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HYPOKALEMIC QUADRIPLEGIA AS A RARE MANIFESTATION IN SJOGREN'S SYNDROME

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ABSTRACT

Background: Sjogren's Syndrome is an immune system disorder characterized by dry eyesands dry mouth. It can be difficult to diagnose because the signs and symptoms vary from person to person and can be similar to those of other diseases. Here we report one such patient who presented with quadriparesis due to hypokalemic periodic paralysis. **Case Information:** 34 year old lady presented to the ER with insidious onset weakness of all four limbs over 2 days. Serum electrolytes revealed Serum Pottasium levels of 1.9 MEq/dl and Arterial blood gas revealing Normal Anion Gap Metabolic Acidosis suggestive of Renal Tubular Acidosis .type 1. Suspicion of Sjogren's by history of dry mouth and repeated episodes of exposure keratitis was confirmed by lip biopsy suugeestive of Sjogren's Syndrome. **Conclusion:** This report is to highlight the various extra-glandular manifestation of Sjogren's Syndrome with Renal Tubular Acidosis being one among them.

KEYWORDS: case report, sjogren's, renal tubular acidosis, rta type 1, hypokalemia.

INTRODUCTION

Sjogren's syndrome (SS) is a chronic, multisystem autoimmune disease characterized by lacrimal and salivary gland inflammation, resulting in dryness of eyes and mouth and sometimes glandular enlargement. In addition, a variety of extra-glandular manifestations including fatigue, rashes, musculoskeletal involvement, pulmonary, renal, hepatic and neurological manifestation. They can be associated with Rheumatoid arthritis, Systemic Lupus Erythematosus (SLE), or even systemic sclerosis.

Renal prevelance of Sjogren's range from 2-67%. Renal manifestations include Interstitial Nephritis, Distal Renal Tubular Acidosis, Nephrogenic Diabetes Insipidus, glomerular disease like MPGN and Membranous Nephropathy. Here we evaluated a case of Hypokalemic Periodic Paralysis due to Distal Renal Tubular Acidosis diagnosed later as Sjogren's syndrome.

Narrative

Case Report 1: A 34 year old female patient was brought in our emergency department as a case of Quadriparesis of 2 days duration with difficulty in lifting her hand above the head and flexing the hips. She gives history of similar illness in the past. no history of fever, seizures, involuntary micturition or defecation. No sensory deficit noted. History of multiple opthalmological consultations for exposure keratitis are present. No history loose stools or vomiting seen. History of attacks which are triggered by rest after rigorous household works. She also gives history of multiple renal calculi and episodes of renal colic.

On Examination: Patient was conscious, oriented, afebrile, mild dehydration. No pallor, no icterus, no cyanosis, no clubbing, no lymphadenopathy noted.

Systemic Examination
CVS: S1 S2 heard, no murmurs.
RS: NVBS+ No added sounds.

CNS: Bulk of muscles Normal, Tone decreased in all four limbs, Power 2/5 in all four limbs noted. cranial nerve examination normal. no sensory deficits noted. no signs of meningeal irritation. bilateral plantar: No response noted.

P/A: soft, BS+, Non tender, no organ enlargement noted.

CBC, RFT, LFT investigations were within normal limits. Serum electrolytes revealed $S.K^{+}$ value of 1.9

mEq/L and serum Calcium level of 5.0 mg/dl.

Urine routine Investigations revealed Urine pH of 7.0, Urine Sodium 60mEq/L, Urine Anion Gap 35 mEq/L, Urine osmolal gap <150mosm/kg, Urine Calcium levels high.

Viral markers Negative, COVID RT-PCR Negative.

ABG: Normal anionic gap Metabolic Acidosis with bicarbonate levels of 15mEq/L. CSF analysis done revealing no abnormality.

CT Brain: No abnormality detected.

MRI brain with whole spine screening +MRA and MRV: No abnormality noted.

USG KUB showed multiple renal calculi in bilateral kidneys with largest measuring 6*8 mm.

With the above investigations, our diagnosis was narrowed down to Renal Tubular Acidosis- Distal type (Type 1).

Treatment was started with IV bicarbonate therapy (~120mEq per day) and Iv pottasium chloride which drastically improved the symptoms.

Further evaluation of the cause of RTA and history of dry eyes pointed towards the possible diagnosis of Sjogren's Syndrome.

Opthalmology consulatation sought revealing Schirmers test positivity in both eyes. Immunological workup showed strong positive ANA of 1:640.

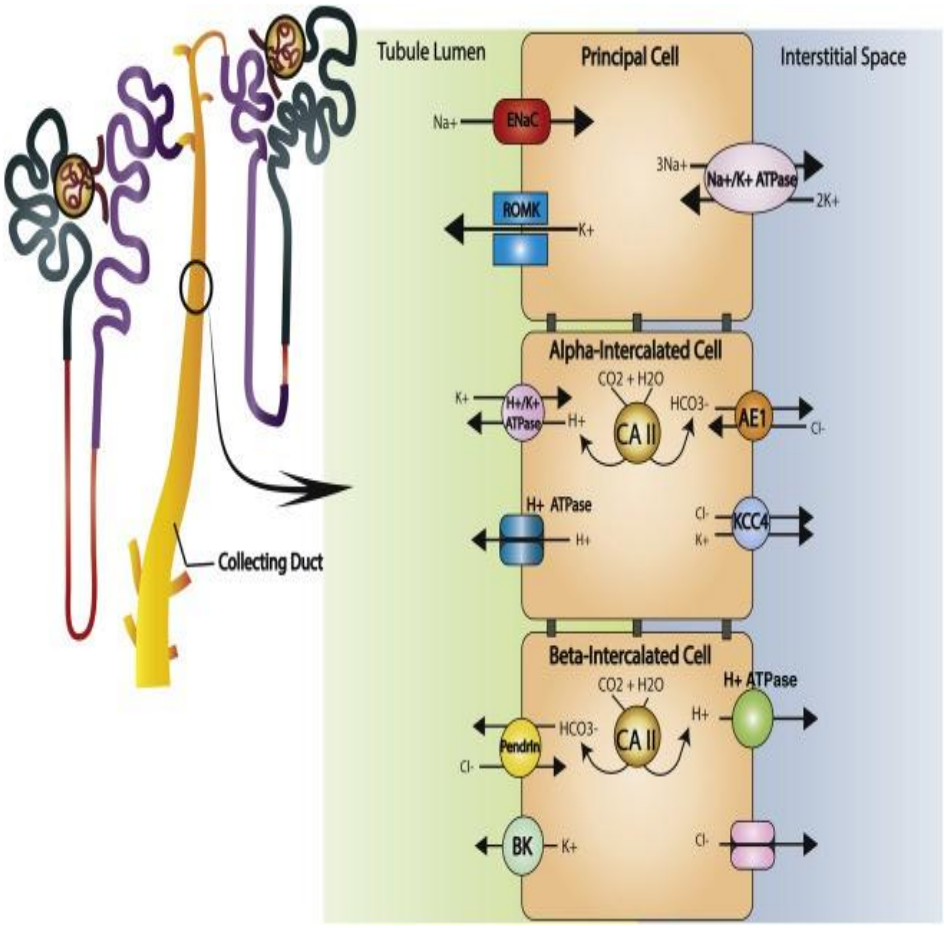
Subsequently, Lip biopsy was taken showing Atrophy of minor salivary glands suggestive ofSjogren's Syndrome.

Sjogren's Antibody testing doen revealing values of Anti -SS-A (RO) >8 AI

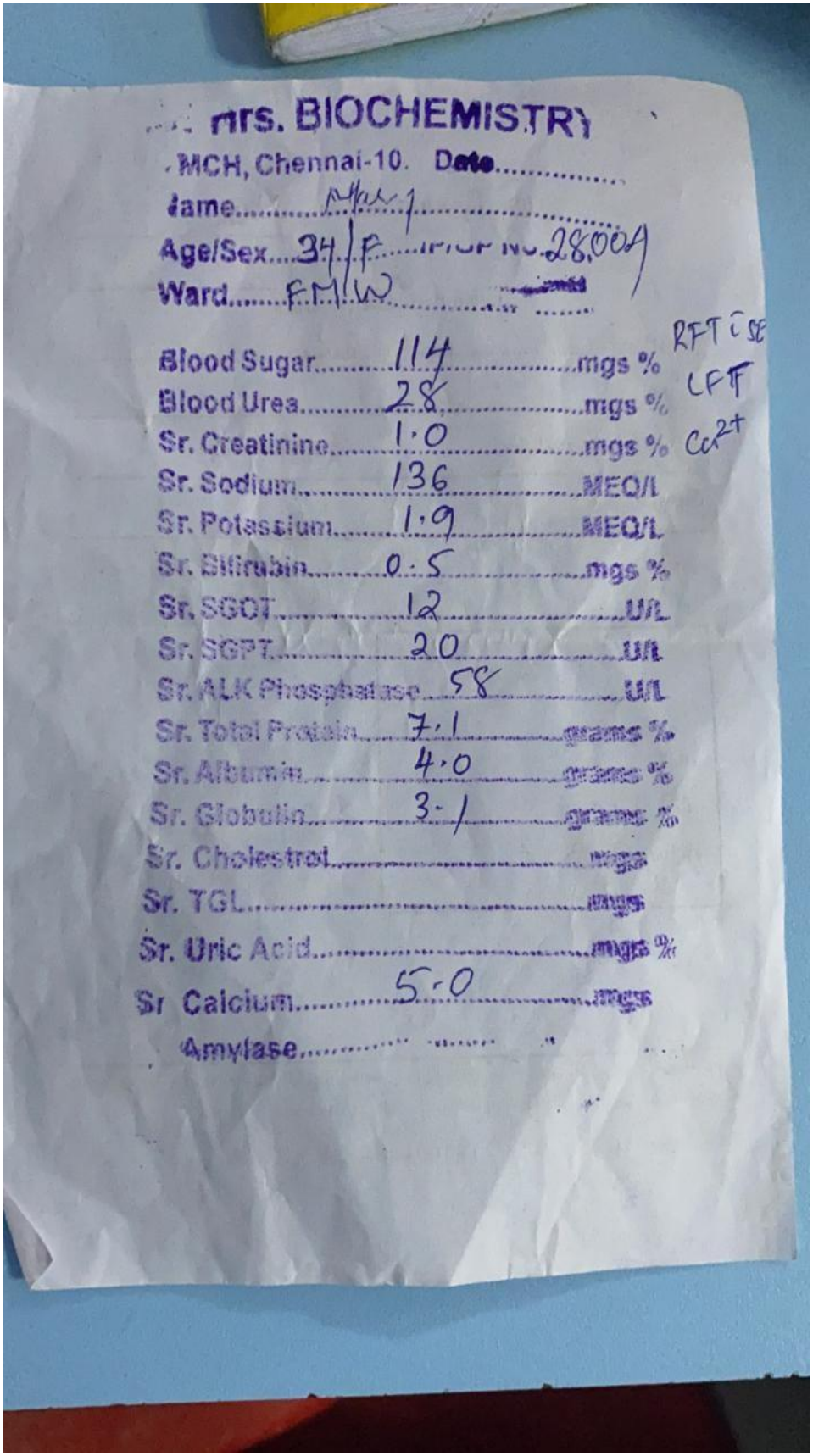
Diagnostics

DATE	TYPE	VALUE	UNIT
2018-06-22	Calcium	5	mg/dL
2018-06-22	Potassium	1.9	mEq/L

Attachments



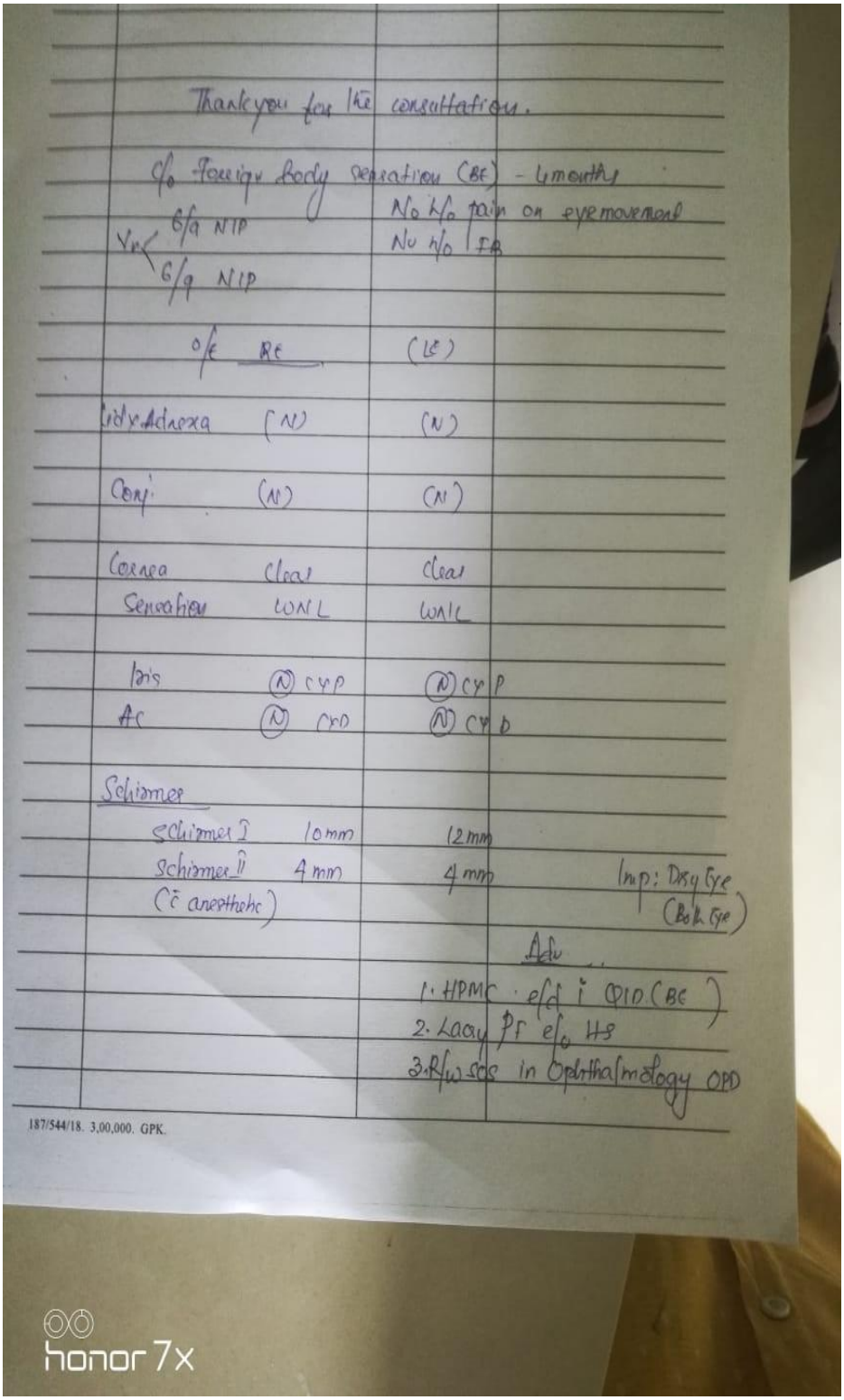
Mechanism of Distal Renal Tubular Acidosis (TYPE 1 -RTA) leading to Hypokalemia



mrs. BIOCHEMISTRY
MCH, Chennai-10. Date.....
Name.....
Age/Sex....34/F.....IP/OP No.28004
Ward.....F.M.W.....

RFT ; SE
LFT
Ca^{2+}

Blood Sugar	114	mgs %
Blood Urea	28	mgs %
Sr. Creatinine	1.0	mgs %
Sr. Sodium	136	MEQ/l
Sr. Potassium	1.9	MEQ/L
Sr. Bilirubin	0.5	mgs %
Sr. SGOT	12	U/L
Sr. SGPT	20	U/L
Sr. ALK Phosphatase	58	U/L
Sr. Total Protein	7.1	grams %
Sr. Albumin	4.0	grams %
Sr. Globulin	3.1	grams %
Sr. Cholestrol		mgs
Sr. TGL		mgs
Sr. Uric Acid		mgs %
Sr Calcium	5.0	mgs
Amylase		



Schirmer's test positivity

DISCUSSION

Sjogren's syndrome should be suspected in individuals with persistent symptoms of dry eyes/mouth, parotid gland enlargement, or abnormal results of specific serological test eg anti -Ro/SSA and /or anti-La/SSB antibodies, rheumatoid factor and hyperglobulinemia. They may also have involvementof many organs beyond the exocrine glands. Skin involvement like xerosis, purpura, associated with vascular or hematological defects, Raynaud's phenomenon, cutaneous vasculitis. SS may be associated with both joint and muscle manifestations, like arthralgia, myopathies. Evidence of thyroid disease like structural, hormonal, or thyroid autoantibody abnormality are also seen in about 10% patients. About 10-20% of patients have thin walled

cysts of lung, with features of cough, and dyspnea with abnormality of pulmonary function test or chest radiograph. Other manifestations include, hematological manifestations like cytopenias, like mild anemia and leukopenia. Immune mediated renal disease, including interstitial nephritis, renal tubular acidosis (Distal RTA) are also manifestations of Sjogren's Syndrome. Distal RTA is commonly associated with Hypokalemia due to renal Potassium wasting. Rarely it is sever enough to produce muscle paralysis (below 2mEq/L). The excretion of Hydrogen ions into the lumen of distal tubule is accomplished by type A intercalated cells (alpha) via luminal H-ATPase pump and, to lesser extend via H-K-ATPase pump.

Impaired H^+ ion secretion into the lumen due to decreased net pump activity leads to reduced K^+ ion uptake. This leads to elevated urinary pH. Immunocytochemical analysis of renal biopsies of patients with Sjogren's Syndrome has shown complete absence of H-ATPase pumps in Intercalated cells. Hypokalemic periodic paralysis is a rare neuromuscular disorder, due to defect in muscle ion channel, with episodes of painless muscle weakness, triggered by rest after vigorous exercise, stressor carbohydrate rich meal. During disabling attacks, prophylactic treatment with Carbonic anhydrase and potassium sparing diuretics are recommended. Here we diagnosed the condition to be a case of hypokalemic quadriplegia due to Distal RTA secondary to Sjogren's Syndrome.

CONCLUSION

Quadriparesis is a condition characterized by weakness of all four limbs. Other symptoms include limp muscles that lack firmness. lack of motor control, inability to walk, depressed reflexes. It can be due to many conditions like infections eg poliomyelitis, enterovirus, flavivirus, HIV, Hepatitis C,Lyme disease.

Many metabolic causes can cause Quadriparesis. One among them being HypokalemicQuadriparesis.

Here we diagnosed a case of Hypokalemic Qaudriparesis due to Distal RTA secondary to Sjogren'sSyndrome.

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