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Case Report

Sjögren's syndrome presenting as hypokalemic paralysis

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Introduction

Distal renal tubular acidosis (dRTA) is a known cause of hypokalemia, which may rarely be severe enough to present as hypokalemic paralysis. Sjögren's syndrome presenting for the first time with hypokalemic paralysis due to dRTA in a patient with no sicca symptoms is even rarer. The only evidence of underlying Sjögren's syndrome in our patient was positive serology and salivary gland histopathology.

Case report

A 39 year old female patient presented with acute onset painless, progressive proximal muscle weakness of all four limbs of one day duration. There were no features of sensory or

autonomic dysfunction. There was no history of diuretic or steroid use, preceding fever, diarrhea or animal bite. She denied dryness of eyes or mouth, history of renal stones or fractures. There was no similar history of paralysis in the past. She gave no past history of irradiation to head and neck. She had regular menstrual cycles and her two pregnancies had been uneventful. On examination, she was normotensive. She had areflexic flaccid quadriplegia with no cranial nerve deficit, sensory loss or autonomic dysfunction. She did not have goiter, enlarged parotid or lacrimal gland. On investigation, she had hypokalemia (S. Potassium 1.2 mEq/l), normal anion gap (11.5, Normal 8–16) metabolic acidosis (pH 7.3, HCO_3^- 12.3 mEq/L) with alkaline urine pH (8.0). Urinary potassium was 16 mEq/l, which in the setting of hypokalemia suggested renal potassium loss. Serum 25-OH Vitamin D levels was 34 ng/ml ($N > 30$ ng/ml). Serum Magnesium was 2.8 mEq/l (1.8–2.8 mg/dl). Blood urea was 28 mg/dl and creatinine 0.7 mg/dl. Estimated glomerular filtration rate (GFR) by MDRD formula was 99 ml/min. Ultrasound examination showed normal kidneys with no nephrocalcinosis or stones. Diagnosis of distal RTA was considered in view of normal anion gap metabolic acidosis, alkaline urine and hypokalemia. Urine biochemistry showed normal calcium, uric acid 16.3 mg/dl, phosphorous 19.60 mg/dl and no abnormal aminoaciduria. Serum ceruloplasmin was 29.9 mg/dl ($N = 27–37$ mg/dl). Slit lamp examination was negative for Kayser Fleischer (KF) rings. Autoimmune disease screening showed positive ANA, negative Anti-dsDNA and strongly positive Anti-Ro (SS-A) 93.7 U/ml ($N < 3.0$), and Anti-LA (SS-B) 75.87 U/ml ($N < 3.0$) suggestive of possible Sjögren's syndrome. Serology for Hepatitis B surface

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antigen (HBsAg), anti-Hepatitis C virus (HCV) and Human immunodeficiency virus (HIV) antibody were negative. Schirmer's test was normal suggesting normal tear secretion. Lip biopsy was done which showed lymphocytic infiltration commensurate with chronic sialadenitis (Fig. 1).

Diagnosis of Sjögren's syndrome was made based on the newly devised Sjögren's International Collaborative Clinical Alliance (SICCA) guidelines wherein 2 of 3 criteria were positive (Serology for Anti SS-A and SS-B antibody plus salivary gland biopsy in absence of symptoms).

She was treated with parenteral potassium followed by oral potassium citrate (2 mMol/Kg body weight) with good response. Her muscle weakness improved over the next 2 days with normalization of serum potassium and blood pH. A diagnosis of hypokalemic paralysis due to dRTA was made. Serology for Sjögren's syndrome was strongly positive although clinical and laboratory evidence for sicca syndrome was negative confirming the diagnosis of Primary Sjögren's syndrome.

In view of renal tubular involvement in the setting of Sjögren's syndrome, she was treated with oral steroids (Prednisolone 1 mg/kg/d) in anti-inflammatory doses for 4 weeks after which it was tapered off to a dose of 10 mg daily. She was followed up over the next 24 months when she remained asymptomatic and her serum potassium was normal. Her requirement of potassium citrate reduced over this period from the initial 2 mMol/kg to normokalemia without potassium replacement suggesting complete resolution of the tubular dysfunction. Dose of Prednisolone was tapered off to minimum of 5 mg daily with good response and complete remission of disease activity.

Discussion

This female patient presented with hypokalemic paralysis due to dRTA as the first and only manifestation of Sjögren's syndrome. Although asymptomatic for sicca symptoms, she had strongly positive serology for Sjögren's syndrome and lip biopsy for salivary gland histopathology was suggestive of the same etiology. Clinically and serologically, there was no evidence of underlying cause for secondary Sjögren's syndrome. She showed good response to replacement with potassium citrate and has shown almost near resolution of tubular defect after a course of immunosuppressive therapy with steroids. This case highlights the need to consider the possibility of Sjögren's syndrome in a case of RTA even without any other manifestation of the syndrome and response to immunosuppressive therapy in complete resolution of renal tubular dysfunction.

Distal renal tubular acidosis is a disorder of the distal nephron, which cannot lower the urine pH normally.¹ The underlying cause is due to the excessive back-diffusion of hydrogen ions from the lumen of the collecting duct to blood or there is inadequate transport of hydrogen ions. It is diagnosed by a normal anion gap metabolic acidosis with a simultaneous urine pH greater than 5.5. Hypokalemia may also occur because urinary concentration and potassium conservation also tend to be impaired. Severe hypokalemia causing paralysis as first presentation of dRTA secondary to

Fig. 1 – Photomicrograph of the submucosal salivary glands obtained by lip biopsy showing lymphocytic infiltration around the acini and the ducts.

Sjögren's syndrome is a rare occurrence. Similar cases have been reported as isolated case reports in literature.^{2–4}

RTA is a frequent extraglandular manifestation of primary Sjögren's syndrome with an incidence of about 30%.⁵ It may also be the first clue in the identification of an underlying autoimmune disorder, particularly Sjögren's syndrome.⁶ The pathogenesis of dRTA in Sjögren's syndrome is not very clear. Patients with Sjögren's syndrome with dRTA have interstitial nephritis with high levels of anti-carbonic anhydrase antibodies which affect the function of carbonic anhydrase in cortical collecting ducts.⁷ The acidification defect was the result of a lack of intact H⁺-ATPase pumps in the intercalated cells. Daily life long alkali replacement in a dose of 1–2 mEq/kg prevents acute hypokalemia as well as the chronic complications of osteomalacia and nephrolithiasis. Use of corticosteroids or other immunosuppressive drugs used for the primary disease have been shown to reverse the renal tubular defect, as seen in our patient. We used a course of steroid in this case although the dRTA was the only manifestation in this patient and she showed a good response to this treatment. She will be followed up for recurrence of tubular dysfunction and other systemic manifestations of Sjögren's syndrome.

Conflicts of interest

All authors have none to declare.

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