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Oral Pathology

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November 30, 2022

Sjögren’s Syndrome

**Overview**

Sjögren’s Syndrome (SS), also known as Sicca Syndrome, is a chronic autoimmune disease that affects the exocrine glands. Examples of exocrine glands are sweat glands, lacrimal glands, salivary glands, mammary glands, and digestive glands in the stomach, pancreas, and intestines. They secrete substances such as saliva, tears, milk, and digestive juices. The inflammatory lymphocytic cells (B and T cells), activated by the person’s own immune system, damage the exocrine glands as they infiltrate. This causes the person to experience dry mouth, eyes, polyarthritis, etc. (Gupta, 2019).

SS can be classified as primary or secondary. Primary Sjögren’s Syndrom (pSS) is characterized by the absence of other diseases, while secondary Sjögren’s Syndrome (sSS) is characterized by the association with underlying autoimmune diseases like lupus erythematosus, systemic sclerosis, or rheumatoid arthritis. (Negrini, 2021)

**Etiology**

The underlying cause of SS, like many other autoimmune diseases, remain unknown. However, based on numerous studies, it can be concluded that this syndrome has a “multifactorial pathogenesis” (Tian, 2021). Some individuals are genetically predisposed while others are more susceptible due to exposure of certain environmental factors.

SS genome analysis was used in some studies to identify risk sites and susceptibility genes (Tian, 2021). Risk sites and susceptibility genes found are those that regulate the development and production of inflammatory mediators in the body. Therefore, individuals with SS have increased amounts of cytokines related to B cells, T cells, and natural killer (NK) cells in their immune system affecting autoimmunity (Negrini, 2021).

Environmental factors such as, infectious agents (viruses), are potential SS triggers – in particular, Epstein-Barr virus (EBV). EBV activates innate immune responses, and is known for its tropism for B cells (Negrini, 2021). With the increased amount of B cells present in the body from SS, the severity of the disease has the potential to increase. Although there is an association between SS and EBV, there is no substantial evidence that EBV causes SS. Other infections like Hepatitis C virus, cytomegalovirus, and human T-lymphotropic virus-1 were found to cause persistent infection of salivary glands leading to organ destruction and presenting sicca symptoms in the oral cavity (Utomo, 2020).

**Clinical presentation**

Not all patients with SS present with the same symptoms, but some include:

Objective:

* Dry mouth
* Dry/skin/ skin rash
* Corneal ulceration
* Swollen salivary glands
* Abnormal liver function tests

Subjective:

* Dry mouth/eyes
* Pain when swallowing/chewing
* Fatigue
* Skin sensitivity
* Continuous coughing
* Vaginal dryness
* Joint paint
* Heart burn/reflux
* Brain fog

**Demographic**

The incidence of SS increases with age (Tian, 2021), but has a predilection for women of middle age. The female-to-male ratio, 9:1, is found highest in Asian patients and lowest in black/African American patients. Non-European backgrounds also have a twofold higher prevalence compared to those with a European background.

It is usually diagnosed in the fifth decade of life (51 – 62 y.o). (Negrini, 2021)

**Biopsy/histology/radiographs**

Labial salivary gland biopsy is a key factor in diagnosis by combining haematoxylin-eosin staining with immunohistochemical staining. The histopathological features in SS presents parenchymal and ductal changes. A decrease/disappearance in acini, lymphocyte infiltration, and proliferation of lining cells is also seen. (Liao, 2022)

**Differential diagnosis**

SS is frequently undiagnosed/misdiagnosed because not all symptoms are always present simultaneously. Different providers from different specialties may treat each symptom individually and not recognize that an autoimmune disease is present.

The symptoms can be similar to:

* Menopause
* Drug adverse effects
* Allergies
* Lupus
* Rheumatoid arthritis
* Fibromyalgia
* Multiple sclerosis
* Chronic fatigue syndrome

(Sjögren’s Foundation, n/a)

**Treatment**

There is currently no curative treatment for SS, but medications are prescribed to alleviate sicca symptoms and to improve quality of life. Topical therapy is the first approach if SS does not affect major organs. If the disease is more severe, systemic steroids are administered to control the disease – immunosuppressives can also be used as steroid sparing agents. For those with severe and refractory systemic disease, B-lymphocyte targeted medications (rituximab, epratuzumab, and belimumab) are used. The goal is to preserve and/or substitute secretions while reducing inflammation in the body. (Negrini, 2021)

For patients with severe sicca symptoms, lifestyle changes may be necessary in addition to medications. Patients with dry eyes should avoid eye stimulating activities for long periods of time, such as spending too much time fixed on a screen. Patients with xerostomia should avoid smoking, alcoholic drinks, and caffeine as they can further dry out the oral cavity. Instead, frequent intake of water or saliva-stimulating agents (sugar-free candies/gum) is recommended to maintain oral lubrication. (Negrini, 2021)

**Prognosis**

Most people with SS carry on a normal life that is unaffected while managing sicca symptoms with eye drops or frequently drinking water. However, depending on the severity of the disease, the person should be regularly monitored for complications like lymphoma, pulmonary, kidney and liver disease. (American College of Rheumatology)

**Professional Relevance**

One of the main clinical manifestations of SS is xerostomia, which can cause a plethora of issues to the oral cavity. When there is not enough salivary flow, oral clearance is minimized leading to increased biofilm/calculus buildup, carious lesions, gingival inflammation, and candidiasis. Because the patient’s oral health is at risk, it is important for the dental hygienist to emphasize proper and consistent. Oral rinses, like Bioténe or ones without alcohol, and fluoride products should be recommended. Most importantly, it would be ideal to visit the dental office at least twice a year or every three months depending on the individual’s oral health.

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