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Case Report

A Rare Case of Recurrent Hypokalemic Paralysis Associated with Distal (Type1) Renal Tubular Acidosis Due to Primary Sjogren's Syndrome with Type 3 Cryoglobulinemic Vasculitis with Autoimmune Hypothyroidism

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Abstract

Tubulointerstitial nephritis (TIN) is the main renal involvement associated with primary Sjogren syndrome (pSS). TIN can manifest as distal renal tubular acidosis (RTA), nephrogenic diabetes insipidus, proximal tubular dysfunction, and others. Distal renal tubular acidosis (RTA) may develop in a large population of patients with Sjogren's syndrome (SS), but most of the subjects are asymptomatic. We report a case of 37-year-old woman who presented with acute quadriparesis with acute hypercapnic respiratory failure. She had history of recurrent episodes of limb weakness which was attributable to hypokalemia and initially labelled as hypokalemic periodic paralysis. Later on, she was found to have metabolic acidosis rather than alkalosis which pointed towards the diagnosis of renal tubular acidosis (RTA) in the absence of apparent gastrointestinal tract loss. Once the diagnosis of RTA was established, an attempt to search the etiology revealed that she was having primary Sjogren's syndrome (PSS) though she did not have any symptom at the time of diagnosis. She was found positive for ANA, anti-SSA and anti-SSB. Schirmer's test confirmed presence of dry eye. A concomitant existence of autoimmune hypothyroidism was a noteworthy association. Continuous potassium replacement produced rapid and complete recovery from quadriplegia and respiratory failure without requirement for mechanical ventilation. Presentation of this case reminds the importance of vigilance while managing a case of recurrent hypokalemia which might be a rare presenting feature of PSS.

Keywords: Distal renal tubular acidosis; Primary Sjogren's syndrome; Quadriparesis; Hypercapnic respiratory failure

Introduction

Sjogren's syndrome is an autoimmune disease with glandular (salivary and lacrimal) and extra glandular (neurologic, renal, hepatic, respiratory, vascular, and cutaneous) manifestations. Tubulointerstitial nephritis (TIN) is the main renal involvement associated with primary Sjogren syndrome (PSS). TIN can manifest as distal renal tubular acidosis (RTA), nephrogenic diabetes insipidus, proximal tubular dysfunction, and others [1], of which RTA is the main clinical presentation [2]. RTA has been reported in 4.3 to 9% of PSS patients; it is more common in middle-aged women, and two-thirds of them will develop symptoms [2,3]. Hypokalemic paralysis is the initial symptom in seven percent of patients with Sjogren's syndrome [4].

Renal tubular acidosis (RTA) is a group of disorders in which renal excretion of acids is reduced, out of proportion to any reduction in glomerular filtration rate. RTA is characterized by hyperchloremic metabolic acidosis with a normal serum anion gap. There are multiple forms of RTA, depending on which aspect of renal acid handling has been affected (Table 1) [5]. In Type 1

or distal RTA (dRTA), the distal nephron does not lower urine pH normally because the collecting ducts permit excessive back diffusion of H+ from lumen to blood. Urinary concentration and potassium conservation also tend to be impaired, so polyuria and polydipsia occur. With the stress of an intercurrent illness, acidosis and hypokalemia can be life-threatening1. Acute hypokalemic paralysis is an uncommon cause of acute muscle weakness, and the reported cases of quadriplegia and acute respiratory failure secondary to hypokalemia are rare [6-9]. We report a case of dRTA who presented with hypokalemic muscular paralysis with acute hypercapnic respiratory failure who did not require ventilation because of fast respiratory function improvement by appropriate treatment with rapid potassium infusion where further workup lead to a diagnosis of primary Sjogren's syndrome.

Table 1: Comparison of different forms of renal tubular acidosis.

Finding	Type 1 RTA	Type 2 RTA	Type 4 RTA
Normal anion gap acidosis	Yes	Yes	Yes
Minimum urine pH	>5.5	<5.5	<5.5
% Filtered bicarbonate excreted	<10	>15	<10
Serum potassium	Low	Low	High
Stones/ nephrocalcinosis	Yes	No	No
Daily acid excretion	Low	Normal	Low
Urine anion gap	Positive	Positive	Positive
Daily bicarbonate replacement needs	<4 mmol/kg	>4 mmol/kg	<4 mmol/kg

Case Report

A 37 years old pleasant, married, Muslim, Bangladeshi housewife, known case of hypothyroidism (on 100 mcg of levothyroxine daily) for 16 years, not known to have any other co morbidities presented to us with the complaints of sudden weakness of both upper and lower limbs for 1 day, shortness of breath for 6 hours and unable to speak for 2 hours. Weakness was sudden in onset and increasing and became so severe that she was unable to move without the help of others and her weakness progressed to the point that she was completely bed ridden. All the proximal and distal muscles and both upper and lower limbs were equally involved but there was no muscle pain. Weakness started in both upper and lower limbs simultaneously. On query her attendant informed that she developed her symptoms after taking heavy meal. There is no history of trauma, pain in the neck or dysphagia, sensory complaints like paresthesia. She also developed breathlessness for 6 hours which was sudden and progressively increasing but not associated with chest pain, sputum, or hemoptysis. For last two hours she was unable to speak but can understand speech and can follow commands like closing her eyes. She denied any history of fever, headache, visual complaints in the form of blurring of vision, diplopia, convulsion, urinary and fecal incontinence, recent vaccination. There was also no history of recent previous infection like respiratory tract infection, urinary tract infection or diarrhea. Patient's attendant also complained of occasional rash about which they could not elaborate. She has history of similar type of attack about 1 year back which was managed conservatively in a local hospital. There was no similar type of illness in her family. She has three children, and all are in good health. She had no history of abortion; no bad obstetric history and all of her pregnancy was uneventful. On physical examination, she was conscious, oriented, but a bit drowsy with GCS 14/15 and had areflexic quadriplegia. Power was graded 0/5 in the legs and in the arms. Plantar responses were absent bilaterally. All modalities of sensation are intact, sings of meningeal irritation were absent. Cerebellar signs, stance and gait could not be evaluated. She had palpable rash over both lower limbs (Figure 1), buttocks and over both palms. Her pulse was 60 beats/min, blood pressure 110/70 mmHg, respiratory rate 30 breath/min. Her chest was clear on auscultation, oxygen saturation was 97% on 5 L of oxygen. The rest of physical exam was unremarkable.

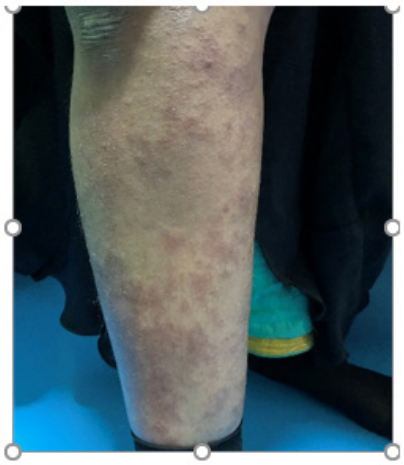

Figure 1: Showing palpable vasculitic rash over lower limb.

On initial laboratory investigations, complete blood count, random blood sugar, SGPT, serum creatinine, CPK, chest x ray revealed normal results except ESR was 68 mmin 1st hour. Serum Electrolyte showed sodium 138 mmol/L, potassium 1.7 mmol/L, chloride 111 mmol/L, bicarbonate 16.5 mmol/L. Anion gap was normal (12.2 mmol/L). Urinary potassium was elevated with 101mmol/L. TSH, serum calcium, serum magnesium level was normal. Electrocardiogram showed sinus bradycardia, diffuse ST depression, T inversion, U wave in precordial limbs and prolonged PR interval (Figure 2).

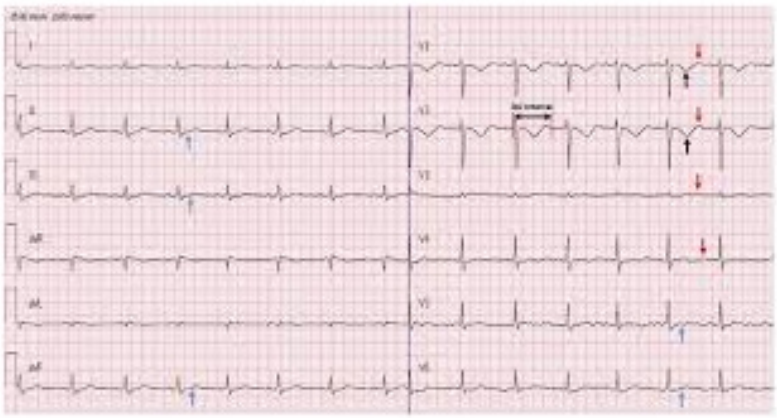

Figure 2: Electrocardiogram showing sinus bradycardia, diffuse ST depression, T inversion (black arrow), U wave (red arrow) in precordial limbs and prolonged PR interval.

Arterial blood gas (ABG) analysis revealed mixed respiratory and metabolic acidosis with hypercapnic respiratory failure (pH 7.03, PO2-69 mm, PCO2- 64 mm, HCO3-16 mmol/L). She was started treatment with intravenous sodium bi carbonate along with intravenous and oral potassium chloride and showed dramatic clinical improvement in 1 day. On next day, her muscle power improved to 4/5 in both upper and lower limbs, respiratory rate was 16 breaths/min and can speak well. SaO2 was 96% in room air. Repeat ABG in room air revealed resolved hypercapnic respiratory failure (pH 7.40, PO2-88 mm, PCO2- 30 mm, HCO3-14 mmol/L). Repeat serum electrolyte revealed sodium 147 mmol/L, potassium 2.9 mmol/L, chloride 109 mmol/L and bicarbonate 18 mmol/L. Despite of correction of respiratory acidosis her metabolic acidosis persisted. Urine routine examination showed pH 7.2, trace proteinuria without any RBC, pus cell or any casts. So, type 1 or distal renal tubular acidosis(dRTA) was suspected and for further evaluation of underlying cause for dRTA, on query, she revealed that she has been suffering from occasional rash for last 5 years which was more prominent in her legs and buttock area and also involved her palm sparing her trunk and face. Rashes care initially itchy and then became painful. She developed these rashes occasionally for last 5 years and resolved spontaneously. She denied any history of joint pain, oral ulcer, muscle pain, hair fall, and weight loss. She also complained of dryness of mouth for last 15 years, that was increasing and became so severe that she cannot swallow her food without water. She also developed dry eyes for same duration and

there is no tear during her emotional event like crying. Bedside Schirmer's test was done and found positive (1 mm wet on filter paper in 5 minutes- Figure 3). She also complaints of bluish discoloration of her fingers and pain on cold exposure and she has been suffering from fatigue and weakness for last 15 years and dry cough and worsening shortness of breath on exertion for last 2 years. With these symptoms, she visited a number of physicians including ophthalmologists and was managed symptomatically without any improvement.

Figure 3: Positive Schirmer test showing 1 mm wet on filter paper after 5 minutes.

Further investigations revealed: ANA was strongly positive on Hep2 cell with fine speckled pattern. Anti-dS DNA, lupus anti-coagulant, anti-cardiolipin antibody was negative. ENA profile showed positive anti SSA(Ro) and anti SSB(La). RA test was positive in high titer, Serum total protein was 77 gm/L, albumin 35 gm/L and globulin 42 gm/L, albumin globulin ratio was 1: 1.42. Anti TPO was > 1300 U/ml (positive >60 U/ml).C4 level was reduced to 0.1 g/L (normal 0.15-0.45 g/L). Protein Electrophoresis showed polyclonal hypergammaglobulinemia with prominent IgG and IgM. Pulmonary function tests showed mild restrictive defect as evidenced by low FEV1(2.02 L) and low FVC (2.05L) with normal FEV1/FVC (94.5%). DLCO was moderately reduced with 55% of predicted value. KCO was normal suggestive of parenchymal restrictive disease. Patient refused for lip biopsy. So final diagnosis of recurrent hypokalemic paralysis due to distal (type 1) renal tubular acidosis secondary to primary Sjogren's syndrome with type 3 cryoglobulinemic vasculitis with autoimmune hypothyroidism was made. She was discharged with sodium bicarbonate, potassium citrate, hydroxychloroquine (400 mg daily) and artificial tear. After 1 month, on outpatient door follow up she was doing well.

Discussion

Approximately, 98% of total body potassium is located intracellularly. Sixty percent of intracellular potassium is within skeletal muscle, which may explain the predominance of muscular symptoms in disorders producing hypokalemia [10,11]. Although, the effects of hypokalemia are multisystemic, potassium deficiency most seriously affects the neuromuscular system, and symptoms related to hypokalemia are typically muscular. Serum potassium concentrations of 3.0 to 3.5 mmol/L may be associated with mild muscle weakness and myalgia. Serum potassium concentrations of 2,5 to 3.0 mmol/L are associated with the development of clinically significant muscle weakness. The muscle weakness generally is limited to the limbs and limb girdles. The respiratory or cranial musculature are rarely involved and characteristically spared in muscle weakness secondary to hypokalemia. When severe, hypokalemia can impair respiratory function leading to hypoventilation, and acute respiratory failure [6-9,12,13]. Hypokalemic muscle weakness may simulate Guillain -Barre syndrome [14-16]. When the serum potassium level falls below 2.5 mmol/L, rhabdomyolysis may occur [11,17,18]. The lowest serum potassium level reported was 1.1 mmol. /L this was associated with areflexic quadriplegia, coma, and acute respiratory failure managed with IPPV [12].

Hypokalemia is a common clinical problem and is the most frequent electrolyte disorder encountered in clinical practice. The major causes of hypokalemia are: Inadequate potassium intake, excessive skin, renal (diuretics, adrenal insufficiency, salt losing nephropathy), or gastrointestinal losses (diarrhoea, vomiting), and increased entry into cells by a variety of mechanisms including increased availability of insulin, elevated beta-adrenergic activity and elevation in extracellular pH (metabolic or respiratory alkalosis). RTA is characterized by hyperchloremic metabolic acidosis with a normal serum anion gap. There are multiple forms of RTA, depending on which aspect of renal acid handling has been affected (Table 1).

The presence of severe hypokalemia (K+ 1.7 mmol/L) with normal anion gap metabolic acidosis and urinary pH >5.5 suggested the diagnosis of Type 1 dRTA. Although quadriplegia is a well-known complication of severe hypokalemia, acute respiratory failure due to dRTA is rarely reported [19]. A thorough search of the available literature (Medline) revealed only six cases of dRTA leading to hypokalemic respiratory failure requiring mechanical

ventilation [20-25]. Paralysis of the extremities suggested Guillain-Barré syndrome, however the presence of hypokalemia, metabolic acidosis and an alkaline urine confirmed the diagnosis, and lumbar puncture was not performed. Moreover, there was a significant improvement in muscle power after K+ supplementation.

The pathogenesis of classic hypokalemic dRTA is not yet clear. The hallmark is the inability to acidify the urine to pH <5.5; there is no impairment in bicarbonate reabsorption in proximal tubules [5]. In the majority of patients, there is a defect in H+ secretion by the H+-K+-ATPase pump in tubular cells which causes hypokalemia, decrease in NH4+ excretion, hyperchloremic metabolic acidosis and volume depletion. Occasionally, strikingly severe hypokalemia, metabolic acidosis and hypocalcemia may require immediate therapy. This constellation of findings in an emergency setting has been labeled as a "crisis of dRTA" [22]. Clinical presentation in our patient was also quite catastrophic, with quadriparesis, acute respiratory failure which could have proved fatal without prompt and effective management. Chronic positive acid balance causes Ca2+, Mg2+ and PO43- wasting and there is an increased incidence of nephrocalcinosis. A prominent feature of dRTA is abnormal Ca2+ metabolism leading to musculoskeletal complaints. Hypercalciuria, alkaline urine and low levels of urinary citrate result in calcium phosphate stones [26].

The majority of patients with dRTA have it in association with a systemic illness such as Sjögren's syndrome, hypergammaglobulinemia, chronic active hepatitis (CAH) or lupus. The frequency of dRTA in Sjögren's syndrome has been reported to be 25 to 40% [26]. Ohtani et al. and Poux et al. have described hypokalemic quadriplegia and respiratory arrest due to dRTA in patients with primary Sjögren's syndrome [22,23]. Koul and Saleem reported a case of CAH with dRTA which presented with hypokalemic muscular paralysis requiring respiratory assistance [37]. Sjogren's syndrome is an autoimmune condition characterized by abnormal lymphocytic infiltration to exocrine glands like salivary and lacrimal glands caused by autoantigens Ro/SSA and La/SSB resulting in sicca syndrome. [27-29] It may occur primarily or secondary to other inflammatory conditions like rheumatoid arthritis. Clinical manifestations are not limited to dry eyes or dry mouth only. Non exocrine organs are affected in varying degree ranging from 30- 40% [30].

The diagnosis of Sjögren's presents a challenge to clinicians, particularly when the initial presentation differs from the exocrine manifestation of dry eyes and mouth. The American-European Consensus Classification Criteria has recently revised classification criteria for diagnosis of Sjogren's syndrome that requires four of six criteria, including: ocular or oral symptoms, objective ocular or oral signs, histopathology from a lip biopsy and the presence of autoantibodies [31]. Our patient had subjective symptoms of sicca syndrome and also had evidence of severe dry eye on schirmer's test. She was positive for autoantibodies too. Thus, she fulfilled the criteria of Sjogren's syndrome. One important feature of our case is coexistence of autoimmune thyroid disease and pSS. This association has been reported in the literature often ranging between 10 to 30% [32-34]. A recent meta-analysis also found increased risk of thyroid disorder in patients with Pss [35]. Whether they should be considered as manifestations of same pathophysiologic mechanism or poly autoimmunity is not settled yet. Both pSS and AITD (auto immune thyroid disease) are linked with increased risk of development of lymphoma. [36] Thus, this association might significantly influence long term morbidity or outcome in such patients.

The diagnostic criteria of Sjogren's syndrome were satisfied by the findings of xerostomia, high titer of anti SSA and anti SSB antibodies. The frequency of dRTA in Sjogren's syndrome has been reported to be about 25-40%. There are many case reports of Sjogren's syndrome associated with hypokalemic quadriplegia. Tsuboi et al. described that the periodic paralysis was observed in almost 40% of Sjogren syndrome cases associated with dRTA [37]. However only three cases were reported that respiratory arrest depends on severe hypokalemia associated with renal tubular acidosis due to various causes [36-38] and there is only one case report describing respiratory failure due to Sjogren's syndrome [38]. There is no significant difference at the level of serum potassium between respiratory arrest group and quadriplegia alone group (1.0-2.8 mmol/L). These results suggests that the progress for respiratory arrest may be influenced by interindividual differences in the sensitivity towards hypokalemia (and presumably also transmembrane K gradient) of respiratory muscles. Even patients with hypokalemia <1.7 mEq/l were liable to develop respiratory arrest. The above cases, including our own, were all female, and showed good prognosis with immediate respiratory support and potassium supplementation. However, Nimmannit et al. [39] described fatal cases of hypokalemic respiratory failure and ventricular fibrillation due to endemic RTA in Thailand. This condition occurred in otherwise healthy young males and sometimes resulted in nocturnal death which is different from the present case. Treatment of dRTA involves correction of metabolic acidosis by administration of alkali in an amount sufficient to neutralize the production of diet-derived acids. This is usually equal to 1-3 mmol/kg/. day of sodium bicarbonate in adults. In children, a large amount of bicarbonate must be administered to correct acidosis and maintain normal growth. Sustained correction of metabolic acidosis usually suppresses renal Na+ and K+ wasting with attendant correction of extracellular fluid volume and hypokalemia. Therefore, in most patients with dRTA, K+ supplementation is not necessary. However, if the patient presents with hypokalemic respiratory paralysis, the K+ deficit should be replaced without delay before initiating bicarbonate therapy [26], because if bicarbonate is given alone, it may further worsen the already existing hypokalemia. The rate of infusion of KCl should be up to a maximum of 0.25 mmol/kg/hr. These patients are often volume depleted and treatment should include correction of the fluid deficit if necessary guided by central venous pressure monitoring.

Management of pSS is tailored to individual patients' symptoms. Our patient presented with severe hypokalemia which was managed with IV and oral supplementation. Muscarinic agonists are used to stimulate residual salivary gland function and artificial

tear drops to alleviate dry eyes. [36] In case of marked systemic features corticosteroids, hydroxychloroquine and methotrexate are used. Biologics like Rituximab is reserved for disabling systemic symptoms despite use of DMARDs. [40] At present there is no cure. Our patient responded well to potassium supplementation, hydroxychloroquine, and other lifestyle measures. Over the last 3 months follow up she did not have any further attack of severe hypokalemia. So far, she has not required any systemic agents.

Conclusion

This case report highlights the fact that the dRTA with severe hypokalemia should be kept in mind in any patient presenting with respiratory failure. Secondly, all patients with dRTA must be investigated for any associated systemic illnesses and put on lifelong alkalinizing agents. In pSS, RTA is commonly asymptomatic. However, if such a state persists, patient may develop quadriplegia. Furthermore, if adequate treatment is not received, muscle paralysis may progress to respiratory arrest. Although respiratory arrest associated with Sjogren's syndrome is very rare, this complication is very severe and can be fatal. It is important to pay attention to the occurrence of severe hypokalemia with metabolic acidosis and provide adequate treatment for this combination in patient with pSS.

Conflict of interest

None declared.

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None.

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