



Speech-language pathology findings in patient with Sjögren's syndrome after multiple strokes

Achados fonoaudiológicos em caso de síndrome de Sjögren após acidentes vasculares cerebrais múltiplos

Hallazgos de fonoaudiología en el caso del síndrome de Sjögren después de múltiples accidentes cerebrovasculares

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Abstract

This study aimed to describe the speech-language pathology findings of a patient with Sjögren's Syndrome, who suffered multiple strokes. A report of a case diagnosed with Sjögren's syndrome and multiple strokes was conducted to this end. The speech-language pathology assessment included neurological anamnesis, application of the MBGR Protocol, Protocol for the Assessment of Apraxia of Speech, Dysarthria Assessment Protocol, Cognitive Screening Battery and the Activities of Daily Living Protocol. The evaluation of the orofacial structures allowed noticing an increase in the height and width of the tongue, dryness of the mucosa of the lips, cheek and palate, poorly adapted dental prosthesis and deep hard palate; however, it was not possible to observe the posterior pharyngeal wall and the soft palate. During swallowing, it was possible to verify the presence of anterior oral escape, reduced anteriorization and laryngeal elevation, presence of noise when swallowing, as well as coughing, throat clearing, wet voice and intraoral stasis of food after swallowing. The assessment of speech apraxia and

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dysarthrophonia revealed distortions and substitution of phonemes, as well as changes in phonation, articulation and resonance. In turn, the memory test showed changes in verbal fluency and recognition. Finally, the clinical evaluation found changes of orofacial structures, neurogenic oropharyngeal dysphagia, and speech and voice disorders.

Keywords: Sjögren's Syndrome; Stroke; Speech-language and Hearing Sciences; Swallowing; Stomatognathic System; Dysarthria.

Resumo

Este trabalho objetivou descrever os achados fonoaudiológicos de uma paciente com Síndrome de Sjögren, vítima de múltiplos Acidentes Vasculares Cerebrais. Para isso, foi realizado um relato de um caso diagnosticado com Síndrome de Sjögren e AVC Múltiplos. A avaliação fonoaudiológica foi realizada por meio da anamnese neurológica, aplicação do Protocolo de MBGR, o Protocolo para Avaliação da Apraxia da Fala, Protocolo de Avaliação da Disartria, Bateria de Rastreo Cognitivo e o Protocolo de Atividades de Vida Diária. Observou-se, na avaliação das estruturas orofaciais, uma língua com altura e largura aumentada, ressecamento da mucosa dos lábios, bochecha e palato, prótese dentária mal adaptada, palato duro profundo, não sendo possível visualizar a parede posterior da faringe e o palato mole. Durante a deglutição constataram-se presença de escape oral anterior, diminuição na anteriorização e na elevação laríngea, presença de ruído durante a deglutição, bem como tosse, pigarro, voz molhada e estase alimentar intra-oral após a deglutição. Na avaliação da apraxia da fala e disartria, foi possível observar distorções e substituição de fonemas, assim como alteração na fonação, articulação e ressonância. Alteração na fluência verbal e no reconhecimento, referentes ao teste de memória. Conclui-se que, por meio da avaliação clínica, foi possível constatar alteração das estruturas orofaciais, disfagia orofaríngea neurogênica, alterações na voz e fala.

Palavras chave: Síndrome de Sjögren; Acidente Vascular Cerebral; Fonoaudiologia; Deglutição; Sistema Estomatognático; Disartria.

Resumen

Este estudio tuvo como objetivo describir los hallazgos del habla y el lenguaje de un paciente con síndrome de Sjögren, víctima de múltiples accidentes cerebrovasculares. Para esto, se hizo un informe de un caso diagnosticado con el síndrome de Sjögren y múltiples accidentes cerebrovasculares. La evaluación del lenguaje hablado se realizó a través de la anamnesis neurológica, la aplicación del Protocolo MBGR, el Protocolo para la evaluación de la apraxia del habla, el Protocolo de evaluación de la disartria, la Bateria de detección cognitiva y el Protocolo de actividades de la vida diaria. En la evaluación de las estructuras orofaciales, una lengua con mayor altura y anchura, sequedad de la mucosa de los labios, mejillas y paladar, prótesis dental mal adaptada, paladar duro y profundo, no fue posible visualizar la pared faríngea posterior y el paladar blando. Durante la deglución, hubo presencia de escape oral anterior, disminución de la anteriorización y elevación laríngea, presencia de ruido durante la deglución, así como tos, limpieza, voz húmeda y estasis intraoral después de la deglución. En la evaluación de la apraxia del habla y la disartria, fue posible observar distorsiones y sustitución de fonemas, así como cambios en la fonação, articulación y resonancia. Cambio en la fluidez verbal y el reconocimiento, refiriéndose a la prueba de memoria. Se concluye que, a través de la evaluación clínica, fue posible verificar cambios en las estructuras orofaciales, disfagia orofaríngea neurogênica, cambios en la voz y el habla.

Palabras clave: Síndrome de Sjögren; Accidente Cerebrovascular; Fonoaudiología; Deglución; Sistema Estomatognático; Disartria.

Introduction

Despite being one of the most common chronic rheumatic inflammatory diseases, Sjögren's syndrome (SS) is still a little-known condition, with a higher prevalence in females, affecting at a rate of 9:1. Sjögren's syndrome is an autoimmune systemic condition, which generates lymphoplasmacytic infiltration, and consequent dysfunction in several important structures, such as the lacrimal and salivary glands^{1,2}.

Xerophthalmia and xerostomia are the most common symptoms of this condition whose onset is more frequent between 40-50 years of age. Patients may also have musculoskeletal, respiratory, urogenital, skin and psychiatric changes, resulting in muscle fatigue, shortness of breath, dry nasal and vaginal mucosa, vaginal itching, and skin rashes, respectively; in addition to anxiety, depression and personality disorders¹.

SS patients also have vascular disorders and are often diagnosed with vasculitis and venous thrombosis¹. This misdiagnosis can increase the risk of strokes, neurological disorders caused by changes in cerebral blood flow, either by obstruction or rupture of a blood vessel^{3,4}.

In both stroke and SS cases, patients may develop disorders that limit their communication, including changes in orofacial motricity, speech articulation, and voice and language quality. Food intake may also be limited, either by changes in chewing or swallowing^{1,4}.

Speech disorders as a result of a neurological injury can be divided into apraxia of speech and dysarthroponia. Apraxia is the inability to plan an action, movement or sequence of movements of the orofacial structures, which impairs the correct positioning of the structures for spontaneous speech production. In turn, dysarthroponia is characterized by changes in five motor bases: breathing, phonation, resonance, articulation and prosody, thus weakening oral communication due to disturbances in the muscular control of the mechanisms involved in speech production^{5,6}.

Dysarthroponia can also cause swallowing disorder, or dysphagia, which is a speech-language disorder and a major cause of death in stroke patients, which can also be present in SS^{1,4,6}. However, there are only a few studies that describe speech-language pathology findings in SS, or that relate

the communication disorders in patients with both pathologies.

Thus, in order to increase the scientific knowledge and prepare the speech-language pathologist for this specific case, the study aims to describe the speech-language pathology findings of a patient with Sjögren's syndrome after multiple strokes.

Presentation of the clinical case

Procedures

This study was approved by the Research Ethics Committee of the institution, under the process no. 2.2.259.004. The legal guardian signed an Informed Consent Form agreeing with the conduct of the research and the dissemination of its results.

This study included a single patient diagnosed with Sjögren's syndrome and recurrent ischemic stroke, with involvement of the frontal, temporal and right parietal lobes, as well as the right hemisphere of the cerebellum. The patient was a 62-year-old woman, who had completed high school, complained of difficulties in speech, chewing and swallowing and was being treated at the Clinical School of Speech-Language Pathology at a Higher Education Institution.

At first, the clinical evaluation included the application of an anamnesis directed to cases of communication disorders associated with neurological disorders⁷. Then, the speech-language pathology evaluation was performed and included: assessment of the orofacial structures and respective functions, based on the MBGR⁸ protocol; assessment of apraxia of speech⁵; assessment of dysarthroponia⁵; cognitive screening⁹; and investigation of daily life activities¹⁰.

Although a screening was performed to detect signs of aphasia, including the skills associated with oral speech, naming, repetition and understanding, no changes were detected. It should be noted that the absence of evident language difficulties is explained by the cortical lesion in the right hemisphere, and that the patient had no complaints of deficit in this function. Finally, despite the patient not having hearing complaints, a questionnaire developed by the institution for the detection of hearing complaints was applied.

Speech-language pathology findings

The history of the patient with Sjögren's syndrome, which was diagnosed in 2016, was

presented during the anamnesis. The patient used artificial saliva frequently to treat the symptoms of the syndrome. The patient also associated SS with her weight loss, increasingly frequently, with the difficulty of eating, as she choked a lot, and reported odynophagia and chewing problems. In addition, she complained of changes in speech.

The patient had a sad face throughout the interview, avoiding communicating with the researcher, which meant that the main data were reported by her daughter, who detailed the emotional problems of the patient, in addition to motor difficulties, hemiparesis, and impaired balance.

The following changes were identified through structural assessment of oromyofacial functions: dryness of the entire intraoral mucosa; facial and cervical muscle flaccidity, with no reports of pain on palpation; tongue with increased height and width, which prevented a more detailed visualization of the soft palate and posterior pharyngeal wall; ogival/hard deep palate; use of poorly adapted dental prosthesis; reduced mobility of lips, tongues, cheeks, as well as of the opening of the oral cavity. There was no reduction in tactile, thermal and intraoral taste sensitivity.

The following characteristics were observed during the functional evaluation of swallowing, with liquid-fine, honey, nectar, pudding and solid food: inadequate lip sealing, with anterior food leak; predominantly unilateral left-sided chew-

ing, with slow jaw movements and consequent increased oral transit; poor bolus formation; decreased anteriorization and laryngeal elevation, compensatory movement of the head upwards when swallowing; need for fluids to facilitate chewing and swallowing; and signs suggestive of dysphagia, such as: cough, throat clearing, and intraoral and pharyngeal food stasis after swallowing, as noticed through cervical auscultation and the presence of a wet voice.

The signs of dysphagia were present in all food consistencies, except pudding. Since the patient only fed on a single food consistency, as the others required the use of a food thickener, her dysphagia was classified as moderate. The report that the patient also had a history of progressive weight loss in the last six months, without any nutritional monitoring, also corroborated to the clinical characteristics of dysphagia.

The Protocol for the Assessment of Apraxia of Speech⁵, which evaluates both verbal and non-verbal praxis, was used to assess the apraxia of speech. The patient was able to speak all words and sentences, but with unintelligibility in speech, and six phoneme substitutions and distortions, with inconsistency in the production of sounds, resulting in verbal apraxia.

The patient obtained a score of 67 in the assessment of non-verbal praxis skills, which indicates moderate non-verbal apraxia (Figure 1).

| DESCRIPTION OF THE RESPONSE | NUMBER PERFORMED | INDIVIDUAL SCORE | TOTAL SCORE |
|--|------------------|------------------|-------------|
| Correct and immediate answer with no hesitation | 2 | 5 | 10 |
| Correct answer after a few attempts | 6 | 4 | 24 |
| Response with decreased amplitude, suitability and speed of movement | 11 | 3 | 33 |
| Incorrect answer or no answer | 1 | 0 | 0 |
| TOTAL | 20 | - | 67 |

Figure 1. Results of non-verbal apraxia test

In turn, the evaluation of dysarthrophonia found oronasal breathing with pneumophonoarticulatory incoordination, rough vocal quality, low voice intensity, hypernasality, articulatory

imprecision, slow mandibular movements, with inconsistent and irregular movements during tongue movement sequences. The patient was also unable to properly perform the oral diadochokinesis.

The Brief Cognitive Screening Battery⁹ was applied in order to assess memory skills, in which the patient had positive results, except for verbal

fluency and recognition, as shown in Figure 2. The patient was unable to draw a clock.

| SKILL | MAXIMUM SCORE | SCORE OBTAINED | REFERENCE |
|---------------------|---------------|----------------|-----------|
| Naming | 10 | 10 | 9 |
| Incidental memory | 10 | 5 | - |
| Immediate memory | 10 | 8 | 5 |
| Learning | 10 | 9 | 7 |
| Verbal fluency | - | 6 | 13 |
| Delayed memory (M5) | 10 | 6 | 6 |
| Recognition | 10 | 8 | 9 |

Figure 2. Results of the cognitive screening test

Katz Index¹⁰ of Independence in Activities of Daily Living was applied to assess the quality of life based on the following criteria: Patient dependent, independent or requiring assistance. According to the results obtained, the patient required assistance to perform two daily activities - continence and feeding - and had total independence in other activities.

Discussion

Given the lack of SS cases associated with multiple strokes in the scientific literature, the study conducted a detailed speech-language pathology assessment, covering data related to orofacial motricity, swallowing, speech, voice and performance in daily activities.

The study showed changes in most aspects analyzed, related to the assessment of orofacial structures, swallowing, speech and voice, with no impairment in memory and activities of daily living. These results are in line with the literature^{1,3-6}.

The results of the evaluation of the orofacial structures obtained in the study justified the change in the oral phase of chewing, swallowing and speech. Based on the characteristic of xerostomia^{1,2,11}, which causes dryness of the oral mucosa, decrease/absence of saliva and changes in the oral structures, the findings in the observation of the oral cavity are compatible with the Sjögren's syndrome. This study is also in line with the literature as for the expected findings for stroke patients, which reports the impairment in the functioning of the orofacial

and sensory motor-oral structures, both implying changes in chewing, swallowing and speech¹².

The absence of saliva impairs the lubrication and protection of the oral mucosa against physical, chemical and biological agents. In addition, dryness in the lips, tongue and pharynx results in changes in stomatognathic functions. Therefore, the use of fluids and artificial saliva is indispensable for this patient¹². A similar finding is described in the speech-language pathology evaluation of this patient, which identified decreased saliva, dryness of the oral mucosa and constant use of fluids to assist chewing and swallowing.

Consequently, the study describes changes in the physiology of swallowing that are in line with the literature, as they indicate a moderate neurogenic oropharyngeal dysphagia^{4,12}.

Oropharyngeal dysphagia is a common complication due to stroke and is associated with increased mortality and morbidity in these patients. However, the management of this symptom is often very neglected¹³, which means that many patients, such as the patient enrolled in this study, are not properly monitored by a multidisciplinary team, especially a nutritionist, who is essential for dysphagic individuals¹⁴.

Some studies indicate changes related to muscle flaccidity, areflexia, hemiplegia, hemianesthesia and cognitive alterations⁴ in a stroke patient. The patient enrolled in this study showed muscle flaccidity and hemianesthesia on the right side, which may impair the functioning of the stomatognathic system.



Also in accordance with the literature related to speech disorder, the study noticed changes in verbal and non-verbal apraxia, as well as dysarthrophonia, which can be associated with both stroke and SS, since both cases are from neurological disorders, which usually result in changes in communication^{5,6}.

Dysarthrophonia is characterized by disorders in the mechanisms of voice and speech production, which may affect breathing, phonation, resonance, articulation and prosody¹⁵. This study detected the presence of weakness, slowness or incoordination of the laryngeal musculature, hypernasality, changes in breathing, phonation, resonance, articulation and prosody.

Dysarthrophonia is also a predictor of oropharyngeal dysphagia¹⁶, which is another high prevalence change observed in patients after stroke and that was also observed in the patient in this study.

The patient had lower than expected verbal fluency in the analysis of cognitive data by means of a memory screening battery, which identified difficulties in producing words. However, she showed good attention and concentration during the test, as well as throughout the evaluation process.

The verbal fluency test is performed through the production of words within a subcategory, which may be affected by several conditions, such as impairment in the temporal and frontal lobes, causing changes in executive functions, verbal memory processes and cognitive aspects. As the psychomotor slowness of speech may reduce the process of producing lexical items, the psychomotor speed is a significant cognitive factor in the performance of verbal fluency. Thus, this recovery process and the semantic lexicon may be intact, but the psychomotor speed explains the impaired performance observed in this study¹⁷.

It is also possible to note the relationship between the praxis and verbal fluency skills. Praxis changes may decrease the movement of articulators and psychomotor and articulatory speed, which may result in a reduced speed in the production of words, as required in the verbal fluency test. As shown by a study¹⁸, that reported a relationship between apraxia and impaired verbal fluency.

Although some studies^{1,5} report a limitation of patients due to emotional changes and decline in general health, the patient enrolled in this study showed independence in the assessment of activities of daily living. Finally, the anamnesis found a

decline in the patient's general health condition, due to the lack of interest in eating, which is caused by the dysphagia and emotional problems that were affecting the weight loss. However, these findings were not sufficient to prevent the patient from carrying out her daily activities.

Thus, the overall aspects observed in this clinical case stress the need for a speech-language pathology intervention aimed at these patients, as well as the importance of an interdisciplinary monitoring in order to promote a more effective health care and contemplate the multiple skills/needs in stroke and SS patients, and their families.

Final Considerations

The patient with recurrent ischemic stroke and Sjögren's syndrome had changes in the orofacial structures, as well as in swallowing, speech and voice, showing a neurogenic oropharyngeal dysarthrophonia and dysphagia.

Since the literature is limited to this type of case, this study may contribute to the promotion of speech-language pathology assessment in cases of stroke and Sjögren's syndrome, as well as guidance to speech-language pathologists to set the actions in order to attenuate the changes and provide a better quality of life.

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