A CASE REPORT OF SJÖgren’S SYNDROME

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ABSTRACT

Sjögren’s syndrome is an autoimmune disease. It is a chronic and slowly progressive disease. The main pathology being, lymphocytic infiltration of the exocrine glands, resulting in xerostomia and dry eyes. Though dryness of eyes, dryness of mouth and parotid enlargement are considered as triad of sjogren’s syndrome, rarely it can present as hypokalemic periodic paresis with renal tubular acidosis. Here, is a case report of patient having recurrent hypokalemic paresis, metabolic acidosis, type 1 renal tubular acidosis occurring secondary to sjogren’s syndrome.

Keywords: Sjögren’s Syndrome, Recurrent Hypokalemicparesis, Metabolicacidosis, Type 1 Renal Tubular Acidosis

INTRODUCTION

Sjogren’s syndrome is a multisystem involving autoimmune disease. Middle aged females are commonly affected, female to male ratio being 9:1. When the disease presents alone it is called primary Sjogren’s syndrome. Secondary Sjogren’s syndrome is associated with other autoimmune rheumatic diseases (Moutsopoulas and Tzioufas, 2015). Systemic or extraglandular features are seen in one third of patients with Sjogren’s syndrome. Renal involvement is one of the rare extraglandular manifestations of primary Sjogren’s syndrome. It usually presents as distal and proximal renal tubular acidosis, chronicinterstitial nephritis, tubularproteinuria (Goules et al., 2000).

CASES

38 years old female referred from private hospital on ventilator support secondary to quadriparesis with respiratory failure. On detail history taking she had history of recurrent quadriparesis. On examination she was drowsy and was responding to verbal commands in the form of eye opening. She had hypotonia in all four limbs, power was grade two in all four limbs, diminished reflexes and mute plantars. On admission BP was 110/90 mm Hg, HR was 85/min, RR was 26/min and saturation was 99% on ventilator SIMV mode. Since she had spontaneous respiratory efforts, she was taken on CPAP mode and gradually extubated in next 2 days. On admission blood biochemistry revealed Na+ 143 mmol/l; k+2.2mmol/l; RBS 116mg/dl. liver function test; renal function tests and complete blood counts were normal. ABGA revealed metabolic acidosis with Ph. of 7.13. ECG showed prominent u wave. She was treated with intravenous potassium correction. Patient recovered completely in next 3 days. On enquiring she had history of dryness of mouth, features s/o sicca syndrome. Auto antibodies test were done and she was Ro/SS-A-positive and La/SS-B-Positive. She had positive schirmers test.

Figure 1: Dryness of Oral Cavity

Figure 2: Dryness of Eyes

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DISCUSSION
Sjogren’s syndrome usually presents with features such as xerostomia, keratoconjunctivitis sicca, and parotid enlargement. The extra glandular manifestation includes, Arthralgia/arthritis-60%, Raynaud’s phenomenon-37%, Vasculitis-11%, Renal-9%, Lungs/lymphadenopathy-6%, Splenomegaly-3%, Peripheral neuropathy-2% and Myositis-1% (Moutsopoulas and Tzioufas, 2015). The first such case report is as early as Raskin et al., (1981). India’s first such case was reported in JAPI by Thomas et al., (1966) from CMC Vellore. Rao et al., (2006) studied 31 cases of hypokalemic periodic paralysis where 3 cases had sjorgen’s Syndrome. This indicates that 10% of people with hypokalemic periodic paralysis can have Sjogren’s syndrome which if diagnosed early has more management options available. Patient was treated with hydroxychloroquin, bicarbonate and potassium supplement and supportive measures. Rheumatologist, nephrologist, dermatologist, and gynecologist opinion taken and done accordingly. Patient recovered and was discharged within 8 days. Sjogren’s syndrome rarely presents with hypokalemic periodic paralysis secondary to renal tubular acidosis with respiratory failure, therefore timely clinical suspicion and early diagnosis is important.

REFERENCE
Case Report

