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Hypokalemic paralysis as a clinical presentation of Sjögren's syndrome: an Interesting Case

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ABSTRACT
Sjögren's syndrome is an autoimmune condition with glandular and extra glandular involvement.Most common presentation of renal involvement is distal renal tubular acidosis.Hypokalaemia is most common electrolyte abnormality in distal renal tubular acidosis.Severe hypokalaemia and paralysis secondary to distal renal acidosis are unusual initial manifestation of Sjögren's syndrome with no sicca symptom. Here, we describe a case of young female admitted in casualty with severe hypokalemic paralysis and diagnosed as Sjögren's syndrome.

I. INTRODUCTION

Sjögren's syndrome is an autoimmune condition with glandular (lacrimal and salivary) typically manifesting as dry eyes and mouth and extra glandular involvement.However, extra glandular organs such as the kidneys are often affected in these patients. Most common presentation of renal involvement is distal renal tubular acidosis.Histologically, the disease is characterised by lymphocytic infiltration with tissue damage in affected organs.

Distal renal tubular acidosis is known cause of hypokalaemia, which may sometime present as hypokalaemic paralysis. Severe hypokalaemia and paralysis secondary to distal renal acidosis are unusual initial manifestation of Sjögren's syndrome with no sicca symptom.

II. CASE REPORT

A 38-year female presented in casualty with complain of acute onset, progressive, painless quadriparesis for 2 days. Quadriparesis involving both upper and lower limb simultaneously. There is no history of fever, trauma, headache, visual abnormality, diarrhea, bowel bladder involvement. There is no history of renal stones. There is no history of similar episode in past.

Patient is known case of hypothyroidism for 10 years and on eltroxin 100µg.

She had normal menstrual cycle and two pregnancies in past which are uneventful.

On examination patient is normotensive, normal pulse and respiratory rate. Reflexes are diminished, flaccid quadriparesis without sensory and bowel bladder involvement. There is no involvement of cranial nerves. Her plantar is bilateral flexor. On ECG U wave is present. On investigation

Normal anion gap metabolic acidosis (PH-7.19, $Hco3^{--}$-18.6mEq/l., Na^{+}-140mEq/l k^{+} 1.3mEq/l cl^{-}-108mEq/l Anion gap-14.7) with alkaline urinary PH and positive anion gap(PH-7.3, $Hco3^{--}$-18.6mEq/l., Na^{+}-101.2mEq/l k^{+} 23.49mEq/l cl^{--}62.2mEq/l urinary Anion gap-62.49). Urinary potassium also suggestive of renal cause. Her renal function test is normal (Blood urea nitrogen-15mg/dl, serum creatinine- 0.9 mg/dl). on USG KUB, her kidney size is normal, and no nephrocalcinosis or stone is seen. Her serum magnesium(2.76mg/dl), serum calcium(8.6mg/dl), serum Vit D(45ng/ml) is normal. Her thyroid profile is normal (T_3-1.66 ng/dl, T_4-117.42µg/dl, TSH-1.3mIU/ml). On basis of normal anion gap metabolic acidosis, hypokalemia, positive urinary gap, alkaline urinary PH, Distal renal tubular acidosis is made.Autoimmune disease screening showed positive ANA, negative Anti-dsDNA, Anti-smith, Anti-Jo, Anti-SCL70, Anti-centromere, U_1RNP and strongly positive Anti-Ro (SS-A) 88.7 U/ml (N < 3.0), and Anti-LA (SS-B) 88.4 U/ml (N < 3.0) suggestive of possible Sjögren's syndrome. Serology for Hepatitis B surface antigen (HBsAg), anti-Hepatitis C virus (HCV) and Human immunodeficiency virus (HIV) antibody were negative. Schirmer's test was normal suggesting normal tear secretion. Her ECHO and PFT is normal study Lip biopsy were done which showed lymphocytic infiltration commensurate with chronic sialadenitis (focus score of 3).Diagnosis of Sjogren's syndrome was made based on the ACR-EULAR 2016 Classification Criteria for Sjögren's (for diagnosis total score ≥4) wherein labial salivary gland with focal lymphocytic sialadenitis

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and focus score of ≥ 1 foci/4mm^2(score-3) and anti SS-A/Ro positive(score-3). A diagnosis of Primary Sjögren's syndrome (extra glandular-distal RTA) is made. A Hypokalaemia and metabolic acidosis were treated with intravenous potassium chloride followed by oral potassium citrate. Her muscle weakness gradually improving within 3 days and U waves also disappear within 1 days after initiation of potassium supplements. Normalisation of serum potassium occur within 2 days and normalisation of metabolic acidosis occur within 6 days. Then, we initiated prednisolone (1 mg/kg/day) for 4 weeks after which it was plan for taper off in follow-up to minimum of 5 mg daily when remission achieved.

Dexa scan is also planned in follow-up after 3 months The patient was discharged and we followed her up in our clinic every month. She was reported to be asymptomatic and normokalaemia with normal PH in blood investigation with the use of potassium citrate and tapering does of prednisolone.

III. DISCUSSION

Sjögren's syndrome is an autoimmune condition which typically involves lymphocytic infiltration of the salivary, parotid and lacrimal glands, resulting in the characteristic symptoms of xerosis (dry eyes) and xerostomia (dry mouth)[1]. This immune process can also affect non-exocrine organs, such as the skin, lungs, gastrointestinal tract and the kidneys.Diagnosis of Sjogren's syndrome was made based on the ACR-EULAR 2016 Classification Criteria for Sjögren's (for diagnosis total score ≥4)[2].

Tubulointerstitial nephritis (TIN) is the main renal involvement associated with primary Sjogren syndrome. TIN can manifest as distal renal tubular acidosis (RTA), proximal tubular dysfunction, nephrogenic diabetes insipidus and others, of which RTA is the main clinical presentation[3]. Distal renal tubular acidosis prevalence fluctuates between 5 and 70% in various studies[4,5,6].

The pathogenesis of distal renal tubular acidosis in Sjögren's syndrome is not very clear. Patients with Sjögren's syndrome with distal renal tubular acidosis have interstitial nephritis with high levels of anti-carbonic anhydrase antibodies which affect the function of carbonic anhydrase in cortical collecting ducts. The acidification defect was the result of a lack of intact H+-ATPase pumps in the intercalated cells[7].

Hypokalaemia is most common electrolyte abnormality in distal renal tubular acidosis. When hypokalaemia is severe, it presents with flaccid paralysis with diminished reflexes with no involvement of cranial nerve, bowel and bladder involvement. on ECG U wave is sometime present.

On basis of normal anion gap metabolic acidosis, hypokalaemia, positive urinary gap, alkaline urine made a diagnosis of distal renal tubular acidosis.Autoimmune disease screening showed positive ANA, negative Anti-dsDNA, Anti-smith, Anti-Jo, Anti-SCL70, Anti-centromere, U_1RNP and strongly positive Anti-Ro/la suggestive of possible Sjögren's syndrome. Serology for Hepatitis B surface antigen (HBsAg), anti-Hepatitis C virus (HCV) and Human immunodeficiency virus (HIV) antibody were negative. Schirmer's test was normal suggesting normal tear secretion. Her ECHO and PFT is normal study Lip biopsy were done which showed lymphocytic infiltration commensurate with chronic sialadenitis (focus score of 3).Diagnosis of Sjogren's syndrome was made based on the ACR-EULAR 2016 Classification Criteria for Sjögren's (for diagnosis total score ≥4) wherein labial salivary gland with focal lymphocytic sialadenitis and focus score of ≥ 1 foci/4mm^2(score-3) and anti SS-A/Ro positive(score-3) (total-6) in our case.

Daily life long alkali replacement in a dose of 1–2 mEq/kg prevents acute hypokalaemia as well as the chronic complications of osteomalacia and nephrolithiasis. Use of corticosteroids or other immunosuppressive drugs used for the primary disease have been shown to reverse the renal tubular defect, as seen in our patient. We used a course of steroid in this case although the distal renal tubular acidosis was the only manifestation in this patient and she showed a good response to this treatment.

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