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A 48 Year Old Female with Quadriparesis Secondary to Sjögren's Syndrome with Atypical Presentation: A Case Report

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Abstract: Sjögren's Syndrome is a autoimmune disease which is characterized by lymphocytic infiltration of exocrine glands, resulting in symptoms related to impaired exocrine gland, particularly lacrimal and salivary gland function such as xerostomia, keratoconjunctivitis sicca and profound B-cell Hyperactivity. The syndrome has unique features since it presents with a wide clinical spectrum from organ specific to systemic disease. The aim of this paper is to present a case of Sjögren's Syndrome, with atypical presentation as Quadriparesis. A 48 year old Female presented with acute onset, progressive muscle weakness of two day duration. On detailed evaluation, she was found to have low Potassium levels, Positive ANA Profile, Positive Schirmer's Test, Distal Renal Tubular Acidosis. In our case, the patient primarily presented with Quadriparesis due Hypokalemic Periodic Paralysis due to distal RTA. Patient incidentally also had other features of Sjogren's Syndrome including Dry eyes (Positive Schirmer's Test), Xerostomia, Positive Rheumatoid Factor.

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I. INTRODUCTION

Sjögren's Syndrome is a autoimmune disease mainly affecting the exocrine glands by lymphocytic infiltration and B- lymphocyte hyperreactivity. The syndrome has unique features as it presents with wide clinical spectrum from organ specific to systemic disease. Middle aged women with female to male ratio of 10-20:1 is primarily affected.

II. COURSE OF DISEASE

A 48 year old female, presented with weakness of medial 3 fingers of the Left Hand on Monday morning for which she visited a local nursing home, following which Injection Calcium Gluconate was given.

The next day, Tuesday the weakness progressed to Left Forearm and also involved the Right Upper Limb. At around 7am, the patient was able to demonstrate finger movements bilaterally ,with proximal muscle weakness of both Upper Limbs. Following which ,around 9 am the weakness progressed to involve bilateral Lower Limb, and the patient was not able to bear weights or get up from supine position.

Patient was taken again to the Nursing Home, where Potassium correction was started in view of hypokalemia. At around 2 pm, the muscle weakness had progressed to involve all four limbs. The patient was able to move the neck, with complete quadriparesis. The patient could neither stand or walk. Following involvement of Respiratory muscle weakness, the patient began desaturating, maintaining 80% on Room air, and was brought to Hospital with complete quadriparesis with heaviness of tongue.

- *On Examination*

BP- 110/80 mm hg, Pulse- 110 bpm , SpO2 - 96% on 10 Liters of O2, Single Breath Count- 4, Neurological examination revealed Power of Grade 1/5 in all 4 limbs leading to Areflexic flaccid paralysis without sensory involvement . Around 4:15 pm the patient was intubated in view of desaturation due to Respiratory muscle involvement. Serum Potassium Level on admission was 1.63 mEq/L. Potassium correction was started intravenously.

- *Past History*

There was no such similar history of muscle weakness in the past. Patient gives a history of dryness of the mouth . She gives history of Chikungunya in 2019. She complaints of Joint Pain involving small and large joints, characteristically more in the morning, and relieving as the day progresses.

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Patient is a Known case of Hypothyroidism since 2020, currently on Tab Thyronorm 100mcg. She is not a case of Diabetes Mellitus and Hypertension. She denies consumption of Alcohol and smoking. She has had irregular menses every 3-4 months since last 1 year.

Patient gives history of 3 pint PCV Transfusion in 2018 in view of decreased Hemoglobin (details not known). Patient underwent Tubectomy in 1996.

III. INVESTIGATIONS

➢ *Laboratory Investigations were as Follows:*

- Hb: 11.9g/dl, TLC: 19,400/mm^3, Platelet Count 335,000 mm^3, Phosphorus: 3.7 mg/dl,
- TSH: 0.49mIU/L, Magnesium: 2.6 mg/dl, Calcium 8.3 mg/dl, Creatinine Kinase: 183 U/L,
- Albumin: 3.4 g/dl, Globulin: 5.0 g/dl. Potassium – 1.63 mmol/L
- ECG on admission – Prolonged PR Interval, ST segment depression in Lead I,II, V2-V6 suggestive of Hypokalemic changes
- Arterial Blood Gas – pH- 7.286, PO_2- 163.6 mmHg ,PCO_2- 26.6 mmHg, HCO_3 -11.4mmol/L
- ANA Profile revealed Strongly Positive – SS-A, SS-B, RO-52.
- Rheumatoid Factor – Positive
- Schirmer's Test: Positive – Right Eye – 4mm, Left Eye – 3mm
- 2D Echo: Normal Resting LV Systolic Function

✓ No Regional Wall Motion Abnormality
✓ Mitral Annular Calcification with Trivial MR
✓ Aortic Valve thickened with Trivial AR
✓ Trivial TR with PPG - 27 mm Hg
✓ Estimated PA Pressure - 32 mm Hg
✓ Mild PAH
✓ Grade 1 Diastolic Dysfunction

- Urine Sodium : 85 mEq/L
- Urine Potassium: 83.5 mEq/L (>40 mEq/L suggestive of Renal Loss of Potassium)
- Urine Osmolality: 282 mOsm/kg (Reference Range: 500-800)

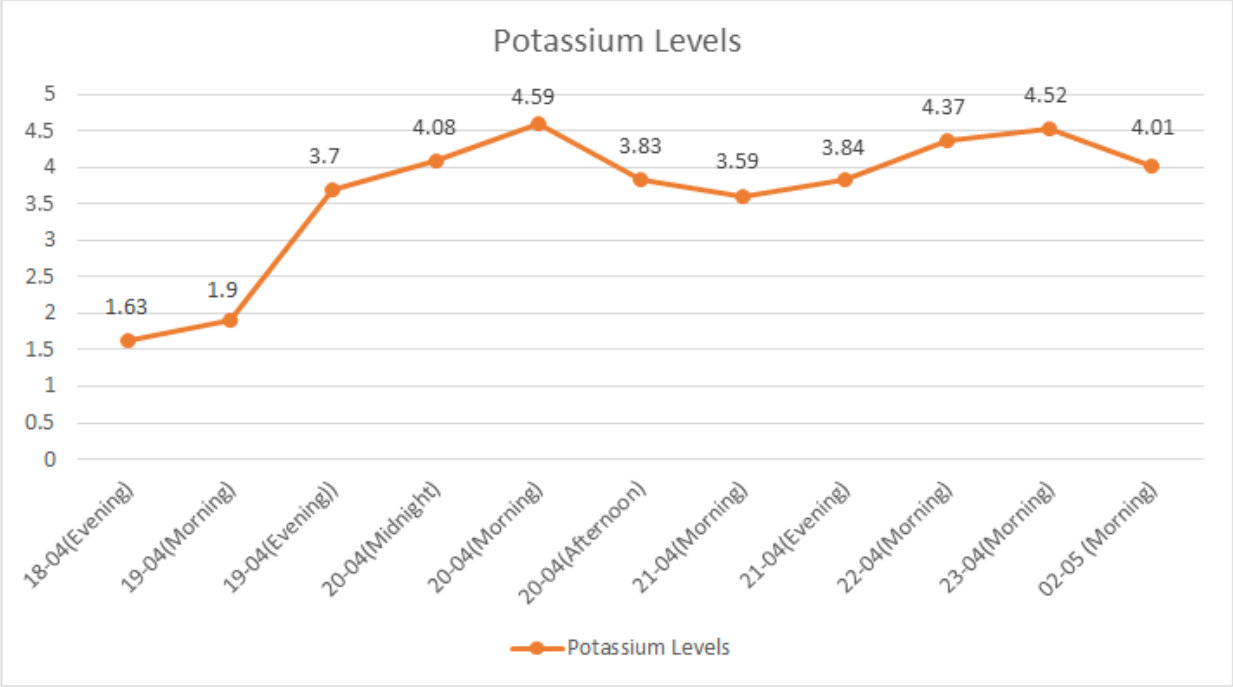

Fig 1 Potassium Levels

IV. DISCUSSION

Sjogren's Syndrome is a rare autoimmune disorder with a prevalence of 0.5-1% which is characterized by lymphocytic infiltration the exocrine glands resulting in Xerostomia and Keratoconjunctivitis sicca. It primarily affects middle-aged women as seen in this case although may occur at any age.

Renal Tubular Acidosis is associated with Sjogren Syndrome. It is associated with Potassium wasting and associated with Metabolic acidosis with hyperchloremia. Patients may present with paralysis due to hypokalemia secondary to Renal Tubular Acidosis.

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Hypokalemic Paralysis is a rare and a potential life threatening condition which may involve respiratory muscles leading to death.

In our case, the patient primarily presented with Quadriparesis. On initial evaluation, Serum Potassium was found to be 1.63, leading to a Preliminary diagnosis of Hypokalemic Periodic Paralysis. On further evaluation, Urine Potassium revealed elevated Potassium excretion (83.5 mEq/L) along with hyperchloremic Metabolic Acidosis (pH:7.2) pointing towards a diagnosis of Distal Renal Tubular Acidosis.

Patient was later screened for Antinuclear Antibodies which revealed strong positivity for SS-A, SS-B, RO-52 and Positive Rheumatoid Factor. Along with this, classical history of dry mouth (xerostomia) and dry eyes (keratoconjunctivitis sicca) proved with Positive Schirmer's Test : Right eye – 4mm and Left Eye – 3mm, female sex, extraglandular involvement in the form of RTA lead to a diagnosis of Sjögren's Syndrome

V. CONCLUSION

Hypokalemic Periodic Paralysis was the presenting manifestation of Sjögren's syndrome in this patient. Although rare, it may precede glandular symptoms. Hence, in all patients presenting as Hypokalemic Periodic Paralysis, patients should be evaluated for Sjogren's Syndrome inspite of lack of sicca symptoms.

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