

Original article

CHALLENGES OF DENTAL MANAGEMENT OF SJOGREN'S SYNDROME IN A NIGERIAN PATIENT: A CASE REPORT AND REVIEW OF LITERATURE

Eweka OM¹, Erinoso OA², Adekunle AA²

¹Department of Preventive Dentistry, College of Medicine, University of Lagos, Idi-araba, Lagos.

²Department of Oral and Maxillofacial Surgery, Lagos University Teaching Hospital, Idi-Araba, Lagos.

ABSTRACT

BACKGROUND: Sjögren's syndrome is a slowly progressive inflammatory disorder that involves the exocrine glands (lacrimal glands and salivary glands) leading to the dryness of the eyes and mouth. It is a rare condition that is seen commonly in females of the middle age.

OBJECTIVE: To present challenges encountered during the management of oral symptoms in a patient with Sjögren's syndrome.

CASE REPORT: A 46-year-old female Nigerian Patient who presented with bilateral facial swelling with complaint of dry eyes, dry mouth, and knee joint pain. Investigations done using sialography with contrast medium of the left parotid and Schirmer's test of both eyes were diagnostic of Sjögren's syndrome. Proper treatment could not be instituted until about one year after the patient's initial presentation due to the non-availability of the required medications in the country.

CONCLUSION: Poor access to the recommended medications for the treatment of patients with Sjogren's syndrome poses a challenge to the clinician, rendering such patients helpless and unsatisfied.

Keywords: Sjogren's syndrome, Salivary gland, Xerostomia, Dry eyes.

Correspondence address:

Dr. Eweka O.M.

Department of Preventive,
Dentistry, College of Medicine,
University of Lagos
Email: tolaeweka@yahoo.com
+2348023203289

INTRODUCTION

Sjogren's syndrome (SS) is a chronic, slowly progressive auto-immune disorder characterized by a triad of xerostomia (dry mouth), keratoconjunctivitis sicca (dry eyes), rheumatoid arthritis (RA) or other connective tissue diseases.

¹ It manifests with exocrine dysfunction and lymphocytic infiltration of exocrine glands. ²⁻³ It is the most common autoimmune disease next to Rheumatoid arthritis primarily affecting perimenopausal and post-menopausal females with a female: males ratio of 9:1. ³⁻⁵

Sjogren's syndrome may be classified into primary and secondary types. Patients with primary SS have keratoconjunctivitis sicca (KCS) and xerostomia, whereas those with secondary SS

have KCS, xerostomia and other underlying autoimmune diseases such as rheumatoid arthritis, systemic lupus erythematosus (SLE) and systemic sclerosis, as well as other connective tissue disorders. ⁵⁻⁶ The prevalence of primary SS in the general population has been estimated to be between 0.5 - 3%, whereas secondary Sjogren's has been observed in approximately 10-20% of patients with rheumatoid arthritis, SLE, and scleroderma. ⁷⁻⁸

These studies were however reported among Caucasian populations example in studies conducted by Bowman's et al in 2004 in the UK, who had a low prevalence of PSS (0.1-0.6%) in the tested population, there were no Nigerian studies citing the prevalence of Sjogrens

syndrome so far.⁹ These proofs that SS remains a rare condition.

Sjögren's syndrome affects any age but commonly adults between the ages of 40 and 60 years.⁵ The etiology and pathophysiology of SS are still unknown. Autoimmunologic factors such as multiple autoantibodies particularly SS-A and SS-B, and a genetic predisposition are associated with SS. Some human leukocyte antigens (HLA) also increase the risk of SS.³ Viral infections may be involved in its induction.^{1, 3, 5} Epstein-Barr virus, human T-lymphotrophic virus-1, human herpes virus, human immunodeficiency virus-1, hepatitis C virus, and cytomegalovirus may also play a role in its aetiology.⁵

The diagnosis of Sjögren's syndrome is based on several clinical and laboratory findings. Early clinical manifestations of SS are primarily decreased tear and saliva secretions, leading to dry eyes and dry mouth syndromes which can exacerbate an increased rate of bacterial infections in the mouth, as well as caries, sialoadenitis and oral mucosal infections.¹⁰⁻¹⁴ Diagnosis of secondary Sjögren's Syndrome requires an established connective-tissue disease and one sicca symptom plus two objective tests for dry mouth and eyes at the time of presentation. Exclusions now include previous radiotherapy to the head and neck, lymphoma, sarcoidosis, graft-versus-host disease, and infection with hepatitis C virus, human T-lymphotrophic virus type I, or HIV. Measurements of tear and saliva flow must be made in the absence of drugs that have anticholinergic side-effects.²

There is no consistently effective or curative treatment for Sjögren's Syndrome. As oral manifestations are common place in these patients, it is important that dentists know how to recognize and manage this condition.⁵ Palliative therapies, such as use of lubricants and artificial saliva, increased fluid intake, and salivary stimulation, are used to relieve the symptoms.

There is a previous Nigerian study that reported on this condition,¹⁵ with no known published literature on the associated oral manifestations of Sjogren's syndrome in Nigerians. This case report therefore highlights the challenges encountered in the management of oral symptoms

seen in a patient with Sjögren's syndrome in a Nigerian population.

CASE REPORT

A 46-year-old female patient presented at the Oral Medicine clinic of the Lagos University Teaching Hospital, Lagos, Nigerian, with a complaint of bilateral, painless facial swelling of three months duration. The associated complaints included dry mouth and difficulty in swallowing of one year duration, dryness of the eyes with associated pepperish sensations and joint pain of five months duration. There was no associated pain, altered taste or burning sensations in the oral cavity.

A positive medical history of blocked ears of one year duration was obtained. The ear problem was being managed by the Otolaryngology unit of the hospital. There was no history of recent drug use and hospitalization.

On clinical examination, there was an obvious symmetrical, bilateral swelling in the parotid region, firm in consistency and non-tender. Intra-oral examination revealed dryness of the oral mucosa, dry and depapillated tongue, a carious cavity on the lower left second molar and retained roots of the upper left third molar.

A provisional diagnosis of Secondary Sjogren's syndrome was made. The following investigations were requested - Full blood count (result was within normal haematological range), SSA/SSB antigen assay (results of which was never obtained due to unavailability of the test), Schirmer's test, and a sialogram of the parotid and submandibular glands. Labial biopsy was not requested.

Symptomatic treatment was commenced to provide relief, aid salivary flow and prevent further complications of oral dryness, in form of biotene saliva stimulant. However, the biotene could not be obtained by the patient due to its unavailability in the Nigerian market. Fluoride therapy was instituted as well as full mouth scaling and polishing. Patient was also advised to chew sugar free/sugarless gums and sip water frequently, but this brought only very little relief to the patient.

On subsequent follow-up visits, Schirmer's test result was obtained from the Ophthalmology unit of the same hospital. Results detailed 3mm in the right eye, while the left was 4mm after 5 minutes. A sialogram of the parotid was done. This sialogram on the left parotid revealed the characteristic "Snow storm" appearance of sialectasia as a result of leakage of contrast medium.



Fig.1. Sialogram of the left parotid gland with Contrast medium: arrow shows the snow storm appearance of the left parotid after injection of contrast medium.

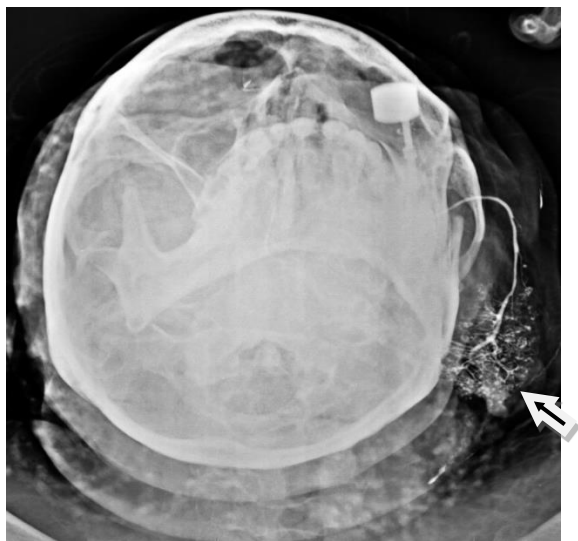


Fig.2. Occipitomenal view with Sialogram showing snow storm appearance with Contrast medium of the left parotid gland.

The results of the Sialogram, the Schirmer's test along with the clinical presentation were diagnostic of Sjögren's syndrome in the patient. The patient was treated symptomatically, considering that there is yet no curative treatment for the condition. She was advised to take frequently sips of water, chew sugarless gum to help stimulate saliva production pending her procurement of biotene. The patient was hereafter referred to the ophthalmology and rheumatology clinics for expert management of other associated symptoms.

DISCUSSION

Sjogren's syndrome is a systemic, autoimmune disease causing secretory gland dysfunction, resulting in the dryness of the main mucosal surfaces such as the mouth, eyes, nose, pharynx, larynx, and vagina.¹⁶ It starts in the salivary and lacrimal glands but eventually includes involvement of multiple organs.¹⁷ Despite extensive study of the underlying cause of Sjögren's syndrome, the pathogenesis remains obscure. In broad terms, the pathogenesis is multifactorial; environmental factors are thought to trigger inflammation in individuals with a genetic predisposition to the disorder.²

The diagnosis of SS could be rather challenging, this is guided by a combination of clinical manifestations, glandular dysfunction, laboratory exams, and minor Salivary Gland biopsy.¹⁸ Many investigators utilize diagnostic criteria for SS based on the revised American-European Consensus Group which include several criteria. Diagnosis is based on meeting three of four objective criteria or four of six total criteria (Table 1).¹⁹

The reported case fulfilled the diagnostic criteria of SS on presentation by meeting 4 of the 6 total criteria. The diagnostic approach for SS starts by ruling out other causes of dry eyes (e.g. allergic conjunctivitis, blepharitis) and dry mouth (e.g. Xerostomia caused by diabetes, chronic viral infections, dehydration, irradiation of salivary glands, and especially, drugs).¹⁶

Table 1. 2002 American-European classification criteria for Sjogren's syndrome

Symptoms	A +ve response to at least one of the following questions
1. Ocular symptoms	a) Have you had daily, persistent, troublesome dry eyes for more than 3 months? b) Do you have a recurrent sensation of sand or gravel in the eyes? c) Do you use tear substitutes more than three times a day?
2. Oral symptoms	a) Have you had a daily feeling of dry mouth for more than 3 months? b) Have you had recurrently or persistently swollen salivary glands as an adult? c) Do you frequently drink liquids to aid in swallowing dry food?
3. Ocular signs	Objective evidence of ocular involvement defined as a positive result for at least one of the following tests a) Schirmer's test for tear function, performed without anesthesia (positive result ≤ 5 mm in 5 minutes) b) Rose Bengal score or other ocular dye score (positive result score ≥ 4 on the van Bijsterveld scoring system)
4. Salivary gland involvement	a) Unstimulated whole salivary flow (<1.5 mL in 15 minutes) b) Parotid sialography with contrast medium showing presence of diffuse sialectasias (punctate, cavitory, or destructive pattern) without evidence of obstruction in the major ducts c) Salivary scintigraphy showing delayed uptake, reduced concentration, or delayed excretion of tracer
5. Histo-pathology	In minor salivary glands (obtained through normal appearing mucosa) focal lymphocytic sialoadenitis, evaluated by an expert histopathologist, with a focus score of 1 (defined as the number of lymphocytic foci (which are adjacent to normal appearing mucous acini and contain >50 lymphocytes) per 4 mm^2 of glandular tissue)
6. Autoantibodies	Autoantibodies to Ro/SS-A or La/SS-B antigens, or both

Patients with Sjogren's syndrome usually presents with a complaint of dry mouth at the dental clinic and a high index of suspicion is required. The most common dry mouth symptoms are reported thirst, saliva that seems thick, stringy, dry, sticky feeling in the mouth, Halitosis (bad breath), dry hoarseness of the throat, dry, irritable and scratchy tongue, burning or tingling sensation of

the tongue, difficulty in speech, inability to chew, swallow or taste food. Other symptoms that may be associated with dry mouth include dry nasal passages, painful sores of the mouth and tongue, chapped lips, increased plaque, dental caries and periodontal disease.²⁰⁻²³ In this case presented, we saw the patient with total dry mouth, which resulted in difficulty in swallowing and speech with associated bilateral swelling of the parotids and pain in the joints.

Furthermore, on examination of the mouth using a tongue depressor, this stuck to the buccal mucosa,²⁴ because of the dry oral mucosa. There was little or no pooled saliva in the floor of the mouth, and the tongue appeared dry with decreased number of papillae, the saliva appeared foamy. Dental caries may be found at the cervical margin or neck of the teeth, the incisal margins or the tips of the teeth,²⁵ as was found in the reported case.

The study of impaired saliva and lacrimal secretions is not routinely available in most dental settings in Nigeria. However, two tests; unstimulated salivary flow measurement, and Schirmer's test—have simple technical requirements readily available in most clinics (graduated tube and a small strip of filter paper, respectively). For salivary flow measurement, the patient is instructed to spit saliva into a graduated test tube every minute; a quantity of <1.5 mL collected over a 15 minutes period indicates impaired saliva secretion, as was done in our case where 1ml was obtained every 5 minutes.

Schirmer's test consists of a strip of filter paper placed in the lower conjunctival corneal sac; lacrimal wetting of <5 mm of the paper after 5 minutes shows abnormality, as seen in the above patient with 3mm after 5mins in the right eye and 4mm in the left eye. Clinical suspicion of Sjögren syndrome should increase when systemic manifestations accompany the sicca complaints, particularly joint pains indicative of rheumatoid arthritis as detailed by the authors in the reported case.¹⁶

Despite the availability of many new imaging procedures, Sialography / sialogram has after decades of use maintained its status as the imaging procedure of choice for evaluating the oral component of Sjögren syndrome (SS). With

expert observers' sensitivity of 87% and a specificity of 84% has been reported.²⁶

In a resource limited environment as Nigeria, the use of more sensitive and specific imaging techniques for diagnosing SS, such as MRI and scintigraphy are not routinely conducted due to the cost implications and scarce availability of radiological centers with these devices.

Treatment of SS is symptomatic. It is directed toward the particular areas of the body that are involved and must be individualized.² Dry mouth can lead to an increased rate of pathologies in the mouth and there may be a need for pharmacotherapy. The management of dry mouth include the use of topical agents such as oral polymer based sprays, so called saliva substitutes, sipping of small amounts of water during the day, elimination of caffeine containing products, chewing of sugar free gum, and elimination of alcohol containing mouth rinses, alongside meticulous oral hygiene and patient motivation.²⁷⁻²⁸

Cholinesterase inhibitors such as pilocarpine are sometimes recommended to stimulate salivary secretion but any benefits must be counter-balanced by side-effects such as nausea, diarrhoea and bradycardia.⁵ Furthermore, in dentate patients, sweet-eating should be discouraged, and a bi-annual dental visit should be advised.²⁹ The patient had challenges obtaining any of the artificial saliva substitutes or stimulants because these medications are not readily available, she had been sipping water which did not give much improvement of the oral dryness.

Dryness of the eyes is treated with artificial tears such as methyl cellulose solution,⁵ after due consultation with the Ophthalmologist. Appropriate referral and consultation was instituted in the multi-disciplinary management of the patient, this multi specialist approach is the key to effective management of SS.

In conclusion, Sjögren's syndrome remains a rare and discomforting autoimmune disorder characterized by dryness of the mouth and eyes, with bilateral swelling of the salivary gland. This condition can be associated with other connective tissue disorders such as rheumatoid arthritis. In the Nigerian environment, management has

always been a challenge due to the scarcity of recommended objective diagnostic and treatment modalities. Poor availability of the recommended medications for the treatment of patients with Sjögren's syndrome poses a challenge to the Clinician, rendering such patients helpless and unsatisfied. Reports in the Nigerian population remain scarce to the best of our knowledge, therefore this case report brings to the fore the clinical presentation, diagnosis, investigation and challenges of treatment in a Nigerian patient with Sjögren's syndrome.

Consent: Written informed consent was obtained from the patient for publication of this case report and the accompanying images.

Conflict of Interest: None declared

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