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A Case of Sjogren Syndrome Presenting As Hypokalemic Paralysis Due to Renal Tubular Acidosis

KEYWORDS: Sjogren syndrome, Renal tubular acidosis, Hypokalemic paralysis

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ABSTRACT *Sjogren syndrome(pSS) is an autoimmune disease characterized by lymphocytic infilteration of exocrine glands mainly salivery and lacrimal.Renal tubular acidosis (RTA) is a common manifestation of pSS. Hypokalemic paralysis is a rare complication of distal RTA. Cases of pSS presenting for the first time as hypokalemic paralysis caused by distal RTA have been rarely reported.We report a rare case of hypokalemic flaccid paralysis due to distal renal tubular acidosis, who was later found to have Sjogren syndrome.*

Introduction

Renal tubular acidosis (RTA) is a disease with impaired reabsorption of the filtered bicarbonate or hydrogen ion (H^+) excretion. This results in a normal anion gap hyperchloremic metabolic acidosis . Plasma potassium (K^+) may be normal, low or high, depending on the type of RTA. There are four types of RTA, among them type 1 (distal) and type 2 (proximal) are common ones. The classic dRTA consists of hypokalemia, normal anion gap hyperchloremic metabolic acidosis, inability to lower urine pH below 5.5, nephrocalcinosis and nephrolithiasis. dRTA can present as either an inherited or acquired condition.Autoimmune disorder like Sjogren syndrome are among the causes of dRTA.Other autoimmune disorders associated with dRTA are rheumatoid arthritis, systemic lupus erythematosus, scleroderma.

The reported incidence of renal involvement in Sjögren's syndrome (SS) patients is 2 to 67 percent. A defect in distal acidification occurs in up to 25 percent of patients with Sjögren's syndrome .

Case Report- A 27years old female patient not a known case of any chronic illness nonsmoker, nonalcoholic, housewife by occupation presented with sudden onset weakness of bilateral upper limbs and lower limbs . On examination she gave history of dryness of mouth and dryness in the eyes since one year. There were no h/o any trauma, fever, backache, neck pain, joint pain, rashes,palpitation,sweating, bowel and bladder incontinence, loss of consciousness. There were no past history suggestive of diabetes/tuberculosis/hypertension. On examination patientt was conscious oriented to time place person. Patient was hemodynamically stable with pulse rate of 78/min and blood pressure 112/66 mm Hg in right arm in supine position. On systemic examination cardiovascular system, respiratory System and per abdomen was within normal limit. Central nervous system examination revealed power of 1/5 in all four limbs with sluggish deep tendon reflexes. Bilateral plantars were flexor. Sensory system was normal.

Laboratory investigations revealed Serum. potassium-2.9meq/dl(Normal3.5to5.5meq/dl), Serum sodium-140meq/l(135-145meq/l) Serum magnesium- 2.10mg/dl(Normal1.70to2.70mg/dl). Arterial blood gas analysis revealed pH 7.302, carbon dioxide (pCO_2) 18.2mmHg, bicarbonate (HCO_3)8.7mmol/L with normal anion gap. Urine acidifaction test did not show fall of urine pH below 5.5. ECG showed prominent U waves . USG Abdomen-Left kidney shows few calculus ,largest of size 15mm in pelvis with grade-1 hydronephrosis, Right kidney show calculus of size 4mm in middle calyx. 24hour Urine Calcium was increased 317.00(N100-300mg/day) Magnesium-42mg/day(N-73-122mg/day),Potassium-69.26meq/day(N,25-125meq/day),Sodium-73.50(N 40-220meq/day)Citrate-4.65mmol/day(N 2-5mmol/day).

Qualitative Anti nuclear antibodies (ANA) was Positive, Anti SS-A/RO-108.95(Positive,N<20.00) Anti SS-B/LA-122.57(Positive, N<20.00). Schirmer test was positive(3mm in right eye and 4mm in left eye at 5min.(Normal 5mm at 5 minute). Lip biopsy show stratified squamous epithelium covered tissue including seromucinous glands revealing mild mononuclear lymphocytic infilterate. Patients was initially managed with intravenous potassium upto 120mEq/ day and sodium bicarbonate 64mEq/day. Vigorous hydration with normal saline was provided. Provision of 15 mEq/day of potassium citrate was followed. Muscle weakness completely recovered five days later. Furthermore serum potassium and bicarbonate returned to normal level. She was discharged with stable condition. Serum potassium and bicarbonate had been tested regularly during routine follow-up and had been maintained within normal limits on 30 mEq of potassium citrate and 16 mEq of oral slow-release potassium chloride per day.

Discussion

In adults dRTA may develop as a consequence of autoimmune disorder such as sjogren syndrome, rheumatic arthritis and systemic lupus erythematous[1,2].

The presence of proximal or distal RTA should be considered in any patient with hypokalemia and unexplained normal anion gap metabolic acidosis.If the value of plasma K^+ is normal or decreased, the demonstration of an inability to lower urine pH below 5.5, either after NH_4Cl loading or after furosemide and fludrocortisone administration, establishes the diagnosis of distal RTA. Patients are diagnosed as primary sojgren syndrome according to 2002 American- European classification criteria[3] which require four out of six criteria . (1) Ocular symptoms (dry eyes every

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day for more then 3 month and/or use of tear substitutes more than 3 times a day)(2) Oral symptoms (daily feeling of dry mouth more than 3 month, recurrent swelling of the salivary glands, or use of liquids to aid in swallowing of dry foods)(3) Ocular signs (positive Schirmer test or a Rose Bengal score of at least 4 of 9)(4) Histopathology with a focus score of 1 in a minor salivary gland biopsy(5) Salivary gland involvement documented by a positive result in salivary scintigraphy, parotid sialography, or salivary flow testing(6)Presence of autoantibodies to Sjögren's syndrome-associated antigen A [SSA (Ro)] and Sjögren's syndrome-associated antigen B [SSB (La)].

The diagnosis of pSS was made in our patient in the presence of dry eyes, dry mouth,ocular sign(Positive schirmer test) and poitive autoantibodies.. There have been few case reports where Sjogren Syndrome has presented primarily with hypokalemia with flaccid paralysis .Reported prevalence of renal involvement in sjogren syndrome is 2 to 67 percent[4]

Managenment of distal RTA, the treatment aim is to provide adequate base to balance H^+ production. A mixture of Na^+ and K^+ salts is advised. In general, total replacement needs 1 to 2 mmol/kg daily. In patients with renal stones, potassium citrate therapy decreases urine calcium excretion and stone formation. Primary distal RTA is a permanent disease, and treatment should be continued throughout life.Prognosis is excellent if associated with dRTA as shown in our patient. Without appropriate therapy, life-threatening metabolic acidosis may occur and require emergency intervention, including respiratory support. Careful monitoring of the physiologic effects of severe hypokalemia, such as arrhythmia, muscle weakness or paralysis, is essential, especially if a faster repletion is used

In conclusion, the patient with hypokalemic normal anion gap metabolic acidosis may hav a diagnosis as complex as dRTA. Adult patient with dRTA should be evaluated for autoimmune disease like Sjogren syndrome Appropriate therapy may then be provided with particular attention paid to the long term adverse effects of metabolic acidosis and severe symptomatic hypokalemia.

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