

A Case of Sjogren's syndrome Presenting as Distal Renal Tubular Acidosis with Hypokalemic Periodic Paralysis

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Abstract: Soren's syndrome (SS) is a chronic slowly progressive autoimmune disease in which there is lymphocytic infiltration of exocrine glands. It may present alone that is Primary Sjogren Syndrome or in association with other autoimmune diseases (Secondary Sjogren Syndrome). Renal involvement causing tubulointerstitial nephritis and distal renal tubular acidosis (RTA) is well known in Sjogren's Syndrome. However, periodic paralysis due to hypokalaemia secondary to distal RTA in primary SS is rarely seen. We report the case of a female who presented with sudden onset flaccid quadriplegia and was found to have hypokalaemia. On further evaluation the diagnosis of Sjogren's Syndrome presenting as distal RTA was made.

Keywords: Hypokalaemia, Periodic Paralysis, Distal RTA, Sjogren's Syndrome.

INTRODUCTION

Sjogren's Syndrome is an autoimmune disease presenting in primary or secondary forms. The clinical hallmarks are keratoconjunctivitis sicca and xerostomia. This immune process can also involve non-exocrine organs including kidneys, lungs, musculoskeletal system and skin. Renal involvement causing tubulointerstitial nephritis and distal renal tubular acidosis (RTA) is well known in Sjogren's Syndrome [1]. However, distal RTA causing hypokalemic paralysis in primary SS is rare [2]. We report a case of SS presenting as distal RTA with hypokalemic periodic paralysis.

CASE REPORT

A female, 48 years old, housewife, without any known comorbidities was referred to emergency department with chief complaints of vomiting and loose motions for four days and weakness of all four limbs for two days. There were three episodes of vomiting per day preceded by nausea and contained food particles. They were non-projectile, non-bilious and non-blood stained. She had loose motions for four days, 4 episodes per day. The motions were large voluminous, watery in nature, yellowish in colour, non-foul smelling, non-blood stained, and not associated with abdominal pain. Weakness of all limbs appeared two days after onset of diarrhoea and vomiting. The weakness was insidious in onset and progressed from lower limbs to upper limbs in one day. There was no history of any sensory complaint, deviation of angle of mouth, snake bite, seizure episode or trauma.

The patient had similar complaints three months prior for which she was admitted and had recovered without any residual weakness. Details were not known. She was treated in a nursing home with IV fluids, antibiotics and was intubated in view of progressive dyspnoea. There was no improvement in two days and therefore she was referred to our hospital for further management.

On examination, she was averagely built and nourished. The endotracheal tube was in situ with T piece and oxygen support and she was maintaining SaO₂ of 94% on 4 litres of Oxygen. She was afebrile. Her pulse and blood pressure were normal, respiratory rate was 28 per minute with shallow breathing. Examination of nervous system revealed a fully conscious patient. There was hypotonia and areflexia of all limbs. Power in all four limbs was grade 2/5 and plantar reflexes were bilateral flexors. Sensory system

examination was normal. There were no localising or lateralising signs and no signs of meningeal irritation.

Investigations showed serum potassium levels of 1.5 mEq/L with normal serum sodium, serum creatinine was 2.2 mg/dL. WBC count was 14000/ μ L with 86% polymorphs. Liver function tests and thyroid function tests were normal. ECG showed U waves with ST segment depression and flattening of t waves in lateral chest leads.

ABG was suggestive of metabolic acidosis with pH of 6.9, bicarbonate-12.4 mEq/L, pCO₂- 63 mmHg, sodium-144 mEq/L, potassium-1.6 mEq/L, chloride-140 mEq/L. Anion gap was 13.8 mEq/L. Ultrasonographic studies of abdomen and pelvis were unremarkable.

In view of the above, a working diagnosis of acute gastroenteritis with acute kidney injury (AKI) with transient flaccid paralysis and hypokalaemia was entertained and the patient was treated with IV fluids, antibiotics, potassium chloride (KCl) infusion, injection sodium bicarbonate and other supportive measures. She improved with this management and was extubated the next day. Her blood counts and serum creatinine became normal. However, her hypokalaemia persisted despite above treatment, necessitating further evaluation. Urine investigations revealed pH 6.5, urinary potassium 18 mmol/day, positive urinary anion gap. These findings were consistent with distal renal tubular acidosis.

History was revisited. She gave history of dryness of mouth and eyes for last 2 and half years. We, therefore, did her tear break up time that was <10 seconds in both the eyes and her Schirmer's test was positive in both the eyes. ANA blot test showed positive SS-A, SS-B and Ro-52. With this, the final diagnosis of Sjogren's Syndrome presenting as Distal Renal Tubular Acidosis with Hypokalemic Periodic Paralysis was made. After recovery, she was discharged on syrup potassium chloride, tab sodium bicarbonate, multivitamins and lubricant eye drops. She was followed up for next 6 months and remained asymptomatic.

DISCUSSION

Sjogren's syndrome (SS) is a chronic slowly progressive autoimmune disease in which there is lymphocytic infiltration of exocrine glands. It may present alone as Primary Sjogren Syndrome or in association with other autoimmune diseases (Secondary Sjogren Syndrome). This immune process can also involve non-exocrine organs, including kidneys, lungs, musculoskeletal system and skin. Reported prevalence of renal involvement in SS has varied widely from 2 to 67 percent[3]. Renal involvement occurs in form of glomerular involvement and tubulointerstitial

disease[4]. Primary Sjogren's syndrome is diagnosed if (1) patient presents with eyes and/or mouth dryness (2) eyes tests disclose keratoconjunctivitis sicca (3) mouth evaluation reveals classic manifestations of syndrome (4) the patient's serum reacts with Ro/SS-A and/or La/SS-B autoantigens[4]. Treatment of dry eyes and dry tongue are done with lubricating eye drops and adequate hydration respectively. Systemic corticosteroids are used for painful debilitating joint disease.

Distal RTA is associated with autoimmune diseases such as primary Sjögren syndrome and systemic lupus erythematosus[5,6]. Distal RTA (type 1 RTA) occurs due to defect in distal acidification of urine in which kidneys are unable to acidify the urine to pH <5.5. So, urinary pH is elevated in the presence of systemic metabolic acidosis. There is impaired hydrogen ion secretion or bicarbonate reabsorption in the distal nephron. Alkaline urine in combination with hypocitraturia and hyperphosphaturia promotes calcium phosphate precipitation leading to nephrocalcinosis and/or kidney stones[7].

Treatment of distal RTA includes alkali replacement of 1-3 mmol/kg per day of bicarbonate in divided doses. The patients, who present later with renal calculi, are advised large fluid intake and sufficient alkali to restore normal acid-base balance, to correct the hypocitraturia, to reduce hypercalciuria and inhibit the formation of new stones.

Hypokalemic paralysis is relatively uncommon but potentially life threatening clinical syndrome. Most cases are due to familial or primary hypokalemic periodic paralysis. Sporadic cases are associated with numerous other conditions including barium poisoning, hyperthyroidism, renal disorders, certain endocrinopathies and gastrointestinal potassium losses. The age of onset, race, family history, medications, and underlying disease state can help in identifying the cause of hypokalemic paralysis. If recognised and treated appropriately, patients recover without any clinical sequelae.

Our patient had second episode of hypokalemic paralysis and on evaluation a diagnosis of distal RTA secondary to Primary Sjogren's Syndrome was made. After starting the treatment, she had no recurrence of hypokalemic paralysis during the period of observation.

This case highlights that acute paralysis is a life-threatening consequence of hypokalaemia. Potassium wasting secondary to distal RTA may be easily prevented. Underline Sjogren's syndrome should be considered in cases presenting with hypokalemic paralysis.

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