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"ABERRANT PRESENTATION OF SJOGREN'S SYNDROME LEADING TO HYPOKALEMIC PERIODIC PARALYSIS – A CASE REPORT"

General Medicine

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ABSTRACT

Tubulointerstitial nephritis (TIN) is the main renal involvement associated with primary Sjögren syndrome (pSS). TIN can manifest as distal renal tubular acidosis (RTA), nephrogenic diabetes insipidus, proximal tubular dysfunction, and others. We present a 50-year-old female with hypokalemic paralysis due to distal RTA (dRTA). She received symptomatic treatment and potassium supplementation with a good response. The evidence suggests that there is an inflammatory background and therefore a potential serious affection to these patients, such as hypokalemic paralysis. For pSS-associated Tubulo-interstitial nephritis, we suggest symptomatic therapy with potassium supplementation and corticosteroids combination in treatment.

KEYWORDS

Tubulointerstitial nephritis, distal renal tubular acidosis, Sjogren's syndrome, hypokalemic paralysis.

INTRODUCTION:

A systemic autoimmune connective tissue illness called Sjogren's syndrome is characterised by lymphocytic infiltration of the exocrine glands, which causes xeropthalmia and xerostomia. [1] Sjogren syndrome can be secondary to autoimmune conditions such rheumatoid arthritis, systemic lupus erythematosus, polymyositis, and systemic sclerosis or it can be primary if there are no other connective tissue disorders present. Although dry eyes and a dry mouth are the predominant clinical hallmarks of primary Sjogren's syndrome (pSS), additional glandular disease symptoms or signs are present in 75% of people with pSS. [2] The diagnosis of SS is dependent on a mix of clinical characteristics and laboratory findings; there is no specific diagnostic test for the condition. The most recent diagnostic standard for Sjogren's syndrome is the 2016 ACR/EULAR classification [3]. This strong criterion is the weighted average of five objective factors, including tests for objective ocular and oral dryness (two ocular tests, including Schirmer's test, and an oral test, each scoring one point), anti-Ro/SSA positivity, lymphocytic sialadenitis on salivary gland biopsy, with a focus score of 1 foci/mm^2, and both. An individual with either ocular or oral dryness with a score of less than 4 points is diagnosed with SS.

Tubulointerstitial nephritis (TIN) is the primary renal involvement associated with primary Sjogren syndrome (pSS). RTA is the most common clinical manifestation of TIN, which can also manifest as proximal tubular dysfunction, nephrogenic diabetic insipidus, and other conditions. [4] RTA has been found in 4.3-9% of pSS patients; it is more common in middle-aged women, and symptoms appear in two-thirds of these patients. dRTA (distal renal tubular acidosis) can be severe enough to cause hypokalemic paralysis in rare cases. It is even more unusual for a patient with Sjogren's syndrome to present with hypokalemic paralysis caused by dRTA. Hypokalemic paralysis is the first symptom of Sjogren's syndrome in 7% of patients. [5]

In this report, we described a rare case of pSS - associated tubulointerstitial nephritis, presenting as RTA. In addition to it, we describe their demographic, biochemical, immunological, and clinical characteristics along with their response to immunosuppression.

Case Presentation:

A 50-year-old female presented to casualty with acute onset flaccid paralysis of all 4 limbs from the previous day. she started experiencing weakness in the Right lower limb 2 days ago followed by the Right upper limb and subsequently involved all 4 limbs with no history of steroid and diuretic use, preceding fever, diarrhoea, or animal bite. No autonomic or sensory involvement. She had no history of renal stones or fractures There was no history of similar complaints in past. On examination she was afebrile, vitally stable conscious oriented did not have a goitre, parotid, or lacrimal enlargement, power was 2/5 in all four limbs bilateral plantar extensors, deep tendon reflexes +1 in all 4 limbs, sensory system intact, no features of the bladder, bowel involvement with no sensory deficits, all cranial nerve function intact, no speech disturbances, no facial deviation was present. On investigating her Sr. electrolytes, she had hypokalaemia Sr. potassium- 2.5 mEq/l, Sr. Sodium 149 mEq/l, her ABG values suggested Metabolic acidosis with urine pH 6.9. There was evidence of K+ loss in the urine (urinary potassium- 276 mEq/l (Normal 22-164 mEq/l) with Anion gap of 33.9 mEq/lit and urine anion gap of 20.6 mEq/l which suggested the possibility of Renal tubular acidosis and ECG showed ST depressions with shallow and broad T waves. NCV and EMG showed bursts of repetitive, spontaneous discharges firing from some motor unit possibility of Myokymic discharges.

We performed an ANA profile which was positive for SS-A and Ro52 antibodies. She was treated with intravenous potassium and syrup potassium citrate. The patient recovered from the treatment the very next day patient and was able to walk. This suggested the possibility of Sjogren's syndrome although she has no complains of swelling over face or dry mouth. Her Schirmer's test showed severe dryness <4mm right eye, <8mm left eye(less than 5 mm of wetting in 5 mins).

She was started on Prednisolone 10mg and potassium citrate syrup TDS and lubricating eye drops her condition improved and thus discharged would be on regular follow-up. dRTA treatment includes potassium restitution before alkali therapy because the last might aggravate hypokalaemia by enhancing the shift of potassium into cells and bicarbonate.

DISCUSSION:

Tubulointerstitial nephritis is the most common manifestation of renal involvement in primary Sjogren's syndrome. [6] There have been numerous case reports of Primary Sjogren's syndrome manifesting as hypokalemic paralysis caused by tubulointerstitial nephritis. [7] Rao et al 2006 investigated 31 cases of hypokalemic periodic paralysis, three of which had Sjogren syndrome. [8] Hypokalaemia as a presenting manifestation of pSS has been reported in 2% of cases. [9] The more common tubulointerstitial nephritis can manifest in a variety of ways, including a mild to moderate decrease in glomerular filtration rate, though progression to end stage renal failure has been reported with pSS TIN. [10]. Proximal tubular dysfunction with low molecular weight proteinuria [11] or a full-blown renal Fanconi syndrome [5] can result from tubular dysfunction (with bicarbonaturia, uricosuria, phosphaturia, glycosuria and low molecular weight proteinuria). Distal tubular disease can result in distal renal tubular acidosis [12] (along with hypokalaemia, nephrocalcinosis/kidney stones, osteomalacia, and a variable metabolic acidosis) or nephrogenic diabetes insipidus (NDI). [13]

Distal RTA is defined in the context of metabolic acidosis as the kidneys' failure to eliminate hydrogen ions from the urine, resulting in

abnormally normal or alkaline urine. The disorder can be identified by normal anion-gap metabolic acidosis and a positive anion gap in the urine. [7] The kidneys lose potassium, resulting in hypokalaemia. Even though distal RTA is common in Sjogren's disease, it frequently goes unnoticed because there are no symptoms. [7] In a study of 130 patients, Ren et al. discovered that distal RTA was present in 70% of those with primary Sjogren's syndrome. [14] Ren et al. supported our patient's clinically proven RTA. The most common electrolyte abnormality in dRTA patients is hypokalaemia. Hypokalaemia can be caused by a decrease in distal tubular Na delivery, secondary hyperaldosteronism, a defective H+-K ATPase, or bicarbonaturia. [15] There are no proven effective systemic immunosuppressive treatments for pSS, and the few randomised trials that have been conducted have been inconclusive or contradictory. The most common treatment for patients with pSS who have joint or skin disease is hydroxy chloroquine or methotrexate. Corticosteroids, anti proliferative agents, calcineurin inhibitors, cyclophosphamide, or B-cell depletion therapy have all been used to treat resistant or extra glandular disease [16]. Corticosteroids are critical in the treatment of pSS-related kidney injury. Corticosteroids (0.5 mg/kg/day) alone were reported to improve eGFR in patients with pSS-associated interstitial injury. [17] With potassium replacement therapy, our patient's symptoms improved. In addition, because her medical history revealed HPP symptoms for a long time and she had overt quadriparesis, we gave her steroids. Her complaints went away after steroid therapy, and they didn't come back after a follow-up. Steroid therapy, which has been shown to be effective in a few cases, should be considered in HPP cases that are unresponsive to replacement therapy and have recurring attacks. However, more detailed studies may be required to better understand the disease mechanism, and large sample multicenter randomised controlled trials should be conducted to assess the efficacy and safety of drugs such as hormones and immunosuppressants in these patients with various types of renal damage.

CONCLUSION:

Although pSS generally causes asymptomatic RTA, rarely, HPP, quadriparesis, and even respiratory arrest and death might ensue. It should be borne in mind that symptoms of SS might be very mild in some subjects and that HPP might be the initial disease manifestation in others. For pSS-associated Tubulo-interstitial nephritis, we suggest symptomatic therapy with potassium supplementation and corticosteroids combination in treatment.

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