

Sjogren's Syndrome: A Case Report

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Abstract:

Sjogren's syndrome is AN AUTOIMMUNE DISORDER associated with hashimoto. Here is a case of 43 years female who had a history of hypothyroidism since 1yr 6months on medication thyroxine 50 mcg. patient had initial complaints of xerostomia and xerophthalmia. Later developed dysphagia and cough.

Keywords: Sjogren's syndrome, xerostomia, xerophthalmia.

Date of Submission: 04-07-2020

Date of Acceptance: 19-07-2020

I. Introduction:

The synonyms for sjogren's syndrome[SS] are "Mikulicz disease or sicca syndrome".It was first described by Swedish ophthalmologist henrick sjogren(1899-1986).⁽¹⁾It is chronic,systemic autoimmune disease which is characterized dry mouth(xerostomia) and dry eyes(xerophthalmia) which results from by infiltration of the lacrimal glands and salivary glands by lymphocyte.⁽⁵⁾It is associatedwith hypergammaglobulinemia,autoantibody production, lung disease,mild kidney disease and eventually lymphoma.⁽⁷⁾ In women it is most common in the fifth decade of life.⁽³⁾ In various populations,it has a prevalence ranging from 0.1 to 4.8%.primary and Secondary are two forms SS may exist, which is associated with other

autoimmune diseases such as rheumatoid arthritis, systemic lupus erythematosus (SLE) and systemic sclerosis. Salivary and lacrimal glands destruction due to infiltration and proliferation of CD4+ T cells, B cells and plasma cells leads to acinar atrophy and results in xerostomia and keratoconjunctivitisicca.⁽⁵⁾ in xerostomia and keratoconjunctivitisicca.⁽⁵⁾

EPIDEMOLOGY:

Approximately 23.5 million people in the United States have some type of autoimmune disorder According to the National Institutes of Health (2012).Some 4 million people currently in this country have Sjögren's syndrome. After the onset of symptoms,approximately half of all persons with Sjogren's syndrome are undiagnosed for several years.⁽⁴⁾

CLINICAL PRESENTATION:

The clinical manifestations of the primary sjogren's syndrome are seen alone,where as secondary sjogren syndrome was associated with another autoimmune disease most commonly rheumatoid arthritis,systemic extraglandular manifestations also occur in third of the cases.⁽¹⁾ Dry eyes and Dry mouth, also referred to as xerophthalmia (or keratoconjunctivitis sicca [KCS]) and xerostomia, respectively are the hallmark symptoms of this disorder.⁽¹⁾In 400 patients with sjogren syndrome ,98 percent presented with xerostomia, and 93 percent with xerophthalmia, in a large prospective cohort study. Dryness, grittiness, pruritus, and foreign body sensation are included under eye symptoms.⁽²⁾ Permanent eye damage can be caused due to lack of tears.⁽¹⁾ Difficulty speaking, eating, or swallowing, are oral symptoms and sips of water frequently may be needed.The patient's eye may show conjunctival injection on physical examination because there may be ocular inflammation independent of lacrimal gland involvement.Clouding of cornea may be seen in more severe cases. Decreased salivary pool and dry mucous membranes,are early oral findings which can progress to erythema, fissuring,and ulceration. multiple dental caries may also have included in the patient as a result of decreased salivary flow. By promoting dental remineralization, providing antimicrobial activity against cariogenic bacteria, and maintaining a physiologic oral pH level,saliva prevents dental caries. Tender or swollen parotid glands may be present.acute suppurative sialadenitis,mumps, tuberculosis, sarcoidosis, and lymphoma are other causes of enlarged parotid gland.⁽¹⁾Extraglandular symptoms may be also present in patients (Table 1).^(1,2)

TABLE:1 EXTRAGLANDULAR SYMPTOMS OF SJOGREN'S SYNDROME^(1,2,8)

CLINICAL SIGNS AND SYMPTOMS	PERCENTAGE(%)
Arthralgia or nonerosive arthritis characterized by tenderness, swelling, or effusion of peripheral joints	37-75
Gastrointestinal symptoms (reflux, dyspepsia, diarrhea,constipation)	54
Autoimmune thyroiditis	15-33
Pulmonary disease (chronic cough, recurrent bronchitis with chronic diffuse interstitial infiltrates on radiography, abnormal spirometry, pulmonary alveolitis or fibrosis on computed tomography)	29
Raynaud's phenomenon	16-28
Cutaneous vasculitis	12
Peripheral neuropathy	7
Lymphadenopathy (enlarged lymph nodes in cervical, axillary, or inguinal region)	7
Renal involvement (proteinuria, renal tubular acidosis,interstitial nephritis, glomerulonephritis, abnormal urinalysis)	6
Fever not associated with infectious process	6

DIAGNOSIS: In patients who present with signs and symptoms of oral and ocular dryness and who test positive for antibodies to the anti-SS-A or anti-SS-B antigen, or who have a positive salivary gland biopsy the diagnosis of primary Sjögren syndrome is strongly suggested.⁽²⁾Table 2 lists the frequency of positive results for common laboratory tests in Sjögren's syndrome.

TABLE: 2 FREQUENCY OF POSITIVE LABORATORY TEST RESULTS IN PRIMARY SJOGREN'S SYNDROME. ^[2]

TESTS	FREQUENCY(%)
Antinuclear antibody	55-97
Anti-SSA(Ro)	16-70
Anti-SSB(La)	7-50
Rheumatoid factor	32 -90

Antibodies to the anti-SS-A and anti-SS-B antigens are not specific to Sjögren syndrome should be noted; they may be present in persons with other diseases(e.g., lupus) and in healthy persons.⁽²⁾The criteria for classification of sjogren's syndrome,which is recent requires a positive minor salivary gland biopsy or a positive anti-SS-A or anti-SS-B antigen test.⁽²⁾The additional autoantibodies in SS to salivary gland protein 1 (SP-1), carbonic anhydrase 6 (CA6) and parotid secretory protein (PSP) were identified.^[7]

Antibodies to SP-1, CA6 and PSP also produced by patients with SS. These antibodies were found in 45% of patients meeting the criteria for SS who lacked antibodies to Ro or La .⁽⁷⁾ Although Sjögren syndrome diagnosis may be suggested by the patient history and physical examination, multiple objective tests are there to help with the diagnosis. In the family physician's office,these tests are not commonly performed.^(1,2)

The Schirmer test or the rose bengal test are used for the evaluation of eye symptoms. The Schirmer test involves placing a sterile filter paper strip for 5minutes beneath the lower eyelid.The test is positive if the moistened area measures less than 5 mm.An ophthalmologist usually performs the rose Bengal test;rose bengal dye 1% is instilled and The integrity of ocular surface is evaluated by quantitatively scoring the staining of the conjunctiva. Devalitized corneal and conjunctival epithelial

cells are stained by rose Bengal dye..KCS is identified by the test when minimal ocular symptoms are present.A diminished tear meniscus can be identified by a routine slit-lamp evaluation.^(1,2)

By nonstimulated whole saliva flow collection,oral dryness can be evaluated objectively,in which the patient spits into a graduated test tube every minute for 15 minutes.In 15 minutes collection of less than 1.5 mL is said to be a positive result.other tests include Contrast sialography and Scintigraphy, which visualizes the salivary glands and ducts via contrast dye injection into the Stensen duct, and which evaluates salivary gland function by measuring sequential uptake and excretion of technetium 99m respectively.^(1,2)

Minor salivary gland biopsy of tissue taken from the patient's lip is not always necessary,although once considered the gold standard for diagnosis of Sjögren syndrome.At least one focus of dense, inflammatory infiltrate containing at least 50 lymphocytes per 4 mm is defined as a positive biopsy. In ambiguous cases or when therapy beyond symptom management is being considered the lip biopsy may be useful.^(1,2)

The confirmatory test ultrasound examination of salivary glands which is simple has added advantage of being noninvasive with no complications. The parenchyma of the gland demonstrates multiple, small, 2-6mm hypoechoic lesions which are representative of lymphocytic infiltrates. By ultrasound, complications of the disease such as extranodal lymphomas can often be detected as larger 1-4cm hypoechoic intraparenchymal masses.⁽¹⁾

For diagnosing sjogren's syndrome a radiological procedure can also be used as a reliable and accurate way. A contrast agent is injected into the parotid duct (of Stensen), which is a duct opening from the cheek into the vestibule of the mouth opposite the neck of the upper second molar tooth. Sjogren's syndrome is indicated by widespread puddling of the injected contrast scattered throughout the gland.⁽¹⁾

The absence of precipitins to DNA and the absence of the "membranous" or "peripheral" pattern of immunofluorescence (associated with anti-DNA antibodies) has been reported in Sjogren's syndrome. In patients with Sjogren's syndrome and renal disease classical hyperglobulinemic purpura occurred frequently, and has been reported previously in association with either Sjogren's syndrome or with renal tubular acidosis. These three features (sicca syndrome, non-thrombocytopenic purpura, renal tubular acidosis) may constitute a clinical triad, and patients with any one of these should be thoroughly evaluated for the other components.⁽⁶⁾

TREATMENT - Non pharmacological treatment- Mouth should be kept well lubricated by sipping water frequently or using sugar free gums.^[8] Lacrimal substitutes such as hypermellose can be used in night for lubrication^[9]

To protect patients from filamentary keratitis soft contact lenses can be used for corneal protection^[9]. Use chlorhexidine mouth wash to free from dental caries^[10]

In patients with Sjogren syndrome 35% of patients also develop Raynaud's phenomenon [skin discoloration] hence few measures should be taken

Wear sunscreen factor 15 or higher if skin is sensitive or if comes out. Exercise and cessation of smoking. Avoid strong soaps if skin is dry and use emollients

Keep eyelids clean to maximize oil secretion. Wear sunglasses and keep rooms with good humidity with plants or bowl of water^[10]

Pharmacological Treatment:

For topical ocular symptoms topical 0.05% cyclosporine improves signs of dryness pilocarpine 5mg orally /QID cevimamine hydrochloride 30mg orally /TID improve xerostomia symptoms^[5,8]

Paraffin based ointments and mucolytic agents such as acetyl cysteine eye drops may be helpful or stickiness of eyes^[10] if dry mouth leads to fungal infections nystatin is recommended^[10]

Atropinic drugs and decongestants are contraindicated as they decrease salivary secretions^[8]

Hydroxy chloroquine 200mg /BD can be used as long term treatment from 6 months to many years^[8,9]

Corticosteroids can be used if systemic complications were present

Table-3^[3]

	mild	moderate	severe
Eye	Patient education, preventive measures, lubricant eyedrops	topical cyclosporin	punctal occlusion, muscarinic agonists
Mouth	patient education, preventive measures, saliva substitutes, gustatory stimulants	muscarinic agonists	
Other location	Patient education and preventive measures, N-acetyl cysteine		

Management:

Postoral oral hygiene is necessary for oral candidiasis and lubricants are useful in treating vaginal dryness^[9]

CASE STUDY-

A female patient of age 43 years 47 kg was admitted with chief complaints of xerostomia, xerophthalmia. She had a past history of hypothyroidism since 16 months on medication levothyroxine sodium. ANA blot capsule revealed positive for ss-A, ro-52, ss-B antibodies. potassium levels were slightly decreased [3.8mmol/L], patient also suffers from metabolic acidosis

Based on subjective and objective evidence it was diagnosed as sjogren syndrome. Her treatment regimen includes syp-protate 15ml with 100ml of waterPO /TID, tab-sobinix-DS po/QID, tab- shelcal -500mg /po /OD, tab thyroxine 100mcg /PO /OD,tab hydroxychloroquine 200mg/ PO/BD. After 2 months of diagnosis patient develop dental caries[systemic symptoms]further prescribed with syrup mucaine gel 2tsp/PO/TID and tab –wysolone 20mg/PO/OD in morning with tapering doses.

After 8months of treatment she further developed

Cough with sputum white, mucoid, non foul smell, and throat pain since 1 week not associated with diurnal and posturnal variations

C/O fever high grade associated with chills since 1 week and rise in temperature at evening

C/O of voice change since 3-4 days

C/Ochest pain associated with cough

C/O of loss of apetite and weight loss

C/O dysphagia and pain while swallowing since 4 days, heart burn and burning sensation in throat. symptoms aggravate with cough

C/O ulcer over right angle of mouth[patient was not cooperative for VLS examination]

She was further prescribed with betadine mouth gargle wash, dentogel ointment, TUSQ lozenges and tab-MONTEC-LC to relive from symptoms

II. Discussion

Sjogren syndrome is chronic ,systemic auto immune disease with dry eyes and dry mouth. its pathophysiology include immune system activation[humoral and cellular]but the exact mechanism is unknown. it involves both T and B cells. It involve organ specific and non organ specific autoantibodies which damage the tissue function. organ specific autoantibodies damage antigens of salivary ducts, pancreas, erythrocytes, gastric mucosa, prostate and nerve cells.non organ specific include rheumatoid factors,antinuclear antibodies and antibodies of RNA protein complexes SS_A[Ro] and SS-B[La]⁽⁵⁾

Hence through serological investigations presence or positive results of SS-A and SS-B confirms the diagnosis of sjogren syndrome.labial gland biopsy is also best confirmative test for sjogren syndrome.

Patient experience xerostomia due to damage of salivary ducts by organ specific auto antibodies. Thus due to less salivary production ulcers in oral cavity occurs. Hematological mild abnormality due to damage for erythrocytes[HCT-27.7]

III. Conclusion

Patient with sjogren syndrome may face systemic complications hence should refer more number of muliti-specality doctors and management should be done patient specific and treated symptomatically .Patient and care taker must be educated with change in lifestyle modifications for complete management of disease.

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B.Navya, et. al. "Sjogren's Syndrome: A Case Report." *IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)*, 19(7), 2020, pp. 08-11.