CASE REPORT

# SJÖGREN'S SYNDROME

Pyar Ali Fazel, Karim Ullah Makki, Husain Haroon, Saira Sheikh

**ABSTRACT**

Sjögren's syndrome is an autoimmune and a chronic inflammatory disorder characterized by the infiltration of the exocrine glands particularly the lacrimal and the salivary glands by the lymphocytes and the plasma cells resulting in xerostomia and xerophthalamia. We report a case of a lady who presented with right sided stroke involving left leg only. On further work up, the patient was found to have Primary Sjögren's syndrome and vasculitis was the cause of the stroke.

**KEY WORDS:** Sjögren's syndrome, vasculitis, stroke.

## INTRODUCTION

Sjögren's syndrome is an autoimmune disorder with both exocrine and non-exocrine features.

Primary Sjögren's syndrome occurs without an underlying disorder whereas secondary Sjögren's syndrome occurs along with other connective tissue disorders such as the SLE , rheumatoid arthritis , progressive systemic sclerosis and dermatomyositis/polymyositis. Other than these features, Sjögren's syndrome is also accompanied by many common and a few rare medical conditions such as primary biliary cirrhosis, chronic active hepatitis, celiac disease and thyroiditis1,2. The present case report describes an uncommon presentation with stroke.

## CASE REPORT

A 60 year old lady (a known case of hypertension for 11 years and hypothyroidism for 8 years) was admitted with weakness of left lower limb for 7 days. There was no history of fits, headache, incontinence, vomiting or altered conscious level. In systemic review, she complained of burning eyes and dysphagia for solid foods along with moderate pain in both knees, ankles and wrist joints for 11 years. On examination, she was hypertensive, mildly anemic with dry skin, dry mouth and bilaterally dry and congested eyes. Examination of abdomen, respiratory and the cardiovascular system was unremarkable. However in the central nervous system examination, there was decreased tone and reflexes, and power was reduced to 0/5 in the left lower limb alongwith the up going plantars. Rest of the CNS examination was normal.

Her hemoglobin was 10.8mg/dl, while TLC, platelets, ESR, blood urea, serum creatinine and electrolytes, urine D/R, LFTs and RBS were within normal limits. Viral markers (HBsAg, Anti HCV) were non-reactive. Chest X-ray and abdominal ultrasound were also normal. CT scan of brain showed a hypo dense area in the right frontal cortex suggestive of an ischemic infarct. Lipid profile was normal and the thyroid profile was consistent with hypothyroidism. However protein electrophoresis demonstrated an increase in the alpha-2 globulin and the gamma globulin fraction suggestive of the polyclonal gammopathy.

Serological reports revealed absence of detectable rheumatoid factor, AMA, anti-smooth muscle, anti-thyroglobulin and anti-thyroid microsomal antibodies in the serum. However high titers of ANA and auto antibodies to Ro/SSA and La/SSB antigens were found. HLA association with HLA-B5, DR 3 and DRw 52 was also present. So the lady was diagnosed as having primary Sjögren's syndrome with vasculitis.

## DISCUSSION

Sjögren's syndrome is more common among females with a female to male ratio of 9:1. Usual onset is in

Department of Medicine, Dow University of Health Sciences, Karachi.

**Correspondence:** Dr. Pyar Ali Fazel, Assistant Professor, Medical Unit II, Dow University of Health Sciences, Karachi, Pakistan.

E-mail: alifazil@hotmail.com

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4th or 5th decade, with overall prevalence being 0.5%-1%. It is associated with HLA-B8, DR 3 and DRw-52. Patients exhibited association with B5, DR3 and DRw-522.

Sjögren's syndrome is an autoimmune disorder of unknown etiology, characterized by the lymphocytic infiltration of the salivary and the lacrimal glands. Common clinical features are keratoconjunctivtis sicca, xerostomia, salivary gland enlargement and arthralgia. Rarer clinical features include anemia, leucopenia, thrombocytopenia, lymphadenopathy, lymphoreticular malignancy, hepatomegaly, vasculitis, neuropathy, myositis, fibrosing alveolitis, glomerulonephritis and renal tubular acidosis3.

Some important serological markers are associated with Sjögren's Syndrome (SS). Anti RO/SSA are found in 80-90% of patients with Sjögren's antibodies syndrome. Anti LA/SSB are found in 50% of patients. Anti nuclear antibodies are found in 90% of patients. Rheumatoid factor is also frequently found. Anti alpha fordin presence is more sensitive and specific for Sjögren's syndrome than anti-LA or anti-RO. Anti acetylcholine receptor antibodies are found in many patients with Sjögren's syndrome. Antibodies to islet cells antigens are currently not recommended but being evaluated to differentiate SLE form SS as it is supposedly more raised in SS versus SLE. Patients with Sjögren's syndrome are found to have a wide range of auto-antibodies against joints, salivary ducts, gastric parietal mucosa, thyroid and a number of other body organs.

Disease is diagnosed by reduced salivary flow rate and the reduced lacrimal secretions demonstrated by the Shirmer's test.

If necessary, diagnosis can be confirmed by demonstrating focal lymphocytic infiltration in the minor salivary glands in a lip biopsy5.

Management includes use of hypermellose and other artificial tear drops for xerophthalmia. Soft contact lenses are worn for corneal protection. Occlusion of lacrimal ducts is occasionally needed. Treatment of xerostomia is more difficult and salivary substitutes are not much effective. Corticosteroids and immunosuppressive agents are only occasionally used to control severe extra-glandular manifestations5.

Prognosis of Sjögren's syndrome is quite good and not life-threatening. It ranges from mild annoyance due to ocular and oral symptoms to risk for blindness and major system involvement.

Major system involvement may remain static, worsen or even regress. It is important to note that there are increased chances of developing lymphoma particularly with the primary form of the disease6.

## REFERENCES


1. Haralampos M Moutsopoulos. Sjögren's Syndrome, In: Harrison's Principles of Internal Medicine, 16th edi: McGraw Hill, USA 2005; 1990-3.

2. Vitale C. Classification criteria for Sjögren's Syndrome: A revised version of European criteria proposed by the American European consensus group. Anne Rheum Dis 2002; 61: 554.

3. Ioannidis J P. Long-term risk of modality and lymphproliferative disease and predictive classification of primary Sjögren's Syndrome. Arthritis Rheum 2002; 46: 741.

4. Kumar P, Clark M. Sjögren's Syndrome and keratoconjunctivitis sicca. 5th ed: W. B. Saunders, 2002; 564.

5. Tierney LM Jr., McPhee SJ, Papadakis MA. Sjögren's syndrome.Current Medical Diagnosis and Treatment. 43rd edi: McGraw Hill, USA. 2004;811-12.

6. Ramos-Caslas M. Primary Sjögren Syndrome, hematologic patterns of disease expression. Medicine (Baltimore) 2002;81:281, [PMID: 12169883].