Sjögren's syndrome (Sicca syndrome)

Sjögren's syndrome: is autoimmune destruction of the **exocrine glands** in which the white blood cells destroy the salivary glands causing **xerostomia** (dry mouth) and Lacrimal glands leading to **Keratoconjuctivitis** sicca (dry eyes) and also other gland (respiratory mucosa and vagina). The disease much more common in Women.

What are the predisposing factors (causes) of SS?

1. **Environmental link** to viruses (Epstein–Barr virus, hepatitis C and rota virus)
2. **Genetic factor (** HLA-DR3 and HLA-A1 haplotype in SS patients).

There are two types of SS.

1- Primary only affected exocrine gland and associated with HLA hplotyping.
2- Secondary associated with other autoimmune disease **(RA,SLE)**

Pathogenesis: Sjögren's syndrome was **self-immune** system-mediated loss of exocrine glands, triggering and activation the infiltration of **lymphocytes**, specifically CD4+ T cells, B cells, and plasma cells, causing dysfunction in the salivary and lacrimal glands.

Diagnosing of Sjögren's syndrome(SS):

What are the Blood tests can be done to determine patient with SS ?

1- **Antinuclear antibody (ANA)**

There are two types of ANA **: SS-A/Ro (Anti-Sjögren's-syndrome-related antigen A) also called anti-Ro) and SS-B/La.**

SSA/Ro is associated with numerous other autoimmune conditions, but are often present in SS. **(SS-B/La is more specific to SS)**

2- IgM Rheumatoid factor RF (because SS frequently occurs secondary to RA),

What the other tests can be use it to diagnosis of SS?

1- Schirmer test : Measures production of tears. a strip of filter paper is held inside the lower eyelid for **5 minutes, Amount of moisture is measured. <5 mm in 5 min is positive.**

2- **The rose bengal test:** measures state and function of the lacrimal glands. Using nontoxic dye rose Bengal on the eyes. Also Lissamine Green dye can be used.

3- **A lip/salivary gland biopsy** .

4- **sialogram, a special X-ray** to see the blockage in the salivary **gland ducts**.

Treatment: (the disease if not treated well may lead to lymphoma)

1-Artifiecial tears substitutions & stimulator of tear production & saliva substitution.

2-Immunosuprassive drugs corticosteroid and immune-modulating drugs like cyclosporine.

Behçet's disease

Define Behçet's disease ? is a rare immune-mediated **small vasculitis** (**but consider auto-inflammation of the blood vessels)**.. Behçet's disease (BD) was named in 1937 after the Turkish dermatologist Hulusi Behçet, who first described the triple-symptom complex of recurrent oral ulcers, genital ulcers, and uveitis.

It is a **systemic disease,** it can also involve visceral organ such as the **gastrointestinal tract, pulmonary, musculoskeletal, cardiovascular and neurological systems.** This syndrome can be fatal due to severe neurological complications. **It is usually severe in men.**

Cause:

The cause is not well-defined. **the risk genetic factor related to BD is associated with HLA class I-B51.** The involvement of **a subset of T cells (Th17)** seems to be important in immune disorder. Also, the secretion of pro-inflammatory mediators or cytokines play role in disease activity like Il-1, Il-18 TNFα.

Signs and symptoms:

Skin and mucosa. :all patients present with painful oral mucocutaneous Painful genital ulcerations usually develop around the anus, vulva, or scrotum and cause scarring in 75% .

Ocular system.

Inflammatory eye disease **can develop early in the disease** lead to permanent vision loss in 20% of cases. **painful eyes, conjuctival redness, and decreased visual acuity,** reduced visual acuity, reduced color vision, swollen optic disc, macular edema.

Bowels

GIT manifestations include abdominal pain, nausea, and diarrhea with or without blood,

Neurological system.

CNS involvement most often occurs **as a chronic meningoencephalitis**.

Diagnosis: There is no specific diagnosis, Behçet's disease has a high degree of resemble to diseases that cause mucocutaneous lesions such as *Herpes simplex* .

Diagnosis of Behçet's disease is based on clinical findings

1-Inflammatory markers such ESR, and CRP may be elevated.

2-Now genetic test for HLA class I-B51 is used.

Current treatment reducing inflammation, and controlling the immune system by **Immunosuppressants** drugs.

High dose **corticosteroid** therapy is often used for severe disease manifestations.

Anti-TNF(Tumor Necrosis Factor) in treating the uveitis . and Interferon alpha for oral ulcer