CASE REPORT

QUADRIPARESIS IN SJOGREN SYNDROME

Nikhil Srivastva[1], Vijay Parashar[2], Praveen Chaturvedi[3], Nilesh Kumar[4]

Author's Affiliations: [1]Junior Doctor, [4]Assistant Professor, Dept of General Medicine; [2]Junior Doctor, Dept. Dental science; [3]Junior Doctor, Dept. of Opthalmology, IMS Banaras Hindu University.
Correspondence: Dr. Nilesh kumar, E-mail: nilesh19arreno@gmail.com

ABSTRACT

Hypokalemic paralysis is a well recognised clinical presentation of Primary sjogren syndrome that occurs due to renal potassium loss caused by interstitial nephritis. However we report a case where a hypokalemic paralysis in a suspected case of sjogren syndrome was associated with high anion gap metabolic acidosis in the presence of a near normal Glomerular filtration rate (RTA) and a failure to acidify urine pH< 5.5 .Such cases represent a minority of distal RTA patients as they characteristically present with an elevated anion gap due to an often unidentified systemic acid load which has lead to precipitation of Renal tubular acidosis and consequent urinary potassium loss and an added metabolic acidosis. The diagnosis often gets delayed due to an elevated anion gap nature of metabolic acidosis but is usually confirmed by hypercalciuria and urine pH > 5.5 in the presence of systemic acidosis**.**

Key words: Sjogren Syndrome, Paresthesia, Hypokalemia

CASE REPORT

A 45 year old female presented to the emergency department with acute onset, rapidly progressive flaccid paralysis of both the upper and the lower limb of 1 day duration .The weakness was bilateral, involving both the proximal and the distal muscles. Patient also had difficulty in moving neck and facial muscles. She denied any pain or paresthesia. Prior to this episode patient was running a low grade fever since last 10 days. There was no history of decreased urine output, diarrhoea , vomitting , visual blurring, altered sensorium, palpitations , shortness of breath, trauma, headache. There was no previous history of Diabetes Mellitus, Hypertension and alcohol intake. Past history was significant for dry mouth and throat since last 12 months for which she was on some non-documented ayurvedic medications with partial benefit. Patient had stopped using these medications during this febrile episode.

On examination Patient was thin built, fully conscious oriented, tachypneic (Respiratory rate-26/min), Blood pressure-110/70 mmHg, Pulse rate-90/min , temperature - Afebrile. Bilateral mild ptosis was present. Oral mucosa was dry. Neurological examination revealed flaccid paralysis of all the limbs. Sensation was intact and Deep Tendon Reflex were slightly diminished (2/4) in all the limbs .Power 0/5 at all joints. There were no enlarged nodes or parotid gland. Other systems were within normal limit. Arterial blood gas analysis showed- low serum K+(1.34) ,metabolic acidosis pH-7.12, Na-149 , Cl-115 ,an elevated anion gap (Anion Gap-23) and a low HCO3 -12.4, pO2-92mmHg and pCO2-38mmHg .Complete blood count showed Hb-12.6, TLC-21,000 N80L16. Amongst renal function Cr-1.5 and urea-44. Other serum chemistry including liver were within normal limit. 3 hours after I.V. potassium supplementation, patients weakness had significantly improved. Patient was also treated with a broad spectrum antibiotic and other conservative measures. Repeat blood count over next few days showed improved white cell count. ANA titere was 5.0(N<1.0) and Anti La-52.8(N<20) . T3, T4 and TSH, serum lactate and magnesium level were normal. 24 hr urinary Ca+ was 435mg(100-300).

Serial ABG analysis during the hospital course showed slowly rising HCO3 (No bicarbonate supplementation was done). A positive Schirmer's test was demonstrated followed by oral mucosal biopsy which revealed inflammation in minor salivary glands.(figure 1). USG abdomen was normal except for cystitis. Renal biopsy was refused by the patient. A provisional diagnosis of

sjogren syndrome with drug induced interstitial nephritis was made. Patient was started on potassium supplementation and artificial saliva substitutes. Two weeks later on follow-up, patient ABG analysis showed complete resolution of acidosis and her HCO3- levels had come to normal. Other electrolytes were also within normal range.

DISCUSSION

Hypokalemic paralysis is an important differential diagnosis in case of an acute onset flaccid paralysis and diagnosis is usually established by demostrating very low plasma K+. EMG and NCV are usually not helpful as it may demonstrate abnormality similar to some variants of GBS.[1] In most cases diagnosis is confirmed by demonstrating marked muscle power improvement within few hours of rapid potassium supplementation.

The etiology of the paralysis is usually evident by a careful history and physical examination. Periodic paralysis except for thyrotoxic must have onset before 25 years of age and thus was excluded in our patient.[2] There was no history suggestive of Gastrointestinal loss, so we concentrated on Renal K+ loss. Other rarer causes like thyrotoxic paralysis and barium poisoning were largely ruled out by a bedside evaluation.

There have been case reports where Sjogren syndrome has presented with hypokalemic paralysis which is due to interstitial nephritis manifesting as renal tubular acidosis.[3] This was considered as the first probable diagnosis in our patient in view of history s/o sjogren syndrome. However, Renal tubular acidosis is chiefly hyperchloremic and thus normal anion gap. Though, hyperchloremia was evident in our patient, it was associated with hypernatremia which more strongly favours loss of hypotonic fluids which in our case was due to interstitial nephritis. Moreover acidosis was non-compensated as expected pCO2 was 27mmHg on ABG analysis as compared to 38mmHg in the patient which presumably reflects respiratory muscle involvement due to hypokalemia.

A second possibility of drug induced interstitial nephritis was considered in view of consistent intake of ayurvedic medications for dry mouth. A dose dependent tubular injury leading to loss of bicarbonate, potassium and fluid (chiefly hypotonic) was considered as the mechanism for causing existing symptoms in our patient. A prior febrile illness precipitated the hypokalemic attack.[4] This was further supported by a slow normalization of body electrolytes (including HCO3-) and acidosis on plain withdrawal of the culprit agent and potassium supplementation. No attempt for bicarbonate supplemenation was done as it would have worsen hypokalemia.

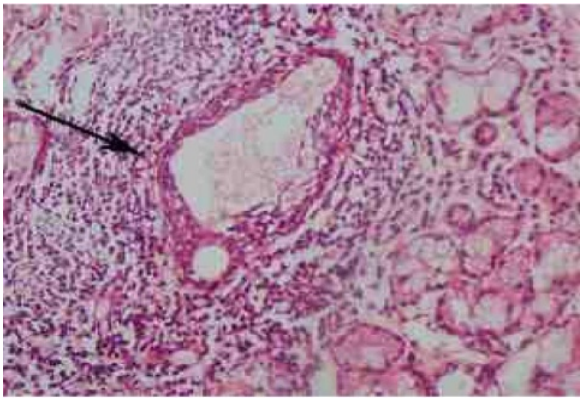

Figure 1: Histopathological Examination of minor salivary glands show Chronic Sialadenitis

REFERENCES

1. Rajshekhar G, Kumar S, Prabhakar S. Reversible electrophysiological abnormalities in hypokalemic periodic paralysis. Indian Pediatr 2008;45:54-5.
2. Kung AW: Clinical review: Thyrotoxic periodic paralysis: a diagnostic challenge. *J Clin Endocrinol Metab* 2006, 91(7):2490-2495
3. Christensen KS. Hypokalemic paralysis in Sjögren's syndrome secondary to renal tubular acidosis.Scand J Rheumatol.1985;14(1):58-60.
4. Ravindra Kumar Garg, Hardeep Sing Malhotra, Rajesh Verma et al. Etiological spectrum of hypokalemic paralysis: A retrospective analysis of 29 patients. Ann Indian Acad Neurol. 2013 Jul-Sep; 16(3): 365–370.