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SJÖGREN'S SYNDROME: PATHOGENESIS, ORAL MANIFESTATIONS, AND DENTAL MANAGEMENT

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ABSTRACT

Methodology: This study is a narrative literature review. A comprehensive search of multiple online databases was conducted to identify relevant literature. The databases used included EMBASE, PubMed, the Cochrane Library, and Google Scholar. Keywords such as "Sjögren's Syndrome," "Pathology," and "Dentistry" were utilized, in combination with Boolean operators "AND" and "OR" to refine the search. The selection process was conducted in three phases: (1) all retrieved records were exported to the Zotero reference manager to eliminate duplicates; (2) titles and abstracts were screened to exclude unrelated studies; (3) full-text reading was performed to assess the final eligibility of the articles based on relevance to the review objective.

Results: The selected studies highlighted the complex etiology of Sjögren's Syndrome, which involves autoimmune mechanisms leading to lymphocytic infiltration of the exocrine glands, particularly the salivary and lacrimal glands. Common oral manifestations include xerostomia (dry mouth), dental caries, mucosal inflammation, candidiasis, and difficulties in mastication and speech. The reviewed literature also discussed diagnostic challenges and emphasized the importance of interdisciplinary care. Therapeutic options ranged from symptom management with saliva substitutes and systemic immunomodulatory agents to interventional therapies such as sialendoscopy and newer biologic drugs under investigation.

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Conclusion: Sjögren's Syndrome is a chronic autoimmune disease with significant oral health implications. Early diagnosis and a multidisciplinary approach are essential to manage its systemic and oral manifestations effectively. Although current treatments focus primarily on symptom relief, ongoing research into immunomodulatory and biologic therapies holds promise for more targeted and effective management in the future.

Keywords: Sjögren's Syndrome. Pathology. Dentistry.

SÍNDROME DE SJÖGREN: ETIOLOGIA, MANIFESTAÇÕES ORAIS E ABORDAGENS ODONTOLÓGICAS

RESUMO

Metodologia: Este estudo é uma revisão narrativa da literatura. Uma busca abrangente em múltiplas bases de dados online foi realizada para identificar literatura relevante. As bases de dados utilizadas incluíram EMBASE, PubMed, Biblioteca Cochrane e Google Acadêmico. Palavras-chave como "Síndrome de Sjögren", "Patologia" e "Odontologia" foram utilizadas, em combinação com os operadores booleanos "AND" e "OR" para refinar a busca. O processo de seleção foi conduzido em três fases: (1) todos os registros recuperados foram exportados para o gerenciador de referências Zotero para eliminar duplicatas; (2) títulos e resumos foram rastreados para excluir estudos não relacionados; (3) a leitura do texto completo foi realizada para avaliar a elegibilidade final dos artigos com base na relevância para o objetivo da revisão.

Resultados: Os estudos selecionados destacaram a etiologia complexa da Síndrome de Sjögren, que envolve mecanismos autoimunes que levam à infiltração linfocítica das glândulas exócrinas, particularmente as glândulas salivares e lacrimais. Manifestações orais comuns incluem xerostomia (boca seca), cárie dentária, inflamação da mucosa, candidíase e dificuldades na mastigação e na fala. A literatura revisada também discutiu os desafios diagnósticos e enfatizou a importância do cuidado interdisciplinar. As opções terapêuticas variaram do manejo dos sintomas com substitutos da saliva e agentes imunomoduladores sistêmicos a terapias intervencionistas, como a sialoendoscopia e novos medicamentos biológicos em investigação.

Conclusão: A Síndrome de Sjögren é uma doença autoimune crônica com implicações significativas para a saúde bucal. O diagnóstico precoce e uma abordagem multidisciplinar são essenciais para o manejo eficaz de suas manifestações sistêmicas e orais. Embora os tratamentos atuais se concentram principalmente no alívio dos sintomas, a pesquisa em andamento sobre terapias imunomoduladoras e biológicas promete um manejo mais direcionado e eficaz no futuro.

Palavras-chave: Síndrome de Sjögren. Patologia. Odontologia.

SÍNDROME DE SJÖGREN: PATOGENIA, MANIFESTACIONES ORALES Y TRATAMIENTO DENTAL

RESUMEN

Metodología: Este estudio es una revisión narrativa de la literatura. Se realizó una búsqueda exhaustiva en múltiples bases de datos en línea para identificar la literatura relevante. Las bases de datos utilizadas incluyeron EMBASE, PubMed, la Biblioteca Cochrane y Google

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Académico. Se emplearon palabras clave como "Síndrome de Sjögren", "Patología" y "Odontología", en combinación con los operadores booleanos "AND" y "OR" para refinar la búsqueda. El proceso de selección se llevó a cabo en tres fases: (1) todos los registros recuperados se exportaron al gestor de referencias Zotero para eliminar duplicados; (2) se examinaron los títulos y resúmenes para excluir estudios no relacionados; (3) se realizó una lectura completa del texto para evaluar la elegibilidad final de los artículos según su relevancia para el objetivo de la revisión.

Resultados: Los estudios seleccionados destacaron la compleja etiología del síndrome de Sjögren, que implica mecanismos autoinmunes que conducen a la infiltración linfocítica de las glándulas exocrinas, en particular las salivales y lagrimales. Las manifestaciones orales comunes incluyen xerostomía (boca seca), caries dental, inflamación de la mucosa, candidiasis y dificultades para masticar y hablar. La literatura revisada también analizó los desafíos diagnósticos y enfatizó la importancia de la atención interdisciplinaria. Las opciones terapéuticas abarcaron desde el manejo de los síntomas con sustitutos de saliva y agentes inmunomoduladores sistémicos hasta terapias intervencionistas como la sialoendoscopia y nuevos fármacos biológicos en investigación.

Conclusión: El síndrome de Sjögren es una enfermedad autoinmune crónica con importantes implicaciones para la salud bucodental. El diagnóstico temprano y un enfoque multidisciplinario son esenciales para el manejo eficaz de sus manifestaciones sistémicas y orales. Si bien los tratamientos actuales se centran principalmente en el alivio de los síntomas, la investigación en curso sobre terapias inmunomoduladoras y biológicas promete un manejo más específico y eficaz en el futuro.

Palabras clave: Síndrome de Sjögren. Patología. Odontología.

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1 INTRODUCTION

Sjögren's Syndrome (SS) is a systemic, chronic autoimmune disorder primarily characterized by lymphocytic infiltration of exocrine glands, especially the salivary and lacrimal glands, resulting in xerostomia and keratoconjunctivitis sicca. First described by Henrik Sjögren in 1933, the disease affects approximately 0.1–0.6% of the population, with a strong female predominance, particularly among middle-aged individuals (Fox, 2005; Mariette & Criswell, 2018). SS may present as a primary condition or in association with other autoimmune diseases such as rheumatoid arthritis or systemic lupus erythematosus (Brito-Zerón et al., 2016). In addition to glandular dysfunction, systemic involvement including fatigue, arthritis, pulmonary involvement, and increased lymphoma risk adds complexity to its diagnosis and management (Mariette & Criswell, 2018).

Oral health professionals play a vital role in the early recognition and management of SS, as oral manifestations are often among the earliest and most debilitating symptoms. Xerostomia significantly compromises quality of life and predisposes patients to dental caries, mucosal inflammation, oral candidiasis, dysphagia, and difficulties in speech and mastication (Villa et al., 2015). The diagnosis of SS relies on a comprehensive evaluation, including clinical examination, serologic testing, minor salivary gland biopsy, and imaging techniques, guided by the 2016 classification criteria proposed by the American College of Rheumatology and the European League Against Rheumatism (Shiboski et al., 2017).

Although there is no cure for SS, various therapeutic strategies are employed to manage its symptoms and improve patient outcomes. These include saliva substitutes, systemic immunomodulatory agents, sialagogues, and newer biologic therapies under investigation (Baer et al., 2020). In dentistry, treatment approaches are evolving to include interventional options such as sialendoscopy and low-level laser therapy, which aim to restore glandular function and enhance patient comfort. A multidisciplinary approach that incorporates both dental and medical expertise is essential for optimal management and long-term care (Fox, 2005; Baer et al., 2020).

2 METHODOLOGY

This study is a narrative literature review; therefore, a wide range of online database searches were necessary to obtain articles that could be included in this review. Thus, searches were conducted in the following databases: EMBASE, PubMed, and the Cochrane Library, in conjunction with Google Scholar. Furthermore, keywords such as "Sjögren's

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Syndrome," "Pathology," and "Dentistry" were used to obtain only results related to the article's topic. Boolean terms such as "AND" and "OR" were also used in the search strategy. The search results underwent three stages to select only articles related to the study topic: The first stage was to export the database results to the Zotero platform to remove duplicate articles. The second stage was to remove studies unrelated to the article's topic by reading their title and abstract. The third and final step was to read the articles in full, removing those that were not relevant to the review, leaving only those relevant to the study. Furthermore, Rother's (2007) study was used to guide the construction of this article, as it differentiates between a systematic review and a narrative review, demonstrating its structure, approach, composition, and characteristics.

3 RESULTS

3.1 HISTORICAL EVOLUTION OF SJÖGREN'S SYNDROME

Sjögren's Syndrome (SS) was first comprehensively described in 1933 by the Swedish ophthalmologist Henrik Sjögren, who observed a unique constellation of symptoms: keratoconjunctivitis sicca, xerostomia, and arthritis, predominantly in middle-aged women (Fox, 2005). Initially interpreted as a localized sicca syndrome, it was later redefined as a systemic autoimmune disorder, particularly after the identification of characteristic autoantibodies and extraglandular manifestations. Over the decades, advances in immunology, molecular biology, and diagnostic imaging technologies facilitated a more nuanced understanding of the disease's multifaceted pathogenesis (Mariette & Criswell, 2018).

The diagnostic evolution culminated in the establishment of the 2016 ACR-EULAR classification criteria, which integrated serological, histopathological, and clinical parameters to improve diagnostic accuracy (Shiboski et al., 2017). These criteria have been instrumental in delineating primary SS from secondary forms that coexist with other autoimmune conditions, such as systemic lupus erythematosus (SLE) or rheumatoid arthritis (RA), underscoring the disease's polymorphic nature (Brito-Zerón et al., 2016). This historical trajectory reflects a broader shift in the conceptualization of autoimmune diseases, from isolated glandular pathologies to systemic immune dysregulations, paving the way for targeted immunotherapies and individualized treatment approaches.

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3.2 PATHOPHYSIOLOGY AND SYSTEMIC IMPLICATIONS OF SJÖGREN'S SYNDROME

Sjögren's Syndrome is characterized by chronic autoimmune-mediated destruction of exocrine glands, especially the salivary and lacrimal glands. The histological hallmark of SS is focal lymphocytic sialadenitis, predominantly involving CD4+ T cells and B lymphocytes infiltrating the glandular epithelium (Mariette & Criswell, 2018). The presence of specific autoantibodies, anti-Ro/SSA and anti-La/SSB, is not only diagnostic but also prognostic, correlating with disease severity and systemic involvement (Shiboski et al., 2017).

The systemic nature of SS extends beyond exocrinopathy. Fatigue, arthralgia, interstitial lung disease, renal tubular acidosis, neurological deficits, and a markedly increased risk of non-Hodgkin B-cell lymphoma are frequently reported (Brito-Zerón et al., 2016). Indeed, lymphoma is the most feared complication, often heralded by persistent glandular swelling, cryoglobulinemia, or monoclonal gammopathy (Baer et al., 2020). From a pathophysiological standpoint, SS involves a complex interplay of genetic predisposition, epigenetic modifications, environmental triggers, and dysregulated immune responses. Interferon signatures, BAFF (B-cell activating factor) overexpression, and aberrant apoptosis of epithelial cells further exacerbate glandular dysfunction and systemic inflammation (Fox, 2005).

Despite these insights, the etiological underpinnings remain incompletely understood, with ongoing studies aiming to unravel the molecular mechanisms that link mucosal autoimmunity to systemic disease manifestations.

3.3 DENTAL IMPLICATIONS OF SJÖGREN'S SYNDROME

The oral cavity is often the initial site where the debilitating consequences of Sjögren's Syndrome become clinically evident, placing dental professionals at the forefront of early disease recognition (Villa et al., 2015). Xerostomia, caused by salivary gland hypofunction, impairs essential oral functions such as mastication, speech, and swallowing. It also creates a permissive environment for opportunistic infections, accelerated dental caries, mucosal atrophy, and periodontal disease (Fox, 2005).

From a clinical management perspective, dental care for SS patients demands a preventive, symptomatic, and interventional approach. Preventive strategies include enhanced oral hygiene protocols, fluoride application, and patient education. Symptomatic relief often involves the use of saliva substitutes, sialagogues, and low-level laser therapy.

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Recently, minimally invasive techniques such as sialendoscopy have shown promise in improving glandular drainage and reducing obstructive symptoms (Baer et al., 2020).

Furthermore, SS patients require lifelong dental monitoring and interdisciplinary coordination with rheumatologists, immunologists, and ophthalmologists. The psychological burden of chronic oral discomfort and functional impairment also necessitates psychosocial support, reinforcing the need for holistic, patient-centered care.

The integration of dental professionals into the multidisciplinary care model not only facilitates early diagnosis but also mitigates disease progression and enhances quality of life making the role of dentistry indispensable in the management of Sjögren's Syndrome.

4 DISCUSSION

Sjögren's Syndrome represents a significant clinical challenge due to its multifactorial nature and the wide range of both local and systemic manifestations. The primary involvement of exocrine glands compromises essential functions such as saliva and tear production, directly affecting oral health and the patient's quality of life.

In dental practice, initial symptoms like xerostomia and oral discomfort often lead patients to seek dental care before receiving a formal diagnosis, highlighting the crucial role of the dentist in early identification of the disease. Timely detection not only helps alleviate debilitating symptoms but also enables early intervention that can prevent more serious complications.

Clinical management requires a multidisciplinary approach, involving dentistry, rheumatology, ophthalmology, and other specialties depending on the systemic involvement of the patient. Current treatment remains largely focused on symptom relief, ranging from local strategies, such as the use of artificial saliva to more complex therapies involving immunomodulatory agents. In dentistry, new therapeutic approaches are being explored, aiming to restore glandular function and enhance the patient's comfort during treatment.

Additionally, ongoing and individualized follow-up is essential, as the progression of the disease can alter the clinical picture over time. A solid understanding of the pathogenesis, combined with thorough clinical assessment, allows professionals to develop more effective and patient-centered treatment plans. Patient education and the maintenance of good oral hygiene are also fundamental pillars for managing oral symptoms and preventing secondary complications.

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5 CONCLUSION

Sjögren's Syndrome is a chronic autoimmune disease with significant oral health implications. Early diagnosis and a multidisciplinary approach are essential to manage its systemic and oral manifestations effectively. Although current treatments focus primarily on symptom relief, ongoing research into immunomodulatory and biologic therapies holds promise for more targeted and effective management in the future.

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