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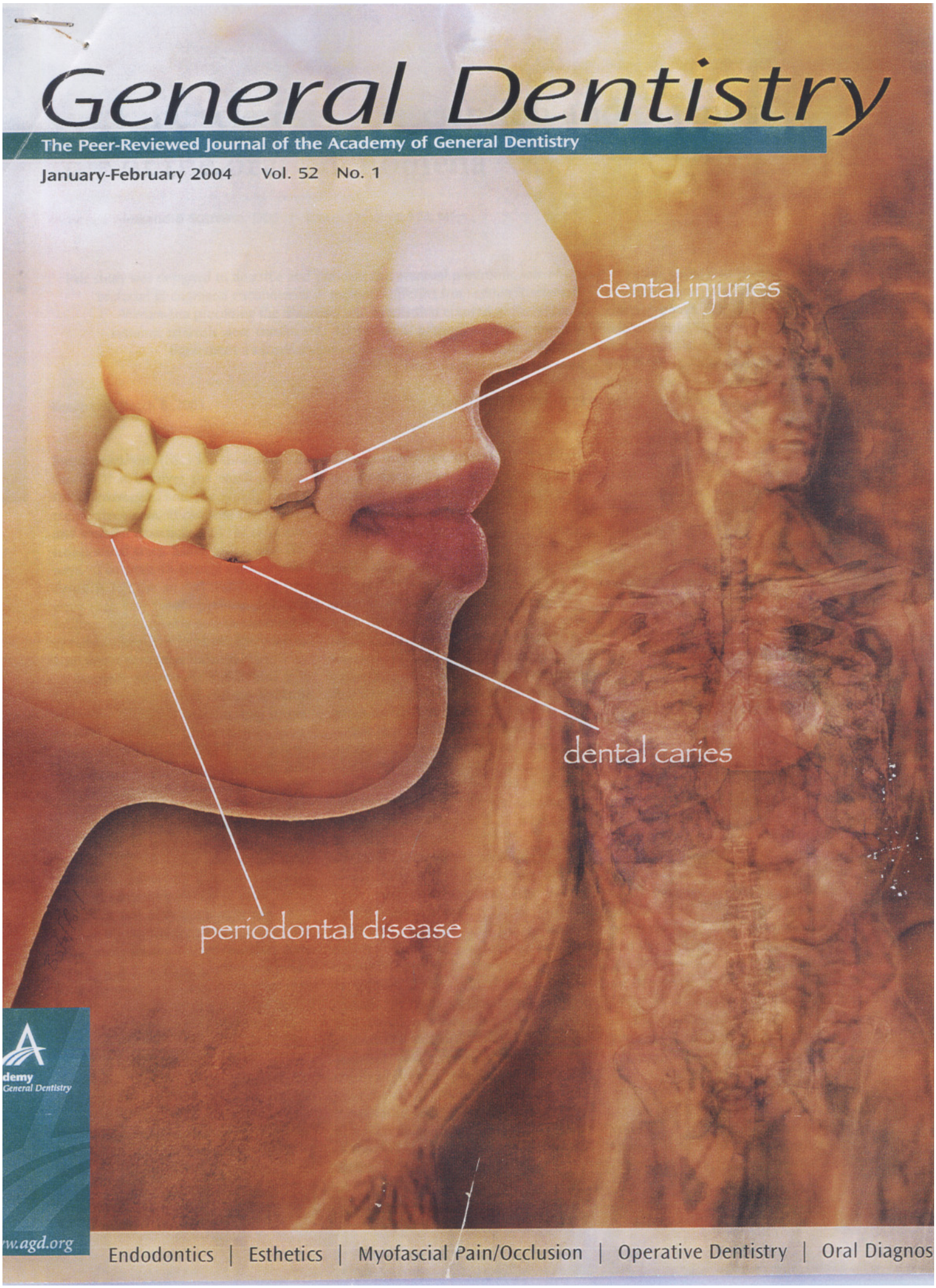

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Systemic sclerosis: Description and diagnostic role of the oral phenomena

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This study was designed to describe and evaluate the eventual prodromic role of the orofacial phenomena encountered in patients suffering from systemic sclerosis. Phenomena preceding the diagnosis and events that occured at the onset of systemic sclerosis were considered. Sclerodermic patients were monitored to implement a secondary preventive plan for controlling oral injuries caused by developing systemic sclerosis.

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Systemic sclerosis (SSc) is a multisystemic disease that involves the small arteries, the microvessels, and the widely spread connective tissue. It can result in fibrosis and vascular destruction of the skin, gastrointestinal tract, heart, lungs, and kidneys. The male-to-female ratio of incidence is 1:3; the annual number of new cases each year ranges between 4–12 per million. SSc currently may be classified as systemic sclerosis or localized scleroderma; both forms have other variants.[1-3]

Systemic sclerosis is subdivided into CREST syndrome and diffuse systemic sclerosis. CREST is characterized by calcynosis, Raynaud's phenomenon, alteration at the esophageal level, sclerodactyliasis, and teleangectasis; it subsequently is subdivided into morphea scleroderma and linear scleroderma. Diffuse systemic sclerosis is rare and acute and results in a severe prognosis for the patient. CREST syndrome occurs more frequently, involving a more restricted area; its prognosis usually is more favorable than that of systemic sclerosis.

The etiological agent that causes SSc remains unknown. Some authors have reported a low family occurrence and have described events involving connective tissue disease among relatives of SSc patients, even when no symptomatology was evident.[4-6]

Hypotheses regarding the pathogenesis of the disease must consider the mechanisms regarding vasal endothelium injuries and extracellular matrix accumulation.[3] Approximately 95% of SSc patients show antinuclear antibodies, the most sensitive and specific of which are the anticentromere antibodies. The antitopoisomerase I antibodies appear to be the most recent autoimmune serological indication of SSc.[7,8]

It has been suggested that sclerodermic injuries may derive from several factors acting together synergically, even though it it is unknown whether the production of autoantibodies is an epiphenomenon pertaining to the disease exclusively or whether the antibodies play an important role in lesion forming and cell activation. Recent research hypothesizes that SSc patients have a genetic predisposition toward the disease.[9]

SSc first manifests with general feeling of general fatigue, arthralgia, Raynaud's phenomenon, and slight fever. Some weeks later, both the fever and inflammatory symptoms disappear and the sclerotic induration stage begins 8–12 weeks later. Sclerotic induration begins suddenly in 10–15% of cases; the widespread cutaneous involvement progresses quickly while visceral involvement (the leading morbidity and mortality factor) appears early and develops into the functional deficit.[9] Orofacial phenomena recorded during SSc include head and neck torpidity; injuries at the perioral cutis and oral mucous membrane; microstomia followed by a reduction of the interincisal and intercommisural distance; fibrosis at the hard and soft palate level; telangiectasis and chromatosis, corresponding to the cutis of both the facial area and the oral mucous membrane; xerostomia; frequent association with Sjögren's syndrome; a widening of the periodontal ligament; bone absorption; and trigeminal neuropathy.

Facial cutaneous damage is the most evident effect of the disease. In its clearest phase, the disease can result in sclerodermic facies.[6] For localized SSc (acrosclerosis), the only clinical phenomena displayed often are found at perioral level; these injuries may be ribbon-like but primarily are sabre-cut shaped.[4]

Sclerodermic facies are typical of diffuse SSc: the sclerodermic process alters the patient's countenance so that the face becomes expressionless. In addition, 80% of patients with sclerodermic facies display microcheilia and relative microstomia with reduced ability to open the mouth. These symptoms may precede the onset of the systemic pathology and portend the manifestation of SSc. The most frequent orofacial phenomena that precede SSc appear as trigeminal sensory neuropathy, a widening of the periodontal ligament space, condylar resorption, resorption of the angle of the jaws, and the presence of a hypomimic facies (microstomia and microcheilia).[4,10-17]

Trigeminal sensorial neuropathy is a sensory disorder that may involve all of the trigeminal branches; based on the literature, it appears to be the most frequent orofacial phenomenon to precede SSc. In 1980, Teasdall et al described 10 sclerodermic patients. In five of those cases, trigeminal neuropathy was the first symptom of the illness; in two others, facial paresthesia directly preceded the onset of SSc by six months and one year, respectively.[10] That same year, Vincent and Van Houzen described a case of overlap SSc in which neuropathy arose prior to the systemic pathology.[11] In 1987, Lecky et al described 22 cases of sclerodermic patients. Nine of the cases showed trigeminal neuropathy as first symptom of the illness, which led the authors to suspect an underlying pathology of the connective tissues.[13]

Rabey described the lysis of the condyles and of the coronoid process for the first time in 1977, when he found four cases of condyle inflammation; the origin

of this inflammation was unknown.[18] In recent years, many other sclerodermic cases with condylar inflammation and open bite have been described. In some of these cases, these alterations were the first and only symptoms of SSc. The presence of a remarkable open bite is the clinical characteristic of condylar inflammation during SSc. Among young patients, for whom the stages of the disease pass quickly, this open bite may constitute the first and primary clinical sign of SSc.[19,20]

Gray et al first hypothesized in 1976 that mandibular bone absorption could be an early symptom of SSc when other clinical symptoms were not apparent.[21] According to Ryatt et al, the absorption at the angle of the jaw occurs twice as frequently as the coronoid process; both of these symptoms (in addition to periodontal ligament thickening) often are associated with bone absorption at the level of the jaws' angle.[22] Wood and Lee pointed out the association between SSc and mandibular erosion, indicating a higher degree of systemic involvement and the extent of oral cavity opening in sclerodermic patients.[2] In 1996, Rout et al confirmed that bone absorption is related to the later stages of SSc.[19] The involvement of the deeper muscular and skeletal planes results in an increase of both bone absorption and alveolar crest deformations.[15]

The alterations that occur at the capillary level in the labial zone of sclerodermic patients demonstrate the microcirculatory system's characteristic pattern of diffuse disorganization. Labial capillaries are short and distributed irregularly, showing variable morphological alterations with a high degree of heterogeneous forms. These alterations are visible via the recently developed microscope technique known as *labial mucous membrane capillarioscopy*.[23-25] Many authors have hypothesized a correlation between the extent of capillarioscopic alteration and the clinical development of SSc; they put forward the idea of diverse biomicroscopic stages of SSc on the basis of a parallelism between microvascular anomaly progress and skin involvement connected with SSc itself.[25] Research supports the hypothesis that capillary anomalies during SSc may serve as predictive criteria for an early diagnosis, since they are a primary phenomena of the disease itself.[24]

Wood and Lee's 1988 study indicated that caries and periodontal diseases were more frequent among SSc patients. This higher incidence was ascribed to xerostomia, which inhibits the salivary action of protection and cleansing and reduces the patient's ability to open the mouth, which interferes with normal oral cleaning.[2]

This study examined a group of 35 sclerodermic patients and described their orofacial phenomena. The potential anticipatory role of these phenomena in the development of SSc was considered, as were treatment approaches.

Materials and methods

Between July 1998 and July 2002, 35 sclerodermic patients were examined at the Department of Oral Medicine of Palermo, Italy. Thirty-two of the patients were women; the mean age of the patients was 61.8.

At the time of this study, all 35 patients fulfilled the diagnostic criteria for scleroderma. These criteria include the presence of Raynaud's phenomenon linked to typical skin lesions and to a visceral involvement, capillaroscopic disorders typical of the nail plate; and a diagnostic evaluation of rheumatic factor positivity and sieric antibodies direct to the nucleus, centromer, and topoisomerosis I.[26] None of the patients took any drug that could affect their salivary secretion. All pharmacological treatment was stopped 24 hours before the clinical test began.

Each patient underwent careful clinical testing for alterations in oral soft tissue, possible xerostomia and correlation with Sjögren's syndrome, possible microstomia, possible *Candida* infections, possible restrictions of mouth opening, possible oral mucosa sores in patients wearing prostheses, DMFS, periodontal health (by means of the uletic-bleeding index), bone absorption state, and laboratory tests.

Visual evaluation was performed to detect oral soft tissue changes such as atrophying oral mucosa, teleangiectasia and retracting sclerosis at the perioral tissue level, and oral mucosa sores. Salivary hypofunction was determined according to Bertram; a secretion rate of less than 0.1 mL/minute was regarded as a sign of salivary hypofunction.[27] Diagnoses of Sjögren's syndrome were based on histologic positivity of accessory salivary glands associated with conjunctivitis, xerostomia, and autoantibody positivity.[9,28-32] Secondary *Candida* infections were confirmed by performing cytologic abstraction and relative culture tests.

Table 1. Grading system of tooth mobility.

Grade	Symptoms
0	No movement with lateral pressure
1	Mild movement; < 1.0 mm of lateral movement
2	Severe rotational movement; depressible/lateral movement > 2.0 mm

Regarding any restriction for opening the mouth, the interincisal distance between the incisal edge of the maxillary and mandibular first incisors was measured. Because of the high number of cases (29) with missing incisors, the distance between the nearest points of the two vermilion borders also was measured in the sagittal plane. The interincisal distance was considered to be decreased if it was less than 40 mm; similarly, a distance less than 45 mm between the nearest points of the two vermilion borders was considered to be decreased.[33-35]

Using the gingival bleeding index with a World Health Organization periodontal probe, the periodontal condition of each patient was examined. Tooth mobility for each patient was assigned one of three grades (see Table 1). DMFS was determined by a clinical examination and confirmed with radiography of the complete dental arch; the stage of bone absorption also was examined using radiographs made by means of ortopantomography.

Radiographs were performed using the same techniques and same exposure procedure for each patient. Involvement of the temporomandibular joint (TMJ) was appraised by radiographic examination and by clinical tests that evaluated indications of involvement, including the presence of pain and clicking and mandibular deviation upon opening the mouth.[36] Emetic withdrawals and related laboratory analysis were performed to evaluate the presence of antibodies antinucleus and eritrosedimentation velocity.

In addition, the patients were instructed to follow a specific home-treatment management plan that included

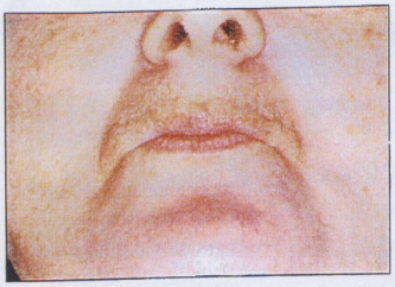

Fig. 1. A face affected by the sclerodermic process.

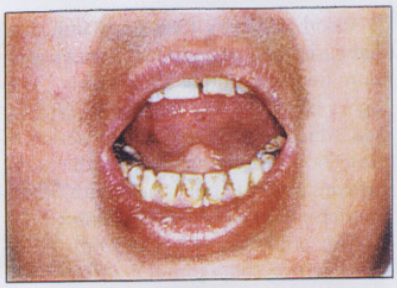

Fig. 2. Teleangiectasis of the oral mucosa.

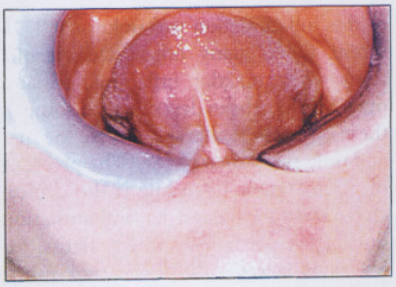

Fig. 3. Sclerosis of the frenulum.

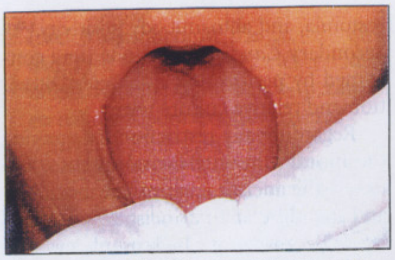

Fig. 4. A patient with a *Candida* infection.

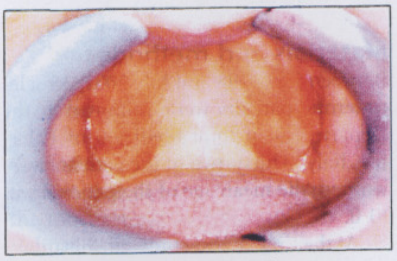

Fig. 5. The rapid bone absorption process and the subsequent nonadaptation of the prosthesis results in sores appearing in the mouth.

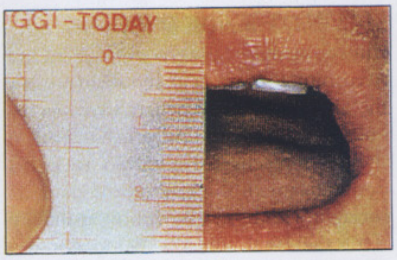

Fig. 6. Microstomia reduces the patient's range when opening the mouth.

checking for plaque by using chlorhexidine in gel and checking for signs of caries by means of fluoride in gel. The orofacial conditions of 45 healthy, sex-matched controls also were investigated. The mean age of the control group was 59.8.

Results

SSc was linked to other autoimmunitary pathologies in 15 of the 35 patients: 7 were diagnosed with diabetes mellitus II, 5 with rheumatoid arthritis, and 3 with systemic lupus erythematosus (SLE). All 15 patients displayed the typical sclerodermic facies (Fig. 1).

Remote pathological anamnesis revealed that seven patients had an episode of trigeminal neuropathy, which manifests as a slow and gradual torpidity in the facial muscles leading directly into pain and paresthesia. Fifteen patients reported an induration of the facial skin (particularly in the cheeks), followed by microstomia. These indurations occured one year before the appearance of symptoms that could be ascribed to the incipient sclerodermic process.

Regarding the alteration of the soft tissues, 15 patients reported diffuse telangiectasia in the oral mucosa membrane while 16 patients reported retracting sclerosis at both the frenulum of the tongue and the perioral tissues. Eight patients displayed atrophying oral mucosa (Fig. 2 and 3).

Eight patients had secondary *Candida* infection; of those, one totally edentate patient was diagnosed with an angular cheilosis state (Fig. 4) in addition to *Candida* at the superior and inferior retromolar arcs and the tongue level.

Fourteen patients were totally edentulous and wore dentures; of these, 7 displayed sores resulting from the rapid bone absorption process and the subsequent misfitting of the prosthesis (Fig. 5). Patients who were not edentulous showed a positive gingival bleeding index. All of the patients showed tooth mobility.

All interincisal distances less than 40 mm were considered below standard; based on that criteria, all patients showed a significant reduction in oral opening compared to the control subjects ($p < 0.001$) (Fig. 6). Salivary hypofunction (a salivary secretion rate less than 0.1 mL/minute) was detected in 15 patients and in 1 of the control subjects. Fourteen of the 15 patients with objective signs of xerostomia complained of subjective xerostomia.

Among the 15 patients with salivary hypofunction, 8 indicated a positive Shirmer's test, the presence of a high focusal infiltrate at the level of bioptic casts of the labial salivary glands, antinucleus antibodies replay, and anti-Sjögren's syndrome A and B (anti-SSA and anti-SSB) antibodies, with an increase in VES. These observations made it possible to diagnose Sjögren's syndrome.

Both DMFS and carious lesions were more frequent among patients with scleroderma than in control subjects, particularly among sclerodermic patients with xerostomia ($p < 0.001$).

Each patient's radiograph showed a diffuse bone resorption; for the most part, these were localized at the ramus of the mandible level. Two patients showed considerable resorption at both the coronoid process level and at the condylar head; as a result, the function of the joint was affected. Patients with mandibular erosions had a significantly greater number of organ systems involved and restriction of mouth opening than those without such lesions. Laboratory tests indicated the presence of antibodies antinucleus in 27 patients while 10 patients

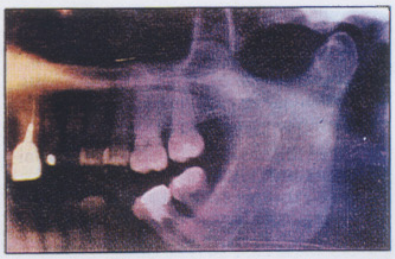

Fig. 7. A radiograph shows a broadening of the periodontal ligament on the mandibular left second premolar.

(and 3 members of the control group) displayed an increasing erythrosedimentation velocity. One female patient had an enlarged periodontal ligament on the mandibular left second premolar; this enlargement could not be ascribed to an ongoing occlusal trauma (Fig. 7). Table 2 presents a complete list of oral symptoms found among patients in this study.

Table 2. Oral symptoms in 35 patients with systemic sclerosis.

	Patients (n = 35)	Control (n = 45)
Autoimmunitary pathologies	15 (42.8%)	0
Trigeminal neuropathy	7 (20%)	0
Facial induration	15 (42.8%)	0
Telangiectasis	15 (42.8%)	5 (11.1%)
Retraction of oral frenulum	16 (45.7%)	0
Atrophy of oral mucosa	8 (22.8%)	3 (6.6%)
Candida infections	8 (22.8%)	2 (4.4%)
Sores	7 (20%)	0
Gingival bleeding index positive	21 (60%)	14 (31.1%)
Tooth mobility	21 (60%)	5 (11.1%)
Interincisal distance (in mm)	31.76 (SD ±3.87)	49.47 (SD ±4.17)
Two vermilion borders distance (in mm)	32.07 (SD ±5.4)	51.66 (SD ±4.06)
Salivary hypofunction	15 (42.8%)	1 (2.2%)
Lymphocitar infiltrate of minor salivary glands	8 (22.8%)	0
Shirmer's test positivity	10 (28.5%)	2 (4.4%)
Antinucleus antibodies	27 (77.1%)	0
Anti-SSA and anti-SSB antibodies	8 (22.8%)	0
VES increasing	10 (28.5%)	3 (6.6%)
DMFS teeth	79.24 (SD ±20.1)	35.80 (SD ±24.8)
Bone resorption (mandible's ramus)	4 (11.4%)	0
Coronoid process–condylar head resorption	2 (5.7%)	0
Periodontal width	1 (2.8%)	0

Table 3. The secondary preventive plan designed to control oral injury resulting from SSc.

Oral hygienic conditions must be kept satisfactory with periodic scaling and root planing.
Dentists must constantly review status of radiographs to determine the extent and long-term development of bone absorption and the extent of periodontal ligament enlargement.
Appraisal of xerostomia conditions with the possible complications they may cause, such as periodontal pathologies, development of caries, and *Candida* infection.
Review of the extent to which the patient can open the mouth.
Analyze labial capillary conditions by performing capillarioscopy of the labial mucous membrane, which should allow evaluation of the conditions of ischemia and atrophia of the vascular system.

Discussion

A careful analysis of literature indicates that orofacial events occur during SSc.[1,2,4-6,10-12,14,17-19,21,28,31,37] These events may be considered phenomena that predict the systemic pathology associated with the secondary events that occur after the onset of illness.

Among the so-called primary orofacial phenomena, a trigeminal sensorial neuropathy arises. The prodromal nature of this neuropathy is well-documented in the literature.[10-13,38,39] Its symptomatology consists of slow and gradual torpidity in the facial muscles, which leads directly to pain and paresthesia. Other clinical symptoms of the neuropathy include a feeling of torpidity around the mouth, a deficient sense of taste, direct ageusia, and a loss of sensitivity at the oropharynx level. This neuropathy not only injures the trigeminus but also causes deficits at the glossopharyngeal, acoustic, facial, and abducens nerve levels. Sensory neuropathy often is diagnosed incorrectly in sclerodermic patients; its symptoms sometimes are confused within the wider status of sclerotic stiffness that results from SSc.

Seven of the 35 patients in this study experienced an incident of paralysis involving the trigeminal nerve; the paralysis regressed after an unspecified therapy. The expansion of the periodontal ligament space is another phenomenon connected with SSc and one that some authors consider to be valid for predicting the illness.[14] More frequently, it is a secondary phenomenon of SSc connected to a fibrous thickening (pachynsis) of the periodontal ligament; according to some authors, it occurs with a frequency of 37%.[14] The expansion of the periodontal ligament space is unique because the broadening of dentoalveolar space uniformly involves the entire radicular surface without descending toward the coronoapical zone, which generally occurs in cases of periodontal disease. The expansion of the periodontal ligament space is both a primary and secondary phenomenon in SSc, since it also acts as a diagnostic indicator. This study provided radiographic evidence that one female patient displayed a broadening of the periodontal ligament on the mandibular left second premolar that could not be ascribed to an ongoing occlusal trauma.

According to Marmary et al, the bone resorption that results from SSc (particularly the alterations that occur at the angle of the jaw) is unique, since such resorption never has been found in association with other systemic disease.[4] Bone resorption that occurs in the orofacial zone during SSc consists of osteolytic lesions that are localized almost exclusively at the mandibular level; the sites affected most are the gonial region, the condyle, the coronoid process, and the ascending branch. Resorption may be mono or bilateral and is not related to the patient's age or gender or the duration of disease. Articular pain, head stiffening, and articular swelling due to tendonitis and synovitis are the first symptoms of disease.[17] Radiographic examinations performed during this study demonstrated considerable bone resorption among all of the patients; resorption was found most often at both the angle of the jaw and the gonial region. In addition, all of the edentulous patients displayed an alveolar crest resorption.

Among the alterations that can predict the onset of SSc are the microstomia and microcheilia that are always present in the sclerodermic process; as evidence of the incipient sclerodermic process, 15 of the 35 patients displayed facial skin induration with subsequent microstomia.

This study found a significant correlation between SSc and Sjögren's syndrome, suggesting that a clinical examination always be performed to identify the possible association between the two pathologies.

Determining the fundamental role of certain orofacial phenomena in predicting the onset of SSc was one of the most important results of this study. Seventeen patients reported a trigeminal nerve involvement and facial stiffness years before the systemic pathology was diagnosed. This finding suggests that such pheneomena must be analyzed properly to guarantee a timely diagnosis of the disease.

A specific therapeutic line of treatment was prepared based on the supposition that a distinction could be made between so-called primary and secondary phenomena of SSc. An effective SSc therapy would allow dentists to realize the benefits of diagnosing SSc early, either to prevent its development (and therefore limit its consequences) or to monitor the evolution of the pathology.

By providing a specific therapy, dentists may be able to improve the lives of sclerodermic patients who otherwise might suffer psychological trauma due to the difficulties typically caused by the disease. Odontostomatological hospitals could address the early phenomena of the sclerodermic pathology by carrying out an initial preventive therapy (utilizing radiographs) that would consider the patient's level of pain; any remarkable, early absorption at either the condyle region or the ramus mandibulae (with no periodontal pathologies present); and an enlarged radiograph of the periodontal space that would indicate that neither periodontal pathologies nor pathologies with occlusal origin were present.

These elements are more significant when they are associated with a case history that heightens the suspicion that an underlying connective disorder exists, particularly when it is supported by a positive personal and/or family history or other autoimmune pathologies.

Of the 35 patients, 15 showed a positive pathological anamnesis for other autoimmune pathologies, 7 suffered from diabetes mellitus II, 5 from rheumatoid arthritis, and 3 from SLE. The diagnosis may be confirmed by applying the oral mucous membrane capillarioscopy, an instrumental test currently in development that is useful for diagnosing early alterations of the vascular microcirculatory system and indicating the onset of the sclerodermic process.[23-25]

To start a secondary preventive plan able to control oral injuries resulting from developing SSc, the monitoring patterns of sclerodermic patients were traced (see Table 3). The results will be discussed in a future manuscript.

Conclusion

It is important that dentists can recognize the oral manifestations of SSc, as some of them are predictive of systemic disease. Early diagnosis may improve the expectations and lifestyle of patients and an early therapeutic protocol may slow the evolution and consequences of the disease.

Author information

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