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CASE REPORT **OPEN ACCESS**

Sjogren's Syndrome Presenting as Hypokalemic Quadripaersis

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A B S T R A C T

Primary Sjogren's Syndrome which is an autoimmune disorder can rarely involve kidney resulting in Distal Renal Tubular Acidosis (RTA type I) which can result in severe hypokalemia resulting in flaccid Quadriparesis/Quadriplegia. However Sjogren's Syndrome primarily presenting with hypokalemic Quadriparesis/Quadriplegia is rare. We report a case of 60 year old lady who presented with rapidly progressive hypokalemic Quadriparesis which responded to treatment with potassium salt was later confirmed to be suffering from Sjogren's Syndrome.

Keywords: Sjogren's Syndrome, Renal Tubular Acidosis Type I, hypokalemia, Quadriparesis, Quadriplegia.

INTRODUCTION

Primary Sjogren's Syndrome is an autoimmune disorder characterised by Lymphocytic infiltration of exocrine glands like lacrimal and salivary glands. Prevalence of the disorder varies from 0.01 to 3% [1]. It most often affects women in the age group of 50-60 years and presents with sicca symptoms such Xerophthalmia (dry eyes), Xerostomia (dry mouth) and parotid gland enlargement. Numerous extra glandular features can occur in Sjogen's syndrome such as arthralgia, myalgia, arthritis, Raynaud's Phenomenon, Lymphadenopathy, Neuropathy, Pulmonary disease, Gastrointestinal disease, lymphoma and Renal involvement [6]. Renal involvement is reported in upto 10-30% of cases usually as tubulointerstitial nephritis and Distal Tubular Acidosis [7]. Renal Tubular Acidosis can cause hypokalemia and non-anion gap metabolic acidosis. Cases of severe muscles paralysis due to hypokalemia in Diagnosed cases of Primary and Secondary Sjogren's syndrome have been reported but patients presenting as hypokalemic paralysis in an undiagnosed case of Sjogren's syndrome are rarely reported in literature [1, 2]. Here we present a case of 60 year old lady who presented with hypokalemic quadriplegia, was later diagnosed as a case of Sjogren's syndrome.

Case Report

One Sixty year old lady presented to our hospital on 09/08/2024 with the complaint of insidious onset rapidly progressive weakness of all four limbs of one week duration. Initially the weakness started in both lower limbs in the

form of difficulty in climbing stairs and getting up from squatting position associated with profound fatigue. Three days later she developed weakness of both upper limbs in the form of difficulty in raising her arms above the shoulder. The weakness progressed over the next two days making the patient bed ridden prior to admission. However there was no bladder and bowel involvement or sensory impairment. She denied history of fever, vomiting or diarrhea associated with or preceding the episode, neither she had experienced such episode in the past, On examination she was found to be conscious, oriented with stable vitals. Central Nervous symptoms examination showed that she had flaccid quadriparesis with grade 2/5 power in both lower limbs and 3/5 inboth upper limbs. She also had neck muscles weakness. All her deep tenderness reflexes were diminished. The sensory system was intact. The investigations showed that she had severe hypokalemia (1.5meq/lt.) with non anion gap metabolic acidosis, normal renal function, acidic urine (PH6.0) and urine potassium loss, all suggestive of Type I Renal Tubular Acidosis. She was treated with intravenous potassium replacement for 24 hours followed by oral potassium chloride. Her serum potassium rose to 3.1 on second day and became 3.8 on third day. With the improvement in serum potassium her muscle power also started improving. She started walking with support after 24 hours and without support after 72 hours of initiating treatment and was discharged after four days in stable condition with grade 5/5 power in all four limbs. On repeated questening she admitted that she was experiencing a gritty sensation in both her eyes on and off since last six months but had not consulted an Ophthalmologist. Suspecting Sjogren's Syndrome as the cause of Renal Tubular Acidosis in this patient, connective tissue disorder panel was ordered which showed SSA-Ab and Ro-52 Ab to be strongly positive. Her RA Factor, Anti CCP and ANA were negative. An Ophthalmolgy examination done later confirmed that she had severe dry eyes with positive Schirmor'sTest. However she did not have evidence of Xerostomia and biopsy of Submandibular gland was not performed. The clinical presentation and the available investigations confirmed it to be a case of Primary Sjogern's syndrome with Type I RTA with hypokalemic quadriplegia.

Investigations:

Parameters	**9/8/24**	**10/8/24**	**12/8/24**	**14/08/24**
1)**V.B.G**				
pH	7.17	7.30	7.36	
Hco_3	15	20	24	
Na	136	138	137	
K	1.5	3.1	3.8	
Cl	111	105	99	
2)CBC				
Hb	12.8			
TC	8440			
Platelets	2.38 lakh			
3) RFT				
Urea	37			
Creatinine	1.1			
Calcium	10.1			
Phosphorous	3.9			
4) Urine				
Potassium	48.9			
PH	6.0			
5) ECG	QT Prolongation			
6) CT Abdomen			No Nephrocalcinosis	
7) AutoimmunePanel				
SSA=Ab			+++	
RO-52 Ab			+++	
RA factor			Neg	
Anti CCP			Neg	
ANA			Neg	
8)Shirmor'sTest				Positive<5mm

DISCUSSION

Interstital Nephritis is a known extraglandular manifestation of primary and secondary Sjogren's syndrome presenting as Type I Renal Tabular Acidosis. Type I Renal Tabular Acidosis presents with Hypokalemia and Hyperchloremic Non- anion gap metabolic acidosis due to loss of Hydrogen and Potassium in the urine. Sometimes hypokalemia can be severe and cause muscular paralysis [3-5]. Our patient who is a 60 year old female presented with subacute onset of flaccid, hyporeflexicquadriparesis without any sensory deficit or involvement of bladder or bowel. Guillain – Barre syndrome and Hypokalemicquadriparesis was thought of as the possible causes and V.B.G was ordered to confirm/exclude hypokalemia which incidentally showed severe hypokalemia with hyperchloremic non-anion gap

metabolic acidosis.On further investigation it was found that she had acidic urine (pH>5.5) and kaliurea. Although she was not a diagnosed case of Sjogern's syndrome, in view of the history of gritty sensation in eyes for last six months and absence of other cause for RTA, she was investigated for Sjogern's syndrome. The autoimmune panel showed that her serum was strongly positive for SSA and RO antibodies and the Shirmer's test was positive favouring the diagnosis of Sjogren's syndrome. Since she didn't have any evidence of other autoimmune disease to suggest secondary Sjogern's syndrome a diagnosis of primary Sjogren's Syndrome presenting as hypokalemicquadriparesis was made. She had good response to potassium correction and was discharged after four days with normal power in all four limbs and normal serum potassium level with the advise to be on regular fallow up by ophthalmologist and physician.

Our case is one of the those rare cases of Sjogern's syndrome reported in literature who primarily had presented to the cliniciansashypokalemicquadriparesis/quadriplegia [1, 2]. A high index of suspicion is required to diagnose Sjogern's syndrome as the cause of this presentation otherwise the diagnosis may be missed. Female patient in her 5^{th} and 6^{th} decades presenting with hypokalemic paralysis should be investigated for Type I RTA in absence of other obvious cause of hypokalemia and if ABG shows presence of hyperchloremic non-anion gap metabolic acidosis suggesting of Type I RTA, Sjogern's syndrome should be considered as a diagnosis and further investigations should be done to confirm it.

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