*Received: 07th September 2014 Accepted: 25th December 2014 Conflict of Interest: None*

*Source of Support: Nil*

**Case Report**

# Sjögren's Syndrome: A Case Study

***Santosh Jadhav[1], Abhijeet Jadhav[1], Shameeka Thopte[1], Swati Marathe[2], Prakash Vhathakar[3], Pankaj Chivte[4], Amol Jamkhande[5]***

***Contributors:***

[1]Assistant Professor, Department of Oral Medicine and Radiology, Bharati Vidyapeeth Deemed University Dental College & Hospital, Pune, Maharashtra, India; [2]Professor and Head, Department of Oral Medicine and Radiology, Bharati Vidyapeeth Deemed University Dental College & Hospital, Pune, Maharashtra, India; [3]Assistant Professor, Department of Periodontics, Bharati Vidyapeeth Deemed University Dental College & Hospital, Pune, Maharashtra, India; [4]Associate Professor, Department of Periodontics, Saraswati Dhanwantari Dental College & Hospital, Parbhani, Maharashtra, India; [5]Associate Professor, Department of Public Health Dentistry, Bharati Vidyapeeth Deemed University Dental College & Hospital, Pune, Maharashtra, India.

***Correspondence:***

Dr. Jamkhande A. Department of Public Health Dentistry, Bharati Vidyapeeth Deemed University Dental College & Hospital, Pune, Maharashtra, India. Phone: +91-9823185830. Email: dr.amolj@gmail.com

***How to cite the article:***

Jadhav S, Jadhav A, Thopte S, Marathe S, Vhathakar P, Chivte P, Jamkhande A. Sjögren's syndrome: A case study. J Int Oral Health 2015;7(3):72-74.

***Abstract:***

Sjögren syndrome is chronic, systemic autoimmune disease characterized by lymphocytic infiltration of the exocrine glands. It is an elaborate involvement of the lacrimal and salivary glands, which eventually lead to keratoconjunctivitis sicca and xerostomia. It may occur in two forms - Primary and secondary, which is associated with another autoimmune disease, most commonly rheumatoid arthritis. Numerous criteria were proposed for diagnosis of Sjögren syndrome. Most widely accepted are American and European group developed international classification criteria for Sjögrens syndrome. These criteria include ocular symptoms, oral symptoms, ocular signs, histopathology, salivary gland involvement and sialography. The classification requires four of the six items, one of which must be positive minor salivary gland biopsy or a positive antibody test. Early diagnosis is important to prevent further complications. The aim of this paper is to emphasis on oral changes, advanced diagnosis, and management of Sjögren's syndrome.

***Key Words***: Autoimmune diseases, salivary gland, Sjögren's syndrome

## Introduction

Sjögren syndrome is chronic, systemic autoimmune disease characterized by lymphocytic infiltration of the exocrine glands.[1] Thus, an elaborate involvement of the salivary and lacrimal glands leads eventually to, keratoconjuctivitis sicca and xerostomia.

Swedish ophthalmologist Henrik Sjögren first described Sjögren syndrome in adults. Sjögren syndrome may occur in two forms:

Primary Sjögren syndrome and secondary Sjögren syndrome. The secondary syndrome is associated with another autoimmune disease, usually rheumatoid arthritis.[1]

Numerous criteria were proposed for diagnosis of Sjögren syndrome. Most widely accepted are American and European group developed international classification criteria for Sjögrens syndrome.[2] These criteria include six different criteria:

Ocular symptoms - (minimum one of the following points) - daily, persistent, troublesome dry eyes for not <3 months. Oral symptoms - (minimum one of the following symptoms) - Daily filling of dry mouth for not <3 months, recurrent salivary glands swelling, needs to drink water persistently.

Ocular signs – Schirmer's test, Rose Bengal dye test.[3,4]

Positive histopathology. Salivary gland involvement[5] – Whole salivary flow collection when not stimulated (<1.5 ml in 15 min,) Sialography – Shows the presence of diffuse sialectasia, salivary scintigraphy shows uptake is delayed, reduced concentration of tracer and its delayed excretion. Antibodies to anti-SS-A, anti SS-B antigens are present. The classification requires four of the six criteria, one of which must be positive – biopsy of minor salivary gland or antibody test.

## Case Report

A 55-year-old female patient reported to the Bharati Vidyapeeth Deemed University Dental College and Hospital Pune, Department of Oral Medicine and Radiology on 28/8/11 complaining of:

1. Inability to eat completely due to loss of teeth.
2. Along with that patient also complained of dryness of mouth, since 1 year, and dryness of eyes since 7-8 years.

Extraoral examination showed bilateral parotid gland enlargement present on the right (Figure 1) and left side of the parotid region (Figure 2).

On an inspection – swelling on right and left of the parotid gland, measuring about 2.5 cm × 2 cm in diameter. Surface

texture over the swelling was normal. Swelling extending superior up to the inferior border of the mandible at the angle region. Ear lobe was everted on the right side.

On palpation it was diffuse, firm, non-movable, warm, tender on palpation, overlying surface texture was normal, along with that dryness of eyes, fever was also noted. Bilateral submandibular lymph nodes were palpable.

Intra-oral examination showed upper and lower well-formed edentulous alveolar ridges. Diffuse black pigmentation present on right and left buccal mucosa and palatal mucosa. Depapilation was present on the anterior 2/3rd and lateral border of the tongue (Figure 3). Buccal mucosa was thin and friable. After milking of the left side of the parotid gland pus discharge was coming through parotid duct (Figure 4).

Culture and sensitivity test showed organism isolated *Escherichia coli* grown. Complete hemogram showed increased erythrocyte sedimentation rate. Schirmer test and Rose Bengal dye test was positive. Serum immunoglobulin – SS-A RO positive for Sjögrens syndrome. RA factor was positive for rheumatoid arthritis. Ultrasonography shows bilateral submandibular and parotid gland enlargement with multiple hypoechoic lesions within showing very high vascularity, likely to present systemic disorder like Sjögren's syndrome. Sialography shows diffuse foci of sialectasis (Figure 5). Incisional biopsy of lower lip was not done because the patient was not ready for the biopsy.

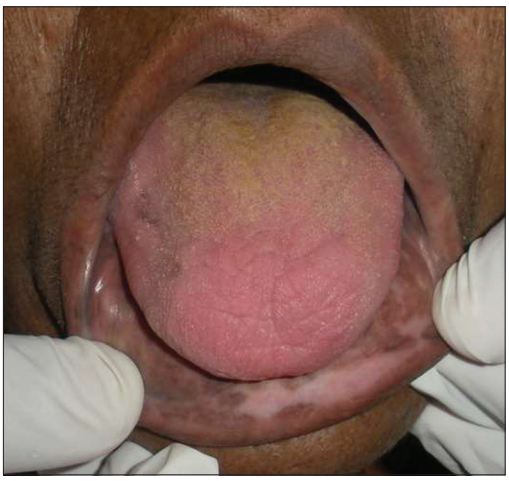

**Figure 3:** Depapillation on lateral border and anterior 2/3rd of tongue.

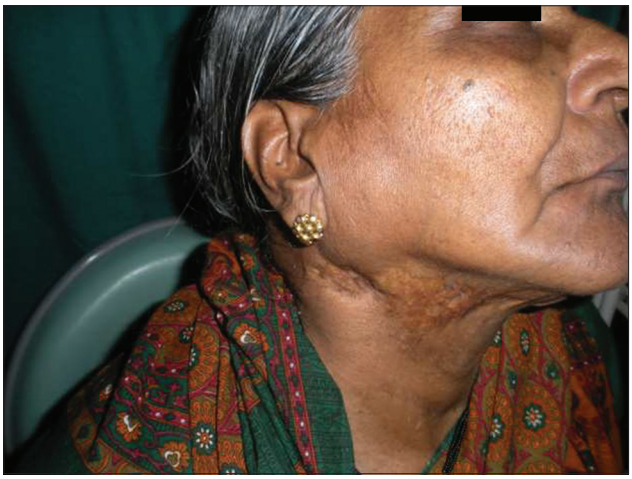

**Figure 1:** Right parotid gland enlargement.

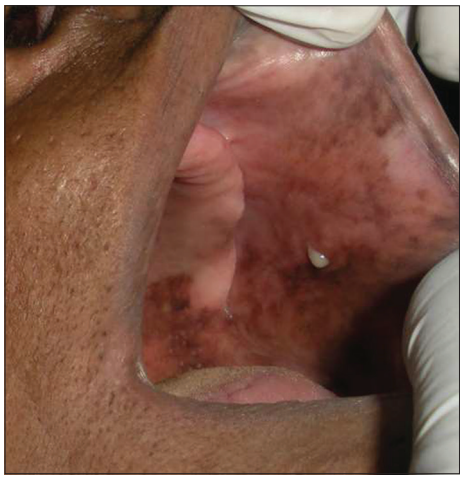

**Figure 4:** Pus discharge through the parotid duct.

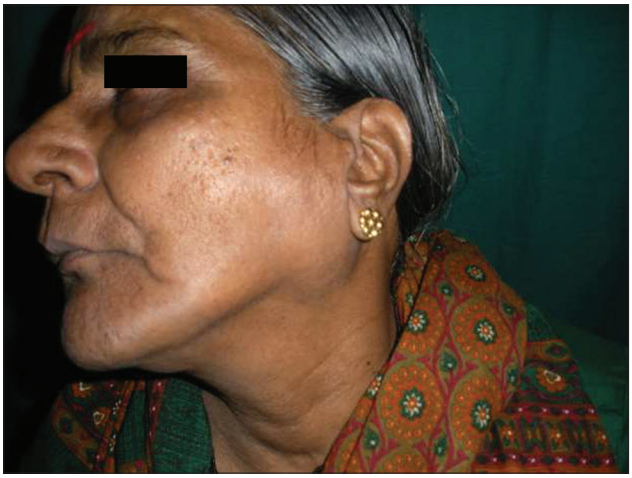

**Figure 2:** Left parotid gland enlargement.

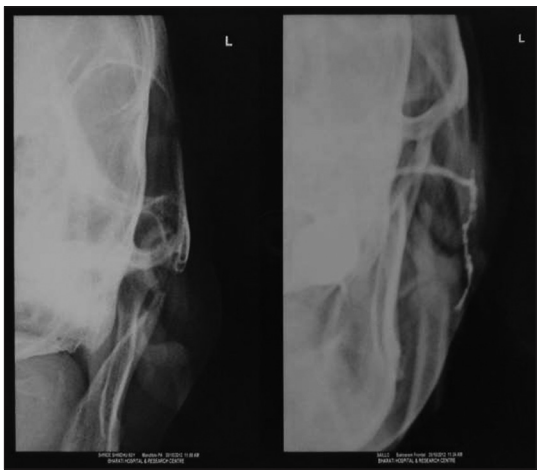

**Figure 5:** Sialography showing diffuse foci of sialectasis.

Based on history, clinical presentation of the patient, and above investigatory findings, a confirmatory diagnosis of primary Sjögren syndrome was given.

**Discussion**

Sjögrens syndrome is a defined as a clinical symptom complex. It is an autoimmune destruction of exocrine glands (primary salivary and lacrimal)that produces the clinical manifestations of dry mouth, dry eyes (keratoconjuctivities sicca), and in more than 50% of cases, parotid gland enlargement. Primary Sjögren syndrome is diagnosed when the syndrome is limited to this pattern of involvement. However, this pattern of involvement may be a manifestation of another -well-defined autoimmune disease such as rheumatoid arthritis, systemic lupus erythematosus, or primary biliary cirrhosis. In this context, it is referred to as secondary Sjögren syndrome.[6,7]

Sjögren syndrome primarily affects women older than 40 years. However, newer diagnostic techniques such as parotid biopsies and antibody identifications have shown it in children and teenagers.[8] The parotid enlargements are usually asymmetric and painless.

Complete physical examination of our patient at presentation and during the follow-up period of three months failed to reveal evidence of any associated connective tissue disease.

Treatment of Sjögren's syndrome depends on the extent and severity of the clinical manifestations and is better instituted through a multidisciplinary approach. Symptomatic treatment includes artificial tears, salivary substitutes to relieve the symptoms and prevent local infectious complications like conjunctivitis and corneal inflammation, development of caries and periodontal disease, a thorough dental preventive program should be implemented in all cases. Corticosteroid treatment should be reserved for all the cases showing evidence of organ damage, significant leukopenia or severe clinical symptoms.

Pathogenesis of Sjögren syndrome is complex and uncertain, but thought to be similar to that of the benign lymphoephtlial lesion. 6-10% of cases undergo transformation to lymphoma, because of infiltration of lymphocytic cell.[9]

The patient reported for the first and second visit shows value added scale (VAS). VAS scale-5 for burning sensation with hot and spicy foods. Pus collected from stensen's duct send for culture and sensitivity. Drainage of pus like the exudate after milking of the left parotid gland. Prescribed medicine was tablet doxycycline 100 mg – on first day BID, followed by OD for 5 days. Tablet paracetamol 500 mg BID for 1 week. Continuous sipping of water throughout the day, orbit VAS scale-4 for burning sensation of hot and spicy foods. After second week, drainage of pus like the exudate through left side Stensons duct after milking of the parotid gland. Tablet augmentin 625 mg – OD for 7 days. Tablet paracetamol 500 mg sos. Continues sipping of water throughout day, orbit chewing gums 3-4 times per day. Aquet spray (lubricating and moisten spray) – 3-4 times per day. Maintenance therapy was continued for next 3 weeks. Capsule menopause was also advised after consultation with a gynecologist from Bharati Hospital for about 1 month.This hormonal replacement therapy shows 10-20% reduction in xerostomia.

**References**

1. Baldini C, Talarico R, Tzioufas AG, Bombardieri S. Classification criteria for Sjögren's syndrome: A critical review. J Autoimmun 2012;39(1-2):9-14.
2. Fujibayashi T, Sugai S, Miyasaka N, Hayashi Y, Tsubota K. Revised Japanese criteria for Sjögren's syndrome (1999): Availability and validity. Mod Rheumatol 2004;14(6):425-34.
3. Cho P, Yap M. Schirmer test. I. A review. Optom Vis Sci 1993;70(2):152-6.
4. Soto-Rojas AE, Kraus A. The oral side of Sjögren syndrome. Diagnosis and treatment. A review. Arch Med Res 2002;33(2):95-106.
5. Stewart CM, Bhattacharyya I, Berg K, Cohen DM, Orlando C, Drew P, *et al*. Labial salivary gland biopsies in Sjögren's syndrome: Still the gold standard? Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2008;106(3):392-402.
6. Schortinghuis J, Pijpe J, Spijkervet FK, Vissink A. Retention of lipiodol after parotid gland sialography. Int J Oral Maxillofac Surg 2009;38(4):346-9.
7. Vitali C, Tavoni A, Simi U, Marchetti G, Vigorito P, d'Ascanio A, *et al*. Parotid sialography and minor salivary gland biopsy in the diagnosis of Sjögren's syndrome. A comparative study of 84 patients. J Rheumatol 1988;(15):262-7.
8. Gomes PD, Juodzbalys G, Fernandes MH, Guobis Z. Diagnostic approaches to Sjögren's syndrome: A literature review and own clinical experience. J Oral Maxillofac Res 2012;3(1):13.
9. Gotoh S, Watanabe Y, Fujibayashi T. Validity of stimulated whole saliva collection as a sialometric evaluation for diagnosing Sjögren's syndrome. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2005;99(3):299-302.