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SJÖGREN'S SYNDROME: A RARE CASE REPORT

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ABSTRACT

Background: Sjögren's syndrome (SS) is a form of autoimmune exocrinopathy that affects the lacrimal and salivary glands. This causes the glands to gradually degrade, resulting in the disease's two signature symptoms, dry eyes and dry mouth. Rarely, a few cases of Sjögren's syndrome (SS) have been reported from the Indian subcontinent. As a result, this condition is not frequently encountered in our region. Even though the disease is uncommon, early detection is crucial to preventing future consequences. The aim of this paper is to emphasize the treatment protocol, diagnostic measures, and management of Sjögren's syndrome. **Case Presentation:** A 40-year-old female was brought to St. John's Medical College Hospital with the complaint of hypothyroidism for 2 months, and she was taken T. thyronorm 100 mcg OD. Currently presented with fever, chills, and rigors since 2 months, at around 4-5 pm, then developed a high-grade fever, which was resolved on T. Dolo, followed by profuse sweating, asymptomatic between fever spikes. Previously presented with generalized mayalgia and cough since 1 month with moderate whitish expectoration and no diurnal or postural variation, which was resolved 1 week ago. The patient also complained about a weight loss of 5 kg over 2 months and bilateral calf pain after climbing stairs. Further examination revealed that anti-SSA and RO52 antibodies were positive, and the ocular SICCA with polyarthralgia was also present, which supported the diagnosis of Sjögren's syndrome. **Outcome:** The prevalence of Sjögren's syndrome is rare in the hospital, even in tertiary care rheumatology clinics. Therefore, more research is needed to have a better understanding of preventive and curative approaches regarding Sjögren's syndrome.

KEYWORDS : Sjögren's syndrome, Case report, rare disease

INTRODUCTION

Sjögren's syndrome is a chronic autoimmune condition that causes glands in the mouth and eyes to generate less moisture. It is named after Swedish ophthalmologist Henrik Sjögren, who first identified the disorder. [1] Sjögren's syndrome is also more susceptible to late diagnosis because of its subtle symptoms, which can take 6 or more years to get diagnosed. [2]

The disease can show a variety of extra-glandular symptoms in addition to an autoimmune exocrinopathy. These can impact the neurological system, musculoskeletal system, pulmonary, gastrointestinal, hepatobiliary, and hematological systems. One of the highest female-to-male ratios (9:1) is found in Sjögren's syndrome (SS) when compared to other autoimmune rheumatic diseases. Additionally, people with Sjögren's disease are particularly susceptible to lymphocytic malignancies. [3] Systemic lupus erythematosus (SLE), rheumatoid arthritis (RA), and systemic scleroderma (SScl) are a few examples of autoimmune disorders that can co-occur with Sjögren's syndrome. Moreover, it can also arise on its own as a primary process (primary SjS). [4] A trigger event can activate the immune system, which starts the syndrome. The immune response is also dysregulated, which results in the development of antibodies that attack healthy cells in the lacrimal and salivary glands locally. Some people's antibodies may also attack different body parts. For some, these antibodies can also attack other areas of the body. [5]

Sjogren's syndrome is mild and does not significantly increase mortality when compared to other autoimmune rheumatic disorders like rheumatoid arthritis (RA) and systemic lupus erythematosus (SLE). [2] Therefore, Sjogren's syndrome is a disease of morbidity rather than mortality, and as a result, it does not get the same level of clinical or scientific attention as its sister disorders. Due to its unawareness and low mortality, Sjogren's syndrome is often referred to as one of rheumatology's "Cinderella" disorders. It is frequently left untreated and undiagnosed, and up until recently, treatment was largely symptomatic. With new attempts to treat Sjogren's syndrome with disease-modifying medications, understanding of its systemic nature and significant morbidity has received increased attention. [6]

Patient Identification

A 40-year-old female was admitted to St. John's Medical College Hospital on July 21, 2023, with the complaints of dyspepsia since 2 months, fever since 2 months, weight loss of 5 kg over 2 months, cough since 1 month, which was resolved 1 week ago, and claudication pain since 1 month in bilateral calf.

Past Medical & Surgical History

The patient had been suffering from hypothyroidism for 2 months and was on T. thyronom 100 mg. On June 1, 2023, the patient had generalized body aches, joint pain, and swelling. RA factor: 87; anti-CCP: negative; ANA: +ve nuclear speckled; ESR: 121; CRP: 57.9. Hence, the patient was diagnosed with rheumatoid arthritis with SAAZ, HCG, and Pred met = developed B/L lower limb pain and mild foot swelling. Therefore, the patient went to another hospital (Manipal) for further consultation, where she stopped all rheumatoid tablets.

Family History

There are three members in the family. The type of marriage is a non-consanguineous marriage. All other members of the family were not having any health issues, except the patient.

Physical Examination

There are not many abnormalities found in the head-to-toe examination. During the physical examination, the patient was conscious, cooperative, and oriented to time, place, and person. The vitals were also checked, and the parameters were normal. (BP: 130/80 mm Hg, PR: 100 bpm, RR: 18 cpm, spo2: 99%, JVP: normal, temperature: afebrile.) There were signs of pallor, icterus, cyanosis, clubbing, edema, generalized lymphadenopathy, and also normal spine, breast, and thyroid functions.

Systemic Examination

During systemic examination, R5 was clear, and normal vesicular breath sounds were heard with equal lung air entry. The cardiovascular system was clear. S1 and S2 were heard with no murmurs. Per abdomen was soft and non-tender; there was no organomegaly and bowel sounds present. During the examination of the central nervous system, the patient was conscious, cooperative, and oriented to time, place, and person. Human milk fortifiers were also normal.

Clinical Finding

Severe dry eyes, breathlessness and polyarthralgia.

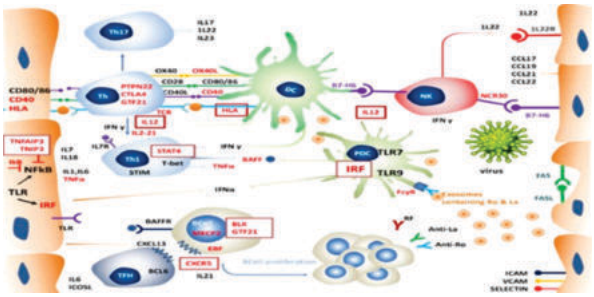

Fig-1: Pathophysiology Of Sjögren's Syndrome

Diagnostic Assessment

Blood culture: negative, procal: negative, Neck/Axilla USG: B/L reactive lymphnodes, USG A+P: normal, Echo: EF 67%, PASP: 30 MMHG, There is no evidence of infective endocarditis and no RWMA. Bone marrow aspiration was normal, but bone marrow culture was noted to show Acinetobacter species. The ENMG report shows demyelinating and axonal motor neuropathy with evidence of conduction block in the lower limb (left>right). The chest x-ray and USG abdomen were normal. A neck USG was also done, which shows reactive cervical and bilateral axillary lymph nodes. Moreover, the immunology report shows SSA: 3+, RO52: 3+, and SSB: 1+, and schemer was 2mm on the right side and 2mm on the left side.

Therapeutic Intervention

PTA + tympanometry; Xylomist nasal drops x TID; T. Montek LC x OD; Refresh liquigel drops x QID; Lacrigel eye ointment x BD; T. HCQ 300 mg P/O x OD; C. Orofer XT 200 mg x OD; T. Folicitrax 15 mg; T. Folic acid 50 mg; Tab Ciprofloxacin 750 mg x BD; Tab Thyronorm 100 mg x OD; Tab Eliwel 10 mg x OD. Tab Pan 40 mg x OD, Tab Dolo 650mg x OD

DISCUSSION

Sjögren's syndrome is a serious disease. However, in a positive sign, most of the affected patients will not die from this, which could be a sense of relief. All Sjogren's patients, including those who are seronegative (SS-A negative), are at risk of significant complications. Regardless of the patient's risk profile, monitoring is critical. Patients with a high risk of serious disease require more frequent monitoring. [7] Sjögren syndrome mostly affects women over the age of 40. However, some newly developed diagnostic techniques, such as parotid biopsies and antibody identifications, show promising results in children and teenagers. The enlargements of the parotid glands are often asymmetric and painless. [8]

There are mainly two forms of Sjögren's syndrome: primary Sjögren's syndrome, which develops on its own, and secondary Sjögren's syndrome, which develops in addition to other autoimmune diseases like rheumatoid arthritis, lupus, and psoriatic arthritis. [9] In this case, we reported that the patients had a history of rheumatoid arthritis, which might aggravate the symptoms of Sjogren's syndrome. Sjögren's syndrome treatment relies on the extent and severity of clinical manifestations and is best implemented through a multidisciplinary approach. Symptomatic treatment includes artificial tears and salivary substitutes to relieve the symptoms and prevent local infectious complications like conjunctivitis and corneal inflammation, the development of caries, and periodontal disease. A thorough dental preventive program should be implemented in all cases. Corticosteroid treatment should be reserved for all cases showing evidence of organ damage, significant leukopenia, or severe clinical symptoms. [8]

The classic symptoms of Sjogren's syndrome are dry eyes, dry mouth, and swollen parotid glands, and in this case, the patients had severe dry eyes and polyarthralgia. However, a study done in the USA [9] shows that the patient was suffering from extraglandular involvement of the cutaneous and xeroderma frequently. Along with this, the patients also complained about gynecological problems like vaginal dryness, fetal loss, and painful intercourse. However, in this present study, the patient did not have any similar symptoms. This can show that there can be an atypical presentation of the symptoms as well. It is the duty of healthcare professionals to approach every patient with an open mind and open eyes. The doctors and nurses need to take every aspect of the health symptoms into account when making a diagnosis. Therefore, it becomes very important for the nursing and medical fraternities to always look for the associated signs. [10]

CONCLUSION

Overall, Sjögren's syndrome patients experience higher morbidity and can also expect a normal lifespan. However, awareness is also one of the most important factors in improving the condition. Continuous monitoring and early intervention can improve outcomes. At the end, to minimize the risk factors, preventive measures should be implemented, like up-to-date immunizations, cervical cancer screening for women, and reducing cardiovascular risk factors. More research on Sjögren's syndrome is indeed important to consider.

Consent: As per international or university standards, the patient's written consent has been collected and preserved by the author(s).

Competing Interests: None declared

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