

Characterization of the frenulum and aspects of the tongue of individuals with systemic sclerosis

Caracterização do frênulo e dos aspectos da língua de indivíduos com esclerose sistêmica

Caracterización del frénulo y de los aspectos de la lengua de individuos con esclerosis sistémica

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Abstract

Systemic sclerosis is a progressive, systemic autoimmune disease of unknown and relatively rare etiology. It is characterized by excessive deposition of collagen in the connective tissue, by the thickening of the skin, involvement of internal noble organs and the stomatognathic system. **Objective:** To characterize the frenulum and aspects of the tongue of individuals with systemic sclerosis. **Method:** This is a clinical, observational, case-report exploratory study. A clinical study of the frenum and aspects of the tongue was carried out with a study group composed of eleven subjects, of both sexes from the Rheumatology Department of the University Hospital in partnership with the study group on orofacial motor, both from the Federal University of Sergipe, and a control group matched in number, age and sex, without rheumatic diseases, selected for convenience. The data collection period was three months (from September to December 2017). **Results:** The study group sample revealed clinical changes in the thickness, size and color of the lingual frenum, as well as in the language aspects of individuals with

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Authors' contributions:

VFS – Performed data collection, substantial contribution to the conception and design of the scientific work, literature review, writing participation, critical review of the work and final approval of the content to be published; LFA - participated in the evaluation of the cases, the critical review of the work and final approval of the content to be published; CPHARC - participated in the critical review of the work and final approval of the content to be published; SEZMB - substantial contribution to the design and idea of the scientific work, participation in data collection in the literature survey, writing, critical review of the work and final approval of the content to be published.

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systemic sclerosis, such as hypertonia, altered tongue tip shape when elevated, telangiectasia among others with lesser occurrence. **Conclusion:** The altered characteristics of the frenulum and the tongue of the subjects with this affection evidences the impairment of the stomatognathic system and the importance of Speech, Language and Hearing Sciences performance in Rheumatology.

Keywords: Scleroderma Systemic; Speech Language and Hearing Sciences; Clinical diagnosis; Lingual frenum.

Resumo

A Esclerose Sistêmica é uma doença autoimune sistêmica, progressiva, de etiologia desconhecida e relativamente rara. Caracteriza-se pela excessiva deposição de colágeno no tecido conjuntivo, pelo espessamento da pele, comprometimento de órgãos nobres internos e do sistema estomatognático. **Objetivo:** Caracterizar o frênulo e os aspectos da língua de indivíduos com esclerose sistêmica. **Método:** Trata-se de um estudo exploratório clínico, observacional, do tipo relato de casos. Foi realizada avaliação clínica do frênulo e dos aspectos da língua com um grupo de estudo composto por onze sujeitos, de ambos os sexos, oriundos do serviço de Reumatologia do Hospital Universitário em parceria com o grupo de estudo em motricidade orofacial, ambos da Universidade Federal de Sergipe, e de um grupo controle pareado em número, idade e sexo, sem doenças reumáticas, selecionados por conveniência. O período de coleta dos dados foi de três meses (de setembro a dezembro de 2017). **Resultados:** A amostra do grupo de estudo revelou alterações clínicas na espessura, no tamanho e na coloração do frênulo, como também nos aspectos da língua de indivíduos com esclerose sistêmica, tais como hipertonia, alteração do formato da ponta da língua quando em elevação, telangiectasia entre outros de menor ocorrência. **Conclusão:** As características alteradas do frênulo e da língua dos sujeitos com essa afecção evidenciam o comprometimento do sistema estomatognático e a importância da atuação fonoaudiológica na Reumatologia.

Palavras-chave: Escleroderma Sistêmico; Fonoaudiologia; Diagnóstico clínico; Frênulo da língua.

Resumen

La Esclerosis Sistémica es una enfermedad autoinmune sistémica, progresiva, de etiología desconocida y relativamente rara. Se caracteriza por la excesiva deposición de colágeno en el tejido conjuntivo, por el espesamiento de la piel, comprometimiento de órganos nobres internos y del sistema estomatognático. **Objetivo:** Caracterizar el frenillo y los aspectos de la lengua de individuos con esclerosis sistémica. **Método:** Se trata de un estudio exploratorio clínico, observacional, del tipo relato de casos. Se realizó una evaluación clínica del frenillo y de los aspectos de la lengua con un grupo de estudio compuesto por once sujetos, de ambos sexos, oriundos del servicio de Reumatología del Hospital Universitario en asociación con el grupo de estudio en motricidad orofacial, ambos de la Universidad Federal de Sergipe y de un grupo control pareado en número, edad y sexo, sin enfermedades reumáticas, seleccionados por conveniencia. El período de recolección de datos fue de tres meses (de septiembre a diciembre de 2017). **Resultados:** La muestra del grupo de estudio reveló alteraciones clínicas en el grosor, el tamaño y la coloración del frenillo, así como en los aspectos de la lengua de individuos con esclerodermia sistémica, tales como hipertonia, alteración del formato de la punta de la lengua cuando en elevación, telangiectasia entre otros de menor frecuencia. **Conclusión:** Las características alteradas del frenillo lingual de los sujetos con esa afeción evidencian el deterioro del sistema estomatognático y la importancia de la actuación fonoaudiológica en la Reumatología.

Palabras claves: Esclerodermia Sistémica; Fonoaudiología; Diagnóstico clínico; Frenillo Lingual

Introduction

Scleroderma, more commonly known as systemic sclerosis (SS), is one of the most fearsome among rheumatic diseases, ever since the masterful narration of Carlo Curzio in Napoli (1752). This condition is part of the progressive systemic autoimmune disorder of unknown etiology and is relatively rare, with an incidence of two to ten new cases per million inhabitants per year¹. It predominantly affects women in the proportion of 3:1, between 30 and 50 years old, being infrequent in children and in men less than 30 years old¹. It is characterized by excessive deposition of collagen in the connective tissue, in addition to skin thickening and early involvement of internal organs, including the cardiovascular system, lungs, gastrointestinal tract and kidneys², as well as affecting the stomatognathic system^{3,4,5}.

In the Speech, Language and Hearing Sciences area, there are few SS studies on the stomatognathic system. Almeida et al. (2016) and Baldrighi et al. (2014; 2016)^{3,4,5}, showed changes, such as: the presence of restriction of mouth opening, difficulty in chewing, swallowing disorder, altered posture, tonus and mobility of phonoarticulatory organs^{3,4,5}. Other findings such as microstomia, xerostomia and loss of gingival mucosa have also been described and cited by physicians and dentists^{6,7,8} and, more recently, thickening and shortening of the sublingual frenulum have been observed, but the prevalence of such abnormalities and clinical changes are not yet known⁹.

It is believed that the low incidence and severity of the disease impose limits for conducting studies related to the condition. Systemic SS has a great impact on the orofacial region^{3,4,5}. However, orofacial involvement still remains underdiagnosed and the corresponding symptoms are often obscured by severe systemic manifestations¹⁰.

Due to the scarcity of researches aimed at the characterization of the tongue frenulum in individuals with this condition, this study was proposed.

The objective of this research was to characterize the frenulum and aspects of the tongue of individuals with systemic sclerosis, in order to broaden the scientific contribution on the subject.

Method

This is an exploratory, observational, case-series study. It was held in a room of the Grupo de Estudos de Motricidade Orofacial (GEMO), in partnership with the Ambulatory of Rheumatology, both from the University Hospital of the Universidade Federal de Sergipe (UFS).

The study involved research with human beings, and was, therefore, carried out in compliance with the ethical precepts of autonomy, non-maleficence, beneficence and justice of the Resolution 466/12 of the National Health Council. It was approved by the UFS Research Ethics Committee under the number CAAE 76721317.0.0000.5546.

The subjects were divided into:

- Study Group (SG): adults or elderly (29 to 74 years) of both sexes, who had previous medical diagnosis of SS, who had not previously performed speech therapy and who presented cognitive ability were included to participate in all stages of the study.
- Control Group (CG): adults or elderly (from 29 to 76 years old) of both sexes, free from rheumatic diseases, without complaints related to oral motor aspects were selected in the same environment as the study group.

The exclusion criterion for the SG was to have one or more associated rheumatic diseases and not to accept participating in the research. However, for the CG, it was to present myofunctional orofacial complaints and to give up participating in the research in any of its stages.

The period of data collection lasted three months (September to December 2017), and counted on the participation of 11 individuals in the SG and 11 individuals in the CG, matched for sex and age.

At first, the subject was invited by the evaluator to participate in the study. After the acceptance, the consent form informing all the procedures was read. After awareness and consent of it, data collection began.

The evaluation was based on clinical protocols, one being national¹¹ and another international⁹.

Initially, a brief anamnesis was conducted. It was related to the general issues of identification of the subject and presented specific questions about the condition, such as time of onset of the disease, difficulties related to chewing, swallowing and speech¹¹. Specific examination investigated,

through visual observation and tongue mobility, the sublingual frenulum thickness, the frenulum length, paleness of the sublingual buccal mucosa, and the presence of telangiectasia⁹. The clinical classification of the frenulum was described as: normal, dubious or altered. The fixation of the frenulum was classified as: anteriorized, short, short and anteriorized or with ankyloglossia¹¹.

During the Speech, Language and Hearing Sciences evaluation, photographic documentation and video recording were performed. The labial retractor and the digital caliper were not used due to the restriction of the mouth opening present in the majority of SG subjects.

For a better analysis, the image of the frenulum was obtained by means of a Sony Cyber-Shot DSC-W710 camera in 16.1 Mpx resolution, positioned on a professional X ZHANG STC-360 tripod, within thirty centimeters of the chair where the subject was found. The tripod was set at a height proportional to the level of the face of each participant and the camera framed the images of the intraoral cavity, face and neck of each subject.

Participants were seated in a chair without head support, not limiting their movements and with their feet properly resting on the floor, according to the recommendation of the literature¹².

At the end of the evaluation, individual de-convolution was given to each subject and those who required Speech, Language and Hearing Sciences therapy were referred to the GEMO group.

The results were tabulated in the Excel spreadsheet software (Microsoft® Office package) for descriptive statistical analysis.

Results

The results were presented in tables for better visualization, with the first table showing the characteristics of the CG as age, sex and time of onset of the disease; the evaluated aspects regarding the tongue are found in Table 2 and for the frenulum in Tables 3 and 4.

In the SG, age ranged from 29 to 74 years old (mean: 54.27 ± 14.56 years old) and the SG from 29 to 76 years old (mean: 54.81 ± 15.30).

Table 1. Characterization of the study group regarding the sex, age and time of systemic sclerosis installation

Subject	Age	Sex	Disease onset time
1	29	M	- 05 anos
2	41	F	+ 10 anos
3	42	F	- 05 anos
4	43	F	- 05 anos
5	48	F	+ 10 anos
6	56	F	+ 10 anos
7	61	F	+ 10 anos
8	65	F	+ 10 anos
9	68	F	- 05 anos
10	70	F	- 05 anos
11	74	F	-05 anos

Legenda: F= feminino; M= masculino; (-) menos de; (+) mais de.

Table 2. Results of the evaluation of the tongue aspects of the studied groups

Evaluated Items - Tongue / Groups Studied		SG	CG
Complaint of dry mouth		n=9	n=1
Altered tongue tip format in elevation		n=3	n=0
Telangiectasia		n=4	n=0
Fungal infection		n=2	n=0
Palpation pain		n=1	n=0
Complaint about alteration of taste		n=1	n=0
Presence of tremor		n=4	n=2
Tension alteration		n=5	n=0
Mobility	Protract / Retract	n=6	n=1
	Touch the upper lip with the apex	n=4	n=0
	Touch the bottom lip with apex	n=0	n=0
	Touching the labial commissures (right and left)	n=0	n=0
	Vibrate	n=7	n=7
	Sucking the tongue on the palate	n=4	n=1
Common Tongue Position	Adequate	n=0	n=11
	It was not possible to visualize or the subject did not know	n=9	n=0
	Anteriorized	n=0	n=0
	Interdentalized	n=2	n=0
	Floor of the mouth	n=0	n=0

Subtitle: SG - Study Group; CG - Control Group; n = number of cases.

Table 3. Characterization of the sample regarding the aspects and fixation of the tongue frenulum in both groups

GS	Color	Thickness	Mounting - floor of the mouth visible from		Fixation - lower face of the tongue		
	Altered		Sublingual Caruncles	Lower alveolar crest	Middle Part	Middle part and apex	On the Apex
SG	n=5	n=7	n=2	n=0	n=6	n=3	n=0
CG	n=1	n=0	n=9	n=2	n=0	n=0	n=0

Subtitle: GS - Group of Subjects; SG - Study Group; CG - Control Group; n = number of cases.

Table 4. Final result of the classification of the frenulum in the groups studied

Frenulum Classification / Studied Groups	Normal	Dubious	Altered	Classification of Changes		
				Anteriorized	Short	Short and Anteriorized
SG	n=2	n=6	n=3	n=1	n=0	n=2
CG	n=10	n=1	n=0	n=0	n=0	n=0

Subtitle: SG - Study Group; CG - Control Group; n = number of cases.

Discussion

In this condition, women are affected three to four times more than men and the mean age of onset of the disease occurs at the age of 50 years old¹⁵, usually between 30 and 50 years of age¹⁶, confirming the results obtained in this study, as shown in Table 1.

As to sex, of the 11 subjects with the disease, only one was male, totaling a ratio of 10: 1 - a fact that corroborates with the literature¹⁷.

Regarding the time of onset of the disease, studies show that the first symptoms of SS appeared on average between the first five years of the disease with a progression of 7.40 ± 8.19 and 9.9 years of the installed pathology^{9, 18}. In this study, six of the eleven SG individuals reported being diagnosed with SS for more than five years and five reported a diagnosis of more than ten years.

The complaint of xerostomia is related to the decrease in salivary flow, which is characterized by a subjective sensation of dry mouth, which becomes an unpleasant experience for the individual¹⁹.

The involvement of the salivary gland, which can lead to xerostomia, is a common feature of SS with a prevalence ranging from 25% to 71.2%^{20,21}. The consequence of the reduction in the amount of saliva produced was one of the complaints reported by the individuals in this study. The salivary flow of the SS patient was evaluated by researchers^{20,21}. But the largest cohort study (with 163 subjects with SS versus 231 controls), recently conducted by the Canadian Scleroderma Research Group, found that the disease was associated with reduced production of saliva (63.19 mg per minute vs. 147.52 mg / minute)²².

In this study, nine SG individuals presented dry mouth complaint, totaling the majority of the sample. This sensation also occurred due to the use of some medications (anti-inflammatories, immunosuppressants, antihypertensives) to treat the complications of SS²³. While in the CG population, only one subject presented this symptom. This can be explained by the age of some individuals, the consumption of medications for continuous use (as antihypertensive) and menopause¹⁹.

Another manifestation was telangiectasia, which is a common cutaneous vascular malformation in the oral mucosa in SS subjects. It is characterized by small red spots originated from the dilation of capillaries that appear on the hands,

arms, face and body²⁵. They develop mainly in exposed cutaneous regions of the hands and face and can also be found in the oral mucosa, as in lips and tongue (Figure 1).

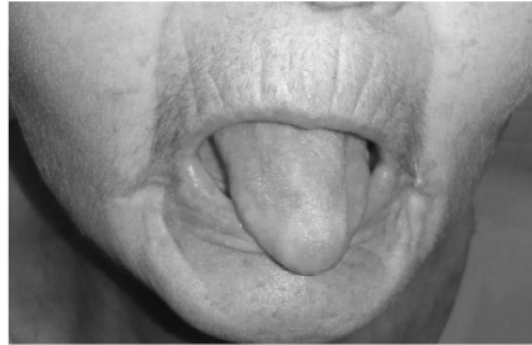


Figure 1. Telangiectasia on the face, lips and tongue

The presence of telangiectasias in the tongue were observed in four individuals of the SG and in none of the CG.

Fibrosis in SS (including oral mucosa and tongue) can occur due to the accumulation of collagen, resulting in loss of elasticity and, consequently, a decrease in the mobility of this region. Simultaneously, muscle atrophy impairs the performance of essential functions, such as chewing and swallowing, promoting additional speech, palate and swallowing difficulties²⁶.

During the evaluation, other intraoral alterations, such as palpation pain and palate alteration, were only observed in the SG (Table 2). Marcucci and Abdala (2009)²⁶ mentioned palate changes in individuals with SS, but no pain was found on palpation.

Changes in the mobility, in the tone of the phonoarticulatory organs and in the habitual resting posture in patients with SS were cited in the literature^{4,5,27}. However it is believed in the hypothesis that these changes occur due to the decrease in the function of the musculature of the face, including the tongue. In this study, five subjects from the SG showed increased tongue tension, which may contribute to the reduction of the amplitude of mandibular movements, the movement of the food bolus, and the swallowing of saliva and food, favoring impairments in stomatognathic functions³.

In four individuals of the SG and in two of the CG, the presence of tongue tremor was evident when the mouth was fully opened. The tongue format observed when raising the tongue inside

the mouth without touching the palate was altered in three individuals of the SG, and of these, two presented the tip of the tongue in heart shape¹¹ (Figure 2), probably due to the frenulum alteration^{9, 11}.



Figure 2. Tongue tip in heart format, thick and short frenulum

The position of the tongue, at rest, during the evaluation in most of the SG subjects could not be observed because they kept their mouths closed, except for the subject S3, who kept the tongue at rest between the teeth laterally, and S5, who rested it between the lower teeth. In the CG, it was not possible to visualize the posture of the tongue due to the lip seal at rest.

As for the praxis tests, these are fundamental, such as tongue protrusion and retraction, tongue lateralization on the labial commissures and on the upper and lower lips (cardinal points), tongue tip vibration and tongue suction on the palate. In this study, all subjects were able to perform most of the mentioned functions, however, six subjects (S2, S3, S4, S5, S9 and S11) presented altered protrusion capacity when compared with other individuals. Proof of tongue mobility involving the cardinal points, only touching the upper lip with the apex of the tongue, was performed with difficulty by four SG subjects and none from the CG.

The vibrating tongue tip was performed with difficulty or not performed by seven SG subjects. It was also observed in seven CG individuals. As it is a refined movement, we can think of the difficulty

in its accomplishment in both groups. In addition, it can be justified in the SG by the increase in the deposit of collagen that these subjects possess, which can cause a hardening in the region of the jugal mucosa, being able to interfere in the mobility of the tongue to carry out the vibration movement²². For the same reason, it can be explained that sucking the tongue on the palate has been performed with difficulty by four SG subjects and by only one of the CG.

Frech *et al.* (2016)⁹, developed a semiquantitative evaluation (with scores) for sublingual abnormalities in individuals with SS, calling it the Sublingual Anomalies Index (SAI). Relevant items of the protocol included the thickness, length and color of the sublingual frenulum, with the presence of oral telangiectasia, being scored and resulting in individual scores. In the study cited, 21 individuals with SS and eight control individuals were evaluated. Individuals with SS presented telangiectasias in the mouth and / or at least one of the following abnormalities in their frenulum: thickening, shortening and pallor⁹.

Compared to the aforementioned study⁹, which guided our work, it was possible to confirm the relevant incidence of changes in the frenulum of SG individuals in SS, totaling seven individuals. The shortening was observed in six individuals, and was not observed in the CG.

Regarding the aspect of coloration, five SG subjects presented pallor (whitish tone). And in seven individuals, the increase in thickness was observed (Figure 3a). The literature⁹ pointed out that the presence of sublingual clinical abnormalities in SS suggests that changes in the tongue frenulum, such as pallor, thickening and intermediate to severe shortening may be late manifestations of the condition, however the authors⁹ warn for the need for more detailed investigation.

In the present study, the individuals with these characteristics were (S2, S5, S7, S8, S9, S10 and S11) and, of these, four had the disease for more than ten years, and the other three, for the last five years. Figure 3a, below, shows the coloring, shortening and thickening of an SG subject compared to the normality pattern of a CG subject.

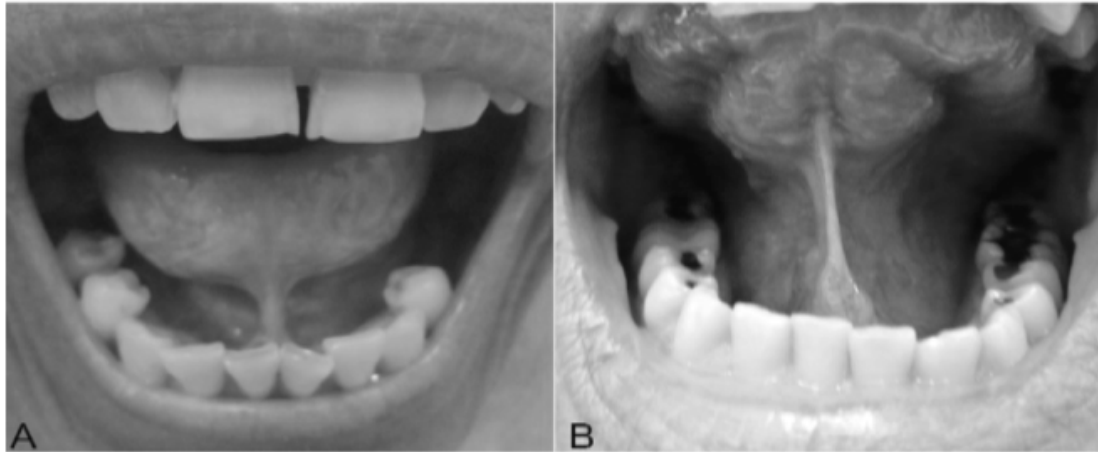


Figure 3. Frenulum during tongue suction proof: A = short tongue frenulum, thick and white on the AG subject. B = tongue frenulum with normal extension and coloring of the subject CG

Regarding the anatomical fixation of the frenulum¹¹, there is a consensus that feeding functions and speech production suffer greater influences from the frenulum alterations. However, in this research, the CG remained in normality with visible frenulums from the sublingual caruncles. In SG, only two of the eleven individuals had their insertion between the sublingual caruncles, six of them presented fixation of the frenulum in the lower middle part of the tongue and three in the middle part reaching the apex of the tongue.

In order to minimize the impact of these changes on the quality of life of these subjects, mainly related to chewing, swallowing and speech, there is a need to study more individuals to better understand the findings and to evaluate the implications and limitations that the changes in the tongue and in its frenulum result in patients with SS.

Conclusion

This study revealed clinical alterations in the thickness, size and color of the frenulum in most SS subjects, as well as in the morphofunctional aspects of the tongue of these individuals.

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