Acute Flaccid Quadriparesis Because of a Rare Systemic Cause

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Abstract

Sjogren’s syndrome is a chronic slowly progressive autoimmune disease characterized by lymphocytic infiltration of exocrine glands resulting in xerostomia and dry eyes. The syndrome has wide clinical spectrum from organ specific exocrinopathy to systemic manifestation. The disease can present alone or with other autoimmune diseases like RA, SLE, Scleroderma, autoimmune thyroid disease etc. Prevalence of primary Sjogren’s is 0.5–1% and of secondary Sjogren’s is 5-20%. Renal involvement is rare and can either be tubulointerstitial or glomerular. Based on biopsy reports in the available literature, tubulointerstitial nephritis (TIN) is the most common histological abnormality, followed by glomerulonephritis as a distant second.¹ Distal Renal tubular Acidosis is the most common manifestation of TIN.

We report a case of a 35 year female with acute onset motor weakness (quadriparesis) with hypokalemia with NAGMA with distal RTA. Patient was diagnosed with Secondary Sjogren’s and managed accordingly.

Introduction

Distal RTA is usually associated with a genetic defect or anatomic abnormality of the urinary system.² dRTA indicates a failure of the intercalated cells in the kidney collecting ducts to secrete hydrogen ions.³,⁴ If the secretion of protons is severely impaired, the secretion of other cations, including potassium, is increased to maintain electroneutrality. This explains why complete dRTA is

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often accompanied by hypokalaemia due to renal potassium loss, which may even result in hypokalaemic paralysis. Renal involvement is not commonly defined in Sjogren syndrome but if involved it mostly causes TIN which may present as Distal RTA with associated diurnal variation (vide Myasthenia Gravis). There was no history of sensory loss or bladder involvement. No history of trauma, backache, fever, vomiting, diarrhea, headache, convulsions, speech difficulty, facial weakness, abnormal body movement, root pain or exposure to STD. On repeated enquiry, history of painful swollen bilateral wrist joint was elicited. The joint pain persisted throughout and was only relieved after medication. Also on and off history of dry mouth and swelling of both jaw region since 2 years was noted. No history of similar episodes in the past, tuberculosis, diabetes, hypertension, thyroid disorders, seizure disorder and surgical intervention was present. The patient was vegetarian by diet and is non alcoholic and non smoker and had good appetite. No similar complaints were present in other family members.

On examination, the patient was conscious but confused, blood pressure was 110/70 mmHg, respiratory rate was 20/min and pulse rate was 50/min. General physical examination revealed pallor and dry oral cavity. Clubbing, cyanosis or lymphadenopathy was absent. From the aforementioned history, I examined the Neurological system first. Neurological examination revealed decreased tone in all four limbs with the power of 1/5 in all limbs and bilateral plantar reflex was mute. Both superficial and deep tendon reflexes were absent. No sensory and cranial nerve involvement, autonomic disturbance, or involvement of bladder was there. On P/A examination, abdomen was soft, nontender but bowel sounds were decreased. The rest of the systemic examination was within normal limits. After history and clinical examination, we sent the patient for routine blood investigations and NCCT head. The CT report was WNL and blood investigation revealed Hypokalemia. From the above mentioned reports we suspected hypokalemia as a cause of the patient’s acute motor weakness and the patient was further evaluated for the same. Arterial blood gas shows pH of 7.2, HCO3- 11 mmol/L, pCO2 17mmHg,
Na+ 140mmol/L, K+ 1.9 mmol/L, Cl- 114 mmol/L and anion gap of 14 mmol/L. The blood and urine parameters of the patient are tabulated under Table 1.

So after getting the ABG report, diagnosis of Normal anion gap hyperchloremic metabolic acidosis with severe hypokalaemia was made. Potassium replacement was started. Gradually (after 2 days) patient’s power started improving (Patient was able to sit and walk without support).

Further urine evaluation revealed urinary Na+124mmol/L, K+18mmol/L and Cl- 114mmol/L with urine anion gap of 28mmol/L(highly positive) s/o Distal RTA (type 1).Further evaluation revealed S.TSH 15.3 uU/ml, FT3 2.79 pg/ml and FT4 0.65 ng/dl s/oAITD and patient was started on L-Thyroxine 50ug OD. Patients RF came out to be positive and ANA quantitative index value was 5.43(Positive). After the above investigations, Reflex to ENA profile was sent (suspecting autoimmune cause for Distal RTA). The profile revealed speckled pattern with strongly positive SS-A, Ro-52 and SS-B/La. Schirmer’s Test revealed tear flow of 15mm in RE and 17mm in left eye (Normal Schirmer’s Test). Final diagnosis of Hypokalemia quadripareisis with distal RTA with secondary Sjogren syndrome associated with autoimmune thyroid disorder was made. The patient was started on sodium bicarbonate and KCl infusion resulting into improvement of hypokalemia and metabolic acidosis. This condition might have been precipitated by underlying urinary tract infection, which may increase the bicarbonate requirement and cause volume depletion and potassium loss.

**Discussion**

Sjögren’s syndrome is one of many fascinating, pluriform autoimmune entities whose underlying pathophysiology remains incompletely understood. This case illustrates presentation of severe symptomatic hypokalemia in the context of distal RTA associated with underlying Sjögren’s syndrome. Our report emphasizes that although Sjögren’s syndrome is most often associated with chronic sicca symptoms, it may present for the first time with extraglandular manifestations which may be life threatening conditions. In distal (type 1) RTA the nephrons lack the ability to secrete H+ ions and hence acidify the urine normally during spontaneous or induced metabolic acidosis. Distal RTA can be inherited or acquired. Inherited forms include autosomal-dominant, autosomal-recessive, or X-linked recessive, of which autosomal-dominant form causing mutations in the basolateral chloride-bicarbonate exchanger (AE1) has been identified as the most common form of inheritance. Acquired causes include hypergammaglobulinemic states, such as hypergammaglobulinemic purpura, cryoglobulinemia, fibrosing alveolitis, Sjögren syndrome, lupus, chronic active hepatitis, thyroiditis, Graves’ disease, primary biliary cirrhosis; disorders of calcium metabolism, e.g., primary hyperparathyroidism, vitamin D intoxication, idiopathic hypercalcuria, familial absorptive hypercalcuria, medullary sponge kidney. Tubulointerstitial diseases include leprosy, hyperoxaluria, chronic pyelonephritis, obstructive uropathy; and genetic diseases like Ehler Danlos syndrome, hereditary eliptocytosis, and genetic diseases like South Asian ovalocytosis, sickle cell disease, carbonic anhydrase II deficiency.

In distal RTA, the urinary ammonium excretion is inappropriately low for the level of acidosis as the defect in acidification decreases ion trapping required for ammonia secretion. Hence urinary anion gap (UAG = urinary Na + K - Cl) is positive. This differentiates from chronic diarrhea in which the UAG is negative due to enhanced renal ammonium excretion. In distal RTA, there is a tendency for renal calcium formation, nephrocalcinosis due to hypercalcuria, and hypocitraturia. Severely depressed plasma bicarbonate levels with a corresponding inappropriate urinary pH >5.5 differentiates from type 2 RTA. Finally the requirement of the patient to maintain plasma bicarbonate levels near normal was less than 1mEq/kg body weight which pointed towards distal RTA.

**Conclusion**

Hypokalemic periodic paralysis as a presenting complaint of Sjogren’s syndrome is an uncommon entity. Sjogren’s syndrome should be considered as a strong differential diagnosis while evaluating hypokalemic paralysis associated with metabolic acidosis. Early treatment along with proper evaluation is essential to prevent recurrence of episodes of hypokalemic paralysis which can be life-threatening. Our case report adds to the existing literature on renal involvement in a patient with Secondary Sjogren’s syndrome.

**Consent**

Informed written consent was taken from the patient’s husband for case report writing.

**References**