"The Diagnostic Maze of Sjögren's: A Lesson in Early Recognition"

Abstract:

Background: Sjögren's syndrome is a chronic autoimmune disorder primarily affecting the exocrine glands, with oral manifestations often preceding systemic involvement.

Aim: To highlight the importance of early oral signs in the diagnosis of Sjögren's syndrome and the role of dental professionals in identifying systemic autoimmune conditions.

Materials and Methodology: A 30-year-old female presented with dry mouth, burning sensation, and difficulty in swallowing for two years. Clinical and intraoral examinations were performed, followed by serological testing, labial salivary gland biopsy, and skin biopsy.

Results: Examination revealed dry, chapped lips, fissured tongue, multiple carious lesions, and lower limb hyperpigmentation. Serology showed strong positivity for SS-A/Ro, SS-B, and ANA. Labial biopsy confirmed focal lymphocytic sialadenitis, while skin biopsy revealed leukocytoclastic vasculitis, indicating systemic involvement.

Conclusion: This case emphasizes the significance of oral manifestations as early indicators of Sjögren's syndrome and underscores the vital role of interdisciplinary collaboration for timely diagnosis and effective management.

Key-words: sjogrenssyndrome, hyperpigmentation ,candidiasis, vasculitis

Introduction:

Sjögren's syndrome is a systemic autoimmune disorder primarily targeting the exocrine glands, mostly the salivary and lacrimal glands, leading to hallmark symptoms of xerostomia and keratoconjunctivitis sicca. However, SS is a clinically heterogeneous disease, frequently extending beyond glandular involvement to include a wide range of extraglandular manifestations. Among these, cutaneous vasculitis (CV) is a significant and sometimes underrecognized feature, occurring in approximately 5% to 10% of cases and often signaling more severe systemic disease activity.[1]

SS is classified into two main forms: primary SS, which occurs independently, and secondary SS, which develops in conjunction with other autoimmune conditions such as rheumatoid arthritis or systemic lupus erythematosus.[2]

In addition to systemic signs, intraoral and ocular manifestations are often the earliest signs of Sjögren's syndrome. Common oral findings include dry mouth, rampant caries, oral candidiasis, and tongue depapillation, while ocular symptoms involve dryness, photophobia, and a gritty sensation.[3]

Beyond xerostomia and rampant caries lies a broader landscape of oral candidiasis, dysgeusia, and even salivary gland swelling clinical hallmarks that are often overlooked or misattributed. Understanding the diagnostic subtleties from

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positive anti-Ro/SSA serology to the characteristic histopathology of minor salivary gland biopsies—demands an interdisciplinary approach where rheumatology and oral medicine intersect.[4]

This case highlights how a routine dental visit led to the diagnosis of primary Sjögren's syndrome—an often-overlooked autoimmune condition. It underscores the critical role oral healthcare practitioners play as frontline detectors of systemic diseases.

Case Report:

A 30-year-old female reported to the Department of Oral Medicine and Radiology with a chief complaint of pain in the upper left posterior teeth region for the past 15 days.

On general examination, the patient appeared malnourished, though vital signs were within normal limits. Hyperpigmented macular lesions were observed over the bilateral lower legs, below the knees(fig 1B).Extraoral examination revealed dry, chapped lips(fig 1A). Intraoral examination showed bilateral blanching of the buccal mucosa. The dorsum of the tongue presented with scrapable white patches interspersed with erythematous, depapillatedareas(fig 2A), and the patient reported a burning sensation upon intake of spicy foods. Mild erythema was observed on the posterior palate(fig 2B). Dental examination revealed multiple carious lesions and cervical abrasions(fig 2C)

An orthopantomogram (OPG) revealed generalized periodontal ligament widening, likely secondary to extensive caries(fig 3)

Upon detailed questioning, the patient reported persistent dry mouth and dry eyes for over a year. She described sensations of sand or gravel in the eyes and required frequent sips of water while eating dry foods. There was no history of joint stiffness, arthralgia, or Raynaud's phenomenon.

Based on clinical suspicion, a provisional diagnosis of primary Sjögren's syndrome was made. Laboratory investigations were advised:

- **CBC:** Revealed anemia and neutropenia.
- **ANA Profile:** strong positivity for SS-A (Ro60, Ro52) and SS-B antibodies.
- **Lip Biopsy:** Demonstrated focal lymphocytic sialadenitis with two foci per 4 mm^2, consistent with Sjögren's syndrome.

The patient met the 2016 ACR/EULAR classification criteria for pSS with a total score of 6, confirming the diagnosis.

A skin biopsy from the hyperpigmented areas of the legs revealed features consistent with Sjögren's syndrome-associated vasculitis.

The patient was referred to a multidisciplinary team for systemic management. Treatment included:

- **Hydroxychloroquine** 200 mg twice daily
- **Prednisolone** 5 mg once daily

For oral candidiasis involving the dorsum of the tongue, topical antifungal therapy was initiated. The patient was advised to use lubricating mouthwashes thrice daily and encouraged to maintain hydration through frequent water intake. Dental referrals were made to the Department of Endodontics for restoration of carious lesions and management of cervical abrasions. An ophthalmology consultation was arranged for evaluation and treatment of dry eye symptoms.

Discussion:

Primary Sjögren's syndrome (pSS) is a chronic autoimmune disorder that can impact multiple organ systems. While its hallmark features include dry mouth and dry eyes resulting from impaired salivary and lacrimal gland function, about one-third to half of affected individuals also experience systemic involvement.[5]

SS significantly elevates the risk of dental caries due to reduction in salivary flow and changes in saliva quality. Saliva is vital for maintaining oral health through its roles in acid neutralization, mechanical cleansing, and antimicrobial action. When saliva production is compromised, the oral environment becomes more acidic and conducive to bacterial overgrowth. As a result, caries in SS patients often appears on tooth surfaces that are generally less vulnerable in individuals with normal salivary secretion—such as the smooth surfaces, particularly cervical regions of anterior teeth, and the incisal edges.

Patients with SS exhibited a higher prevalence of oral lesions (OL) compared to individuals without the condition. Among these, angular cheilitis was the most commonly reported lesion, followed by atrophic glossitis, candidal infections, oral ulcerations, and fissures or grooves on the tongue. The latter two are especially indicative of the oral dryness commonly associated with SS.[6]

Although SS affects around 2% of the adult population, more than half of the cases remain undiagnosed. A wide range of symptoms may appear before the classic signs of ocular and oral dryness, including skin manifestations—among which vasculitis is relatively common. The prevalence of CVin SS is estimated at 5% to 10% and is often linked with high levels of rheumatoid factor and hallmark autoantibodies such as anti-Ro and anti-La. Clinically, CV may present as a single episode or follow a relapsing pattern. The most frequently observed skin lesions are palpable and/or non-palpable purpura, occurring in about 88% of cases, typically affecting the lower limbs.Histopathologic evaluation of vasculitic lesions reveals leukocytoclastic vasculitis (LCV) in approximately 90% of cases, primarily affecting the small vessels of the skin.[7] Although vasculitis is recognized in some patients with primary Sjögren's syndrome, detailed histopathologic characterization and clinicopathologic correlation of these vasculitic lesions have not been thoroughly explored—unlike in patients with SLE and RA, where such analyses are more extensively documented.[8]

Conclusion:

Primary Sjögren's syndrome may present subtly with oral and ocular symptoms. Clinicians should maintain a high index of suspicion in patients with chronic xerostomia, mucosal changes, and systemic complaints. A multidisciplinary approach is essential for early diagnosis, effective management, and improving patient quality of life.

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Figure Legend

FIGURE 1A : Extraoral photograph showing dry, chapped lips

FIGURE 1B: Clinical photograph showing hyperpigmented macular lesions on the bilateral lower legs

FIGURE 2A : Intraoral photograph showing the dorsal surface of the tongue with white scrapable plaques interspersed with erythematous, depapillated regions.

FIGURE 2B : Clinical photograph depicting subtle erythematous changes on the posterior palatal mucosa.

FIGURE 2C: Clinical photograph depicting extensive dental caries involving cervical regions, along with cervical abrasions.

FIGURE 3 : OPG demonstrating generalized PDL widening, possibly related to advanced carious lesions.