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SJOGREN'S SYNDROME PRESENTING WITH HYPOKALEMIC PERIODIC PARALYSIS DUE TO DISTAL RENAL TUBULAR ACIDOSIS:- A CASE REPORT

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ABSTRACT

Hypokalemic Periodic Paralysis is one form of Periodic Paralysis, a rare group of disorders that can cause of sudden onset weakness. We Report a Case of a 59 Year Old Female Who Presented With Sudden Onset Flaccid paralysis Predominantly Involving Proximal Upper Limb and Lower Limb. She had difficulty in holding neck and difficulty in opening mouth. The episode occurred 1 day before she presented to us. She gave previous history of Muscular pain, Dryness of mouth and Dryness in the eyes. There was no Significant Sensory, Bladder or Bowel Involvement. At the time of admission she was Found to have Hypokalemia (1.57mmol/L). Potassium supplementation in form of potassium chloride infusion and oral potassium were started. Muscle strength completely recoverd five days later. Diagnosis of Distal Renal Tubular Acidosis was made in view of an alkaline urinary Ph (>5.5) in setting of metabolic acidosis and further Diagnosed as a case of Sjogren 's syndrome.

Key words: Sjogren's Syndrome, Distal Renal Tubular Acidosis, Hypokalemic paralysis.

INTRODUCTION

Sjogren's syndrome is a chronic autoimmune disorder with systemic involvement. The disease most commonly affects the exocrine glands; almost any systemic organ may be affected. The sicca (dryness) component is the most frequent presentation [1]. Rare and more severe presentations include vasculitis, glomerulonephritis, and neurological involvement [2].

A very rare presentation that has reported a few times in literature is distal renal tubular acidosis (RTA) that can occur as a complication of Sjogren's syndrome. Distal RTA that may lead to hypokalemia, which could result in periodic paralysis, is a severe presentation that needs intensive care and management [3].

Hypokalemic periodic paralysis (HPP) usually presents as an acute flaccid paralysis secondary to decreased serum potassium levels. This decrease may be due to the intracellular shift of potassium ions or due to loss of potassium ions [4].

CASE DESCRIPTION

A Case of 59 Year Old Female Who Presented With Sudden Onset Flaccid paralysis Predominantly Involving Proximal Upper Limb and Lower Limb without significant Bulbo- respiratory involvement on admission. The weakness was more prominent in the lower limbs and it was more severe in the proximal muscles of both the lower and upper limbs. She had normal extra occular and facial movements. She had difficulty in holding neck and difficulty in opening mouth. The episode occurred 1 day before she presented to us. She gave previous history of Muscular pain, Dryness of mouth and Dryness in the eyes. There was no Significant Sensory, Bladder or Bowel involvement.

On physical exam, she was afebrile with a pulse rate of 78/min and BP 118/80 mm/Hg. On general inspection the patient was not in any obvious discomfort.

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There were no obvious signs of dehydration. There was no jaundice, lymphadenopathy, thyroid and parotid glands enlargement or clubbing. The patient was not apparently anemic.

On neurological exam, the patient was conscious oriented in time, place and person. The power in the lower and upper limbs was 1-2/5. The weakness was more obvious in the proximal muscles. The tone was normal. The deep tendon reflexes were also normal. There was no sensory loss of any modality. There were no fasciculation or obvious muscle atrophy.

Routine chemistry, liver enzymes and complete blood count were normal except for a potassium level of 1.57mmol/L. ECG showed a normal rate and rhythm but there was flattening & inversion of the T waves. Her serum sodium level was normal (149mmol/L). Further, an arterial blood gas analysis revealed a blood pH of 7.24, signifying acidosis. This decrease in pH, along with an elevated transtubular potassium gradient indicates in the diagnosis of distal RTA, with HPP as its complication. Potassium supplementation in form of potassium chloride infusion and oral potassium were started. Muscle strength completely recovered five days later.

Autoantibody screen revealed positive antinuclear antibody and RF. Anti ds DNA, anti-Sm antibodies and anti U1 Sn RNP were negative. Thyroid function tests and aldosterone level were normal. Anti-Ro (SS-A) and anti-La (SS-B) antibody were strongly positive in very high titer of 34 AU/ml and 82AU/ml respectively.

These reports and distal RTA raised a high index of suspicion of Sjogren's syndrome Presenting with Hypokalemic Periodic Paralysis**.** A salivary gland biopsy was refused by patient. Patient could not afford salivary gland scintigraphy and sialography. The patient was discharged on low dose oral corticosteroids and potassium supplements, following which she has improved symptomatically.

DISCUSSION

Our patient, 59 years old female, presented with sudden onset flaccid paralysis without sensory involvement. A differential Diagnosis of AIDP and HPP was considered. Hypokalemia favoured the second diagnosis which was strengthened by immediate improvement of Muscle strength by potassium supplementation. Further, metabolic acidosis and raised TTKG indicated towards Distal RTA. Distal RTA may be primary or more commonly secondary to paraproteinaemia, medullary sponge kidney, nephrocalcinosis, obstructive uropathy and autoimmune disease [5].

Hypokalemia due to dRTA is usually a late manifestation. However it has been reported in < 2% cases of Sjogren syndrome (SS) as a presenting manifestation [6]. The systemic manifestations necessitate corticosteroids, cytotoxic agent or both.

CONCLUSION

To conclude, we would like to emphasize on the fact that hypokalemia should not just be corrected but also investigated for the cause. Sjogrens syndrome presenting as HPP is very rare, but it should be borne in mind when investigating a case of HPP secondary to distal RTA.

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CONFLICT OF INTEREST

None

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