Hypokalemic quadriparesis in Sjogren syndrome

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ABSTRACT

We report a 58-year-old woman who presented with acute quadriparesis, bulbar weakness, atrial fibrillation, ventricular ectopics, and distal renal tubular acidosis with severe hypokalemia. She recovered completely on potassium and alkali supplementation. The patient also had hypergammaglobulinemia with probable primary Sjogren syndrome.

Key words: Bulbar weakness, distal renal tubular acidosis, hypergammaglobulinemia, hypokalemic quadriparesis, Sjogren syndrome

Introduction

Renal tubular acidosis (RTA) is a disorder of renal acidification out of proportion to the reduction in glomerular filtration rate characterized by normal anion gap (AG) metabolic acidosis.

Type 1 RTA can cause hypokalemia; however, initial presentation of type1 RTA with hypokalemic quadriparesis with bulbar weakness and cardiac arrhythmia is extremely rare. Only 14 cases of distal RTA with hypokalemic muscle paralysis have been previously reported in association with Sjogren syndrome.[1]

Case Report

A 54-year-old postmenopausal female patient presented with history of intermittent low-grade fever 8 days back followed by weakness of lower limbs (difficulty in getting up from squatting position associated with dull aching pain in both buttocks and thighs) 2 days prior to admission. The weakness rapidly worsened by next day to involve the trunk, upper extremities, and neck muscles such that the patient was barely able to move in bed by herself or lift her head. On the day of admission, the patient also complained of difficulty in swallowing solid food and inability to speak loudly. There was no history of visual blurring, diplopia, facial asymmetry, sensory, bladder, or bowel complains. She had a clear sensorium with no history of seizures, involuntary movements, root pains, or band-like sensation over the trunk. She did not give history of prior drug intake, colicky abdominal pain, recent vaccination, or similar episodic weakness in the past. Her past medical and family histories were uneventful. On examination, patient was conscious, cooperative, and oriented. Vital signs revealed pulse of 86/min irregular, BP=110/70 mmHg, RR=16/min thoracoabdominal. Patient looked pale and dehydrated. Cardiovascular system examination revealed irregular rhythm. Respiratory system examination was normal. Bowel sounds were sluggish with no organomegaly. On central nervous system examination, higher function examination was normal. Cranial nerve examination showed weak gag reflex bilaterally. Tone was decreased in all the limbs. The lower limb muscles were moderately tender. Power was reduced in all limbs, proximal muscles (grade 1/5) being more severely affected than distal ones (grade 3/5). Neck and trunk muscles were weak (grade 2/5). Superficial reflexes were absent. Plantar reflexes were flexor bilaterally. Muscle stretch reflexes were normal at all joints except that bilateral ankle jerks were depressed. All modalities of sensation were normal.

In view of the irregular pulse, ECG was obtained,

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which showed intermittent atrial fibrillation with multiple multifocal ventricular ectopics with conduction abnormalities.

Investigations revealed that serum sodium was 130mEq/L, potassium 2.0mEq/L, chloride 110mEq/L, creatinine 1.5mg/dL, magnesium 1.95mg/dL, mild leucocytosis with ESR of 40 mm at 1 h. Random blood sugar was 89mg/dL, thyroid function tests being within normal limits. Serum protein electrophoresis showed total protein of 8.9g/dL, serum albumin 3.36g/dL, serum globulin 5.54g/dL, A/G ratio 0.61, gamma globulin 4.27g/dL, and diffuse band in gamma region suggesting polyclonal hypergammaglobulinemia. Arterial blood gas (ABG) analysis showed pH=7.22, pCO_2=18.4 Kpa, pO_2=76.3 Kpa, HCO_3=7.4mmol/L, SO_2=90.7%. AG was normal (12). The urinary pH was 7.0; there was 2+ proteinuria and microscopic examination revealed 7–10 WBCs/hpf without casts. Trans tubular potassium gradient (TTKG) was 12.5, suggestive of renal potassium loss. Distal RTA with hypokalemic quadripaersis was diagnosed.

Patient was given intravenous Ringer's lactate along with potassium supplements through Ryle's tube with serum potassium monitored 12 hourly. ECG was repeated which showed reversion to normal sinus rhythm.

Following improvement in gag reflex and ability to take orally, oral sodamint tablets (sodium bicarbonate) and potassium chloride syrup was started. Patient's weakness showed significant improvement in 24 h and complete recovery within 48 h of admission.

HIV ELISA, HBsAg, and Anti HCV antibody were negative. Ultrasound of the abdomen showed no evidence of nephrocalcinosis.

Urine for Bence Jones proteins was negative. Serum antinuclear antibody (ANA) was positive++ titer (1:320). Serum anti SS-A/Ro: positive (above 200) and serum anti SS-B/La: positive (157) suggesting the possibility of Sjogren syndrome. Schirmer I test was negative in both eyes (>5mm at 5 min). Minor salivary gland biopsy showed lymphocytic infiltrate in submucosa.

Finally the definitive diagnosis was hypokalemic quadriparesis with bulbar weakness with distal (type 1) RTA with hypergammaglobulinemia with probable Sjogren syndrome.

Patient was discharged on sodamint, two tablets four times a day and potassium citrate syrup two teaspoons thrice a day.

Discussion

In distal (type 1) RTA the nephrons lack the ability to secrete H^+ ions and hence acidify the urine normally during spontaneous or induced metabolic acidosis.[2] Distal RTA can be inherited or acquired. Inherited forms include autosomal-dominant, autosomal-recessive, or X-linked recessive, of which autosomal-dominant form causing mutations in the basolateral chloride-bicarbonate exchanger (AE1) has been identified as the most common form of inheritance. Acquired causes include hypergammaglobulinemic states, such as hyperglobulinemic purpura, cryoglobulinemia, fibrosing alveolitis, Sjogren syndrome, lupus, chronic active hepatitis, thyroiditis, Graves' disease, primary biliary cirrhosis; disorders of calcium metabolism, e.g., primary hyperparathyroidism, vitamin D intoxication, idiopathic hypercalciuria, familial absorptive hypercalciuria, medullary sponge kidney. Tubulointerstitial diseases include leprosy, hyperoxaluria, chronic pyelonephritis, obstructive uropathy; and genetic diseases like Ehler Danlos syndrome, hereditary eliptocytosis, South Asian ovalocytosis, sickle cell disease, carbonic anhydrase II deficiency.[2]

In distal RTA, the urinary ammonium excretion is inappropriately low for the level of acidosis as the defect in acidification decreases ion trapping required for ammonia secretion. Hence urinary anion gap (UAG = urinary Na+KCl) is positive. This differentiates from chronic diarrhea in which the UAG is negative due to enhanced renal ammonium excretion. Also TTKG in diarrhea is <4 due to renal conservation of potassium, hence excluded in our case. In distal RTA, there is a tendency for renal calculi, nephrocalcinosis due to hypercalciuria, and hypocitraturia.[2] Severely depressed plasma bicarbonate levels with a corresponding inappropriate urinary pH >5.5 differentiates from type 2 RTA.[3] Finally the requirement of the patient to maintain plasma bicarbonate levels near normal was less than 1mEq/kg body weight pointed toward distal RTA.

The interesting feature of this case was the presentation with severe hypokalemia, causing acute flaccid quadriparesis with bulbar weakness with cardiac arrhythmia which is extremely rare. This might have been precipitated by underlying urinary tract infection, which may increase the bicarbonate requirement and cause volume depletion and potassium loss. Sudden life-threatening hypokalemia with muscle paralysis is the most serious clinical consequence of distal RTA.[4] Warning symptoms of muscle weakness, psychic apathy,

and lethargy are often present for months or years before muscle paralysis occurs.[5]

To our knowledge, only 14 cases of distal RTA with hypokalemic muscle paralysis have been previously reported in Sjogren syndrome.[6,7] Except for one patient[8] all others had underlying primary Sjogren syndrome. Delays of anywhere from 3 months[9] to 5 years[5] from the onset of initial clinical manifestation have been reported before a diagnosis of Sjogren syndrome is established. Urolithiasis and distal RTA can precede subjective Sicca symptoms, and patients with dRTA may have Sjogren syndrome in the absence of subjective Sicca symptoms. Anti-SS-A antibodies are common in patients with urolithiasis and distal RTA. At least six patients with distal RTA and objective signs of Sjogren syndrome, but without subjective Sicca symptoms, have been reported. Thus, Sjogren syndrome can be diagnosed in dRTA patients even in the absence of subjective Sicca symptoms.[10,11] Although the diagnostic criteria for Sjogren syndrome were not satisfied and that our patient was not having signs and symptoms of xerophthalmia or xerostomia, whether distal RTA could be a manifestation of evolving Sjogen syndrome is a matter of debate.

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