 2018. 915-916
3. Mason AM, Golding PL. Renal tubular acidosis and autoimmune thyroid disease. Lancet; 1970. 1104-1106

All correspondence to:
Dr. Razib Kumar Saha
Assistant Registrar,
Department of Medicine,
Rajshahi Medical College Hospital
Email: razibrmc@gmail.com