

Autoimmune Resource & Research

Centre
ARRC



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Sjögren's Syndrome

Introduction

Sjögren's syndrome is a disease of unknown cause in which the immune system attacks the lacrimal and salivary glands resulting in a decreased production of tears and saliva. This causes the most common symptoms of Sjögren's syndrome namely dryness of the eyes and mouth, the so-called "SICCA complex". These symptoms often occur in association with other connective tissue diseases, such as rheumatoid arthritis and systemic lupus erythematosus, where Sjögren's syndrome is referred to as secondary. Primary Sjögren's syndrome occurs without other connective tissue diseases and although the SICCA complex is often the only symptom, other features as detailed below may also occur.

History

The first report of Sjögren's syndrome was made in 1888 by Johann Mikulicz. He described a 42 year old farmer with parotid and lacrimal gland enlargement. In 1933 a Danish ophthalmologist, Henric Sjögren, described the detailed clinical features of Sjögren's syndrome in a series of patients, most of whom had associated rheumatoid arthritis (secondary Sjögren's Syndrome). In 1953 Morgan and Castleman defined a subset of patients who had symptoms of Sjögren's syndrome in association with other features that suggested a more generalised disease process, which has come to be known as Primary Sjögren's syndrome.

Clinical Features of Sjögren's Syndrome

Glandular Manifestations

The major symptoms of Sjögren's syndrome are a result of inflammation of the exocrine or secretory glands.

- a. **Lacrimal gland** . Inflammation of the lacrimal or tear gland causes dryness of the eyes due to a reduction in tear production. This is the most frequent symptom of Sjögren's syndrome. The dryness or grittiness usually develops gradually over a period of several years but the symptoms vary in severity from day to day. Other symptoms can include irritation, itching, sensitivity to the light and accumulation of thick, ropery mucus filaments, which are present particularly on waking. This dryness may lead to increased susceptibility to infection and also the development of damage to the corneal surface of the eye.
- b. **Parotid or Salivary gland** . Inflammation of the parotid gland causes an under production of saliva and hence a dry mouth. Other problems that may reflect under-secretion of saliva include trouble with swallowing, food sticking to the inner surface of the mouth, changes in taste and difficulty speaking for prolonged periods of time. The dryness of the mouth may lead to the development of dental decay as well as bad breath. Sometimes superficial infection in the mouth may also occur.
- c. **Glandular enlargement** . The inflammation of the parotid glands, found just in front of the ears, and of the lacrimal gland, found in the upper outer corner of the orbit, can

cause swelling of the glands. The swelling is usually episodic but can be quite marked with associated pain tenderness and redness of the overlying skin. Persistent swelling of these glands needs to be checked by your doctor regularly as there is a very small chance that you may develop a malignant condition called lymphoma. This is a rare complication of Sjögren's Syndrome.

- d. **Dryness of the upper airways.** Dryness of the upper airways will lead to complaints of dryness of the nose and posterior pharynx and larynx, as well as a persistent dry cough. This dryness can also predispose to infection.
- e. **Vaginal dryness.** Vaginal dryness is common and may cause pain on intercourse. It also predisposes to the development of infections such as thrush.

Non-Glandular Manifestations

- a. **Skin manifestation.** A number of different skin problems can be seen in Sjögren's syndrome. These include dry skin with associated itching. Inflammation of the small blood vessels in the skin can lead to a condition called "vasculitis" which causes crops of small lesions, usually the size of a pinhead. Occasionally these can ulcerate.
- b. **Lung disease.** As well as dryness of the upper airways, occasionally a condition known as pulmonary fibrosis can develop in patients with Sjögren's syndrome. This leads to reduced elasticity of the lungs and although often unnoticed by the patient, it can lead to shortness of breath on exertion.
- c. **Gastrointestinal features.** A number of problems can develop with the gastrointestinal system in people with Sjögren's syndrome. Perhaps most frequently is alteration of the large bowel function, leading to features of abdominal pain and alteration in bowel habit. It is likely that this is caused by a reduction of intestinal secretions. Rarely the liver can become inflamed with Sjögren's syndrome, but it is unusual for this to be a major problem.
- d. **Nervous involvement.** Occasionally the nerves can become inflamed in people with Sjögren's syndrome. This leads to a condition called sensory neuropathy where altered sensation and loss of muscle power can develop. Very rarely patients with Sjögren's syndrome develop inflammation of the central nervous system. This causes symptoms similar to a stroke or multiple sclerosis.
- e. **Psychiatric disorders.** Subtle changes, including a reduction in the ability to think clearly and complaints of poor memory and poor concentration, occur occasionally. Depression is more common in people with Sjögren's syndrome.
- f. **Arthritis.** Arthritis in people with Sjögren's syndrome is quite common. It usually involves the small joints of the hands and feet. The arthritis is usually intermittent and does not lead to deformity. Treatment of arthritis includes practical management such as the use of heat and cold therapies. If necessary, your doctor may prescribe non-steroidal anti-inflammatory medications to relieve pain and inflammation.
- g. **Myositis.** Inflammation of the muscles causing pain and weakness, may occur in people with Sjögren's syndrome.
- h. **Non-specific systemic features** . It is extremely common for people with Sjögren's syndrome to complain of tiredness, lethargy and a sense of general unwellness. Sometimes the tiredness follows a cyclical course during the day and may be worse premenstrually. The fatigue may be due to the development of a sleep disorder usually due to disruption of sleep patterns. Fatigue may also be caused through chronic inflammation.

Diagnosis

Diagnosis of Sjögren's syndrome can sometimes be made purely on clinical grounds but often further investigation is required.

The typical features of dryness of the eyes and mouth in association with glandular swelling are often all that is required to make the diagnosis.

A reduction in tear production can be confirmed by the performance of a Schirmer's test, where a small piece of blotting paper is inserted under the lower eyelid for a period of five minutes. The degree of wetness is then measured.

A number of techniques have been described to demonstrate reduction of salivary production, such as chewing an absorbent sponge for a set period of time or cannulation of the main salivary duct. In practice, inspection inside the mouth is all that is required.

Occasionally biopsy of the minor salivary glands of the lower lip is performed. This is a relatively simple procedure performed under local anaesthetic. Characteristic inflammation of the salivary glands is seen under the microscope.

A number of different blood tests are helpful in making the diagnosis of Sjögren's syndrome. This includes a full blood count, which may show abnormalities of the haemoglobin with anaemia and occasionally a reduction in white cell count numbers. The erythrocyte sedimentation rate or ESR test is usually elevated and reflects the degree of inflammation occurring. Biochemical studies are usually performed to look for involvement of the liver, the kidneys and muscle.

Abnormal immunological tests are often noted, particularly in primary Sjögren's syndrome. The ANA test is usually positive and particular antibodies referred to as SSA and SSB are often abnormal. In addition abnormalities of the blood proteins can occur causing what is referred to as a polyclonal gammopathy.

For further information about Sjögren's see [Sjögren's Syndrome Foundation, USA](#).

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