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A CASE OF PROBABLE SJOGRENS SYNDROME PRESENTING WITH HYPOKALEMIC PARALYSIS AND INCIDENTAL CHRONIC HEPATITIS B VIRUS INFECTION

Rheumatology

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ABSTRACT

Sjogren's syndrome is relatively rare in males, with female to male incidence ratio of 16:1. It is an autoimmune disorder characterized by lymphocytic infiltration of exocrine glands. However, the disease may be associated with extra glandular manifestation affecting multiple organs. We report one such uncommon presentation where a 19-year-old male was brought to our emergency department with chief complaint of weakness of all four limbs. He had hypokalemia at admission. Upon further evaluation, we diagnosed this case as probable Sjogren's syndrome with distal renal tubular acidosis (dRTA) which caused the hypokalemia. Incidentally he was found having chronic hepatitis B virus infection. This association of hepatitis virus infection and Sjogren's syndrome is rarely described in the literature.

KEYWORDS

Sjogren's Syndrome, distal Renal Tubular Acidosis, hypokalemic paralysis, Hepatitis B.

BACKGROUND

Sjögren syndrome is an autoimmune condition characterized by lymphocytic infiltration into moisture secreting glands, manifesting in dry eyes and dry mouth (sicca syndrome). Sjögren syndrome is one of many fascinating, pluriform autoimmune entities whose underlying pathophysiology remains incompletely understood. The association of hepatitis virus infection and Sjogren's syndrome is rarely described in the literature. The causal role of hepatitis infection triggering Sjogren's syndrome is under debate.

CASE REPORT

A 19-year-old male was brought to the emergency department with chief complaint of weakness of all four limbs for 1 day. There is no history of trauma, fever or headache preceding the weakness. The vital signs were within normal limits at presentation. Neurological examination revealed a power of 2/5 in all four limbs and the reflexes were normal.

Initial lab investigations revealed low serum potassium of 1.6mEq/L, serum sodium of 143mEq/L, serum calcium of 9.9mg/dL. An arterial blood gas showed a pH of 7.37, bicarbonate of 17.1 mmol/L, and a CO2 of 24.8 mm Hg.

The patient was given intravenous and oral potassium supplementation over the next 24 hours and his symptoms gradually improved.

The Trans tubular potassium gradient (TTKG) was 7.54, suggesting renal potassium loss.

Urine anion gap was positive; suggesting distal renal tubular acidosis (dRTA).

Anti SS-A(Ro) antibody was 58.4 (positive) Units and Anti SS-B(La) Antibody was 27.3 Units (positive).

Upon routine viral screening, we found that he was positive for Hepatitis B surface Antigen (HBsAg), but negative for IgM anti-HBc Antibodies and HbeAg.

However, the patient did not have sicca symptoms upon further probing into history.

So, we diagnosed the case as probable Sjogren's syndrome with distal renal tubular acidosis with hypokalemic paralysis with HBsAg infection.

DISCUSSION

Sjögren syndrome is an autoimmune condition characterized by lymphocytic infiltration into moisture secreting glands, manifesting in dry eyes and dry mouth (sicca syndrome). Estimated prevalence of Sjögren syndrome in the general population ranges from 0.03% to 0.08%. (1) Sjögren syndrome is diagnosed based on a combination of clinical and laboratory findings, such as the classification criteria by the 2012 American College of Rheumatology criteria (2) and the American-European Consensus Group revised criteria.

Although classification criteria can help guide diagnosis, the gold standard for diagnosing Sjögren syndrome remains clinical judgment. Renal involvement occurs in 16% to 67% of Sjögren patients (3); the reported large range reported being due to different definitions of the disease, and the inclusion of secondary Sjögren in some studies. The most common renal presentations are tubulointerstitial nephritis and distal renal tubular acidosis (RTA). Prevalence of tubulointerstitial nephritis in Sjögren syndrome with renal involvement can be as high as 65% to 71%. (4) This may present with hypokalemia, elevated creatinine, a relatively bland urine sediment with leukocyturia, features of proximal tubule dysfunction (Fanconi syndrome), or nephrogenic diabetes insipidus.

Distal acidification defects can occur in up to 40% of all Sjögren patients, (5) and usually exists in the setting of tubulointerstitial nephritis, but can also present in isolation. Distal RTA can result in severe hypokalemia and development of paralysis, cardiac arrhythmias, bulbar weakness, and respiratory arrest, all of which have been reported in Sjögren patients. (6, 7, 8, 9). The pathophysiology of the relationship between HBV and autoimmunity is not clear. It is known that infections can trigger autoimmune diseases and that HBV has been associated with the development of autoimmunity through immune complexes composed of hepatitis B antigens and antibodies and molecular mimicry mechanisms. The causal role of hepatitis infection triggering Sjogren's syndrome is under debate. (10)

CONCLUSION

Sjögren syndrome is one of many fascinating, pluriform autoimmune entities whose underlying pathophysiology remains incompletely understood. This case illustrates a presentation of severe symptomatic hypokalemia in the context of distal RTA associated with underlying Sjögren syndrome.

Our report emphasizes that although Sjögren syndrome is most often associated with chronic sicca symptoms, it may present for the first time with extra glandular manifestations which may be life threatening. The causal role of hepatitis infection triggering Sjogren's syndrome is under debate.

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