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Case Report

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Sjogren's Disease: A Case Report on the Multifaceted Disease

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Abstract

Sjogren's Disease is one of the most prevalent autoimmune diseases that affects multiple systems of the body. Autoimmune diseases are not always easy to identify, in fact, it can take years to diagnose. Once a diagnosis is made, treatment options can help with the symptoms, however, there is no cure for Sjogren's. It is important to address the symptoms with Sjogren's early, so that proper treatment can be given to prevent further damage. The aim of this paper is to emphasis the importance of early diagnosis and the need for multiple specialists (healthcare team) to help with the treatment of each symptom associated with Sjogren's.

Keywords: Sjogren's Disease, autoimmune disease, healthcare team.

Educational Objectives:

Upon completion of this article, the healthcare professional should be able to:

- Understand the symptoms that are related to Sjogren's Disease
- Bring awareness to the health impact patients face with Sjogren's Disease
- Learn the importance of a healthcare team since Sjogren's is a multifaceted disease
- Gain a better understanding of treatment options for each ailment associated with Sjogren's Disease

Introduction

Sjogren's Disease, formerly known as Sjogren's Syndrome, is a chronic, systemic autoimmune disease in which the immune system attacks the exocrine glands [1,2]. As of September 2024, the International Sjogren's community officially changed Sjogren's Syndrome to Sjogren's Disease "to better indicate the serious and systemic nature of the disease" [1]. Sjogren's has a distinct pattern of histopathologic features and clinical symptoms that helped push the need for it to be labeled as a disease [3]. Typically, a patient with Sjogren's Disease initially presents with clinical manifestations of xerophthalmia (dry eye) and xerostomia (dry mouth). The disease can fluctuate over time with flares (exacerbation of symptoms) and remissions (temporary recovery), as well as progressing to other disease processes, such as shortness of breath, fatigue, or muscles aches to name a few [3]. Moreover, Sjogren's Disease can be associated with other systemic autoimmune diseases [1]. "Approximately 30% of patients with rheumatoid arthritis, 10% of those with lupus, and 20% of those with scleroderma also suffer from Sjogren's Disease" [4]. Sjogren's Disease is the second most common autoimmune disease following rheumatoid arthritis [5]. Sjogren's affects approximately 1 in 200 people and primarily affects middle aged women, however, it has been seen in younger patients as well [5,6]. Additionally, Sjogren's has a high incidence of 9:1 in females in comparison to males [7]. Furthermore, there is a higher prevalence of Sjogren's Disease in the Caucasian population [3].

Symptoms

Initially, most patients will develop dry eyes and/or dry mouth which can then manifest into additional symptoms. These additional symptoms can include: dysphagia (difficulty swallowing), keratoconjunctivitis (eye inflammation of the cornea and conjunctiva), fatigue, joint pain, skin rashes, muscles aches/weakness, acid reflux, vaginal dryness, swelling of the salivary glands, difficulty sleeping, memory issues, neuropathy, and shortness of breath to name a few [8]. Fatigue is seen in approximately 70% of Sjogren's patients, which is often reported as the most debilitating symptom [9]. Moreover, other clinical manifestations such as cutaneous, musculoskeletal, pulmonary, renal, hematologic, and neurologic involvement can be seen in Sjogren's patients [3]. It is unclear what causes this autoimmune disease, but it is believed that genetic or environmental factors could play a role in this disease [3].

Diagnosis

Early diagnosis is important to prevent further complications [2]. There is not a single test to determine Sjogren's Disease. The American-European Consensus Group (AEGG) criteria is accepted as a standard reference for Sjogren's diagnosis [8,9]:

1. Ocular symptoms (dry eye)
2. Oral symptoms (dry mouth)
3. Ocular tests

a. Schirmer-I test. This test checks tear production.

b. Rose Bengal Score. This test checks for ocular surface epithelial damage.

4. Evaluation of salivary gland function through sialography
5. Histopathological examination of the salivary glands, such as a lip biopsy for glandular inflammation
6. Autoantibodies-bloodwork-inflammatory markers of autoimmune disease [10]

a. Anti-SSA/Ro or Anti-SSB/La (Sjogren's Antibodies A & B) Blood test when positive is highly suggestive for Sjogren's Disease

b. ANA (Antinuclear Antibody)
Blood test when positive checks if immune system is attacking healthy cells

c. ENA (Extractable Nuclear Antigen)
Blood test when positive identifies connective tissuee diseases

d. RF (Rheumatoid Factor)
Blood test when positive is an indicator for rheumatoid arthritis and other autoimmune diseases

Treatment

Unfortunately, there is no cure for Sjogren's Disease, so treatment focuses on relieving the patient's symptoms and slowing or preventing further damage [3,8]. Treatment options can vary depending upon each individual's symptoms. Some of the common treatments for Sjogren's Disease are:

- Artificial tears such as Restasis to treat dry eye [8].
- Saliva production stimulators such as Biotene to treat dry mouth [8].
- DMARDS (disease-modifying anti-rheumatic drugs) such as hydroxychloroquine (Plaquenil) or methotrexate to treat arthralgia, myalgia, and fatigue [9]. This is considered the first line treatment for Sjogren's patients [3].
- Corticosteroids such as prednisone can be used to treat inflammation, especially during a Sjogren's flare [8].

Case Presentation

A 44-year old female patient presented with several unusual symptoms intermittently over a three-year span: shortness of breath, left shoulder pain, fatigue, blepharitis (eyelid inflammation), joint/tissue swelling, frequent urinary tract infections, muscle weakness, dry mouth, hair loss, hoarseness, throat tightness, and dysphagia. The patient had multiple episodes where the symptoms exacerbated and she would either see her primary care provider or even a provider in the emergency department when the shortness of breath was severe. During those times, the patient would be prescribed a steroid and the severity of the symptoms would subside (sometimes for a few months and sometimes closer to a year). Several providers felt that these symptoms might be stressed induced. But, after a couple of years with the intermittent symptoms, the primary care provider deemed it necessary to refer the patient to multiple specialists due to the numerous systemic manifestations she was experiencing. The patient was referred to a Rheumatologist, Pulmonologist, Neurologist, Dermatologist, Ophthalmologist, and an ENT/Otolaryngologist.

Importance of the Patient Seeing Multiple Specialists

A rheumatologist has expertise in the treatment of inflammatory diseases and autoimmune diseases [11]. Based upon the patient's history of abnormal bloodwork showing inflammation, the rheumatologist decided to have additional bloodwork drawn to look for inflammatory markers that could be associated with autoimmune disease. The bloodwork revealed a positive ENA, positive SSA, low RBCs (red blood cells), low WBCs (white blood cells), low hemoglobin/hematocrit, and low lymphocytes, which can all indicate an infection or inflammation within the body. As a result of these findings combined with the patient's symptoms, the rheumatologist diagnosed the patient with Sjogren's Disease and was started on 200 mg of Plaquenil (DMARD medication). Additionally, due to the patient suffering from blepharitis, it was recommended to use artificial tears and Vaseline to help with the dryness of the eye and eyelid. ***See Figure 1.***

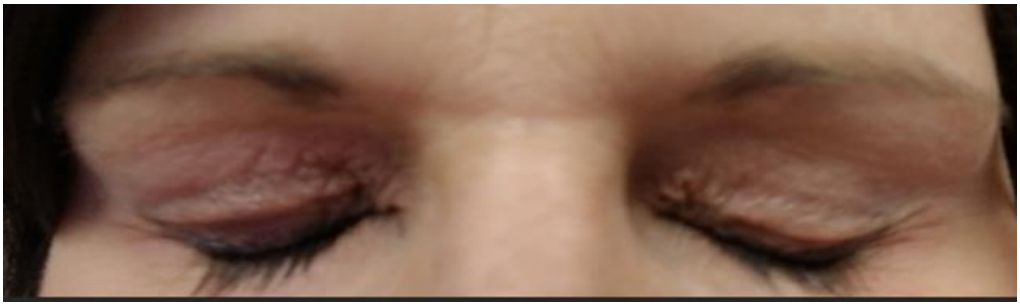

Figure 1: Blepharitis.

Pulmonology's role in patients with Sjogren's Disease is to assist with breathing issues and certain lung conditions linked to Sjogren's, such as interstitial lung disease, which causes inflammation and scarring to the lungs [12]. Due to the patient's complaint of dyspnea, the pulmonologist ordered a pulmonary function test that is utilized to help diagnose or monitor lung disease [12]. The test results revealed a reduction in diffusion lung capacity which can be seen in patients with underlying early interstitial lung disease, obstruction pulmonary disorders such as asthma or emphysema, or pulmonary vascular disease, which is a disease the affects the blood vessels leading to/from the lungs [13]. Diffusion capacity is a measure of capacity to transfer gas from alveolar spaces into the alveolar capillary blood [13]. Additionally, the patient had a CT chest which revealed atelectasis (complete or partial collapse of a lung) with minimal central lobular ground glass opacifications (nodules that can be caused by interstitial fibrosis and inflammation) and vascular indistinctness suggestive of vascular congestion (enlargement of blood vessels within the lungs). As a result of these findings, the pulmonologist felt the patient was in a current Sjogren's flare and had developed asthma-like symptoms. Additional monitoring of the lungs by CT scan and a prescription of Montelukast (anti-inflammatory to prevent allergies and asthma) combined with Prednisone (anti-inflammatory medication) was given to the patient.

A neuromuscular specialist's role in patients with Sjogren's Disease is to aid in impairment of muscle function due to the immune system attacking normal cells by mistake. Neurological involvement of Sjogren's is predominantly those of the peripheral nervous system, such as neuropathy [7]. Small fiber neuropathy is defined by the presence of painful sensory symptoms with normal nerve conduction studies and abnormal neurophysiological tests for small nerve fibers with a skin biopsy [14]. Small fiber neuropathy is divided into two types: non-length dependent (NLD-SFN) and length dependent (LD-SFN). Length dependent small fiber neuropathy tends to involve the longest nerve fibers and mostly the distal lower extremities [14]. Non-length dependent small fiber neuropathy can involve fibers regardless of the length and may affect the proximal and distal legs as well as the arms, face, and trunk of the body [14]. The rheumatologist referred the patient to the neuromuscular physician to assess issues with muscle weakness,

joint swelling, and tingling/numbness/burning feeling in extremities. ***See Figure 2.*** A skin punch biopsy was then ordered to assess for small fiber neuropathy. A skin punch biopsy sample is typically taken at the distal leg and proximal lateral thigh to check for damage to the nerve fibers [15]. Based on symptoms and punch biopsy results, the patient was diagnosed with non-length dependent small fiber neuropathy (NLD-SFN). The patient is currently utilizing an anti-inflammatory diet and regular exercise to help reduce inflammation and nerve irritation.

Figure 2: Normal hand Swollen hand

A dermatologist does not only treat skin conditions that could be associated with autoimmune diseases such as rashes, but also can help with scalp and hair issues. The patient self-referred herself to see a dermatologist due to hair loss. The dermatologist prescribed ketoconazole shampoo and clobetasol propionate (steroid solution) to help with itching and inflammation on the scalp. Minoxidil (a type of hair growth stimulant) is another treatment option for some people. Many times, hair loss can be attributed to an emotional or physical stress that occurred three to six months before the onset of hair loss. In this patient's case, her hair loss would increase about three to six months after an autoimmune flare, which is known as telogen effluvium [16]. Telogen effluvium is a form of nonscarring alopecia where the hair excessively falls out after metabolic stress, hormonal changes, or can be medication related [16]. ***See Figure 3.*** Once the Sjogren's flare has been treated, the hair will usually grow back within a few months to a year [17]. After six months of excessive hair loss, the patient started to see the hair slowly regrow.

Figure 3: Start of hair falling out 1 month later

An ophthalmologist has an important role in the eye health of patients with Sjogren's Disease who take hydroxychloroquine (Plaquenil). As a side effect to the medication, these patients are at a higher risk of retinal damage [18]. Although the patient was not having any problems with her vision, it was recommended by the rheumatologist that she become established with an ophthalmologist. During the initial visit, the ophthalmologist will gain a baseline eye exam to measure the health of a patient's eyes and then evaluate the patient yearly [18]. The patient's baseline eye exam was normal.

ENT/Otolaryngologist's role in patients with Sjogren's Disease to assess the salivary glands and the vocal cords with associated hoarseness and clearing of the throat. Due to the patient's hoarseness, shortness of breath, and dysphagia the patient underwent a laryngeal videostroboscopy. "Videostroboscopy is the most commonly used method to visualize vocal cord vibration and is an essential tool for voice assessment" [19]. ***See Figure 4.***

Figure 4: Laryngeal Videostroboscopy of Vocal Cords During Phonation.

The findings were fairly normal with the exception of some evidence of paradoxical vocal cord motion also known as vocal cord dysfunction. Unfortunately, voice disorders are a result of dysfunctions of the vocal tract due to Sjogren's Disease [20]. Additionally, decreased saliva production can cause esophageal dryness, therefore causing dysphagia [4]. The patient was then seen by a speech pathologist for speech therapy. Speech therapy is one of the most effective treatments for vocal cord dysfunction. It can help coordinate muscle movement around the vocal cords and improve your breathing pattern through breathing and phonation exercises. The patient found these exercises to be quite helpful with the addition of dry mouth moisturizing mouth spray to help with dryness.

Additional providers that can help with Sjogren's patients are:

- Dentist: The loss of saliva production causes an increase in susceptibility to dental issues, which then can lead to loss of teeth [7].
- Urology: Renal involvement is uncommon for Sjogren's patients with the most prevalent disease being interstitial nephritis, which is swelling of the kidney tubules [3].
- Cardiologist: Sjogren's patients are typically associated with greater overall cardiovascular risks, such as pericarditis or myocarditis [21].
- Mental health professionals: Important for improvement to quality of life and mental health

Conclusion

Sjogren's Disease has a myriad of clinical presentations which can make finding a diagnosis difficult [7]. Additionally, autoimmune diseases can take an average of four years before a diagnosis is made [22,23]. Some patients with Sjogren's Disease may only have mild symptoms, while others may undergo good health followed by severe disease [3]. Having a healthcare team can be very important to aid in treatment with the diverse range of symptoms associated with Sjogren's Disease [24]. Additionally, there are some lifestyle changes that can help with Sjogren's symptoms, such as using humidifiers to help with moisture production, extra dental care for dry mouth, moisturizers, getting plenty of rest, eating a balanced diet, and mental health supports are a few examples [8]. Since Sjogren's Disease is a slowly progressive disease that is incurable, it is important to seek medical attention early to help treat the symptoms without the expectation of complete disease remission [3]. A long delay in diagnosis will therefore hinder quick treatment which can lead to insidious progression [25]. As one of the most prevalent autoimmune diseases, proper control over the symptoms associated with Sjogren's could be achieved with the many treatment options that are available, thus the importance of different specialists for this multifaceted disease [10].

Conflict of Interest

The author declares that they have no conflict of interest.

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Declaration of Competing Interest

The author declares that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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