REVIEW

ENT manifestations of Sjogren's syndrome: A narrative review

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Abstract

Sjogren's syndrome (SS) is an autoimmune condition marked by dryness of the mouth and eyes, frequently manifesting as a range of ENT symptoms that have a major negative impact on the quality of life of patients. Dryness and discomfort in the mouth and eyes, brought on by lacrimal and salivary gland dysfunction are ENT symptoms in SS. However, ENT regions are involved in more areas than just the ocular surface and mouth cavity. Due to laryngopharyngeal involvement, patients may experience hoarseness, dysphagia, and the sensation that something foreign is in their throat. Common nasal symptoms include nasal dryness, congestion, and recurrent sinusitis, while Eustachian tube dysfunction can lead to middle ear infections and hearing problems. To improve the quality of life in SS patients, clinicians and healthcare professionals should be careful in identifying and treating these symptoms. More investigations and clinical trials are required to create tailored medicines that can reduce ENT symptoms and enhance long-term outcomes for SS patients.

Keywords: Sjogren's syndrome, ENT manifestations, Autoimmune disease, Systemic lupus erythematosus, Systemic sclerosis

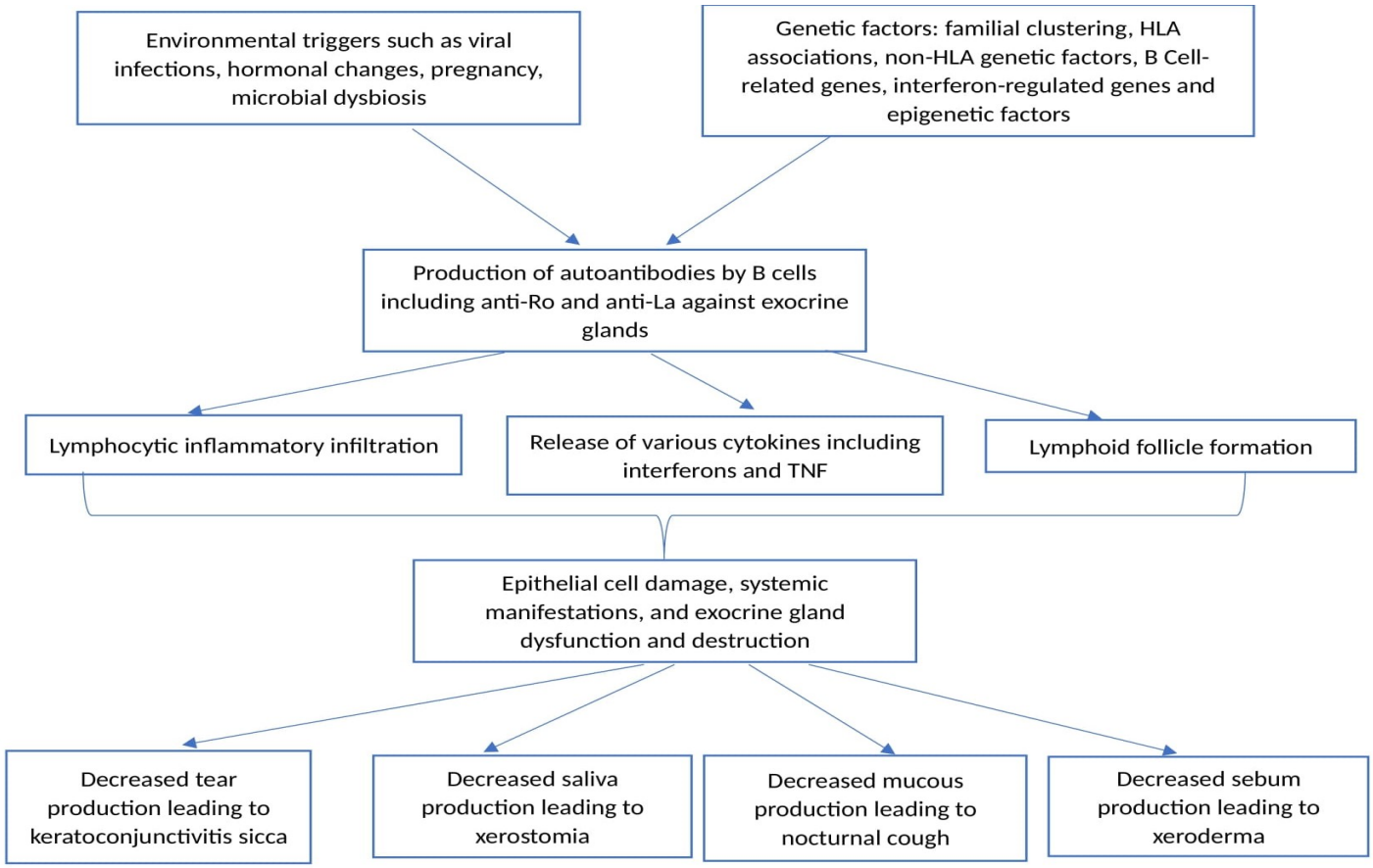
Introduction

Sjogren's syndrome (SS) is a common chronic autoimmune disease characterized by lymphocyte infiltration and inflammation in the exocrine glands, especially in salivary and lacrimal glands, resulting in secretory gland dysfunction. It primarily affects the mucosal surface of the oral cavity, eyes, nose, pharynx, throat, and vagina. SS is broadly categorized into two main subtypes: primary SS, which occurs independently as a standalone condition, and secondary SS, which is associated with other autoimmune diseases, such as rheumatoid arthritis, systemic lupus erythematosus, and systemic sclerosis. It may be more accurate to describe SS as 'overlapping' with other rheumatic diseases. Although sicca symptoms (dry mouth and dry eyes) are among the most common presentations in general medicine practice, SS is often underestimated or misdiagnosed.

The pathophysiology of SS is considered to be multifactorial, involving the interaction between genetic factors and exogenous and endogenous factors that can trigger abnormal autoimmune responses, particularly mediated by T and B lymphocytes. Inflammation increases tissue damage, leading to functional damage of affected organs and a chronic inflammatory environment. There are three recurrent events generally associated with SS: (1) a triggering period induced by environmental factors under specific epigenetic factors, genetic susceptibility, and hormone regulation; (2) dysfunction of normal salivary gland epithelial cells; and (3) chronic inflammation characterized by hyperactivity of lymphocyte B and autoantibodies.

The pathogenesis of SS remains clear with the involvement of multiple factors such as genetic defects, immune mechanisms, and virus infection. Genetic susceptibility to SS plays a role in disease onset. A strong correlation between human leukocyte antigen-DR isotype (HLA-DR) and HLA-DQ alleles (belonging to the major histocompatibility complex [MHC] class II genome) and SS has been observed in different populations, including Caucasians, Japanese, and Chinese. Moreover, some research has analyzed the contribution of epigenetics to SS and autoantibody production (Fig. 1). The epigenetic processes, more closely related to the disease, are DNA methylation, as well as functions of circular mRNA, miRNA, and long non-coding RNA.

Fig. 1: Pathogenesis and clinical findings in SS



Pathophysiology and clinical features of otorhinolaryngological manifestations

SS has varied otolaryngologic manifestations with an estimated global prevalence of 1%-3%.[1] It is known to be the second most common rheumatic autoimmune disorder. The severity of SS increases with age, and it is more prevalent in the female population. The symptoms commonly include glandular, ocular, oral, otologic, pharyngeal, laryngeal, and sinonasal manifestations.

1. Ear involvement

SS affects all the anatomical structures of the ear (internal, external, and middle). Although the pathophysiological effects of primary SS on the human auditory system have not been thoroughly investigated, dryness of the ear canal epidermis, middle and inner ear fluids, and Eustachian tube dysfunction are considered potential causes of auditory symptoms.[2,3] The molecular structure of the perilymph and endolymph fluids can be disrupted by variations in the fluid volume, potentially resulting in hearing loss.

Ototoxic drugs used in the management of autoimmune inner ear diseases alter the molecular concentrations affecting physiological mechanisms. In addition, autoantibodies are produced inadequately in perilymph and endolymph fluids.[2,3] In some patients with SS, pathological changes have been noted in the stria vascularis of the inner ear, and spiral ganglia neurons may also be affected.[4-5]

Symptoms include otalgia, hearing loss, tinnitus, vertigo, dry skin of the ear canal, and dysfunction of the Eustachian tube.[1] Sensorineural hearing loss (SNHL) in primary SS has been noted in many patients. Although clinically substantial abnormalities are uncommon, SNHL that primarily affects the high frequencies is related to primary SS. Retrocochlear disease and greater vestibular involvement are not present in primary SS, and SNHL was linked only to the disease duration.[6,7] The SNHL was found to be the first manifestation of SS in a few patients.[8] When compared to patients without autoimmune illnesses, the probability of sudden SNHL was significantly higher in those with SS.[9]

Patients with SS often experience difficulties in perceiving high frequencies. The frequencies between 9000 and 20,000 Hz are referred to as extended high frequencies, as the human hearing range is up to 20,000 Hz.[10]

Before considering the influence of the medium and low frequencies, which significantly influence hearing capacity, extended high-frequency audiometry (i.e., 10,000, 12,000, 14,000, 16,000, 18,000, and 20,000 Hz) served as a useful technique in the diagnosis of early hearing loss.[11] This information is necessary to comprehend the significance of expanding audiometric evaluation to very high frequencies. It enables the identification of any hearing deficiency before it has an impact on the patient's ability to communicate and leads to irreversible changes in the communication hearing thresholds (500-4000 Hz).

2. Nasal involvement

The nasal manifestations in SS may be attributed to autoimmune-mediated damage to the nasal exocrine gland elements, interference with mucosal muscarinic receptors, and disruption of gland extracellular matrix via metalloproteinases. Additionally, the mucociliary function of the nose also gets affected in these patients.

The presence of a highly viscous, thick outer mucus layer that does not move effectively with periciliary fluid, prolongs mucociliary clearance time. This dysfunction in the tracheobronchial system results in xerotrachea. These destructive processes in SS lead to slow mucociliary clearance and mucus production in the nose, resulting in chronic inflammation.[12] It causes mucosal dryness of the upper respiratory mucosa. Nasal mucosal biopsies of patients with SS have shown severe peri glandular infiltration of plasma cells and lymphocytes. There is also atrophy of glands and peri-glandular fibrosis, which can reflect the severity of the disease.

Certain reports have noted that the nasal effects of SS may manifest with symptoms such as rhinorrhea, congestion, crusting, epistaxis, and olfactory dysfunction.[13] According to electron microscope findings, the submucosa is filled with dense collagen bundles, and these bundles commonly extend through the disrupted basal lamina to the surface epithelium, resembling cirrhosis. The intranasal Schirmer test measures mucosal humidification. Wetting distances between 6 and 18 mm are considered normal in healthy individuals. Dryness of the nose has been observed to be associated with primary SS patients who had lower intranasal Schirmer scores.[14]

Patients experience atrophy of the nasal mucosa, xerorhinia (dry nose), hyposmia (decreased sense of smell), hypogeusia (decreased perception of taste), nasal crusting synechiae formation, nasal obstruction, epistaxis, and nasal septum perforation.[13] Patients with SS have a 2.5-fold greater chance of developing chronic rhinosinusitis.[13] Primary SS is significantly associated with increased risks of overall malignancy, non-Hodgkin lymphoma (NHL), and thyroid cancer.[15]

Abnormal chemosensory perception leading to impaired smell and taste noted in SS patients could be due to reduced mucin (an odorant carrier) and recurrent rhinosinusitis.[16] In addition, recurrent epistaxis, due to dryness, leads to scab formation and nasal blockage that also contributes to reduced smell sensation.

Nasal polyposis has been recently identified as associated with SS. Rhinomanometry, mucociliary clearance determination, or smell discrimination tests have little value in the management of the primary SS. The diagnosis of the disease seems to be very challenging based on nasal or paranasal symptoms and signs. Hence, it can be concluded that nasal glandular involvement in the formation of polyps is mild and insignificant in primary SS.[17]

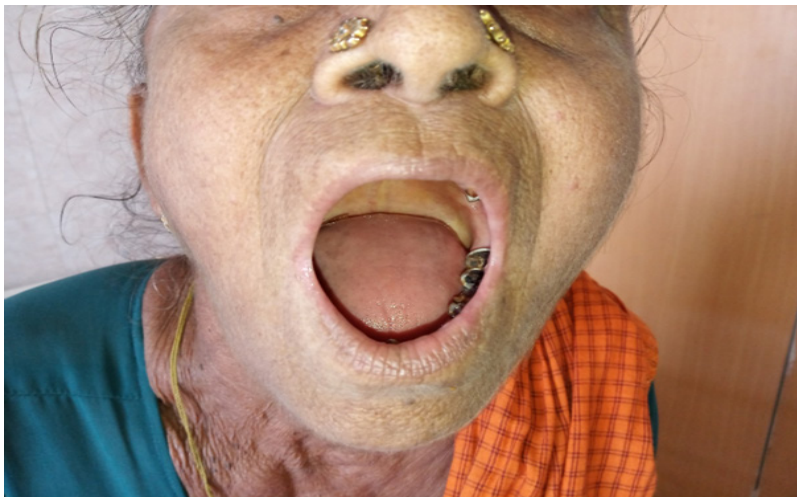
3. Salivary gland involvement

Medications, chronic conditions like SS, and medical procedures like radiation therapy and bone marrow transplants can contribute to xerostomia.[18] In SS patients, xerostomia is caused by a reduction in the volume of the entire saliva rather than the viscous palatal saliva.[19] Parotid glands are the most commonly affected, with isolated submandibular gland enlargement seen in about 2.5% of patients.[20] Diagnosis is typically based on clinical confirmation of dry eyes and dry mouth, along with labial minor salivary gland biopsy. Additionally, the diagnosis is supported by the detection of autoantibodies such as anti-La/anti-SS-B and anti-Ro/anti-SS-A.[20]

Reduced salivary secretion due to progressive infiltration of T and B lymphocytes, which destroy the glandular cells and ductal epithelial cells of exocrine glands, is noted in about 50% of patients with SS.[21] A history of parotid involvement at diagnosis is reported in 82% of patients with SS.[21]

Dry mouth and difficulties in swallowing, speech, and taste are the complaints reported by patients with SS. In certain cases, sialadenitis of the major salivary glands, caused by atypical inflammation or obstruction of Stensen's duct, can be the primary manifestation of SS (Fig. 2). Sialadenitis is

Fig. 2: Dry mouth with dental caries in a patient with Sjogren's syndrome



usually caused by sialolith formation. Although sialolithiasis is generally found in the submandibular gland (72-94%) in SS, it may also frequently involve the parotid gland.[22]

The mechanism behind lithogenesis is not completely understood but is mostly attributed to reduced salivary flow and a high concentration of calcium salts in the saliva. Calculi generally consist of a mixture of various calcium phosphates (carbonate-apatite, hydroxyl-apatite) together with an organic matrix. Calculi formation begins with an organic nidus, followed by progressive apposition of organic and inorganic substances.

The fragments of minor salivary glands are processed completely according to the SS focus score, which grades the degree of inflammatory infiltration in the salivary gland.[23] A focus score of 1 or greater is considered supportive of the diagnosis of SS. Long-term follow-up is important, with an assessment every 3 months.[23] Histopathological specimens show atrophy of acinar cells with lymphoplasmacytic inflammation, myoepithelial islands, and lymphoepithelial lesions. Punctate expansion is seen in peripheral ducts, and a lacy appearance also emerges in the main duct. The parotid gland in SS is usually seen as a 'tree in bud'.[24]

Sialography has traditionally been considered the best modality for detecting early changes, but MR sialography demonstrates similar sensitivity and specificity. The earliest change is punctate sialectasis, characterized by the appearance of numerous 1-mm collections of contrast. Subsequent acinar atrophy leads to the formation of more globular collections of contrast and the development of contrast-filled peripheral cavities, known as cavitatory sialadenitis. In advanced disease, the central ductal system may become dilated.

The staging of SS, as described in 1957, is still commonly applied to MR sialography, and it includes the following stages: Stage I, characterized by punctate contrast collection of 1 mm; Stage II, marked by globular contrast collection of 1 to 2 mm; Stage III, featuring cavitatory contrast collection of 2 mm; and Stage IV, indicating the destruction of the parenchyma of the gland.[20]

Cross-sectional imaging is typically normal in the early stages of the disease, while ultrasound (US) of the gland tends to reveal abnormalities in advanced disease. The gland may appear enlarged, displaying multiple hypoechoic areas, or it may exhibit a reticulated pattern due to the presence of hyperechoic lines. Color Doppler US often reveals increased vascularity. Contrast-enhanced CT findings are nonspecific and frequently include enlarged glands, nodules, punctate calcification, abnormal enhancement, and small low-attenuation cystic areas. In the affected glands, MRI changes reflect the process of atrophy and fatty replacement, resulting in parenchymal heterogeneity, cystic areas (reflecting the globular changes seen in digital sialography), and solid nodules (histologically described as benign lymphoepithelial lesions).[20]

The typical MR appearance on T2-weighted images involves a honeycomb parenchymal pattern that corresponds to the reticular pattern seen in the US. Benign lymphoepithelial lesions (BLEL) can sometimes be mistaken for neoplasia,

and peri parotid adenopathy may also be present. Short tau inversion recovery (STIR) and fat-saturation imaging have revealed that SS is associated with premature fat deposition in the parotid gland and the degree of fat deposition is linked to the loss of secretory function.[25] A recent study utilizing low-field chemical shift MR in the assessment of SS showed a strong correlation between the secretory function of the gland and parenchymal signal heterogeneity, reflecting an irregular distribution of fat, on the opposed phase sequences. Furthermore, images obtained with a low-field magnet lacked the spatial resolution required for MR sialographic grading.[20]

MRI is frequently used to diagnose parotid gland tumors and to detect inflammatory alterations. The 'salt and pepper appearance' or many areas of low signal intensity on T1 weighting, and foci of high signal on T2 weighting are the MRI findings that are indicative of SS (Fig. 3). According to reports, hyperintense foci signify focal lymphocyte aggregation, while hypointense foci denote a dilated ductal system.[26]

The complications of SS include sialadenitis, sialolithiasis, pyogenic infections of the salivary glands, dental caries caused by xerostomia, and corneal ulceration caused by xerophthalmia.[21] Moreover, SS patients have a 40-fold higher risk of developing lymphoma.[26]

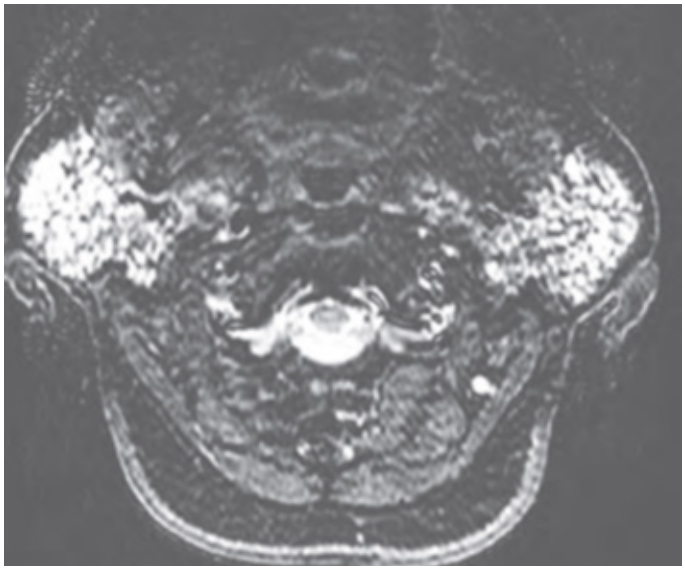
4. Oral cavity involvement

Saliva is vital for protecting and preserving the oral cavity. It provides lubrication, buffering capacity, protection of the teeth, and antibacterial activities. Saliva allows the excretion of toxins and has a possible endocrine function (antigonadotropins). It is made up of proteins, glycoproteins, enzymes, electrolytes, and small organic molecules that are essential for maintaining oral homeostasis. Salivary proteins include lysozyme, mucins proline-rich proteins, Catherine, and histatins.[25]

Patients present with salivary hypofunction: xerostomia is the major oral manifestation. Self-reported xerostomia in population-based samples was analyzed in a systematic review which concluded a range from 0.9 to 64.8%.[26] When the rate of salivary flow is reduced to less than 50%, xerostomia occurs. Common risk factors include female gender, old age, smoking, repeated use of medications, and hospitalization. In this syndrome, there is an alteration of the protein content of the saliva and secretory IgA is decreased which leads to impairment of the antibacterial defense system against caries.[26]

Prolonged dry mouth may cause alterations of functions, such as difficulty in chewing, swallowing, speaking, and symptoms in the mucous membranes, tongue (i.e., geographic tongue), and lips, angular cheilitis, increased risk of dental caries and erosions, and reduced quality of life (Fig. 2). Patients may suffer from bacterial sialadenitis or chronic oral candidiasis due to decreased salivary flow. Clinically, angular cheilitis is manifested as erythema with or without painful fissures and sores at the corners of the mouth. It can occur as a manifestation of a fungal or bacterial infection or due to anemia.

Fig. 3: T1-weighted image with both hypointense cystic structures and hyperintense foci are evident in the parotid gland parenchyma



The most important lesions of the oral mucosa in SS and dry mouth have been investigated in a recent study, which concluded that angular cheilitis was the second most frequent sign, including exfoliative cheilitis, that results in fissured and exfoliative lips with easily bleeding.[27]

Reduced salivary secretion frequently leads to atrophy of the tongue mucosa and the development of fungal infection. The characteristic glossal and mucosal sign is defined as 'crocodile skin' due to mucosal atrophy, dorsal papillary atrophy, and fissuring of the tongue and erythema. When salivary flow is low, younger people may be more likely to feel symptoms of oral dryness, whereas older people may attribute symptoms of dry mouth to a more intricate constellation of circumstances, of which salivary flow is only one element.[28]

5. Pharyngeal, laryngeal, and tracheal involvement

A laryngological examination can reveal viscid secretions on the posterior pharyngeal wall and tenacious mucus over the vocal cords. The most common complaints include soreness and/or dryness of the throat, dysphagia, and hoarseness. Additionally, tracheal dryness may lead to a chronic dry cough and predispose the bronchial mucosa to dryness. Due to mucosa dryness, SS patients may have an increased risk of pharyngeal and tracheo-bronchial infections.[29]

Another important factor in SS patients is gastrotracheal reflux. In SS, the patient can be predisposed to gastro-oesophageal reflux and also to reflux into the trachea, which can mimic upper respiratory tract infection.[29] Along with sicca symptoms, other reported laryngeal manifestations associated with SS are neurological processes such as deviated laryngeal pharyngeal axis and vocal cord palsy, bamboo nodules, esophageal reflux, laryngopharyngeal reflux, and cricoarytenoid arthritis or fixation. Patients also exhibit auditory–perceptual features of dysphonia characterized by hoarseness, breathiness, and strain, with estimates of dysphonia severity ranging from mild to moderate.[30]

The phonation threshold pressure (PTP) represents the energy required to drive small-amplitude oscillation of the vocal cord and it depends on the biomechanical properties including the thickness, elasticity, and viscosity of the vocal cords. It serves as an indicator of ease of phonation and the health of the vocal folds. Theoretically, the hydration of the vocal folds along with the lowering of the viscosity can lead to a reduction of PTP. Dysphonia rates of 41.9% and 59.4% have been reported in SS patients.[31] It has been noted that voice disorders begin gradually, are chronic, and are correlated with SS disease severity independent of age, sex, disease duration, comorbid autoimmune conditions, and the use of SS-related medication.[31]

Evaluation using laryngostroboscope has revealed that straightness and smoothness are affected in SS patients, whereas there is no difference in phase symmetry and amplitude.[32, 33] Dysphonia and articulatory imprecision are potential symptoms of primary SS, usually in a mild-to-moderate form.[34] Due to the lack of structural changes, it is difficult to recognize the clinical significance of laryngeal dryness as a cause of dysphonia. It has been stated that laryngeal adduction could be affected and can result in a lack of periodical vibration. In addition, an imbalance of the vocal cords can also be considered as a reason of dysphonia.[35]

A distinct type of laryngeal lesion, known as the 'bamboo node,' is characterized by a white or yellow transverse lesion in the submucosal region, typically confined to the middle one-third of the vocal fold. Additionally, both granulomatous and non-granulomatous laryngeal nodules have been reported in patients with SS. These patients may also experience intermittent hoarseness, even when no laryngeal pathology is identified during physical examination.[35] Primary lymphoma of the subglottic airway mimicking high laryngotracheal stenosis has been reported in certain patients with SS. In such cases, surgical treatment with single-stage resection and laryngotracheal reconstruction can be preferred.[36]

Musculoskeletal symptoms can occur with a frequency range of 15-90%. The impact of SS on TMJ dysfunction remains unclear.[38-39] Subjective symptoms of the TMJ such as pain on lateral TMJ palpation, pain during mandibular movement, and pain on muscle palpation are significantly higher in SS patients. These findings provide evidence of the effects of SS on the masticatory system.[37]

Certain researchers have reported that taste disorders in SS patients are caused by decreased salivary flow. However, it was found that a small number of individuals with chronic or total loss of function in the major salivary glands did not complain about their sense of taste. In fact, they demonstrated normal taste function in subjective testing.[40] For the examination of taste function, an

electrogustometer (EGM) was employed along with the filter paper discs (FPD) method, which is considered a quantitative evaluation modality for taste function.[41]

In a study involving 29 SS patients, elevated detection and recognition thresholds were reported for sour or bitter stimulants in 90% of the subjects, 76% for salty stimulants, and 38% for sweet stimulants.[42] In a few patients, a correlation was reported between salivary flow and taste function, while in some patients, no significant correlation was observed between the degree of salivary flow deficiency and taste disorders.[43] In addition, there is no correlation between taste function and serum zinc concentration in SS patients.[41]

In conclusion, taste disorders occur in cases with severe deficiency in salivary flow, and the incidence of such disorders is less frequent in SS patients than previously reported.[41] Hence, If a patient presents with dryness of the throat, dysphagia, hoarseness of voice, laryngeal nodules, TMJ arthralgia, and taste disorders, SS should be considered as a part of the differential diagnosis and disease workup.

6. Esophageal involvement

Many autoimmune diseases may have dysphagia as the presenting complaint. Both oropharyngeal and esophageal swallowing disorders have been reported in patients with SS. Dysphagia is often attributed to factors such as sicca of the mouth and throat, loss of dentition, neuropathy, esophageal abnormalities, and gastroesophageal reflux. The prevalence of swallowing disorders reported in SS ranges between 32 and 71%, significantly higher than the general adult population (12%).[44] The severity of SS tenders to be more pronounced in patients with associated swallowing disorders.[45]

Dry and damaged laryngopharyngeal mucosa, resulting from lymphocyte infiltration, can lead to residue formation with solid and semi-solid foods during swallowing. The presence of residue in different areas indicates that multiple factors contribute to the mechanism of SS. Detection of residue in the postcricoid area, particularly with foods like fish crackers or yogurt, can be significant for identifying upper sphincter pathology. Residue in the vallecula or pharyngeal wall is also related to a lack of secretion. Additionally, residue in the pharyngeal wall with semi-solid food can provide insights into motility. The loss of mucosal wetting, hydration, dilution, and lubrication functions results in poor oral function and raises additional systemic concerns that can reduce the overall quality of life.

A statistically significant difference in dysphagia is observed between solid food and semi-solid food.[46] In addition, changes in taste, chewing, speaking, food bolus formation, and swallowing, as well as trauma to oral tissues, are often experienced by individuals with primary SS. Swallowing difficulties are frequently encountered complaints (32-85%) in primary SS patients.[46]

The residue formation during swallowing increases the risk of laryngeal penetration and aspiration. This indicates that the incidence of aspiration pneumonia might be higher in patients with SS than in the normal population, contributing to increased morbidity and mortality.[46]

Previous research has demonstrated that primary SS patients show signs of both impaired sympathetic and parasympathetic nervous function and there are various mechanisms have been proposed to describe this association. For instance, anti-M3-receptor antibodies and cytokines interfere with neural signaling or inflammation that affects autonomic nerves or ganglia.[47] Since gastrointestinal motility and secretion are influenced by the autonomic nervous system, disturbed autonomic neural signaling may impair the gastrointestinal system functioning.

Indeed, primary SS patients have been reported to have an abnormally slow gastric emptying and anti-M3-receptor antibodies have been suggested to be a possible pathogenetic factor. However, the presence of such antibodies has not yet been demonstrated in some patients. While comparing SS patients with and without dysphagia, a significantly lower excitation–inhibition (E/I) ratio (a parasympathetic index) has been noted in the former group. In addition to regulating esophageal motility, the parasympathetic nervous system also plays a role in controlling exocrine secretion in the esophagus and salivary glands. This suggests that parasympathetic dysfunction could potentially lead to both dysmotility and reduced exocrine secretion in these areas, contributing to the development of dysphagia.

Dysphagia affects the quality of life of approximately 64% of SS patients, with only 27% achieving a complete cure.[45] Esophageal manometry has revealed that around one-third of SS patients exhibit varying degrees of esophageal

dysmotility. While some studies have reported a correlation between dysphagia and esophageal dysmotility. Additionally, esophageal webs have been suggested as a cause of dysphagia and are found in about 10% of primary SS patients.[47]

Various otorhinolaryngeal manifestations of SS have been listed in table 1.

7. Otolaryngeal malignancy and SS

The rare occurrence of primary manifestations of nasopharyngeal involvement, presenting as an infiltrative lesion/malignancy, has been noted in patients with SS. The risk of developing non-Hodgkin lymphoma is estimated to be 44 times greater in SS patients than in the general population, probably due to chronic stimulation of the autoreactive B cells.[15] Reports have also indicated the occurrence of transitional cell carcinoma of the nasopharynx and squamous cell carcinoma of the salivary glands in these patients.[15]

Patients with SS have a 40-fold higher risk of developing lymphoma.[26] Predictive factors for lymphoma in SS include persistent enlargement of the parotid gland, lymphadenopathy, glomerulonephritis, splenomegaly, and mixed cryoglobulinemia. Non-Hodgkin lymphoma is the most common type of lymphoma in SS, with the majority being marginal-zone B-cell lymphomas. The prevalence of non-Hodgkin lymphoma in SS is approximately 4%.[26] It is essential to rule out the possibility of a localized lymphoma in patients presenting with persistent enlargement of the parotid gland.

Regular follow-ups are essential for the early detection of signs suggestive of lymphoma. During each follow-up visit, a clinical examination of the neck should be conducted to check for signs of lymphadenopathy, and the parotid gland should be examined for changes in size and asymmetry. Additionally, associated symptoms, such as recurrent fever, persistent fatigue, unexplained weight loss, and night sweats, must also be assessed. If there is clinical suspicion of malignancy, a repeat MRI followed by fine needle aspiration cytology (FNAC) should be performed.[25]

Conclusion

SS can manifest with a spectrum of ENT symptoms. These may encompass the typical signs, such as dryness and discomfort in the mouth and eyes. Furthermore, SS can impact the throat, leading to symptoms like hoarseness, difficulty swallowing, and a sensation of a lump in the throat. Additionally, it can affect the nose and ears, potentially causing congestion, dry nasal passages, and recurrent ear infections. Dysphagia, and pharyngeal, esophageal, and gastroesophageal reflux disease (GERD) symptoms are more prevalent in primary SS patients compared to controls, with no significant association between dysphagia and dysmotility. This underscores the importance of comprehensive care and monitoring for individuals living with SS to address the wide array of ENT manifestations and associated complications.

Table 1: Otorhinolaryngeal manifestations of SS

Salivary glands	Oral cavity	Ear	Pharyngeal, laryngeal and tracheal manifestations	Nasal and paranasal manifestations
Parotitis Sialolithiasis Sialadenitis Reduced salivary flow rate	Xerostomia Oral candidiasis Glossitis and mucositis Dysgeusia Dysphagia Dental caries and tooth decay Recurrent aphthous ulcers Angular cheilitis Oral lichen planus Enamel erosion	Otitis media Sensorineural hearing loss Vestibular dysfunction Tinnitus Chronic ear infections Eustachian tube dysfunction Autoimmune inner ear disease	Susceptibility to infections Voice disorders Dysarthria Dysphagia Dry cough	Nasal dryness Nasal congestion Epistaxis Sinusitis Anosmia Chronic rhinitis Septal perforations

Competing interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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