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Case Report

Sjogren's Syndrome Presenting with Refractory Hypokalemia and inflammatory peripheral vascular disease: Double Trouble

Rumaisa Ahmed[1*], Prasanna Kumar[2], Tameem Imran[3]

1. Junior Resident, Department Of Medicine, Sri Devaraj Urs Medical College, Sri Devaraj Urs Academy of Higher Education and Research, Kolar
2. Assistant professor, Department Of Medicine, Sri Devaraj Urs Medical College, Sri Devaraj Urs Academy of Higher Education and Research, Kolar
3. Senior Resident, Department Of Medicine, Sri Devaraj Urs Medical College, Sri Devaraj Urs Academy of Higher Education and Research, Kolar

Abstract

Sjogren's syndrome is a slowly progressing autoimmune disease that is characterized by lymphocytic infiltration of the exocrine glands resulting in impaired secretory function. There are varied causes of quadriparesis ranging from cervical cord myelopathy to neuromuscular junction disorders. Even though hypokalemia is a frequent cause of quadriparesis but is usually due to periodic paralysis. Here we present a case in which hypokalemia secondary to Sjogren's syndrome presented with quadriparesis, which progressed, to respiratory paralysis.

Keywords: Sjogren's syndrome, distal renal tubular acidosis, hypokalemia, peripheral vascular disease.

Introduction

Sjogren's syndrome is a slowly progressing autoimmune disease that is characterized by lymphocytic infiltration of the exocrine glands, mainly the lacrimal and salivary glands, resulting in impaired secretory function. Simultaneously, systemic features of cutaneous, respiratory, renal, hepatic, neurologic, and vascular nature often occur. Renal involvement is well-recognized extra glandular manifestation of primary Sjögren's syndrome (pSS). [1] Most common manifestations are related to tubular dysfunction resulting from chronic interstitial nephritis and can manifest as distal renal tubular acidosis (RTA), proximal RTA, tubular proteinuria, and nephrogenic diabetes insipidus.

There are varied causes of quadriparesis ranging from cervical cord myelopathy to neuromuscular junction disorders. Thyrotoxicosis is the most frequent secondary cause of hypokalemic paralysis. [2]

Here we present a case in which hypokalemia secondary to Sjogren's syndrome presented with quadriparesis, which progressed, to respiratory paralysis.

***Corresponding Author**
Dr. Rumaisa Ahmed ,
Final year postgraduate,
Department of General Medicine
Sri Devaraj Urs Medical College,
Sri Devaraj Urs Academy of Higher Education and Research, Tamaka, Kolar-563101
Mobile No : 9482216030
E-mail: arumaisa@ymail.com
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Case History:

40 year old female presented with complaints of inability to move all four limbs which progressed to difficulty in breathing. On Examination, she was pale, she had quadriparesis with hypotonia and hyporeflexia. Patient sustained cardiac arrest following bradycardia and was intubated and revived after 2 cycles of CPCR and shifted to ICU.

Initial evaluation revealed a GRBS of 480 with serum K+ of 1.5 mEq and ECG showed hypokalemic changes. ABG showed high anion gap metabolic acidosis.
IV potassium and IVF was initiated.
1 Pint PRBC was transfused I/V/O severe anemia (HB – 5.4g/dl) – normocytic normochromic anemia
HbA1C (5.8), urine ketones, Lipid profile , PTH levels were normal.
Serum Mg2+(1.9mg/dl), chloride(100mmol/L), homocysteine - 6.3mcmol/L (3.7 - 13.9 mcmol/L) levels were normal.

On day 3, she started developing blackish discoloration of the right limb, 2nd toe of right foot and great

toe tip of left foot. Oedema of right upper limb till elbow and bilateral lower limb till knee present . Allen's test was positive. Doppler was done which showed diffuse atherosclerotic changes with internal luminal thickening noted and flow volume and vessel size irregularities were seen.

Patient was eventually extubated. Serial monitoring of K+ was done which showed refractory hypokalemia. She was on oral and IV K+ correction.
Urine K+ was 16mEq/L suggestive of renal loss, TTKG was 4.
Urine pH was 6.0, urine Na+ - 750 mEq/l, urinary K+ - 16Meq/l, urine chloride- 30 mEq/L, urine Urea- 184mg/dl, U.creatinine- 9mg/dl.
T3 – 0.986, T4 – 4.92, TSH 1.94, Serum Amylase – 68U/l
She further recollected decreased tears, gritty sensation in the eyes and oral secretions since 3 months. Schirmer's Test was 6mm in 5 mins.
ANA PROFILE was done and antibody to SS-A (RO) and SS-B (LA) was positive.
A Diagnosis of Sjögren's Syndrome with Hypokalaemic paralysis secondary to Distal RTA and Inflammatory peripheral vascular disease was made.

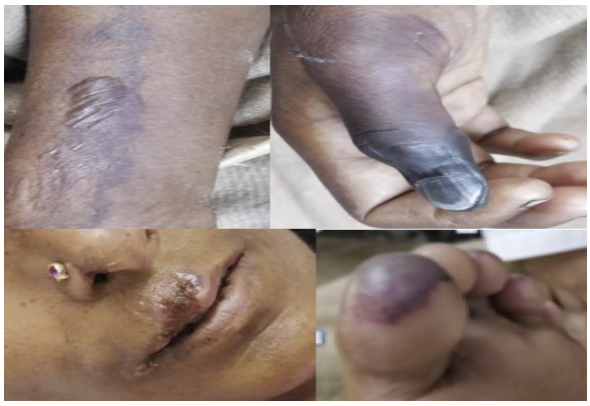

Figure 1 – Blackish discolouration of forearm, right thumb, upper lip and left toe.

Discussion:

Primary Sjogren's syndrome is a disease of exocrine glands presenting with manifestations related to dry eyes and dry mouth. Nonexocrine organ systems may also be involved, including skin, lung, gastrointestinal tract, central and peripheral nervous system, muscular skeletal apparatus, and the kidney. [3]

The reported rate of renal involvement in pSS in literature is variable ranging from 4.2% to 50%. [4] The spectrum of renal disease includes interstitial nephritis, which can manifest as distal RTA, proximal RTA, tubular proteinuria, nephrogenic diabetes insipidus, glomerular diseases, or renal failure. The most common manifestations are related to tubular dysfunction which results from chronic interstitial nephritis.

In our case patient presented with weakness of all four limbs without fever, sensory, bladder and bowel involvement. Due to high blood sugars and hypokalemia with ECG changes and metabolic acidosis, diabetic ketoacidosis was kept in mind and fluid and potassium correction was started. However urine ketones were negative and HbA1C was normal.

Hypokalemia was further evaluated, Urine K+ was 16mEq/L and TTKG was 4 with Non anion gap metabolic acidosis suggesting Distal RTA.
Gangrene of the digits and Doppler study showed diffuse atherosclerosis suggestive of peripheral vascular disease.

Keeping above findings in mind ANA profile was done, which showed Antibody to SSA & SSB to be positive. Ocular sign with ocular and oral symptoms were present. However patient refused salivary gland and renal biopsy. Our patient's score was 4 according to ACR/EULARs classification to diagnose Sjogren's syndrome.
The mechanisms of distal RTA-induced hypokalemia include decreased distal tubular Na delivery, secondary hyperaldosteronism, defective H-K ATPase, and bicarbonaturia. [5]
Hypokalemic paralysis as the initial manifestation of SS is extremely rare.
In a study of dRTA in SS by Chen et al, six out of eight patients (75%) were not diagnosed with SS before dRTA episode. [6]

Patient also had peripheral vascular disease. Peripheral inflammatory vascular disease is seen in patients with sjogrens and they are 2-3 times at greater risk of developing cardiovascular disease.

Conclusions

When a patient presents with hypokalemic paralysis, the notion of labeling it to be a case of periodic paralysis may not always be a correct if other secondary causes are not ruled out. For this, a good history, clinical examination, thorough knowledge of arterial blood gas, and urine analysis are very important so that conditions such as dRTA secondary

to sjogrens are not overlooked or missed. When a diagnosis of dRTA is made a search for an underlying cause including autoimmune diseases, especially SS must be done even in the absence of the Sicca symptoms.

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