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Sjogren's Syndrome

KEYWORDS: Sjogren's syndrome (SS), keratoconjunctivitissicca (KCS), Schirmer'stest, Autoimmuneexocrinopathy

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ABSTRACT *A 55 year old female presented with typical history of dryness of mouth, poor lacrimation, dryness of nose and dysphagia. No obvious parotid enlargement or lymphadenopathy was present.ANA was borderline and RA test was positive. Her lip biopsy wasconclusive of Sjogren's syndrome. Also her ophthalmic evaluation showed positiveSchirmer'stest.Considering the clinical features, positive Schirmer's test and positive lip biopsy, the diagnosis of SS was established. Methyl cellulose 0.5% eye drops were given to the patient. She was encouraged to take liberal amount of water in between meals and during meals.*

Sjogren's syndrome (SS) is a chronic systemic disorder characterized by polyglandular tissue destruction that causes keratoconjunctivitissicca (KCS) and xerostomia (dry mouth). Patients with primary SS have KCS and xerostomia, whereas those with secondary SS have KCS, xerostomia and an autoimmune disease. Sjogren's syndrome can affect any age but the onset is most common in middle age or older, and 2-5% of people aged 60 years and above have primary SS. The prevalence of SS is 4 cases per 100 000 of population with nine times as many females as males affected. The disease affects predominantly middle-aged women in the peri- or post-menopausal period. In addition to the primary syndrome, 30% of patients with rheumatoid arthiritis, systemic lupus erythematosus and systemic sclerosis suffer secondary SS. [1]

CASE REPORT

A 55year oldfemale presented withdrynessof mouth,poor lacrimation and dryness of eyes, joint pain, difficulty in swallowing, occasionalfever&drynessofnose. Patientwas not a known case of diabetes, hypertension ormixedconnectivetissuedisorder. No other members of the maternal side of the familyhadsuchsymptoms. On examination she was conscious, oriented, pulse-82/min, BP-130/80 mm Hg and mild pallor was noted. No obvious parotid enlargement or lymphadenopathy was present. Her laboratory investigations showed Hb-10.6 gm%, TLC-9400/cu.mm, P-62%, L-33%, E-2%, Platelets-4,29,000/cu.mm, LFTs were normal, RFTs were normal, HIV was non-reactive &HBsAg was negative, ANA wasborderline & RA test was positive. Her parotid USG showed smallerglands withincreased echogenicity. Herautoimmune testresults were SSA(Ro) : 1.005 (Negative) and SSB(La) : 0.07 (Negative). Ophthalmic evaluation showed severe dry eyes with Schirmer's test results of 3 mm and 2 mm in right and left eye respectively at the end of 5 min. Lower lip biopsy was done which was consistent with Sjogren's syndrome. The subepithelial tissue showed minor salivary glands and skeletal muscle tissue underneath and majority of these salivary glands showed periductal focal and dense lymphoplasmocytic infiltrates.

Considering the clinical features, positive Schirmer'stest and positive lip biopsy, the diagnosis of SS was established.Methyl cellulose 0.5% eye dropsweregiven to the patient.She was encouraged to take liberal amount of water in between meals and during meals.Steroids were not given as she did not have any evidence of vasculitis. Patient improved symptomatically after the treatment and was advised follow-up every month for first 3 months and 6 monthly thereafter.

DISCUSSION

Sjogren's syndrome (SS) is a chronic autoimmune exocrinopathy characterized by lymphocytic infiltration of lacrimal and salivary glands. Initially labelled as 'KeratoconjunctivitisSicca' (KCS). It has two types – primary (multisystem autoimmune disease) and secondary – (due to SLE; rhumatoid arthritis, systemic scleritis or polymyositis).

Primary Sjogren's syndrome is a multisystemic autoimmune disease with complex interaction between innate and adaptive immunity & salivary gland dysfunction.Environmental factors and viral infections trigger the inflammation in a genetically predisposed individual. There is infiltration of glandular tissue by CD4+T lymphocytes. The glandular epithelial cells express high levels of HLA-DR which presents viral antigens auto antigens to the CD4+T lymphocytes. This sets in process of inflammatory cascades with cytokine production notably interferon gamma (IFN-8 and interleukin2). Two third of patients also have ANA and RA factor positive. **Patient may usually present with the following symptoms :**

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System involved	Symptoms
Ocular	Dryness of eyes due to poor lacrimation can lead to redness of eyes, photophobia and corneal ulcerations.
Oral	Dryness of mouth due to poor salivation requires frequent water intake, dryness may extend upto throat and can be a cause for dysphagia. Dental carries and oral infectious are common due to poor salivation.
Nose	Upto 40% of cases can develop dryness of nose.
Systemic manifestations	a. Musculoskeletal: Arthralgia, Myalgia, Arthritis b.Pulmonary: Xerotraceha can cause dry cough, interstitial lung diseases, recurrent bronchitis. c.Gastrointestinal: Dryness of pharynx and esophagus, gastroesophageal reflex, pancreatitis (Rarely). d. CNS: Myelopathy, optic neuropathy. e. Kidneys: Interstitial nephritis.
Vasculitis	Vasculitis is found in 15% of primary SS patients.Palpable purpura which is seen on standing is a major feature.

Diagnostic criteria (revised international classification) [2,3,4,5]

Ocular Symptoms:	A positive response to at least one of the three validated questions - Have you had daily, persistent, troublesome eyes for more than 3 months ? Do you have recurrent sensation of sand or gravel in the eyes ? Do you use tear substitute more than 3 times a day ?
Oral symptoms :	A positive response to at least one of the three validated questions - Have you had daily feeling of dry mouth for more than 3 months ? Have you had recurrent or persistently swollen salivary glands as an adult ? Do you frequently drink liquids to aid in swallowing dry fruits ?
Ocular signs :	Objective evidence of ocular involvements defined as a positive result to at least one of the following two tests - Schirmer's test performed without anesthesia (< 5 mm in 5 min) Rose Bengal score or other ocular dye score (> 4 according to Van Bijsterveld's scoring system)
Histopathology	In minor salivary glands focal Lmphocyticsialoadenitis, with focus score >1
Salivary gland involvement	Objective evidence of salivary gland involvement defined by a positive result to 1 of the following test : Unstimulated whole salivary flow <1.5 ml in 15 min Parotid sialography Salivary scintigraphy
Antibodies in the serum to Ro/SSA or La/SSB antigens, or both	

Sjogren's syndrome can be considered if any four of six criterias are positively seen.

Treatment

Treatment is symptomatic relief. Deficient tears are to be substituted by ophthalmic preparations (Tearisol; Liquifilm; 0.5% methyl cellulose; Hypo Tears).Corneal ulcerations should be managed by eye patching and boric acid ointments. Avoid diuretics, antihypertensive drugs, anticholinergics and antidepressants which may worsen salivation and lacrimation. For xerostomia best treatment is water. To stimulate secretionsPilocarpine 5 mg thrice daily or Cevimeline30 mg thrice daily can be given orally. Glucocorticoids and immunosuppressants are needed only if vasculitis is observed.

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