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Osmotic Demyelination Syndrome As a Manifestation of Hypokalemia Secondary to Sjogren's Syndrome With Renal Tubular Acidosis

Case Report

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Abstract

Sjogren's syndrome is a female dominated autoimmune disorder with diverse phenotypical expression. For most patients, disease runs an indolent course with sicca symptoms, musculoarticular pain and disabling fatigue. Distal real tubular acidosis is an extra-glandular manifestation of Sjogren's syndrome characterized by failure of kidney to secret hydrogen ions. This results in increased secretion of other cations including potassium with resultant metabolic acidosis and hypokalemia.

We present herewith a case of Sjogren's syndrome with distal renal tubular acidosis who presented with a wide spectrum of neurological manifestations. Further evaluation of this patient along with the brain imaging revealed osmotic demyelination. As patient did not have hyponatremia at any point during hospitalization, we could attribute occurrence of Osmotic demyelination syndrome to hypokalemia only.

Keywords: Osmotic demyelination syndrome; Sjogren's syndrome; Hypokalaemia; Renal tubular acidosis

Introduction

Sjogren's syndrome is a chronic, slowly progressing autoimmune disease characterized by lymphocytic infiltration of the exocrine glands resulting in xerostomia and dry eyes (keratoconjunctivitis sicca). Extraglandular manifestations are seen in one third of patients with Sjogren's syndrome. Renal involvement in Sjogren's syndrome is not uncommon and may precede sicca symptoms. The pathology in most of the cases is tubulointerstitial nephritis causing distal renal tubular acidosis [1-5]. It indicates failure of the intercalated cells in the collecting duct of kidney to secrete hydrogen ions. As a result, secretion of other cations, including potassium, is increased to maintain electroneutrality. Potassium loss can result in hypokalemic paralysis, often presenting as recurrent episodes. However, cerebellar and extra-pyramidal manifestations are uncommon due to primary neurological involvement in Sjogren's syndrome [6,7].

Case History

A 43 years old female, residing in a slum area, a homemaker, presented with chief complaints of loose motions since two days, vomiting since one day and weakness in both lower limbs since eight to ten hours.

On examination, the patient was afebrile, had a pulse rate of 60 beats/min and a blood pressure of 90/60 MM of mercury. No signs of dehydration were present.

Her cardiovascular, respiratory and abdominal system examinations were normal.

On CNS examination, patient was drowsy but arousable. Her speech was slurred with stress on some syllables. Cranial nerve examination was normal. Motor system examination revealed power of grade five in both upper limbs and grade one in both lower limbs. Her deep tendon reflexes were absent in all four limbs. She had an intention tremor and finger nose test was positive on cerebellar examination. Sensory examination was within normal limits. Bilateral plantar responses were flexor. On the next day, she developed weakness in both upper limbs as well with a power of grade three (3/5).

Her lab report showed Serum Sodium- 136.4mEq/L(normal range-:125-145mEq/L), Serum Potassium- 1.19mEqu/L(normal range-:3.5-5.5mEq/L). She received potassium correction in the form of infusion of injection potassium chloride in normal saline. After 24 hours of correction patient's muscle weakness and sensorium improved significantly but serum potassium remained low. Samples for urinary electrolytes and serum magnesium were sent which revealed normal serum magnesium and a positive urine anion gap. ABG revealed PH - 7.388(normal range-:7.35-7.45), Chloride-135 mEq/L(normal range-:102-119mEq/L), Potassium- 1.4mEq/L(normal range-:3.5-5.5mEq/L). The picture was suggestive of distal renal tubular acidosis. Owing to the persistence of cerebellar signs we ordered an MRI of brain with the possibility of CNS vasculitis. MRI revealed features of osmotic demyelination syndrome with extrapontine myelinolysis.

Other lab parameters were as follows-:

Her hemogram showed Hemoglobin of 11.2g/dl(11-18g/dl), Total leucocyte Count-8140/uL(4000-11,000/uL), platelet count-1,21,000/*u*L(1,20,000-5,00,000/*u*L).

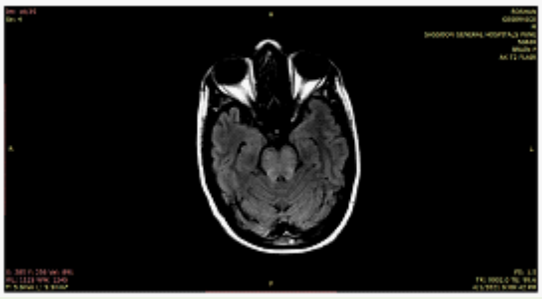

Figure 1: T2W FLAIR image showing hyperintense signal in pons with peripheral fibre sparing.

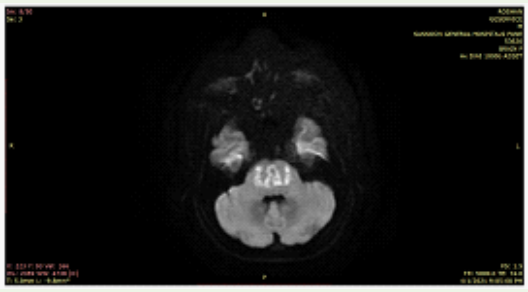

Figure 2: Diffusion weighted image showing restricted diffusion in the pons.

Liver function tests were within normal limits. Ultrasound examination of abdomen was normal.

ECG - Widening of QRS complex, RBBB, ST segment depression with 'T' wave flattening in leads V1 to V6 and generalized U waves.

Taking into consideration, a picture of distal Renal tubular acidosis and MRI suggestive of osmotic demyelination syndrome, we contemplated Sjogren's syndrome to be the culprit and got ANA Blot assayed which revealed SM/RNP-58AU (normal range 6-12AU), SSA/RO60KD-75AU (normal range 6-12AU), SS/RO52KD-69AU (normal range 6-12AU). Patient had no evidence of other rheumatological disorders. The patient satisfied 4 out of 6 criteria for Sjogren's syndrome as per the American-European Consensus criteria and thus, the diagnosis of primary Sjogren's syndrome was made. She was started on oral potassium supplements and Shohl's solution.

Around 6 months ago, she had presented with subacute diarrhea with hypokalemia that responded to symptomatic treatment. The episode was self-limited.

Discussion

We had a case of 43 years old female presenting with acute diarrhea. During the second episode, features of hypokalemia dominated the clinical picture. Presence of additional neurological signs led us to the diagnosis of osmotic demyelination syndrome. On evaluation of etiology, the hypokalemia occurred as a result of distal renal tubular acidosis secondary to Sjogren's syndrome. She had pyramidal with extra pyramidal features on the day of admission itself. These findings were explained by the MRI picture of Osmotic demyelination syndrome involving pontine as well as extra pontine areas. The quadriparesis was secondary to hypokalemia.

We attribute the Osmotic demyelination syndrome findings to hypokalemia only as the patient did not have hyponatremia at any point during hospitalization. Also, the findings were demonstrated before her hypokalemia was corrected, obviating any potential role of Intravenous fluids administered for correction of hypokalemia. Several case reports have been documented in literature regarding hypokalemia as one of the implicated mechanisms in the causation of Osmotic demyelination syndrome. M D Norenberg hypothesized that hyponatremia caused cerebral endothelial injury by osmotic dysregulation, further leading to release of myelinotoxic factors causing vasogenic cerebral edema [1]. A similar mechanism could have resulted in Osmotic demyelination syndrome associated with hypokalemia.

I Dørup et al have documented the correlation between cellular potassium: sodium ratio and concentration of H-ouabain binding sites. Potassium deficiency possibly leads to downregulation of sodium-potassium pumps in skeletal muscles [2]. A mechanism close to this finding could be operational in endothelial or glial cell membranes of the central nervous system as well. Decreased concentration of Na-K ATPase in endothelial cell membrane during hypokalemia may predispose the cell susceptible to injury by slight

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increase in osmotic stress [3]. Therefore, there is a possibility that Osmotic Demyelination syndrome was induced by a slight increase in osmotic pressure attributable to fluid infusion such as of electrolytes in the presence of severe hypokalemia, even if the increase rate was so low as to not induce injury in normal state.

Thus, the findings of Sjogren's syndrome with distal Renal tubular acidosis, manifesting as hypokalemia and Osmotic demyelination syndrome, are of a rare occurrence.

This lady showed resolution of symptoms gradually as her potassium levels were normalised. After day 10 of admission, her serum potassium was 3.7mEq/L and there was no focal neurological deficit on examination. On follow up visit, she had no signs of hypokalemia and was asked to continue oral potassium supplements. In our case, there was a conspicuous absence of hyponatremia. Thus, hypokalemia leading to downregulation of sodium potassium pumps in glial cell membranes of the central nervous system could be the possible mechanism. The cerebellar dysfunction improved after the correction of hypokalemia. The patient followed up with us after one month and on examination showed no residual neurological deficit.

Thus, Hyponatremia is not the only implicating factor in the causation of osmotic demyelination syndrome. Hypokalemia should be corrected and maintained in normal reference range depending on the cause.

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