AN INTERESTING CASE OF HYPOKALEMIC PARALYSIS

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Abstract:
Hypokalemia is usually asymptomatic or may manifest as weakness, fatigue or abdominal distension. It may also present as cardiac arrhythmias, quadriplegia or respiratory paralysis. We report a 34 year old female who presented with acute onset progressive flaccid quadriplegia with respiratory paralysis. The laboratory tests revealed a severe hypokalemia with hyperchloremic metabolic acidosis and abnormally acidified urine. Here urine anion gap was positive, which in the presence of acidosis led to the diagnosis of distal renal tubular acidosis. The patient fully recovered after potassium and alkali replacement. Further investigations revealed Sjogren's syndrome as the underlying cause.

Keyword: Hypokalemia, Distal renal tubular acidosis, Sjogren's syndrome

CASE SUMMARY
A 34 year old woman presented to the emergency department with acute onset rapidly progressive quadriplegia. She had also developed neck muscle weakness, difficulty in speaking, swallowing and breathing difficulty. There was no history to suggest sensory and visual disturbances. There was no history of recent fever, vomiting, diarrhoea or respiratory tract infection. She denied diuretic, laxative or any other drug abuse. There were no recent vaccinations. She had never experienced a similar event in the past and there was no significant family history. She was not a smoker or alcoholic. She had normal menstrual cycles. On examination patient was drowsy, dyspnoeic and unable to swallow or speak. She was hemodynamically stable and body temperature was 36.7°C, the pulse rate - 72 beats per minute with 4 or 5 missed beats, the recumbent blood pressure – 112/74 mmHg and the respiratory rate - 28 per minute. There were no abnormal signs observed in the lungs, heart and abdomen. Except for a sluggish gag reflex, cranial nerve examination was unremarkable. Motor system examination demonstrated a power of in both the upper and lower extremities with reduced tone and neck muscle weakness. Deep tendon reflexes were symmetrically hyporeflexic. Plantar response was bilaterally flexor. Sensory system was normal. Laboratory investigations were as follows: haemoglobin – 8 g/dl, leucocyte count - 5700 cells/mm³,
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eythrocyte sedimentation rate – 46 mm for the first hour, blood sugar - 82 mg/dl, bilirubin - 0.9mg/dl, AST – 30 IU/L, ALT – 23 IU/L, magnesium – 2.6 mg/dl, calcium - 9.37 mg/dl, serum sodium 130 mEq/L, serum potassium -1.0 mEq/L, creatinine phosphokinase – 19 U/L. Blood gas analysis revealed:pH – 7.110, pCO₂ - 44.7 mm Hg, sodium – 137 mEq/L, potassium – 1.0mEq/L, bicarbonate – 11.2 mEq/L, chloride – 114 mEq/L, plasma anion gap – 10.8. Urine examination showed: specific gravity – 1.012, occasional epithelial cells, sodium – 100 mEq/L, potassium – 31 mEq/L, chloride – 105 mEq/L. Urine anion gap was positive (Urinary Na⁺ + K⁺ - Cl⁻= 26 mEq/L). Serial early morning urine pH was more than 5.5 (Day1 – 7.5; Day2 – 7.2; Day3 – 7.0). No organisms were grown in urine culture. Electrocardiogram showed a few premature ventricular ectopics and U waves. Chest radiograph and ultrasonogram of abdomen and pelvis was normal. Urine and blood culture showed no growth. Based on hypokalemia, hyperchloremic normal anion gap metabolic acidosis, renal potassium loss and normal blood sugar a diagnosis of Renal Tubular Acidosis was made. With normal plasma anion gap, positive urine anion gap, serial urine pH >5.5, in the absence of diarrhoea and urinary tract infection the patient was diagnosed as a case of Distal Renal Tubular Acidosis. She was treated with intravenous potassium chloride 20 mEq over 2 hours after which her respiration improved and the patient was able to swallow and talk. After serum potassium normalized, she regained her power and was placed on oral potassium citrate. Since there was no similar family history and her audiogram was normal primary distal renal tubular acidosis was ruled out. On probing history for secondary causes of distal renal tubular acidosis patient denied photosensitivity, myalgia, arthralgia, dryness of mouth or eyes, decreased tears, jaundice or symptoms suggestive of hypo or hyperthyroidism. There was no parotid or thyroid swelling. Free T4 was 1.08 ng/dl (Normal – 0.70 to 1.80 ng/dl) and TSH was 1.77micro IU/ml (Normal – 0.30 to 5.5 micro IU/ml). Schirmer’s test was more than 10 mm in both eyes. Anti-nuclear antibody was positive with 1 in 300 dilution with speckled pattern. Antibodies to SS-A and SS-B were positive. With the lip biopsy showing minor salivary glandular tissue with focal infiltration by lymphoid cells a diagnosis of Sjögren’s syndrome was made. Patient was started on oral administration of prednisolone (30 mg/day) which was tapered gradually. She is being currently maintained on potassium citrate therapy. She is on regular follow up and has not developed systemic signs of Sjögren’s syndrome.

DISCUSSION

Sjögren’s syndrome is an autoimmune disorder belonging to the group of the chronic systemic rheumatic diseases¹. This disease is predominantly seen in middle aged women (M:F = 1:9)². The principal target organs are the exocrine glands, especially the lacrimal and salivary glands, resulting in symptoms that include dry eyes and dry mouth, which together are called the ‘sicca complex’¹. However, several nonexocrine organ systems may also be involved, including skin, lung, gastrointestinal tract, central and peripheral nervous system, muscular skeletal apparatus, and the kidney³. Sjögren’s syndrome may occur alone (primary Sjögren's syndrome), or in association with a variety of connective tissue diseases as well as with autoimmune disorders (secondary Sjögren's syndrome)¹, ³. Diagnosis of primary Sjögren’s syndrome according to the current American European consensus group criteria requires at
least four of the following six items: subjective xerophthalmia, subjective xerostomia, objective test for xerophthalmia, objective evidence of salivary gland dysfunction, presence of either anti–Ro/SSA or anti–La/SSB antibodies and histopathologic criteria for primary Sjögren’s syndrome on minor salivary gland biopsy. One of the four criteria must be either positive serology or positive histopathology. As in our case who presented with extraglandular involvement without sicca symptoms, a diagnosis of primary Sjögren’s syndrome is possible if both a positive serologic test and histologic criteria are met. In renal involvement both tubular and glomerular damage have been described in Sjögren’s syndrome. It is generally accepted that glomerular disease is rare and when it occurs, it is often associated with mixed cryoglobulinemia. The major histological finding in patients with Sjögren’s syndrome and renal disease is a lymphocytic and plasma cell infiltration of the renal interstitium. Interstitial nephritis results in latent or overt tubular disease, which comprises the major renal involvement in Sjögren’s syndrome. It may manifest itself as distal renal tubular acidosis, nephrocalcinosis, nephrogenic diabetes insipidus or even Fanconi syndrome may occur. Several mechanisms might explain the distal renal tubular acidosis found commonly in Sjögren's syndrome, including the absence of H^+ATPase in the intercalated cells of renal tubules or a permeability defect in the collecting tubule resulting in bicarbonate leak. Renal involvement in Sjögren's syndrome may be frequently latent. Clinically evident renal disease is rare in patients with primary Sjögren’s syndrome and that the presence of subclinical renal dysfunction, usually ascribed to tubulointerstitial nephritis of variable degrees, may be detected by means of appropriate tests. Renal disease may precede the onset of the subjective sicca syndrome, considered to be the classical manifestation of Sjögren's syndrome. There are numerous case reports of primary Sjögren’s syndrome presenting as hypokalemic paralysis showing that overt distal renal tubular acidosis can precede the development of sicca complex symptoms as it did in our patient. Involvement of respiratory muscles due to hypokalemia is unusual although a few cases have been reported so far. It is not clear why some patients with hypokalemia develop respiratory paralysis. In one study no difference was noted in serum potassium levels between patients with and without respiratory arrest suggesting, inter individual difference in sensitivity to hypokalemia of respiratory muscle.

Initial therapy for distal renal tubular acidosis always should focus on treating life threatening hypokalemia and correction of acidosis should begin only after the potassium level is normal. Rapid alkalization using sodium bicarbonate alone aggravates hypokalaemia, because serum potassium shifts into the cellular fluid by exchange of H^+ and K^+. Severe, symptomatic hypokalemia not only demands aggressive intravenous replacement but also close cardiovascular and laboratory monitoring. Potassium chloride salt is the preferred formulation for intravenous administration, up to 20 mEq/h. The most important part of maintenance therapy is alkali replacement, usually 1 to 3 mEq/kg per day. A number of different forms of alkali replacement are available including sodium bicarbonate, sodium citrate solution and potassium citrate solution. Replacement of alkali results in normalization of potassium and other electrolyte abnormalities in most patients.
Prednisone is not a first line treatment of distal renal tubular acidosis secondary to Sjögren's syndrome, although it has been used in several patients with some benefit. If normalization of pH and electrolytes is achieved with alkali replacement and no progression of renal insufficiency or systemic features occurs, additional benefit from glucocorticoid therapy is unlikely and patient may be maintained on potassium citrate therapy alone with good response.

**CONCLUSION**

In primary Sjögren’s syndrome, renal tubular acidosis commonly is asymptomatic. However, if such a state persists, patients will develop quadriplegia. Furthermore, if adequate treatment is not received, muscle paralysis may progress to respiratory arrest. Although respiratory arrest associated with Sjögren’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 hypokalaemia with metabolic acidosis and provide adequate treatment for this combination in patients with Sjögren’s syndrome.

**REFERENCES:**


