



Libyan International Medical University
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Complications of Sjögren syndrome

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Abstract:

Sjögren syndrome (SjS, SS) is a long-term autoimmune disease in which the moisture-producing glands of the body are affected, characterized by lymphocytic infiltration and acinar destruction of lacrimal and salivary glands, leading to dry eyes and dry mouth. In about half of the cases the syndrome occurs in association with another autoimmune disease, most frequently rheumatoid arthritis or systemic lupus erythematosus. On this basis the syndrome is divided into: (1) primary Sjögren syndrome (the combination of dry mouth xerostomia), and dry eyes (xerophthalmia or keratoconjunctivitis sicca) or (2) secondary Sjögren syndrome (the triad of xerostomia, xerophthalmia, and an autoimmune connective tissue disease (usually rheumatoid arthritis))

Introduction:

Sjögren syndrome predominantly affects middle-aged females (the female: male ratio is about 9:1). Cause of Sjögren syndrome is unclear. However genetic factors are thought to be important in increasing the susceptibility of an individual to external environmental factors, which then trigger the disease. Sjögren syndrome occurs with increased frequency in patients with particular combinations of the HLA class II major histocompatibility genes, supporting a genetic role, and several viruses, especially Epstein-Barr virus have been suggested as potential trigger factors. Immunological mechanisms leading to destruction of glandular tissue probably involve mainly T cells and their associated cytokines.

Histopathological examination of glands shows lymphocytic infiltration initially around intralobular ducts which eventually replaces the whole of the affected lobules. The infiltration is accompanied by acinar atrophy but, in contrast, the ductal epithelium may show proliferation. The hyperplasia of ductal epithelium eventually obliterates the duct lumen leading to islands of epithelial tissue in a sea of lymphocytes, replacing entire salivary lobules. These appearances are described by the term 'benign lymphoepithelial lesion' (also referred to as myoepithelial sialadenitis). The benign lymphoepithelial lesion is characteristic but not pathognomonic of Sjögren syndrome.

Discussion:

The most commonly reported complications of Sjögren's syndrome are xerostomia and xerophthalmia, both of which can lead to other associated symptoms

Associated symptoms of Xerostomia include:

- Tooth decay, leading to an increased risk of tooth loss.
- Gum line decay.
- Dry cough.
- Burning sensation.
- Difficulty swallowing and chewing.
- Alterations in the sense of taste.
- Hoarse voice.
- Difficulty speaking.
- Repeated fungal infections of your mouth (oral thrush), symptoms of which include the appearance of white, cream-coloured or yellow spots on the inside of your mouth and tongue.

Drinking a lot of sugar drinks to stimulate saliva or maintain moisture in the mouth, these complications can become worse.

Associated symptoms of xerophthalmia include:

- Burning or stinging sensation in your eyes
- Itchy eyes
- Feeling that there is a piece of sand or gravel in your eyes
- Irritated and swollen eyelids
- Sensitivity to light (photophobia)
- Tired eyes
- Discharge of mucus from your eyes

Symptoms can become worse when you are in a windy or smoky environment. Air-conditioned buildings and travelling on aeroplanes can also make symptoms worse.

In addition to Xerostomia and xerophthalmia, Sjogren syndrome can present with a wide spectrum of clinical features involving abnormalities of other exocrine glands and a variety of extraglandular manifestations other exocrine glands including :

- In salivary glands causes swollen (located between your jaw and your ears) .
- In skin causes xeroderma .
- In Respiratory tract causes nasal dryness, sinusitis, tracheitis .
- In Pharynx and GI tract causes dysphagia, atrophic gastritis, pancreatitis .
- In Reproductive system causes vaginal dryness , which can make sexual intercourse painful.

Extraglandular including :

- In Joints causes arthritis .
- In Skin causes purpura, Raynaud's phenomenon .
- In Liver causes primary biliary cirrhosis .
- In kidney causes kidney inflammation or kidney stones.
- Endocrine causes thyroiditis.
- In Neurological causes central and peripheral neuropathies a condition that causes loss of sensation in the hands and feet.
- In Haematological causes anaemia, leucopenia, thrombocytopenia .

In both primary and secondary Sjogren syndrome there is a risk of B cell malignant lymphoma arising within an affected gland.

Conclusion:

Sjögren syndrome (SjS, SS) is a chronic autoimmune disease in which the moisture-producing glands of the body are affected. This results primarily in the development of a dry mouth and dry eyes. Other symptoms can include dry skin, vaginal dryness, a chronic cough , joint pains and thyroid problems. It can occur independently of other health problems (primary Sjögren syndrome) or as a result of another connective tissue disorder (secondary Sjögren syndrome).

References:

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