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Case Report

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CNS Manifestations of Sjogren's Syndrome

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Introduction

Primary Sjögren's Syndrome is a systemic chronic autoimmune disease with a female-to-male predominance of 9:1 and a peak incidence at approximately 50 years of age [1,2]. It is considered to be one of the most common autoimmune diseases with a prevalence thought to be equal or exceed that of rheumatoid arthritis [3]. Sjören's Syndrome often goes undiagnosed for many patients suffering the conditions of exocrinophaties affecting tear and and saliva production but can also have central nervous system (CNS) manifestations, which can present at a psychiatric disorder most commonly as an atypical mood disorder [1,4]. Some investigators have recently documented higher rates of CNS involvement in primary Sjögren's Syndrome (CNS-SS) than previously noted. These CNS complications include focal neurological deficits, diffuse cerebral involvement, and psychiatric disorders [3]. Quoted from a specific reference, "Cox and Hales have suggested that the incidence of mild to moderate psychiatric and/or cognitive impairment may be as high as 80% in patients with CNS-SS" [3]. What's important about the presentation of Primary Sjögren's Syndrome is the early manifestations of the disease, including the development and early manifestations of mental disorders as a response of the autoimmune process, which presumes the direct immunological activity of Sjögren's Syndrome on the central nervous system (by T cells, autoantibodies, cytokines, or apoptosis) [3]. 40-50% of patients suffering from this disease do not show detection of Sjögren's in their bloodwork, which can leave them going undiagnosed for decades. The triad of symptoms include dryness of the mouth and eyes, fatigue and pain, but the first signs of the disease include presentations of oral dryness [1], in most cases. The incidence of Sjögren's Syndrome is being seen in younger individuals as the awareness of this disease continues to rise and an increasing awareness of autoimmune diseases presenting with neuropsychiatric manifestations [1].

Aim

The aim of this case report is to bring awareness of Sjören's Syndrome among dentists. Dental providers are usually the first ones to recognize the symptoms of patients suffering from this disease. Early signs and symptoms include oral dryness, which can result in dental cavities, oral fungal infections, as well as burning mouth and lips. Increasing awareness among dental providers brings sufficient action and direction into getting diagnosis of a disease state that many patients can suffer with for years.

Challenges in Diagnosis

A 30-year-old Caucasian female, who is a self-referral, requests an appointment for evaluation of recurrent decay and parafunctional habit. She stated that she gets stressed over small things and probably that is the reason for parafunctional habit. The patient is not satisfied with her current treatment and she also stated that many times she has been suggested to consider psychiatric medication for anxiety disorder.

The patient's medical history was positive for Endometriosis, Poly Cystic Ovarian Syndrome, Metabolic Syndrome, ADHD, Generalized Anxiety Disorder, Menorrhagia, Over Weight and Hashimoto's Thyroiditis. The medications that she was on are Vyvanse 50 mg daily, Victoza 1.8 mg daily and Synthroid 200 mcg daily.

Her review of systems was positive for emotional instability, depression, paranoia, irritability, brain fog, dry mouth, dry eyes, lethargy, aphasia, tingling in her hands and feet, Raynaud's, random blood pressure spikes, increase in facial hair, loss of sex drive, body aches, painful sex, and dry skin. The patient denied having any allergies.

An intra-oral examination showed good oral hygiene, but the patient has a number of restored teeth. She also has oral dryness with a lack of pooling of saliva in the anterior floor of her mouth. Parafunctional habit was confirmed with the presence of morsicatio buccarum bilaterally on the buccal mucosa and on her lower lip (Figure 1-4).

Saliva measurements were taken and she was low on both unstimulated and stimulated saliva levels. Her fungal swabs came back negative. After discussion with the patient's physician, it was mutually agreed upon to remove Vyvanse from her list of medications. The patient's symptoms improved, but the improvement was limited. The patient's physician was contacted again and it was agreed to decrease her Synthroid medication gradually from 200mcg daily to 150 mcg daily. This reduction in Synthroid was very helpful in reducing a number of her symptoms, which were related to her anxiety but did not take care of all her symptoms. The patient's blood work was ordered for ANA,

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RA, Anti SSA and Anti SSB. Her ANA was positive, but the rest of the test results were negative. Anti SSA and Anti SSB can be negative in up to 40% of the patients with Sjogren's Syndrome. This was discussed with the patient and it was agreed to proceed with minor salivary gland biopsy to confirm the clinical diagnosis. Her biopsy result came back positive for Primary Sjogren's Syndrome (Figure 5-8).

Conclusion

Patient was having these symptoms without getting a diagnosis for around 12 years. In most cases, patients with Sjogren's Syndrome do not get a diagnosis for at least five years after developing initial symptoms of the disease. In most cases patients get diagnosis of conditions like fibromyalgia are told that the symptoms could be due to hormonal changes. Apart from dry mouth and dry eyes, neuropsychiatric manifestations may arise and present as an early sign of Sjogren's Syndrome, which is supported by evidence within the literature. In the case of this patient, it is a possibility that the symptoms of anxiety and ADHD could be neuropsychiatric manifestations of Sjogren's Syndrome. There is need for more research to establish the link between Sjogren's Syndrome and the neuropsychiatric conditions.

We are seeing more cases among younger populations, either related to the disease changing its natural history or clinicians becoming increasingly aware of the disease, enough to diagnose the condition early. Early diagnosis will save the patients' time and healthcare costs by reducing unnecessary referrals and tests, including salvaging the impact it would have on the patients' quality of life. In doing so, the proper treatment and management would be administered to control their symptoms early on. It is of significant importance for dentists to be aware of the presentations of this disease as they may be the first ones

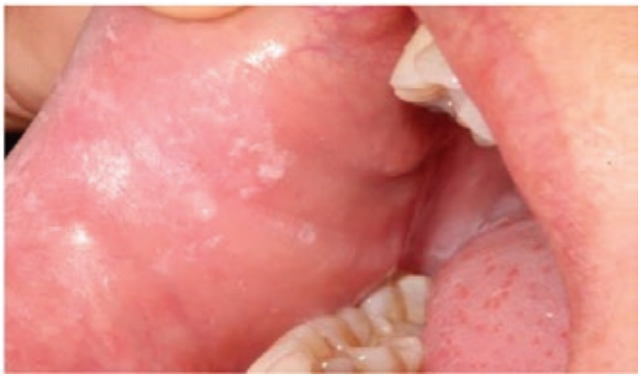

Figure 1: Morsicatio Buccarum on the right buccal mucosa

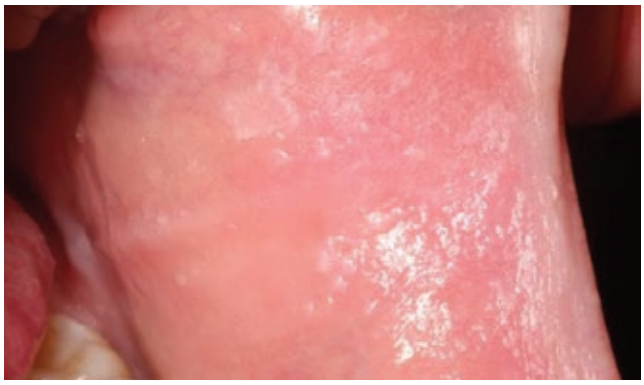

Figure 2: Morsicatio Buccarum on the left buccal mucosa

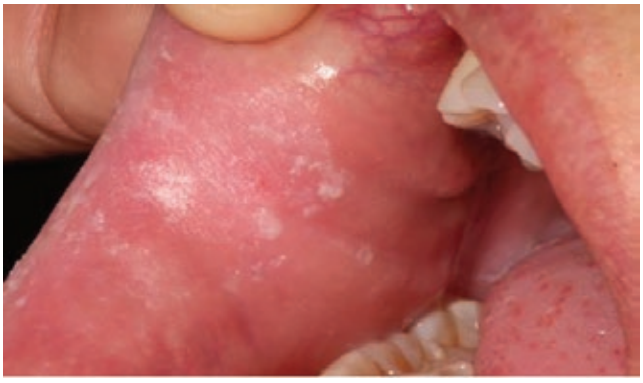

Figure 3: Morsicatio Buccarum on the right buccal mucosa

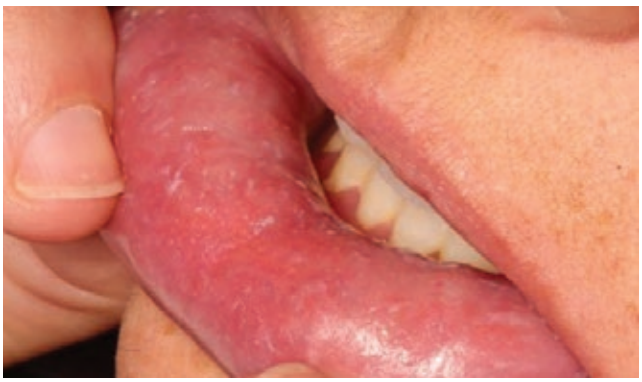

Figure 4: Morsicatio Buccarum on the lower lip

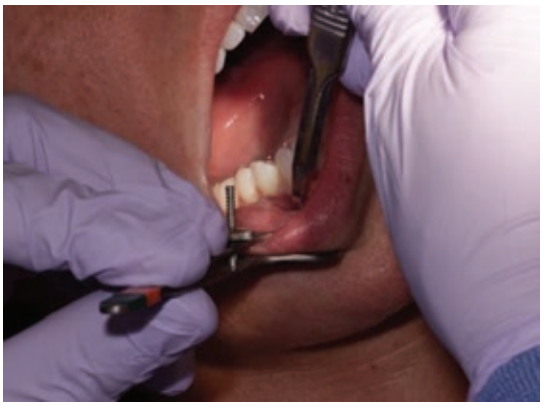

Figure 5: Incision being made on the right lower lip

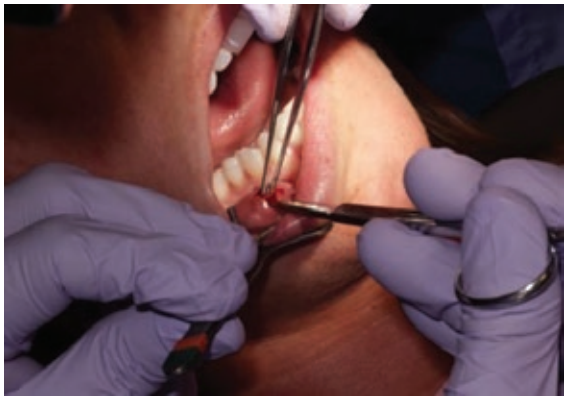

Figure 6: Insicion site being expanded

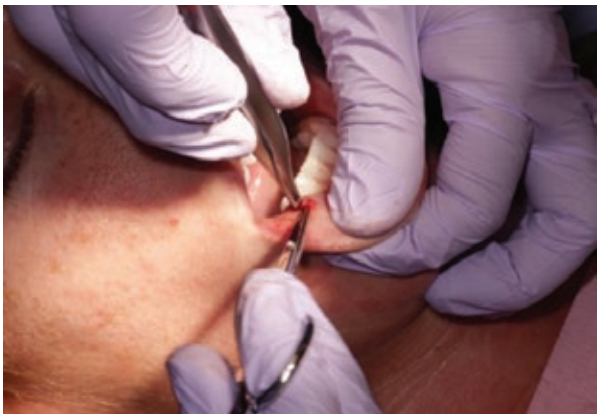

Figure 7: Harvesting the minor salivary glands

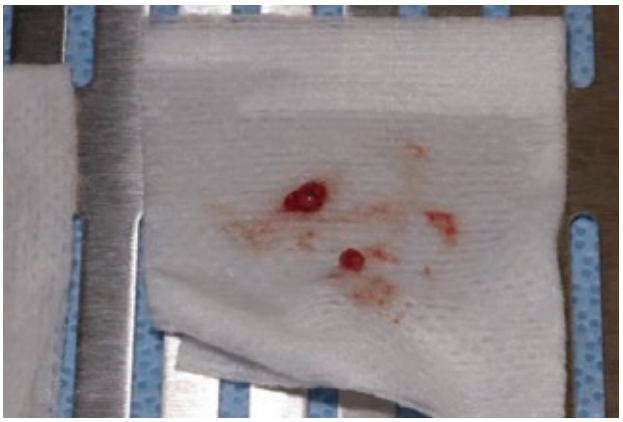

Figure 8: Harvested minor salivary glands

Table 1: 2016 American College of Rheumatology/European League Against Rheumatism Classification Criteria for Primary Sjögren's Syndrome.

	New diagnostic criteria	Score
1	Anti-SSA/Anti SSB antibody positivity	3
2	Focal lymphocytic sialadenitis with a focus score of ≥ 1 foci/4 mm2	3
3	Abnormal ocular staining score of ≥ 5	1
4	Schimer's test result of ≤ 5 mm/5 minutes	1
5	Unstimulated salivary flow rate of ≤ 0.1 ml/minute	1
	Total score of ≥4 for the above items meet the criteria for primary SS	

(The current 2016 American/European diagnostic criteria table for Sjogren's Syndrome shown above)

to come across these symptoms and help with the diagnosis process in a timely and accurate manner by recognition of the disease presentations and referral for further evaluation.

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