Acute Flaccid Quadriplegia as the Presenting Symptom of Sjogren’s Syndrome- A Rare Presentation

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Abstract
Sjogren’s syndrome is a slowly progressive autoimmune disorder characterized by lymphocytic infiltration of the exocrine glands resulting in xerostomia and dry eyes. The syndrome has unique features since it presents with a wide clinical spectrum from organ-specific autoimmune exocrinopathy to systemic disease. A small but significant number of these patients progress to develop malignant lymphoma. Middle-aged women (female-to-male ratio, 9:1) are primarily affected. However, Sjogren’s syndrome may occur at any age, including childhood¹. We present the case of a 42-year-old female who presented with flaccid paralysis of all four limbs and on evaluation was diagnosed to have Sjogren’s syndrome. Primary Sjogren’s Syndrome presenting as quadriplegia extremely rare and the significance of managing hypokalemia due to renal tubular acidosis with potassium citrate instead of potassium chloride is highlighted.

Keywords: Sjogren’s syndrome, Hypokalemic periodic paralysis, distal renal tubular acidosis.

Introduction
One among the many causes of hypokalemia is distal renal tubular acidosis (dRTA), but it is rarely severe enough to present as hypokalaemic paralysis. A patient with Sjogren’s syndrome, with no sicca symptoms, presenting for the first time with hypokalaemic paralysis due to dRTA is very uncommon². Positive serology and salivary gland histopathology was the only evidence of underlying Sjogren’s syndrome in our patient.

Case Report
A 42-year-old female patient with no known co-morbidities was brought to the causality with complaints of acute onset weakness of bilateral upper and lower limb since 1 day. On presentation, her vitals were stable, BP: 126/70mm Hg, PR: 86bpm, RR: 20 cycles/min, and oxygen saturation of 98% on room air. She had bilateral flaccid quadriplegia with diminished reflexes. Power was worse in the lower limb (MRC grade 1) than the upper limb (MRC grade 2). She had diminished deep tendon reflexes with bilateral plantar reflex mute. Sensory system and cranial nerve examinations did not reveal any abnormalities. All other systemic examinations were within normal limits. The patient is a known case of hypothyroidism for 3 years, for which she was on regular thyroid supplements. An MRI brain with whole spine screening was done to rule out any neurological cause of quadriplegia, however, it was reported to show only a diffuse
disc bulge at C4-5 and C5-6 levels indenting the thecal sac with no significant disc bulge (Figure 1,2).

**Figure 1: C5-6-disc bulge**

On further evaluation, her blood reports showed a serum potassium level of 1.75 mmol/L, and a diagnosis of hypokalemic periodic paralysis was made. Serum bicarbonate level was 6.1 mmol/L and urine pH was 6.0. Arterial blood gas analysis showed a pH of 7.25, pCO2 14, pO2 101, and bicarbonate of 10.2, indicative of severe metabolic acidosis. To identify the root cause of the metabolic acidosis, an ANA profile was done. The patient’s ANA profile was positive for SS-A/Ro and SS-B/La suggestive of Primary Sjogren’s syndrome.

A lower lip mucosal biopsy of the minor salivary gland was done to confirm the diagnosis of Sjogren’s syndrome. Histopathology was suggestive of Lymphocytic sialadenitis favoring the diagnosis of Sjogren’s syndrome (Figure 3,4,5). A diagnosis of distal renal tubular acidosis secondary to Sjogren’s syndrome was hence confirmed.

**Figure 2: MRI brain normal study**

**Figure 3: Low power view of salivary Acini with ducts and blood vessels**
Figure 4: A salivary acini with ducts and blood vessels (40x H & E)

Figure 5: Higher power of the salivary lobule with lymphocytic infiltration

The patient was started on oral potassium citrate at a dose of 3mmol/kg/ body weight) and her muscle weakness improved over the next 4 days. Given the renal involvement, she was initiated on oral steroid therapy (oral prednisolone 1mg/kg) for four weeks and tapered slowly. Over a few weeks, the patient achieved normokalemia without oral potassium citrate therapy and there was complete remission of disease activity.

Discussion
Sjogren’s syndrome (SS) is the second most common autoimmune disease affecting mainly middle-aged women. As a result of lymphocytic infiltration and destruction of salivary and lacrimal glands, the main symptom of the disease includes xerostomia and xerophthalmia. The first and only manifestation of Sjogren’s syndrome in our patient was hypokalemic paralysis due to dRTA. Her serology was strongly positive for Sjogren’s syndrome and lip biopsy for salivary gland histopathology was suggestive of the same etiology, even though she had no sicca symptoms. This case brings light to the fact that Sjogren’s needs to be considered in a case of dRTA even in the absence of classical manifestations of the disease.

Renal tubular acidosis (RTA) refers to a group of disorders characterized by defective renal acid-base regulation. The ability for normal urinary acidification is flawed, resulting in net acid retention and hyperchloremic metabolic acidosis. Patients with Sjogren’s syndrome with dRTA have interstitial nephritis with high levels of anti-carbonic anhydrase antibodies. These antibodies affect the function of carbonic anhydrase in cortical collecting ducts. The defect in acidification is due to insufficient intact H+-ATPase pumps in the intercalated cells.

In our patient, daily life-long alkali replacement in a dose of 1–2 mEq/kg will prevent acute hypokalemia and the use of corticosteroids or other immunosuppressive drugs used for the primary disease have shown to reverse the renal tubular defect.

Conflicts of Interest: Nil

References


