

Rare case of Sjögren's syndrome presenting for the first time with hypokalemic paralysis due to dRTA.

¹Dr. Dixitkumar D. Zalavadiya, ²Dr. Megha R. Ninama, ³Dr. Sudhansubhai P. Nayak, ⁴Dr. Subhashchandra K. Gadhavicharan

¹R1-General Medicine, ^{2,3}R2-General Medicine, ⁴Professor
General Medicine
C.U. Shah Medical College & Hospital

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.

CASE REPORT

-> A 33 year old female patient presented with acute onset painless, progressive proximal muscle weakness of all four limbs since one day duration. There were no feature of sensory or autonomic dysfunction. There was no history of diuretic or steroid use, preceding fever. She denied dryness of eyes or mouth. There was no similar history of paralysis in past. She gave no past history of irradiation to head and neck. She had regular menstrual cycles and her three pregnancy had been uneventful.

-> On examination, She was normotensive. She had areflexic flaccid quadriparesis 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 - 1.26 mEq/l). Positive urine anion gap(7.1 mEq/l). Metabolic acidosis (pH - 7.12, HCO₃ - 11.1 mEq/l) with alkaline urine pH. Urinary potassium was 15.16 mEq/l, which in setting of hypokalemia suggested renal potassium loss. Serum magnesium was 2.3 mEq/l. Serum creatinine 0.97 mg/dl. Estimated GFR - 73 ml/min by Cockcroft-Gault formula. Ionic calcium was 4.9 mg/dl, Phosphorus was 1.33 mg/dl (normal 2.5-4.9 mg/dl). Ultrasound showed normal kidneys with multiple left renal calculi. Slit lamp examination was negative for ophthalmic manifestation of vitamin A deficiency. ANA by IFA is positive and shows speckled pattern. BLOT-LINE ANA shows positive anti-RO52(SS-A), anti-RO60(SS-A) and positive anti-LA(SS-B) suggestive of possible Sjögren's syndrome.

-> Dignosis of Sjögren's syndrome was made based on newly devised Sjögren's International Collaboration Clinical Alliance (SICCA) guidelines wherein 2 of 3 criteria were positive. (serology for Anti SS-A and SS-B antibody in absence of symptoms.)

-> She was treated with parenteral potassium followed by oral potassium chloride (20 mEq PO TID) with good response. Her muscle weakness improved on next 2 days with normalisation of serum potassium and blood pH. On discharge we give oral potassium citrate 10ml PO TID and start Tab. HCQ (200mg) OD.

-> Diagnosis of hypokalemic paralysis due to dRTA was made. Serology for sjögren's syndrome was positive although clinical evidence of sicca syndrome was negative confirming diagnosis of Primary Sjögren's Syndrome.

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 positive serology for Sjögren's syndrome. Clinically and serologically, there was no evidence of underlying cause for secondary Sjögren's syndrome. She showed good response to replacement with potassium chloride. 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.

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 alkaline urine pH. 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 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. She will be followed up for recurrence of tubular dysfunction and other systemic manifestations of Sjögren's syndrome.

CONCLUSION

We report case of 33 year old female who presented with areflexic quadriparesis and no sicca symptoms with no significant past clinical history. She was found to have hypokalemia due to distal renal tubular acidosis and further diagnosed as case of Sjögren's syndrome.

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