

HYPOKALEMIA WITH RENAL TUBULAR ACIDOSIS TYPE I AS AN INITIAL PRESENTATION OF PRIMARY SJOJGREN'S SYNDROME

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ABSTRACT

Renal involvement either interstitial nephritis or glomerulonephritis have been seldom noticed in isolated cases of primary Sjogren's syndrome (pSS). In pSS, renal involvement is primarily due to a monolymphocytic tubulointerstitial nephritis (TIN) characterised by a predominance of T lymphocyte subsets and manifested by renal tubular dysfunction. In this instance, we describe a case of primary Sjogren's syndrome presenting with hypokalemia and renal tubular acidosis type I including relevant positive and negative history and laboratory evidence of renal dysfunction. This case shows that primary Sjogren's syndrome may present with systemic manifestations other than classical one.

KEYWORDS: Interstitial Nephritis; Glomerulonephritis; Hypokalemia; Type I Renal Tubular Acidosis; T-Lymphocytes; Sjogren's Syndrome.

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INTRODUCTION

Sjögren's syndrome is associated with a lymphocytic and plasmacytic infiltrate in the salivary and lacrimal glands, causing sicca syndrome. The prevalence of this disease is 0.5-1.0% in general population, middle aged females being affected more than males by a ratio of 9:1. This immune derangement can also affect the kidneys, producing an interstitial nephritis and defects in tubular function. Literature survey depicts the prevalence of extraglandular and renal involvement is about 30 and 9 percent respectively in primary Sjogren's syndrome.^{1,3} Since many patients present with extra-glandular clinical features, laboratory studies also play an important role in diagnosis of Sjogren syndrome, supported by clinical findings according to the American-European Consensus group.¹

We report a case of hypokalemia associated with renal tubular acidosis due to primary Sjogren's syndrome in a 30 year old female presenting with lower limb weakness.

CASE PRESENTATION

A 30 year old female was admitted in a tertiary care institution with weakness of both lower and upper limbs. The systemic examination of the nervous system indicated a decrease in tone and power of all the four limbs but no remarkable change in reflexes. Cerebrospinal fluid examination was found to be normal which excluded meningitis and encephalitis. Absence of any preceding history of viral infections excluded acute demyelinating neuropathy.

At presentation, her serum potassium (K⁺) concentration was 2.1 mmol/ L with an arterial pH of 7.23. Imaging studies of brain and spinal cord were normal. An ion-channelopathy was suspected and she was subsequently corrected for potassium deficit. However, there was no suggestive family history or any history of consanguineous marriage. Her urinalysis showed K⁺ about 26 mmol/ day (>15mmol), low specific gravity of 1005 and trace proteinuria. A normal routine and microscopic urine examination excluded urinary tract infection. There was an associated metabolic acid base disturbance as detected by the ammonium chloride challenge test. Six gm ammonium chloride (body weight 62 kg) was administered orally to the patient, and then the urinary pH was measured. Failure of the urinary pH to reduce below 5.5 established distal renal tubular acidosis (RTA). She also had elevated erythrocyte sedimentation rate and normocytic normochromic

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anaemia. She was thoroughly investigated for the associated renal tubular acidosis type I. Her anti-nuclear antibody (ANA) showed speckled pattern in 1:640 titre. She was strongly positive for the marker antibodies of Sjogren's i.e, Ro/SS-A and La/SS-B. A negative Manteaux test excluded renal Koch's disease. The histopathology of kidney biopsy indicated a diffuse interstitial inflammatory infiltrate in the renal cortex suggesting interstitial nephritis. A minor salivary gland biopsy from the lower lip revealed lymphocytic infiltration which further clinched the diagnosis. She responded dramatically to oral glucocorticoids (methyl prednisolone 1mg/kg body weight for 4 weeks) and was discharged. Sodium bicarbonate was added to her drug regimen to prevent nephrocalcinosis.

The patient in this case did not fulfil the classical symptoms of Sjogren's. Though systemic manifestations are rarely seen in Sjogren's disease, yet complications like hypokalemia, or Fanconi's syndrome may progress to life threatening renal impairment given the progressive nature of the disease. Therefore a very high index of suspicion on the part of the clinician and an appropriate laboratory backup were the cue to diagnosis.

DISCUSSION

Though the presence of renal involvement in primary Sjogren's syndrome has been documented since 1960s, yet estimates of true prevalence of renal involvement have varied widely.³ Studies on small retrospective cohorts and various case series reports have largely contributed to our knowledge of clinical presentation of renal involvement of primary Sjogren's syndrome. Górriz Molino⁴ in their case report have reported renal tubular acidosis type 1 and hypocalcemic paralysis, as the initial clinical manifestation of primary Sjögren's Syndrome. They investigated into the etio-pathogenesis of this and found immunological humoral factor as the inciting factor for nephropathy. Interstitial infiltrate damages the tubules and produces variable clinical manifestations like raised plasma creatinine concentration, Fanconi syndrome, distal (type I) renal tubular acidosis (RTA), nephrogenic diabetes insipidus and hypokalemia.⁵ Fulop and Mackay⁶ in their survey noticed a patient with RTA type had radiological and biochemical findings compatible both with osteomalacia and Sjögren syndrome. A study conducted by Goules and Masouridi⁵ revealed that

10 patients had interstitial nephritis, 8 patients had glomerulonephritis, and 2 patients presented with both entities on percutaneous renal biopsy. Though clinically significant renal involvement is infrequent in pSS but may have a less favorable prognosis.

CONCLUSIONS

Patients with hypokalemia should be carefully evaluated for associated metabolic acid base disturbance. Diagnosis of connective tissue disorder in such cases requires both biochemical and histopathological techniques. Timely diagnosis and treatment can be lifesaving and retard the sequel of Sjogren's disease, as in this case.

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CONFLICT OF INTEREST

Authors declare no conflict of interest.

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None declared.