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SJOGREN'S SYNDROME - An Immune System Disorder Characterised By Dry Eyes And Dry Mouth.

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ABSTRACT:

Sjogren's Syndrome is an auto immune condition that can occur at any age, but it is most common in older women. Symptoms vary in type and intensity but people may able to live normal life. Most treatment of Sjogren's Syndrome is aimed at relieving symptoms of dry eyes and mouth and preventing and treatment long term complications such as infection and dental disease. It is due to immune mediated injury characterised by lymphocytic infiltration of exocrine glands and circulating antibodies. Genetic studies show association of HLA-B8/dr3 in primary Sjogren's Syndrome.

Common complications are dental cavities, yeast infection and vision problems. Prescribed eye drops, artificial tears , and moisture chamber spectales can help to relieve dry eyes. Some are at the increased risk for cancer of lymph glands lymphoma. Thus regular medical care and follow up is important for all patients.

KEY WORDS: Sjogren's Syndrome, auto immune, cancer, dry eyes, lymphoma

SJOGREN'S SYNDROME

Introduction:

It is a chronic autoimmune, inflammatory disease characterized by lymphocytic infiltration of exocrine gland producing dryness of mouth (xerostomia) and dryness of eyes (xeropthalmia). Hence also called kerato conjunctivitis, **SICCA** syndrome. A small number of patients may develop malignant lymphoma.

The disease affects middle aged females (female to male ratio 9:1)

Types:

1. Primary isolated Sjogren's Syndrome

2.Secondary when associated with other auto immune disease such as SLE, RA, Polymyositis, Primary biliary cirrhosis, Autoimmune Hepatitis ,Vasculitis And Myasthenia.

Risk factors

Sjogren's syndrome typically occurs in people with one or more known risk factors, including:

- Age-Sjogren's syndrome is usually diagnosed in people older than 40.
- Sex- Women are much more likely to have sjogren's syndrome.
- Rheumatic disease- It's common for people who have sjogren's syndrome to also have a rheumatic disease such as rheumatoid arthritis or lupus.

Pathogenesis:

It is due to immune mediated injury characterised by lymphocytic infiltration of exocrine glands and circulating antibodies. Auto antibodies in sjogren's syndrome are directed against non organ –specific antigens such as immunoglobulins and extractable nuclear and cytoplasmic antigens.

The epithelial cells of the exocrine glands in sjogren's syndrome act as an antigen presenting cells against whose components antibodies are produced.

When white blood cells attack salivary glands, tear glands and other tissues leading to decrease in tear and saliva production. This can lead to dryness in the mouth, eyes, skin, nose, upper respiratory tract and vagina.Genetic studies show association of HLA-B8/dr3 in primary Sjogren's Syndrome.

Clinical features:

Glandular manifestations: these occur due to involvement of exocrine glands leading to their diminished secretion.

Oral symptoms:

- Dryness of mouth
- Difficulty in swallowing
- Inability to speak
- Burning sensation
- Enlargement of parotid and salivary glands glands

Extra glandular manifestations:

- Arthralgia
- Raynaud's phenomenon, dysphagia
- Pulmonary involvement e.g interstitial lung disease ,lung diffusion defect
- Vasculitis e.g purpura, urticaria, skin ulceration, glomerulonephritis and neuropathy, myositis
- Renal involvement. interstitial nephritis, renal tubular defect
- Non-hodgkin lymphoma-b cell type
- Spleenomegaly, lymphadenopathy
- Fits, Poly neuropathy, depression.
- Skin rashes or dry skin
- Vaginal dryness
- Persistent dry cough
- Prolonged fatigue

Diagnosis:

- ESR is raised. leucocytosis may be seen.
- Anti-RO and Anti-LA antibodies are present in primary sjosren's syndrome. Positive rheumatoid factor positive.
- Secondary sjogren's syndrome be investigated on thevlines of connective tissue disorder by RNA and auto antibodies.
- Positive **schimer's test**: a standard strip of filter paper is placed on the inside of the eye lid wetting for <10mm in 5 minutes indicates defective tear production.(positive test). Schirmer's test ≤5 mm/5 min in at least one eye – weight/score 1
- **Lip biopsy**: (labial minor salivarygland biopsy) shows focal lymphocytic infiltration.
- Rose Bengal staining of the eyes shows punctuate keratitis.
- Sialometry: Unstimulated whole saliva flow rate ≤0.1 mL/min – weight/score 1
- Salivary gland scintigraphy / technetium excretion radionuclide scanning-provide dynamic picture of the function of all major salivary glands.

Treatment

Treatment for sjogren's syndrome depends on the parts of the body affected. Many people manage the dry eye and dry mouth of sjogren's syndrome by using over-the-counter eye drops and sipping water more frequently. But some people need prescription medications, or even surgical procedures.

Medications:

Decrease eye inflammation. Prescription eyedrops such as cyclosporine (restasis) or lifitegrast (xiidra) may be recommended.

Increase production of saliva. Drugs such as pilocarpine (salagen) and cevimeline (evoxac) can increase the production of saliva, and sometimes tears. Side effects can include sweating, abdominal pain, flushing and increased urination.

- **Address specific complications.** If you develop arthritis symptoms, you might benefit from non steroidal anti-inflammatory drugs (NSAIDS) or other arthritis medications. Yeast infections in the mouth should be treated with antifungal medications.
- **Treat system wide symptoms.** Hydroxychloroquine (plaquenil), a drug designed to treat malaria, is often helpful in treating sjogren's syndrome. Drugs that suppress the immune system, such as methotrexate (trexall), also might be prescribed.

TO HELP WITH DRY MOUTH:

- **Don't smoke.** Smoking can irritate and dry out mouth.
- **Increase your fluid intake.** Take sips of fluids, particularly water, throughout the day. Avoid drinking coffee or alcohol since they can worsen dry mouth symptoms. Also avoid acidic beverages such as colas and some sports drinks because the acid can harm the enamel of your teeth.
- **Stimulate saliva flow.** Sugarless gum or citrus-flavored hard candies can boost saliva flow. Because sjogren's syndrome increases the risk of dental cavities, limit sweets, especially between meals.
- **Try artificial saliva.** Saliva replacement products often work better than plain water because they contain a lubricant that helps mouth stay moist longer. These products come as a spray or lozenge.
- **Use nasal saline spray.** A nasal saline spray can help moisturize and clear nasal passages so that you can breathe freely through nose. A dry, stuffy nose can increase mouth breathing.

Diet:

The best option is to choose a balanced diet that is high in fresh fruit and vegetables and low in saturated fats and sugar. Avoid artificial sweetners. lozenges may help to keep the mouth moist. Increase the fluid intake.

Surgery

A minor procedure to seal the tear ducts that drain tears from eyes (Punctal occlusion) might help relieve dry eyes. Collagen or silicone plugs are inserted into the ducts to help preserve tears.

Complications

The most common complications of sjogren's syndrome involve eyes and mouth.

- **Dental cavities.** Because saliva helps protect the teeth from the bacteria that cause cavities, if the mouth is dry.
- **Yeast infections.** People with sjogren's syndrome are much more likely to develop oral thrush, a yeast infection in the mouth.
- **Vision problems.** Dry eyes can lead to light sensitivity, blurred vision and corneal damage.

Less common complications might affect:

- **Lungs, kidneys or liver.** Inflammation can cause pneumonia, bronchitis or other problems in lungs; lead to problems with kidney function and cause hepatitis or cirrhosis of liver.
- **Lymph nodes.** A small percentage of people with sjogren's syndrome develop cancer of the lymph nodes (lymphoma).
- **Nerves**- numbness, tingling and burning in your hands and feet (peripheral neuropathy).

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