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A Case Report On Sjogren's Syndrome

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ABSTRACT:

Sjogren's syndrome is an autoimmune disease characterized by sicca symptoms, usually dry eyes and dry mouth resulting from infiltration of lacrimal and salivary glands. The disease may be primary (idiopathic) or secondary Sjogren's syndrome. The exact etiology is not known. This report focuses on a brief introduction and the case report of a 52 year old female patient who was brought to our hospital for the management of Sjogren's Syndrome presenting with Interstitial Lung Disease. The physician chose to treat her symptomatically however she showed mild improvement and she was discharged.

KEYWORDS: Sjogren's syndrome, Xerostomia, Xerophthalmia, Interstitial Lung Disease

INTRODUCTION:

Sjogren's syndrome (SS) is a chronic autoimmune disease characterized by lymphocytic infiltration of salivary and lacrimal glands that results in dry mouth (Xerostomia) and dry eyes (Xerophthalmia).^[1] It occurs worldwide and in all the ages with a female to male ratio of 9:11. It has a prevalence ranging from 0.1 to 4.8% in various populations and a peak incidence in the fourth and fifth decades of life.^[2] The disease may be primary (idiopathic) or secondary to other diseases like rheumatoid arthritis (RA), systemic lupus erythematosus (SLE), systemic sclerosis (SSc), polymyositis or biliary cirrhosis.^[3] Similar to other systemic autoimmune diseases, Sjögren's syndrome develops in genetically predisposed individuals, who are exposed to various environmental factors resulting in a dysfunctional immune system response to self-antigens.^[4] The exact etiology and pathophysiology of the disease is still unknown. Auto antibodies SS-A and SS-B, genetic

predisposition are believed to result in SS1.^[1] The impaired immune system cascades by activation of innate immune response along with activation of glandular epithelial cells that in turn leads to activation of B and T lymphocytes resulting in eventual damage of the target organs (4). Respiratory involvement has been reported in primary SS in 9% to 75% of cases depending on the sensitivity of the variables studied and the criteria used to define the abnormality.^[5] The prognosis of Sjogren's lung disease varies on the type of pulmonary involvement. Cystic lesions are associated with good survival and few complications as compared with ILD or lymphoid interstitial pneumonia that carries a poor prognosis in terms of not only survival but also quality of life.^[6] The only signs considered markers for Sjogren's are anti-SSA and anti-SSB antibodies, and while anti-SSB is more specific, elevated levels of anti-SSB are actually less common in patients

with Sjogren's syndrome than anti-SSA, leading to some ambiguity in diagnosis.^[7] There is no definitive treatment for SS. Alternative therapies, such as use of lubricants and artificial saliva, increased fluid intake, and salivary stimulation,

CASE REPORT:

A 52 year old female patient was admitted in the Hospital with the chief complaints of dryness of mouth and eyes. Patient had history of morning facial puffiness. Patient also had complaints of cough and joint pain mainly joints involvement like knee, ankle.

On general examination patient was found to be pallor, clubbing. Pitting edema, dry tongue. Systemic examination was found normal, no abnormality was detected. Patients vital were normal whereas blood pressure was found to be (126/60 mmHg).

Investigations revealed that her Hb was 11.1g/dl and ESR 45mm/hour. Her ANA blot (antinuclear antibody) was positive. Her chest X-ray revealed reticulonodular shadows in B/L lower zone. Her CT chest revealed multiple honey comb cysts with peribronchovascular interstitial thickening involving all basal segments.

The diagnosis of Sjogren's syndrome and Interstitial lung disease was made and she was initially treated with NSAID Aceclofenac 100mg orally, Lubricant Carboxy methyl cellulose eye drops and also with antibiotics Cefotaxime 1.5g IV BD and Azithromycin 500mg IV BD. On Day 3 she had complaints of joint pain and she was treated with NSAID Tramadol 100mg IM by which complaints of pain decreased. On examination she was clinically normal and discharged.

DISCUSSION:

Sjogren's syndrome (SS) is named after the Swedish ophthalmologist who first reported on this condition, is a relatively common autoimmune disease, affecting up to 2 million people in the United States.^[7] According to the European

are used to relieve the symptoms of sjogren's syndrome. However, these treatments are palliative could help to relieve the discomfort in the patients.^[8]

Community Study Group, the diagnosis of the Sjogren syndrome should be based on the results of: 1) biopsies of the minor salivary glands and/or lacrimal glands; 2) examination of the oral cavity consisting of sialography (in which a sponge is chewed for 2 min by the patient and then weighed) combined with salivary-gland scintigraphy; eye examination consisting of the Schirmer test plus the Bengal rose test or the fluorescein test; and 4) assays for anti-Ro/SSA or anti-La/SSB antibodies.^[9] In patients with SS, the immune system attacks both the salivary and lacrimal glands, that causes dry mouth and dry eyes. There is no known cause of Sjogren's syndrome, and no preventative treatment is known.^[7] Respiratory involvement has been reported in both primary and secondary forms of SS. Desiccation of mucosa and changes in the bronchial glands, similar to the pathology in salivary glands, was described by Sjogren way back in 1933.^[10] The respiratory involvement in primary SS can be in the form of interstitial lung disease (ILD).^[3] Currently the management of SS is primarily symptomatic.^[1] For dry eyes, artificial tears or methyl cellulose eye drops can be used. A proper oral hygiene has to be maintained and the use of antibacterial mouth wash has to be initiated.^[11] Patients, particularly those with dry mouth, must be regularly monitored for tooth decay, as dental caries are quite common and can progress to severe decay if left untreated.^[7] NSAIDs are often a first-line therapy used in Sjögren's and reduce inflammation which is often high in Sjogren's. NSAIDs reduce the production of prostaglandins that promote inflammation and pain.^[12] The importance of anti-inflammatory agents cannot be overstated because of their efficacy frequently as life-saving drugs in diseases such as cancer, diabetes, arthritis, and rheumatic fever, etc.^[13] More severe systemic symptoms, especially of the internal organs, can be treated

with corticosteroids or other immunosuppressive drugs, but must be used with caution due to the increased risk of infections.^[12]

CONCLUSION:

Sjogren syndrome is a systemic autoimmune disease that affects the exocrine glands. Diagnosis of the syndrome is based on biopsy of the minor salivary glands and/or lachrymal glands, examination of the oral cavity and eyes, and

autoantibody positivity. Early diagnosis and management is important as it may help in the prevention of severe complications. Patient was managed symptomatically based on ANA blot, chest X-ray, CT chest. Patients with Sjogren's syndrome need multidisciplinary management and often need a referral to different specialists such as ophthalmologist and primary care physician. Therefore appropriate referral should be made in order to manage the systemic complications.

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