Sjögren’s Syndrome

Sjögren's syndrome is a multisystem autoimmune disease characterized by dryness due to exocrine gland destruction. Patients complain of xerostomia (dry mouth), xerophthalmia (dry eyes), and have joint involvement in the form of arthritis. Diagnosis of Sjögren's syndrome can be made through various methods. Serologic diagnosis can be made via testing for anti SS-B (La) and anti SS-A (Ro) antibodies, while Schirmer’s test measures tear production. Diagnosis can also be made through salivary gland biopsy. There is no definitive treatment for Sjögren's syndrome, and patients are given symptomatic treatment, alleviating the particular complications of disease. Those with Sjögren's syndrome have a higher prevalence of lymphoma development.



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Mechanism

Autoimmune Exocrine Gland Destruction
Auto-in-moon with XO Gland Destroyed
In Sjögren's syndrome, immune cells destroy salivary, lacrimal and other respiratory and GI tract exocrine glands. It can occur as a primary disease or can develop secondarily to another connective tissue disease, such as rheumatoid arthritis, systemic sclerosis, SLE or polymyositis. The most common demographic affected by Sjögren's syndrome is females between 40-60 years of age.

Symptoms (Classic Triad)

Xerostomia
Zero-spit-mouth
In this disorder, destruction of salivary glands leads to xerostomia, or dry mouth. This can manifest as cough, dysphagia (trouble eating/swallowing) and trouble speaking. Due to decreased salivation in the mouth, which helps carry immunoglobulins, patients are more prone to dental caries. This salivary gland destruction can also manifest as enlarged, painful parotid glands.

Xerophthalmia
Zero-tear-eyes
Xerophthalmia is a condition where the eyes fail to adequately produce tears. It is also known as keratoconjunctivitis sicca (KCS) or dry eye syndrome (DES). In Sjögren's syndrome, destruction of lacrimal glands leads to xerophthalmia, manifesting as dry eyes and conjunctivitis. Patients may specifically complain of feeling like there is sand in their eyes.

Arthritis
King-Arthur
Joint involvement in the form of rheumatoid arthritis is characteristic of Sjögren's syndrome. In cases of secondary Sjögren's syndrome, disease symptoms occur as a result of complications from primary rheumatoid arthritis.

Diagnosis

Anti SS-B (La)
Lawman Ant-tie in SS-B-Bee
There is a strong correlation with Sjögren's syndrome and autoantibody seropositivity. Specifically, autoantibodies to the ribonucleoprotein antigen anti SS-B (anti-La) are present. Though seropositivity is not specific for Sjögren's syndrome, presence of autoantibodies is associated with greater severity and longer duration of disease.

Anti SS-A (Ro)
Rowing Ant-tie in SS-A-Apple
There is a strong correlation with Sjögren's syndrome and autoantibody seropositivity. Specifically, autoantibodies to the ribonucleoprotein antigen anti SS-A (anti-Ro) are present. Though seropositivity is not specific for Sjögren's syndrome, presence of autoantibodies is associated with greater severity and

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longer duration of disease. ANA can also be elevated, but this is not diagnostic, as it can be elevated in the general population and in many autoimmune conditions.

Schirmer's Test

Shrimper Testing-tears

Schirmer’s test measures the output of the lacrimal glands in order to determine if adequate tears are being produced. During this test, paper strips are inserted into the lower eyelids, and tear secretion is measured. Less than five millimeters of moisture collected on the filter paper in five minutes yields results highly suggestive of Sjögren's syndrome.

Salivary Gland Biopsy

Biopsy-needle taking Salivary Gland

In Sjögren's syndrome, there is autoimmune destruction of exocrine glands, and on biopsy, this is demonstrated as lymphocytic infiltration and damage to salivary glands. Salivary gland biopsy is the most accurate test in diagnosing this disorder; however, is not required for diagnosis.

Treatment

Symptomatic Treatment

Treating Symptoms with water and eye-drops

There is no definitive treatment for this Sjögren's syndrome; instead, treatment is supportive and deals with specific symptoms. Pilocarpine or cevimeline, artificial tears, good oral hygiene (to prevent dental caries) and NSAIDs (for joint inflammation) can all be used to mitigate the disease symptoms.

Complications

Lymphoma

Lime-foam

Those with Sjögren's syndrome have a higher rate of developing non-Hodgkin lymphoma. Additionally, MALT lymphomas of the thyroid and salivary glands have been linked to this syndrome. Of note, malignancy is the most common cause of death in patients with Sjögren's syndrome.

Other

Sicca Syndrome

Soccer-ball with dry mouth and eyes

Sicca syndrome is very similar to Sjögren's syndrome; however, arthritis symptoms are not present. Additionally, sicca syndrome also incorporates vaginal dryness and chronic bronchitis. Furthermore, it may affect other organ systems, presenting with myositis, kidney disease, lung involvement, as well as the hepatobiliary system and brain.

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