Autoimmune salivary gland diseases

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Autoimmune salivary gland diseases

- Sjogren’s syndrome
- Sialosis
- Xerostomia
Autoimmune salivary gland diseases

**Xerostomia (Dry Mouth):**
Is clinical manifestation is not a disease by itself.
Xerostomia (Dry Mouth):

Causes of xerostomia:

❖ Organic causes
❖ Functional causes
❖ Drugs
Xerostomia (Dry Mouth)

Organic causes:
- Sjogren's syndrome
- Irradiation
- Mumps (transient)
- HIV infection
- HCV infection
- Sarcoidosis
- Amyloid
- Iron deposition (haemochromatosis, thalassaemia)
Xerostomia (Dry Mouth)

Functional causes:
- Dehydration
- Fluid deprivation or loss
- Haemorrhage
- Persistent diarrhoea
- and/or vomiting
- Psychogenic
- Anxiety states
- Depression
Xerostomia (Dry Mouth)

Drugs:
- Diuretic over dosage
- Drugs with antimuscarinic effects (Atropine, ipratropium, hyoscine and other analogues)
- Tricyclic and some other antidepressants
- Antihistamines
- Antiemetics (including antihistamines and phenothiazines)
- Neuroleptics, particularly phenothiazines
- Some older antihypertensives, (ganglion blockers and clonidine)
- Drugs with sympathomimetic actions
- 'Cold cures' containing ephedrine, etc.
- Decongestants
- Bronchodilators
- Appetite suppressants, particularly amphetamines
Xerostomia (Dry Mouth)

Classification of xerostomia:
1. True primary or permanent xerostomia.
2. Symptomatic or secondary xerostomia.
Xerostomia (Dry Mouth)

**True primary or permanent xerostomia:**
A condition in which there is pathological lesion on salivary gland.

**Symptomatic or secondary xerostomia:**
A condition in which there is no pathological lesion on salivary gland but dry mouth due to certain transient condition such as drug.
Xerostomia (Dry Mouth)

Clinical features:

- Patients with severe xerostomia make no complaint of dry mouth, though they may, for instance, admit that they have difficulty in eating dry food.
- Some complain of an unpleasant taste in the mouth.
- Some who complain of dry mouth have normal salivary flow rates on objective testing and the problem is neurotic in nature.
Xerostomia (Dry Mouth):

Treatment:

- Treat the underlying causes if possible.
- Keep good oral hygiene.
- Paint mucous membranes with glycerin and lemon juice to avoid cracking.
- Administrate pilocarpine to stimulate salivary flow.
- Use artificial saliva.
benign lymphoepithelial lesions:
- Sjogren syndrome
- Micluicz disease (syndrome)
- IgG4 related salivary gland disease

Characterized by intense lymphocytic infiltration of the salivary and lacrimal glands with acinar destruction.
Sjogren’s syndrome:
Is an autoimmune (connective tissue) disease which shows a corresponding variety of immunological abnormalities.

Histologically defined as a chronic autoimmune disease characterized by lymphocytic infiltration and acinar destruction of salivary and lacrimal glands.
Sjogren’s syndrome

Aetiology and pathology:

- Autoimmune.
- Viral (CMV, EBV).
Sjogren’s syndrome

Classification of sjogren’s syndrome:

- **Primary Sjogren's syndrome:** comprises dry mouth and dry eyes not associated with any connective tissue disease.

- **Secondary Sjogren's syndrome:** comprises dry mouth and dry eyes associated with rheumatoid arthritis or other connective tissue disease.
Sjogren’s syndrome

Clinical features:

- Females are affected nearly 10 times as frequently as males.
- Middle age
- Sjogren's syndrome affects 10-15% of patients with rheumatoid arthritis.
- Possibly affects 30% of patients with lupus erythematosus.
- Affects variable proportion of patients with or without other connective-tissue diseases.
Oral effects of Sjogren's syndrome:

- Discomfort
- Difficulties with eating or swallowing
- Disturbed taste sensation
- Disturbed quality of speech
- Predisposition to infection
- Dry fissure depapillated tongue
- Angular cheilitis
- Dry smooth glazy mucosa.
Sjogren’s syndrome

Ocular effects of Sjogren's syndrome:

- Failure of tear secretion
- Failure of clearance of foreign particles from the cornea and conjunctiva (keratoconjunctivitis sicca)
- Gritty sensation in the eyes and inflammation
- Risk of impairment or loss of sight
Sjogren’s syndrome

- Swelling of the parotids is not common, but a history of swelling at some stage may be obtained in about 30% of patients.
- Patient with persistent salivary gland enlargement should be investigated for lymphoma.
Sjogren’s syndrome

Histopathological features:

- Lymphocytic infiltration.
- Acinar atrophy.
- Proliferation of ductal cells forming epimyoepithelial islands.
- Replacement of the whole gland by dense lymphocytic infiltration.
Sjogren’s syndrome

Diagnosis of Sjogren’s syndrome:

• Depend on careful history taking to exclude other causes of xerostomia.
• Careful clinical examination.
• Estimation of normal salivary flow is between 1 and 2 ml per minute but may be reduced to 0.5 ml/min or less.
• Labial salivary gland biopsy showing periductal lymphocytic infiltrate.
• A snowstorm Sialography appearance on sialography (so-called sialectasis) is due to leakage of contrast material through the duct walls.
Diagnostic criteria for Sjogren syndrome:

I. Primary Sjogren syndrome
   A. Symptoms and objective signs of ocular dryness
      1. Schirmer's test less than 8 mm wetting per 5 minutes.
      2. Positive Rose Bengal staining of cornea or conjunctiva to demonstrate kerato-conjunctivitis sicca.
   B. Symptoms and objective signs of dry mouth
      1. Decreased parotid flow.
      2. Abnormal findings from biopsy of minor salivary gland
   C. Serologic evidence of a systemic autoimmunity
      1. Elevated rheumatoid factor > 1:320
      2. Elevated antinuclear antibody (ANA) > 1:320
      3. Presence of anti-SS-A (Ro) or anti-SS-B (La) antibodies
Secondary Sjogren syndrome

A. Presence of characteristic signs and symptoms of primary Sjogren syndrome

B. Clinical features sufficient to allow a diagnosis of:
   Rheumatoid arthritis, systemic lupus erythematosus, polymyositis, scleroderma, or biliary cirrhosis.

C. Exclusions: sarcoidosis, preexisting lymphoma, HIV, hepatitis B or C, primary fibromyalgia, and other known causes of autonomic neuropathy, keratitis sicca, or salivary gland enlargement.
Sjogren’s syndrome

**Management:**
- Salivary gland damage is irreversible.
- Give reassurance and help with dry mouth.
- Ophthalmological investigation for keratoconjunctivitis sicca.
- Refer to specialist if connective tissue disease is untreated.
- Check for any associated drug treatment contributing to dry mouth.
- Improve dry mouth.
Sjogren’s syndrome

- Observe regularly for possible development of ascending parotitis or lymphoma
Mikulicz Disease
Mikulicz syndrome,

- An unusual bilateral painless swelling of the lacrimal glands and all of the salivary glands.
- They represented salivary and lacrimal involvement by other disease processes, such as tuberculosis, sarcoidosis, and lymphoma.
- Histopathologic examination of the involved glands showed an intense lymphocytic infiltrate, with acinar destruction.
Küttner tumor :

a tumor that manifests as a chronic inflammatory disease of the salivary gland, which mostly affects the unilateral submandibular gland. Clinically, it produces afirm swelling of the glands and it may be difficult to distinguish from neoplasia.
Küttner tumor

histopathologic features:

➢ the tumors usually present areas of localized fibrosis characterized by lymphoplasmacytic infiltrate within the glands.

➢ The lobular architecture is preserved and the degree of involvement varies from lobule to lobule.

➢ Reactive lymphoid follicles are frequently present
IgG4-related disease

- characterized by a high serum IgG4 level and tissue infiltration of IgG4-positive plasmacytes.
- Chronic sclerosing sialadenitis (kuttner) may be associated with sclerosing cholangitis, retroperitoneal fibrosis, or sclerosing pancreatitis.
- Some cases of kuttner may be part of IgG4-related plasmacytic disease
- All cases not confirmed as Sjogren and diagnosed previously as Mikulicz’s disease are in fact IgG4-related sialadenitis.
Sialosis (sialadenosis):
A non inflammatory non neoplastic recurrent bilateral salivary gland swelling.
Commonly occur in parotid.
Sialosis (sialadenosis)

Etiology:

Unknown, but has been reported in association with:

- Hormonal disturbances
- Malnutrition
- Liver cirrhosis
- Chronic alcoholism
- Administration of various drug
Sialosis (sialadenosis)

Histopathological features:

- Hypertrophy of serous acinar cells.
- Atrophy of striated duct.
- Oedema of interstitial supporting stroma.
Sialosis (sialadenosis)

**Treatment:**
- No specific treatment
- Treat the suspected underlying cause.