Case Report

Acute flaccid Paralysis as first presentation of Primary Sjogren's Syndrome: A case report from Nepal

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Abstract

Sjögren's syndrome is an autoimmune disease with glandular and extra-glandular involvement. The common neurological manifestation of Sjögren's syndrome are peripheral neuropathy, anterior horn cell disease, transverse myelitis and aseptic meningitis. However, Sjögren's syndrome presenting with acute flaccid paralysis is rare phenomenon. We report a case of a 35-year-old woman who presented with acute flaccid paralysis with respiratory failure who was later diagnosed as Primary Sjogren's syndrome.

Introduction

Sjögren's syndrome is an autoimmune disease with glandular and extra-glandular manifestations.[1] It is associated with lymphocytic and plasmacytic infiltrate in the secretory glands primarily the salivary, parotid, and lacrimal glands and manifest as sicca syndrome. The common neurological manifestations of Sjögren's syndrome are peripheral neuropathy, anterior horn cell disease, transverse myelitis and aseptic meningitis.[2] Acute flaccid paralysis (AFP) is a clinical syndrome characterized by rapid onset of lower motor neuron type weakness, commonly caused by Guillian Barre syndrome(GBS), neuroparalytic snake envenomation and acute transverse myelitis.[3] The acute flaccid paralysis as a first presentation of Sjögren's syndrome is rare phenomenon. We report a 35 year old patient who presented with AFP and was diagnosed as Primary Sjogren's syndrome.

Case Report

A 35 year old patient from terai region of Nepal, with no prior co-morbidity presented to Emergency with complaints of cramps in her right upper hand for 2 days and sudden onset inability to move bilateral upper and lower limbs for 1 day. The weakness was bilaterally symmetrical, gradually progressive which made her difficult to get up from the bed. The limbs were flaccid. She had also difficulty in breathing which is more in supine position. There was no history of speech disturbances, loss of consciousness, fever, bowel and bladder disturbances. She denied alcohol consumption, animal bite or sting, illicit drug use recent herbal or any other medications.

On Examination, her vitals were Blood pressure- 90/60mm Hg, Pulse rate- 82bpm, Respiratory rate- 24/min, Temperature- 98 F, Spo2 88% room air. General physical examination was within normal limits. Respiratory, cardiovascular and gastrointestinal examinations were unremarkable. Single breath count of the patient was 8. On motor examination, power was 1/5 (Medical research council grading) in all four limbs. Deep tendon reflexes were hypoactive. Plantar reflex was bilaterally mute. Higher mental function, Cranial nerves, and Sensory examinations were normal.

Laboratory examinations (table 1. And table 2.) revealed severe hypokalemia with non anion gap metabolic acidosis with respiratory acidosis with alkaline urine. ECG showed prolonged PR and QRS duration with ventricular premature beats. Creatinine kinase was raised. Patient was initially diagnosed as a case of hypokalemic quadriparesis with type 2 respiratory failure secondary to distal tubular acidosis. We looked for the etiology of severe hypokalemia and renal tubular acidosis . Antinuclear Antibody(ANA) and Rheumatoid factor was high. Schirmers test was positive(2 mm bilateral eye).Anti Ro and Anti La was positive.

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Diagnosis of Sjögren's syndrome was made on the basis of ACR/ EULAR Criteria. [Table 3] .

Table 1. Laboratory Investigation of the patient

Laboratory Parameters	Result	Normal value
Hemoglobin (g/dl)	10.1	11-15
Urea (mg/dl)	41	10-15
Creatinine (mg/dl)	1.5	0.3-1.2
Na+ (mmol/L)	140	136-135
K+ (mmol/L)	1.5	3.5-5.0
Mg++ (mmol/L)	0.82	0.8-1.1
Ca Ionized (mmol/L)	1.11	1.1-1.3
Ca Total (mg/dl)	8.4	8-10.5
Ra factor (IU/ml)	192	<12
ANA (IU/ml)	280	<40
CKMB (U/L)	44	5-25
CK Total (U/L)	355	55-130
Urine		
Protein	+++	
Sugar	-	
WBC	1-2	
RBC	1-2	
pH	7.03	
Anti Ro (Units)	131.71	<20
Anti La (Units)	90.84	<20

ANA: Antinuclear antibody, CKMB: Creatinine Kinase MB, CK Total: Creatinine kinase total

Table 2. Arterial blood gas of the patient

ABG parameter	At Admission	Day 1	Day2	At Discharge
pH	6.956	7.117	7.283	7.385
HCO3 (mmol/l)	10	14	17	21
Pco2 (mm Hg)	66	52	49	28
Po2 (mm Hg)	98	96	98	96
Anion gap	10	9	10	11

HCO3: bicarbonate, PCO2:partial pressure of carbon dioxide, PO2: partial pressure of oxygen

Table 3.ACR/ EULAR Criteria for Sjogren's syndrome

SN	Item	Score
1	Labial salivary gland with focal lymphocytic sialedenitis	3
2	Anti Ro/La Positive	3
3	Ocular staining score >5	1
4	Unstimulated whole saliva flow rate <0.1ml/ min	1
5	Schirmer's test <5mm/5minutes in at least 1 eye	1

Score≥ 4 is Diagnostic for Sjogren's syndrome

(Criteria no 2 and 5 was fulfilled in our case)

Patient was initially treated with intravenous Potassium chloride(KCL).Her daily requirement was 100 meq/L of potassium for 3 days after which it was changed to oral potassium supplementation. Her symptoms improved by day 2nd of treatment. Sodium bicarbonate was started after Potassium correction to correct the metabolic acidosis which reduces inappropriate urinary potassium loss. Oral potassium supplementation was gradually tapered and stopped. Aldosterone antagonist was started along with oral steroid (prednisolone 1mg/ kg) at the time of discharge for interstitial nephritis secondary to Sjogrens syndrome which lead to severe hypokalemia.

Patient was followed up after 2 weeks , 1 month and 3month and patient was asymptomatic. Her Potassium level and serum pH were within normal limits. Steroid was gradually tapered on each follow up.

Discussion

In our case, patient presented with acute onset symmetric, progressive flaccid paralysis with respiratory failure .On initial investigation patient had severe hypokalemia with non anion gap metabolic acidosis with respiratory acidosis. On evaluating the cause of non anion metabolic acidosis we found it was type 1 distal tubular acidosis. On further evaluating for cause of type 1 distal tubular acidosis diagnosis of Sjögren's syndrome was made on clinical and serological basis. We initially had GBS, transverse myelitis, and channelopathies as initial differential diagnosis. Transverse myelitis was ruled out as there was no definite sensory level and no bowel and bladder involvement. We ruled out GBS as there was CSF analysis was normal and also there was no cranial nerve and autonomic involvement.

Sjögren's syndrome represents a group of diseases characterized by inflammation and destruction of exocrine glands. As reported, 18.4% to 67% of patients with Sjogren's syndrome has renal involvement.[4] Tubulointerstitial nephritis is one of the common renal manifestation of Sjögren's syndrome which can present with renal tubular acidosis (RTA), and nephrogenic diabetes insipidus.[5] Hypokalemia is the most common electrolyte abnormality, occurring in 28–53% of patient with distal RTA.

The treatment of hypokalemic paralysis is replacement of potassium and treat the cause. In our case hypokalemia was secondary to distal RTA. So we have started alkali therapy after

potassium restitution .It is important to correct hypokalemia before alkali therapy because the alkalosis might aggravate hypokalemia by enhancing the shift of potassium into cells and bicarbonaturia. Alkali therapy corrects the metabolic acidosis which reduces inappropriate urinary potassium loses.[6] Aldosterone antagonist is used as a potassium sparing agent for maintainencetherapy. RTA is not a usual indication for immunomodulatory therapy in Sjögren'ssyndrome[7] Steroid is used for interstitial nephritis secondary to Sjögren's syndrome. Steroid therapy is indicated in cases that are nonresponsive to replacement therapy and in those with recurring or life threatening hypokalemic paralysis attacks.[8] So we have started our patient on oral prednisolone tapering over 3 months. The use of steroid reduces the relapse rate in Sjogren syndrome.[9]

Our case report has one the rare presentation of Sjögren's syndrome presenting with acute flaccid paralysis. The limitation of the case report was that we couldn't perform labial salivary gland biopsy, ocular staining and sialography in this patient.

Conclusion

The cause of Acute flaccid paralysis in our patient was hypokalemia secondary to distal renal tubular acidosis. Although, the cause of distal RTA is unknown, one possibility is Sjogren syndrome which is treatable condition. Our patient recovered well with potassium supplementation, sodium bicarbonate, aldosterone antagonist and steroid. Hence, as a clinician we should be always work up for Sjögren's syndrome as one of the cause of acute flaccid paralysis.

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Conflict of Interest: None

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