Secondary Sjogren's Syndrome and Scleroderma Presenting as Renal Tubular Acidosis

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Abstract

We report a case of distal renal tubular acidosis in a twenty year old female patient of scleroderma and secondary Sjogren's syndrome. This patient presented with two episodes of flaccid quadriplegia which were associated with hypokalaemia and was later found to have an underlying scleroderma with secondary Sjogren's syndrome.

INTRODUCTION

Distal renal tubular acidosis is characterised by a clinical triad of hypokalaemia, hyperchloraemic metabolic acidosis and inability to lower the urinary pH below 5.5.1,5 The basic defect is an impairment of H⁺K⁺ ATPase proton pump at the renal tubules leading on to defects in acidification, urinary potassium loss and hyperchloraemic acidosis.1,4 In Sjogren's syndrome there is an interstitial nephritis which results in tubular dysfunction with defective H⁺ ion secretion and a distal renal tubular acidosis.1,4

CASE REPORT

A 20 years female was admitted on two occasions with acute onset of weakness of all four limbs. The first episode followed a febrile illness. She had symptoms of dry mouth and dry eyes at the time of admission.

There was no history of a similar weakness in any of the family members. On examination, the patient was conscious, her vital signs were stable. She had a pinched nose, pursed lips and sclerodactyly. There was skin atrophy and pitted scars were present on the fingers. There was no enlargement of the parotid or lacrimal glands. The patient had a hypotonic and areflexic paralysis of all four limbs without any sensory abnormalities, cerebellar signs and bladder involvement. There was no difficulty in swallowing or respiratory distress. All other systems were within normal limits.

Investigations prior to treatment

Random blood sugar 110 mg/dl, B urea 20 meq/l, s creatinine 1 mg/dl, S sodium 130 meq/dl, s potassium (day 1) 2.2 meq/dl, (day 2) 1.2 meq/dl, s bicarbonate (day 1) 15 mmol/dl, (day 2) 12 mmol/dl, s chloride 110 meq/l, anion gap 7.2, Urine pH 7.4 (fasting), blood pH 7.25, PaCO₂ 30 mm Hg, PaO₂ 95 mm Hg, 24 hr urine potassium 30 meq, 24 hr urinary calcium 93 mg/24 hr, 24 hr urinary phosphorus 562 mg/dl, urine aminogram normal.

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ANA (Elisa) 47 IU/ml (n<40), antiDs DNA (Elisa) 52 IU/ml (n<40), Rh factor <10 (agglutination) negative, anti Scl 70 (Elisa) 30 EU/ml (normal <20 EU/ml).
Anti Ro SSA antibody (ANA, IF method) positive, anti La SSB antibody (ANA, IF method) positive
CSF study normal, pulmonary functions normal, barium swallow normal.
Schirmer’s test positive with 5 mm length of wetting.
Lip biopsy: salivary gland infiltrated by a moderate lymphoplasmacytes with focal destruction of acinar epithelium. Features suggestive of Sjogren’s syndrome.
X-ray abdomen did not show calcification.
Ultrasound abdomen did not reveal nephrocalcinosis.
Renal biopsy was not done as patient recovered quickly and remained asymptomatic.
The patient made a rapid recovery when the metabolic imbalance was corrected with intravenous potassium followed by oral potassium citrate. She was then placed on oral bicarbonate, potassium and methyl cellulose eye drops. The patient is being followed up and remains asymptomatic.

Post-treatment results
S potassium 3.5 meq/l, S chloride 102 meq/l, urine pH 6.5, S bicarbonate 24 meq/L, S Sodium 135 meq/l, anion gap 12.5.

DISCUSSION
Sjogren’s syndrome described in 1933, is a triad of dry eyes, dry mouth and arthritis with inflammation and destruction of exocrine glands, lacrimal glands and salivary glands. Sjogren’s syndrome can be primary or secondary to other autoimmune diseases like rheumatoid arthritis, systemic lupus erythematosus, scleroderma and chronic active hepatitis.

In 9% of Sjogren’s syndrome patients renal involvement is observed. The commonest lesion is tubulointerstitial disease presenting as Fanconi’s syndrome, distal renal tubular acidosis or impairment of renal concentrating function. Glomerular disease is very rare, but can occur in secondary Sjogren’s syndrome in association with systemic lupus erythematosus or mixed connective tissue disease.

Distal renal tubular acidosis is characterized by inappropriately high urinary pH, hyperchloremic metabolic acidosis and nephrocalcinosis. Pathophysiological abnormality includes impaired H⁺ ion secretion by the distal nephron leading to reduced acidification of urine and a defect in H⁺ ATPase and H⁺ K⁺ ATPase. Hypocitraturia occurs due to increased absorption facilitated by acidosis and hypokalaemia. Hypokalaemia is caused by the defect in H⁺ K⁺ ATPase and secondary hyperaldosteronism due to sodium loss in urine. High urinary pH with hypercalciuria, hyperphosphaturia and hypocitraturia favours nephrocalcinosis. Chronic metabolic acidosis induces loss of bone material and reduced formation of 1, 25 dihydrocholecalciferol levels leading on to rickets and osteomalacia.
Distal RTA is treated with alkali to keep the serum bicarbonate level above 18 meq/dl. Alkali supplementation is as sodium bicarbonate or Shohl’s solution.
Potassium citrate is used if hypokalaemia correction is required, since citrate is metabolized to bicarbonate after absorption.

This patient had clinical and laboratory evidence of scleroderma with secondary Sjogren’s syndrome and hypokalaemic renal tubular acidosis presenting as acute flaccid paralysis. She had positive titres of ANA, anti Scl 70, anti Ro SS A, anti Ro SS B, a positive Schirmer’s test and a lip biopsy suggestive of Sjogren’s syndrome.

This case is being reported as a combination of secondary Sjogren’s syndrome with scleroderma is a rare presentation the usual combination being with rheumatoid arthritis in 33% cases.

REFERENCES