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*Corresponding author

*Dr. Bashir Abdrhman Bashir, Faculty of Medical Laboratory Sciences, Port Sudan Ahlia University, Port Sudan,Sudan

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Sjogren's Syndrome in a Sudanese female: A Case Report

Dr. Bashir Abdrhman Bashir*

Faculty of Medical Laboratory Sciences, Port Sudan Ahlia University, Port Sudan,Sudan

Abstract

Background: Sjögren's syndrome (SS) is a variety of autoimmune inflammatory conditions distinguished by lymphocytic glandular invasion and various extra-glandular manifestations. SS is most likely seen in middle-aged females. Immunological, viral, genetic, ecological, and hormonal etiologic aspects are dubious regarding the etiology of SS, and no aid has been provided.

Case presentation: A 30-year-old female reported indications of dry eyes, diminished salivation, and trouble swallowing for the past two years, as well as a persistent dry cough lasting for 10-15 days and increasing dyspnea lasting for 4-5 days. Anti-Ro/Sjögren's system-A and Anti-La/Sjögren's system-B antibodies and anti-nuclear antibody (ANA) were positive among the profiles. A sub-lingual incisional biopsy was also performed, which confirmed the existence of Sjögren's syndrome. The patient demonstrated substantial improvement after initiating oral glucocorticoids, systemic anti-inflammatory drugs, artificial tear drops, oral iron supplements, and other supportive care.

Conclusion: The main findings of this case report are the conventional positive results of anti-Ro and anti-La antibodies and their histopathological inspection of a lip biopsy in a female adult. She began taking oral steroids as well as other supportive medications. Her overall condition has vastly improved, and she is doing extremely well on regular follow-ups.

Background

Sjögren's syndrome (SS) is a systemic autoimmune ailment marked by lymphocyte infiltration and eventual execution of the exocrine glands, which include those in the nose, ears, skin, vagina, pulmonary, and gastrointestinal systems [1]. Clinically, salivary gland hypofunction and sicca keratoconjunctivitis result from the involvement of the salivary and lacrimal glands [1,2]. Dry eyes, dry mouth, fatigue, muscle and skeletal soreness, and enlargement of salivary glands are typical symptoms. To enhance consistency in its diagnosis, many cooperative initiatives have been made to categorize Sjögren's Syndrome correctly [2-5].

To identify and categorize people with Sjögren's syndrome, various distinct classification and diagnostic criteria sets have been published. However, a multispecialty international panel of experts has created consensus criteria for primary Sjögren's syndrome utilizing the American College of Rheumatology (ACR) and the European League Against Rheumatism (ELAR) recommendations [6]. Both professional associations have now formally adopted these criteria, which were verified in various cohorts of patients [6,7]. These standards were created chiefly to categorize homogeneous patient populations for clinical trials and other types of research. [8]

The majority of SS treatments are symptomatic and experimental. Parasympathomimetic and artificial tears are frequently recommended for ocular problems. Remedy for oral signs is made available by the use of artificial saliva, sugarless gums, and mucolytic pharmaceuticals. Nonsteroidal Anti-inflammatory Drugs (NSAIDs) are recommended to minimize edema in the salivary glands. Oral

Table

Variables	Patient results	Normal or control
White blood cells **x10^9/l**	8.7	4 – 10
Red blood cells **x10^{12}/l**	4.29	3.5 – 5.5
Hemoglobin **g/dl**	11.9	12 – 16
Hematocrit **%**	36.2	35 – 47
Mean Corpuscular Volume **fl**	84.5	78 – 98
Mean Corpuscular Hemoglobin **pg**	27.8	26 – 35
Mean Corpuscular Hemoglobin Concentration **%**	32.9	30 – 36
Absolute lymphocytes count **x10^9/l**	2.9	1.0 – 4.3
Absolute neutrophil count **x10^9/l**	5.1	1.5 – 7.0
Absolute monocytes count **x10^9/l**	0.52	0.1 – 1.0
Platelet count **x10^9/l**	352	150 – 400
Erythrocyte sedimentation rate, 1st hour, **mm/h**	90	Up to 20
Erythrocyte sedimentation rate, 2nd hour, **mm/h**	116	Up to 30
Bleeding time, **min**	3.49	2 – 7
Clotting time, **min**	7.39	5 – 15
Clot retraction, **%**	60	30 – 120
Prothrombin time, **s**	13.1	12 – 16
Partial thromboplastin time, **s**	28.4	26 – 43
Thrombin time, **s**	12.2	8 – 18
Antithrombin, **%**	93	78 – 126
D-Dimer, **mg**/l	< 0.1	Up to 0.3
Creatinine**, mg/dl**	0.6	0.4 – 1.6
Blood Urea**, mg/dl**	21.0	10 – 50
Blood Urea Nitrogen**, mg/dl**	9.0	7 - 21
Uric Acid**, mg/dl**	4.3	3.4 – 7.0
Total Bilirubin**, mg/dl**	0.46	0.2 – 1.3
Direct bilirubin**, mg/dl**	0.09	Up to 0.25
Total protein, **g/dl**	6.9	6.6 – 8.3
Albumin, **g/dl**	3.7	3.5 – 5.5
Alanine transaminase, **U/l**	19	Up to 41
Aspartate transaminase, **U/l**	20	Up to 40
Alkaline phosphatase, **U/l**	66	Up to 115
Glycosylated Hemoglobin**, %**	5.3	4.5 – 6.5
Thyroid-stimulated hormone, **µIU/ml**	0.61	0.27 – 4.2
TT4, **ng/dl**	5.8	4.9 – 12
TT3, **ng/dl**	1.5	0.79 – 2.58
Hepatitis C virus screening (HCV)	Negative	—
Hepatitis B virus screening (HBV)	Negative	—
Human Immunodeficiency Virus Screening (HIV)	Negative	—
Hepatitis C virus screening (HCV)	Negative	—
Cytomegalovirus (CMV)	Negative	—
Complement C3, **mg/dl**	112	75 – 135
Complement C4, **mg/dl**	25	9 – 40
Anti-streptolysin O titer	55	< 200
C-reactive protein (quantitative), **mg/l**	20.0	< 10.0
Rheumatoid factor, **IU/ml**	14.3	Up to 20
Anti-cyclic citrullinated peptide, **U/ml**	7.0	Up to 20
Antinuclear factor antibody (ANA)	1/320	< 1:100
Anti-double stranded DNA (dsDNA), **IU/ml**	< 30	30 - 70
Anti-Sjogren syndrome-related antigen A (Ro), **U/ml**	299.2	Up to 20
Anti-Sjogren syndrome-related antigen B (La), **U/ml**	33	Up to 20
Anti-Smith antibodies, **EU/ml**	12.5	< 20
Anti-neutrophilic cytoplasmic antibodies **AU/ml**	7.2	< 20
Anti-cardiolipin antibodies (IgG), **AEU/ml**	6.9	Up to 10
Anti-cardiolipin antibodies (IgM), **AEU/ml**	4.7	Up to 8
Beta-2-Glycoprotein antibodies (IgG), **SGU/ml**	0.4	< 20
Beta-2-Glycoprotein antibodies (IgM), **SMU/ml**	0.5	< 20
Lupus anticoagulant, **s**	37	26 – 45

corticosteroids and immunosuppressants are prescribed to treat symptoms throughout the body [6]. This report presents a case study of a patient with SS, including their clinical presentation, paraclinical examinations, alternative diagnoses, and treatment plan based on recent research.

Case presentation

Sjögren's syndrome was identified in a 30-year-old woman. The primary problems were dry eyes (xerophthalmia) and a two-year history of recurrent sensation of sand in the eyes, both of which the ophthalmologist treated with supportive medicine without success. In addition, this patient had a prolonged dry cough for 10 to 15 days, rising dyspnea for 4 to 5 days, and signs of dry mouth (xerostomia). There is no known family history of a medical problem. Red eyes were also discovered during the examination, and vital signs were recorded as BP 108/81 mmHg, pulse rate 92 beats/min, and body temperature 36.4 C°. Laboratory tests looked at usual renal and liver profiles, a full hemogram, and typical coagulation markers (Table 1). Thyroid function tests, random blood glucose levels, and anti-streptolysin O results were normal. The erythrocyte sedimentation rate was grossly high. Complement (C3) and (C4), double-strand DNA (ds-DNA), antineutrophil cytoplasmic antibodies, and all other parameters were normal. Antinuclear factor (ANA) titers were abnormal, and autoantibodies of anti-SSA/Ro and anti-SSB/La were positive. Additionally, the patient expresses exhaustion and polyarthralgia. There was a negative Rheumatoid factor. Furthermore, negative results came from screening tests for syphilis, cytomegalovirus (CMV), human immunodeficiency virus (HIV), hepatitis B, and hepatitis C viruses (Table 1).

The diagnosis of Sjögren's syndrome was also proved by lymphocytic infiltration in an oral incisional biopsy. The Schirmer test was run on both eyes, soaking the test strip after 5 minutes with 3mm for the left eye and 4mm for the right. For almost three months, patients have been receiving treatment with Resochin, Imuran, and Medrol. The therapy was changed after a thorough clinical

assessment and paraclinical studies. Folic acid, artificial tears, Medrol 1 tablet daily, and Methotrexate 15 mg once a week were prescribed. On the day that methotrexate was taken, folic acid was skipped. The problems in the mouth and eyes have greatly alleviated after seven weeks. Regular paraclinical exams and appointments with a rheumatologist, endocrinologist, and neurologist are strongly advised.

Discussion

SS is a pathologic disorder that typically manifests as the so-called Sicca syndrome, which includes a dry mouth and eyes as well as extraglandular expression. In roughly 20 to 30% of instances, reduced function and hypertrophy of the glands also coexist with vasculitis, pulmonary or renal problems, tiredness, and polyarthralgia [6,9]. The etiology of Sicca syndrome, which can be split into glandular and non-glandular causes, is numerous and well-known (or extraglandular). Neurologic and emotional changes, dehydration, oral sensory impairment, mouth breathing, and unexplained risk factors are all included in the non-glandular etiology. The most major glandular reasons were discovered to include immunodeficiency syndrome, diabetes, sarcoidosis, end-stage renal disease, radiotherapy, trauma, and gland tumors [7]. People treated with beta-blockers, tricyclic antidepressants, antihistamines, or diuretics may experience xerostomia and keratoconjunctivitis sicca. These details ought to be taken throughout the workup, and it is imperative to learn as much as you can about the patient's overall health [6]. The medical history must be carefully reviewed to identify the appropriate course of treatment and establish the correct diagnosis.

When performing a clinical examination on a patient, SS should always be anticipated, especially in middle-aged women who exhibit symptoms associated with mouth and enlarged salivary glands. Additional interrogation will check the existence of further indications, especially keratoconjunctivitis sicca and arthritic problems. According to the updated international classification criteria outlined by Carson et al [10], the diagnosis of primary SS is made in this case. In the current case, the patient disclosed using NSAIDs and painkillers with just momentary effects. Treatment for SS is still debatable. For the proper control of the indications, conservative treatment, regular accurate clinical examinations, and paraclinical diagnostics are likely and of utmost importance.

Conclusion

The key conclusions of this case study include the typical favorable outcomes of anti-Ro and anti-La antibodies and their lip biopsy. Together with additional supporting drugs, she started taking oral steroids. Eventually, there has been a significant improvement, and regular follow-ups show excellent results.

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