

# UNVEILING THE ROLE OF ORAL PHYSICIAN IN THE DIAGNOSIS AND MANAGEMENT OF SECONDARY SJOGREN'S SYNDROME

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## ABSTRACT

### UNVEILING THE ROLE OF ORAL PHYSICIAN IN THE DIAGNOSIS AND MANAGEMENT OF SECONDARY SJÖGREN'S SYNDROME

Sjögren's syndrome is a chronic autoimmune inflammatory disease, which principally attacks the lacrimal glands and salivary glands. The damage that occurs to these glands in Sjögren's syndrome leads to a decrease in the volume of secretions (tears or saliva) and a change in their quality. This results in the symptoms that characterize the disorder, including dry eyes or dry mouth. When it occurs with rheumatoid arthritis, the condition is said to be secondary to rheumatoid arthritis and is called Secondary Sjögren's syndrome.

The present case is of a 50 year old female patient with a history of burning sensation of the oral cavity; she was previously diagnosed with rheumatoid arthritis. A positive immunological assay of SS-A and SS-B antibodies confirmed a diagnosis of Secondary Sjögren's syndrome. The aim of this case report is to highlight the early diagnosis and treatment planning of Secondary Sjögren's syndrome including effective management of oral symptoms by the oral physician. This can definitely pave the way for a comfortable life style and delay the disease process for the patient.

**KEY WORDS:** Secondary Sjogren's Syndrome, Oral Physician, Oral Cavity, Xerostomia

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## INTRODUCTION

Sjogren's syndrome (SS) is a chronic autoimmune disorder of the exocrine glands associated with lymphocytic infiltration and immunological hyper reactivity. It involves several organs like lungs, kidneys, and nervous system.<sup>[1]</sup> Glandular involvement is seen more commonly which involves the salivary and the lacrimal glands. This leads to progressive dryness of the mouth called Xerostomia and of the eye called as, Xerophthalmia.<sup>[2]</sup> Dr. Henrick Sjogren, an ophthalmologist, first described this condition in 1933.<sup>[1]</sup> SS can be classified as primary and secondary. Primary Sjogren syndrome which occurs by itself and is not associated with any other diseases. Secondary Sjogren's syndrome develops in association of an underlying autoimmune disease like, Rheumatoid arthritis, Systemic lupus erythematosus, Poly arteritis nodosa and Scleroderma.

The etiology of SS is said to be controversial and is multifactorial. The pathogenesis is however complicated and is not completely understood. It causes a multifaceted activation of the immune system where there is B-lymphocyte deregulation and hyperactivity.<sup>[3]</sup>

Few events like, the initiation by an exogenous factor, disruption of salivary gland epithelial cells, T-lymphocyte migration and lymphocytic infiltration of exogenous glands and production of rheumatoid factor and antibodies to Ro (SS-A) and La (SS-B) are postulated in the

pathogenesis of SS.<sup>[2]</sup> The hall mark of this disorder is, xerophthalmia (Keratoconjunctivitis sicca) and xerostomia.<sup>[2]</sup> The most serious complication of Sjogren's syndrome is salivary gland and gastrointestinal tumors like, B-cell lymphoma which develop in approximately 5% of patients.<sup>[1]</sup>

The diagnosis criteria are reassessed so that no patient can be missed having the condition. Most widely accepted are the American and European group developed international classification criteria for Sjögrens syndrome.<sup>[4]</sup> The symptoms and signs in SS patients are considered to be very vague and can be easily misdiagnosed by a general and an oral physician. Lack of awareness of this entity leads to a delayed management by the attending physicians, ophthalmic surgeons and dentists which may be one of the factors for the suffering experienced by most of these patients.

The present case report highlights the role of the oral medicine specialist in accurately diagnosing the condition and rendering an appropriate treatment to the patient, thus improving her quality of life. The patient was treated by a rheumatologist and an ophthalmologist but an oral physician had effectively treated the patient successfully. The effective management of oral manifestations and minimization of oral diseases result in an improved general well being of the patient. This can certainly pave the path for a comfortable lifestyle to the patient and an effective diagnosis can delay the disease process.

## **CASE REPORT**

A 50-year-old female patient reported to the department of Oral Medicine and Radiology with the chief complaint of burning sensation, oral dryness and with a history of difficulty to eat spicy food past 2 years. Her complaint of dryness in the mouth started 3 years ago with coexistent arthralgia, followed by the dryness of the eyes since 5 years. She had given a history of presence of a foreign body like sensation in the eyes and was advised to use Refresh tear drops (carboxy methyl cellulose) by an ophthalmologist 5 years ago.

Her medical reports revealed a Positive rheumatoid factor (44 IU/ ml) suggestive of Rheumatoid arthritis, an increased ESR (51H) and also Antibody to Sm, SS-A, SS-B was positive. Patient was under the following medication for rheumatoid arthritis: Methotrexate once daily and prednisolone twice daily prescribed by the general physician. She discontinued this medication and since 1 year she was on Ayurveda medicine and was not aware of the name of the medication.

On general examination, she appeared fit and healthy with the vital signs within the normal limits. On intraoral examination there was lack of pooling of saliva with sticking of one end of mouth mirror to the surfaces of oral mucosa suggestive of dry mouth. At the corners of the lip there were fissures and bleeding spots seen, suggestive of angular cheilitis and also depapillation seen on the dorsum of tongue [FIGURE 1, 2]. On hard tissue examination all teeth appeared to be hypoplastic and extreme sensitivity was elicited by the patient even on drinking normal water [FIGURE 3]

**FIGURE 1: DRY AND CRACKED LIPS**



**FIGURE 2: ANGULAR CHEILITIS & DEPAPILLATION OF TONGUE NOTED**



**FIGURE 3: EROSION OF TEETH NOTED SCHIMMER TEST**



**FIGURE 4: MODIFIED**



Considering the present and the past medical history, patient was provisionally diagnosed as secondary Sjogren's syndrome associated with Rheumatoid arthritis. An ultrasound was performed, which revealed both the sub mandibular and parotid glands to be normal in size, heterogenous echo patterns were noted with some cystic spaces within the glands, which were consistent with the changes seen in SS.

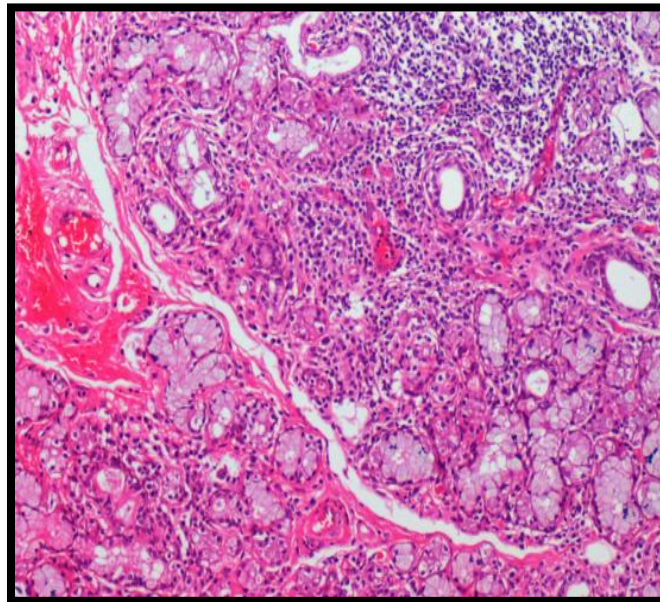
Spitting method and Modified Shimmer's test [FIGURE 4] was performed to test the amount of saliva and the degree of xerostomia. The test revealed at the end of 1min, 2min, and 3 min a collection of 0 ml of saliva noted, which suggested of severe xerostomia. Minor salivary gland biopsy was performed on the lower labial mucosa to confirm the diagnosis as shown in [FIGURE 5a]. Under local anesthesia an incision of around 1.5 to 2 cm was made between midline and the commissure, through the mucosa with penetration of the epithelium. The histopathological report revealed mononuclear infiltration with periductal or perivascular distribution of tissue. Sections were stained by hematoxylin-eosin and considered positive when a focus of 50 lymphocytes/4 mm<sup>2</sup> was found. Histopathological report showing multiple

lymphocytic foci and intact acinar units (original magnification x100) was noted [FIGURE 5b]. The biopsy report confirmed the diagnosis of SS.

**FIGURE 5a: MINOR SALIVAR GLAND BIOPSY**

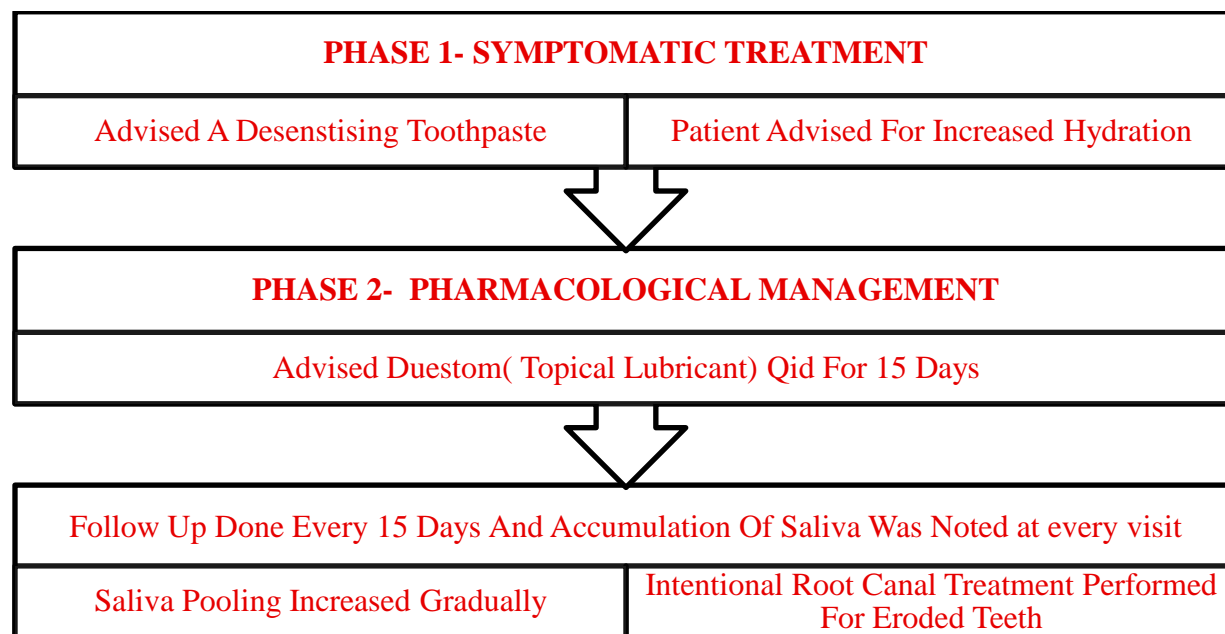


**FIGURE 5b: HISTOPATHOLOGY SLIDE APPEARANCE**



Combining the clinical findings, investigations performed and biopsy reports, a final diagnosis of Secondary Sjögren's syndrome was rendered which was according to the Sjögren's International Collaborative Clinical Alliance (SICCA). The treatment was carried out to relieve the symptoms of the patient in order to lead a comfortable and productive life. The treatment protocol followed is denoted in a flow chart [TABLE 1].





## DISCUSSION

Management of oral diseases with underlying medical conditions requires a thorough knowledge in dentistry, medicine and pharmacology. Several oral mucosal diseases either due to any systemic diseases present or the diseases per se are very commonly encountered by an oral physician. Several individuals visit the dentist more than a general physician for the treatment of their conditions. The oral physician will certainly be the first one in the chain to diagnose any systemic disease which the patient is unaware of. They serve as an essential link between dentistry and medicine and they possess a thorough knowledge of the basic physiological and pathological mechanisms that contribute to the health and disease.

Autoimmune diseases are a group of disorders where, the body's immune system not only fights the infection but also attacks its own tissues<sup>[1]</sup>. SS is a chronic autoimmune disease characterized by lymphocytic infiltration of lacrimal and salivary glands resulting in sicca like symptoms. It has diverse manifestations ranging from mild glandular changes to severe extra glandular disease including vasculitis, neuropathy and glomerulonephritis. The disease is poorly studied and is reported to have a varied prevalence of 0.1 to 4.8% in various populations with an estimated female to male ratio of 9:1<sup>[5]</sup>

Secondary SS has an increased risk of developing non-Hodgkin's lymphoma which is approximately 44 times greater than the risk of the general population. The patients are put to a 1000-fold increased risk of parotid gland marginal zone lymphoma and diffuse large B-cell and follicular lymphomas. The risk of lymphomas is closely related to B-cell hyper reactivity<sup>[1]</sup>.

Xerostomia makes swallowing of food and even the speech difficult for these patients. The sudden development of pain in the mouth is due to angular cheilitis and depapillation of the tongue. Dryness of the mouth cannot simply be attributed to the total destruction of the gland, the

local environment of the inflamed gland leads to dysfunction of the residual glandular units owing to release of cytokines, metalloproteinases, and autoantibodies.<sup>[1]</sup> The patient often have dry cracked lips, sore mouth, depapillation of the tongue, unpleasant taste as seen in the present case. Intra orally, the mucosa is pale and dry, minimal salivary pooling can be noted and the saliva present is thick and ropy.<sup>[6]</sup> Xerostomia predisposes the oral cavity to dental caries, oral mucosal inflammation, erythematous mucosa, and traumatic ulcer.

The pathological hallmark of Sjogren's syndrome is a chronic inflammatory infiltrate in the exocrine glands, mainly constituted by activated T- and B-cells. In the early stages of disease, focal aggregates of lymphocytes appear in the glandular lobules.<sup>[7]</sup> The management of dry mouth aims to prevent infections, periodontal disease and dental caries<sup>[6]</sup> adequate hydration remains the simplest yet the most effective means to treat xerostomia. Frequent sips of water not only rehydrate the oral cavity but also cleanse and reduce microbial load. Salivary stimulants are available in both topical and systemic forms. Oral pilocarpine has also been recommended. Fluoride carriers and re mineralization solutions are necessary for caries control.

## CONCLUSION

Sjogren syndrome is a common and an under diagnosed inflammatory disease with a significant impact on oral health. Sjogren's syndrome features increases with age due to normal aging process or due to systemic illness or medication. Oral physicians can be the first health care providers to encounter this disease at an early stage and can certainly improve patient's quality of life. Therefore, oral physicians should be familiar with the manifestation of the disease and be prepared to take an active role in diagnosis, management and treatment of oral complications which are associated with the disease. A team approach to multisystem disorders is always prudent, as Sjogren's syndrome can be easily overlooked or misinterpreted, and diagnosis can be delayed. The present case report clearly reflects

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