Case Report

Primary Sjogren's Syndrome with Renal Tubular Acidosis with Central Pontine Myelinolysis - A rare case report

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Abstract

A middle aged female patient, a case of Primary Sjogren's Syndrome with renal tubular acidosis as revealed by severe hypokalemia along with normal anion gap, metabolic acidosis and acidic urinary pH had brain stem lesion which presented as quadriplegia, dysphagia and dysarthria. Laboratory tests revealed that anti-nuclear antibodies (ANA) and anti Ro/SSA antibodies were positive. MRI showed hyper intense lesion in T2W images in middle pons, typical characteristic of central pontine myelenolysis. So, patient was diagnosed as Primary Sjogren's Syndrome with renal tubular acidosis with central pontine myelinolysis. She recovered with correction of hypokalemia, intravenous methyl prednisolone and cyclophosphamide.

Key words

Primary Sjogren's Syndrome, Renal Tubular Acidosis, Central Pontine Myelinolysis.

Introduction

Primary Sjogrens Syndrome (pSS) is a chronic autoimmune disease seen in middle aged females, presenting with most common symptoms as dry mouth and dry eyes. Sjogrens syndrome can also cause renal tubular acidosis leading to hypokalemia, nephritis, hypothyroidism, thyroiditis, optic neuritis, brain stem syndromes like central pontine myelinosis or intra-nuclear ophthalmoplegia, myelopathy and peripheral neuropathy as well. Here we report a very rare case of a female of Primary Sjogrens Syndrome with association of Central

Pontine Myelinolysis (CPM) and hypokalemia , presenting as acute flaccid quadriplegia [1-9].

Case report

A 45 year old female patient, housewife, presented in emergency with history of rapid and progressive weakness of all 4 limbs, difficulty in respiration, difficulty in speaking and swallowing and altered sensorium since 48 hours. On further inquiry, there was no preceding history of fever, rash, polyuria, arthralgia, myalgia, recent vaccination, diarrhoea or flu. There was an acute onset of weakness in both lower limbs which progressed to both upper limbs as well in 48 hours. Patient also complained of difficulty in speaking and difficulty in swallowing for past few days. From 2 days patient developed shortness of breath and derranged sensorium. Patient had significant past history of hypertension since 8 years and was on regular medication for it. Patient had a persistent complaint of dry eyes and dry mouth since 4 years which was progressive despite local hospitals' treatment. Addiction and allergic history were unremarkable. Patient had menopause 2 years back and obstetric history revealed G2P2A0L2. Due to quadriplegia and shortness of breath patient was shifted to ICU and was put on elective mechanical ventilatory support.

Preliminary examination showed that BP-150/90mmhg , pulse was 84 beats per minute, regular in all 4 limbs without any delay , RR-14 on SIMV mode with Spo2 99% with Fio2 0.4 General physical examination revealed no pallor, no icterus, no cyanosis, no clubbing, no lymphadenopathy, no thyroid enlargement, no carotid enlargement.

Neurologically examination showed that patient was dull and drowsy with altered sensorium. Cranial nerves were normal. Pupils were 3mm in size normally reacting to light in both eyes. Sensory examination for pain and crude touch was unremarkable. Motor system evaluation showed normal bulk of muscles but power grade 0 in all 4 limbs with normal deep tendon jerks.

Investigations showed hypernatremia (164 meq/l), severe hypokalemia (1.9 meq/l). ABG showed metabolic acidosis (ph- 6.93) with bicarbonate level of 16mmol/l. Urine analysis showed 1+ albuminuria and urine ph of 6.5 suggesting acidic urine. In view of metabolic acidosis, hypokalemia and hyperchloremia, possibility of RTA was thought and patient was worked up for immunological disease. Investigations revealed hypernatremia and severe hypokalemia , positive anti Ro-SSA antibody (196), positive ANA with speckled pattern with 1;320 titre, mild leukocytosis (15000/cumm), high 24 hour urinary protein excretion (869 mg/d) , with high serum osmolality (349 mosm/kg) and low urine osmolality (453 mosm/kg), high LDH (1183IU/l). NCV suggested mild axonopathy of bilateral common peroneal nerves and rest normal conduction for all the nerves. As patient had spastic dysarthria and dysphagia, MRI and CSF study was done. CSF study was normal and MRI Brain showed abnormal T1W hypointense and T2W/flair/diffusion hyperintensity involving middle part of pons showed possibility of central pontine myelinolysis and extra pontine myelinolysis.

From the above list of investigations patient was thought to be of Primary Sjogren's Syndrome, renal tubuar acidosis, hypokalemia, central pontine myelinolysis and quadriplegia.

Patient was treated with antibiotics, hypotonic saline and correction of potassium for next 3 days by intravenous potassium chloride and patient showed improvement in dullnesss and motor weakness. Patient was given intravenous methyl prednisolone 1g/day for 5 days on which patient responded. Pulse therapy of intravenous cyclophosphamide was also given. Patient was discharged after 2 weeks of treatment and she was able to walk with support.

After one month at follow up, patient improved significantly and was ambulatory with residual

neurological deficit in form of only right lateral rectus palsy.

Discussion

Primary Sjogren's Syndrome (pSS) is a chronic autoimmune disease characterized by destructive lymphocyte infiltration in exocrine glands [8] especially lacrimal and salivary glands so it is also called dry eyes-dry mouth syndrome. If Sjogren's syndrome occurs without any associated connective tissue disorder is termed as primary Sjogren's syndrome (pSS). It is seen in middle aged with male:female ratio of 1:9. In addition to dry eyes and dry mouth, Sjogren's syndrome can also cause renal tubular acidosis or nephritis or hypothyroidism or thyroiditis or atrophic gastritis or pulmonary and liver diseases, peripheral and central nervous system involvement.

Diagnosis of PSS according to American-European consensus group (AECG) [1] criteria require at least 4 out of 6 criteria to be positive

- Subjective xerophthalmia
- Subjective xerostomia
- Objective test for xerophthalmia
- Objective test for salivary gland dysfunction
- Either anti-Ro/SSA or anti- LA/SSB positive
- Histopathology criteria for PSS on minor salivary gland biopsy

In our patient, xerophthamia, xerostomia, positive anti-Ro/SSA, supported the diagnosis of pSS. Renal damage [7] in Primary Sjogren's Syndrome is also common. pSS involves urinary concentrating capacity [4] due to damage in renal interstitium. Glomerular damage is infrequent and mild and often associated with other connective diseases in conjunction. Renal tubular acidosis is main cause of hypokalemia with normal anion gap metabolic acidosis. Renal involvement [5, 6] in pSS is seen in 27% of cases. It is frequent site for extra glandular involvement after peripheral nerves and CNS.

Spectrum of neurological disorder in pSS is broad. Peripheral nervous system involvement is more common than central nervous system. Peripheral neuropathy is often the presenting feature [9]. Distal sensory or sensory-motor neuropathy is most common, painful sensory neuropathy, autonomic neuropathy, trigeminal sensory neuropathy, mononeuritis multiplex or involvement of multiple cranial nerves can occur. Biopsy of involved peripheral nerve is likely to show epineural vascultits [9]. In CNS, Sjogren's syndrome may cause focal brain lesions which may present as stroke like episode, Optic neuritis, focal paresthesias, brain stem syndrome like CPM [2] and intra nuclear ophthalmoplegia and myelopathy can occur. Recently a report showed that patient with CPM showed considerable improvement after iv immunoglobulins means immune mechanism may be involved in CPM [3].

The neurological manifestations in our patient were mainly due to central pontine myelinolysis. Both hypokalemia and CPM contributed to quadriplegia.CPM is also associated with alcoholics and liver diseases with rapid correction of chronic hyponatremia. However CPM in Sjogren's syndrome patients is rarely reported. The clinical feature of CPM includes confusion, altered sensorium, quadriplegia, locked-in syndrome and dysarthria with dysphagia. CPM on MRI is characteristically a symmetric lesion in basal pons hyper intense in T2W images. In our patient, changes in consciousness level could not have been caused by hypokalemia alone, since it only reduces the muscle power in the limbs. The neurological symptoms in our patient were mainly due to central pontine myelinolysis and both central pontine myelinolysis and hypokalemia contributed to quadriplegia. Our patient of CPM responded with pulse therapy of cyclophosphamide and methyl prednisolone.

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