Speech Language Pathology and Audiology Therapy Implications of Dermatomyositis: Case Report

Implicações Fonoaudiológicas da Dermatomiosite: Relato de Caso

Implicaciones Fonoaudiológicas de aa Dermatomiositis: Reporte de un Caso

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Abstract

The Dermatomyositis is a rare idiopathic inflammatory myopathy, which main symptom is muscle weakness. **Objective:** To describe the speech therapy findings in a case of dermatomyositis. **Description of the history and procedures:** From patient chart analysis, a female, 68 years old, with confirmed diagnosis of dermatomyositis, an interview and a clinical examination were performed, after free and informed consent. **Results:** Restriction in mouth opening, of mobility of the speech organs, difficulty in swallowing in a solid consistency, with episodes of choking, dyspnea, a need for multiple swallowing, burning sensation (lips and tongue), presence of wet voice after swallowing and muscle weakness widespread, indicating the severity of the condition. **Conclusion:** speech therapy performance in patients with dermatomyositis should be as early as possible, considering that changes in mobility and in execution of oral functions, in particular swallowing, can interfere significantly in nutritional condition and in the well-being of affected subjects.

Keywords: Speech Language Pathology and Audiology; Rheumatology; Deglutition disorders; Evaluation

Resumo

A dermatomiosite é uma miopatia inflamatória idiopática rara, cujo principal sintoma é a fraqueza muscular. Objetivo: Descrever os achados fonoaudiológicos em um caso de dermatomiosite. Descrição do histórico e procedimentos: A partir de análise de prontuário de paciente do gênero feminino, 68 anos, com diagnóstico confirmado de dermatomiosite, foi realizada entrevista e avaliação clínica fonoaudiológica, após consentimento livre e esclarecido. Resultados: Limitação na abertura de boca, de mobilidade dos

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órgãos fonoarticulatórios, dificuldade na deglutição para consistência sólida, com episódios de engasgos, dispneia, necessidade de múltiplas deglutições, ardência (lábios e língua), presença de voz molhada após a deglutição e fraqueza muscular generalizada, indicando gravidade do quadro. Conclusão: a atuação fonoaudiológica em pacientes com dermatomiosite deve ser o mais precoce possível, tendo em vista que alterações na mobilidade e na execução das funções orais, em especial, da deglutição, podem interferir significativamente na condição nutricional e de bem-estar dos sujeitos acometidos.

Palavras-Chave: Fonoaudiologia; Reumatologia; Transtornos da Deglutição; Avaliação.

Resumen

La dermatomiositis es una miopatía inflamatoria idiopática rara, cuyo síntoma principal es la debilidad muscular. Objetivo: Describir los hallazgos fonoaudiologicos en un caso de la dermatomiositis. Descripción de la historia y procedimientos: Desde la análisis de registros médicos de una paciente del género femenino, 68 años, con diagnóstico confirmado de dermatomiositis, se llevó a cabo entrevista y evaluación clínica fonoaudiológica, después de consentimiento libre e informado. Resultados: Restricción en la apertura de la boca, en la movilidad de los órganos fonoarticulatórios, dificultad para tragar consistencia sólida, con episodios de asfixia, falta de aire, (la) necesidad de múltiples degluciones, ardor (labios y lengua), (la) presencia de (la) voz húmeda después de tragar y debilidad muscular generalizada, lo que indica la gravedad de la condición. Conclusión: la actuación fonoaudiología con pacientes con dermatomiositis debe ser lo más precoz posible, teniendo en cuenta que los cambios en la movilidad y en la ejecución de las funciones orales, en particular, la deglución, pueden interferir significativamente en el estado nutricional y en el bienestar de los sujetos afectados.

Palabras clave: Fonoaudiología; Reumatología; Trastornos de deglutición; Evaluación.

Introduction

A Dermatomyositis (DM) is part of the idiopathic inflammatory myopathies (IIM), a heterogeneous group of chronic systemic autoimmune myopathies associated with high morbidity and functional disability. It presents epidemiological, histological, immunohistochemical, pathological, clinical and evolutionary characteristics[1]. It is a rare disease, characterized by progressive proximal muscle weakness and typical rash [2].

It occurs more frequently in women, with low prevalence, and the diagnosis confirmation is performed by dermatologic clinical evaluation, serum muscle enzymes dosage, electromyography and biopsies (skin and muscle). As common complaints, besides the report of muscle weakness, there is dysphagia, both associated with skin lesions[3].

The DM presents geographically variable incidence, with about one case per 100,000 inhabitants. Its physiopathology is unknown and among the most likely mechanisms, it is suggested muscle injury mediated by viruses or microvascular insult by muscle autoantigens[4].

The main symptom is muscle weakness that leads to diagnostic suspicion, revealing itself as symmetrical, bilateral, involving the four limbs, and the dermatological changes characteristics are the heliotrope and the Gottron's papules and rash in the areas of photoexposure[5]. In severe cases, the weakness can be generalized [6], and some patients may present alterations of respiratory muscles and dysphagia[7]. In these cases, dysphagia is high and results from decreased pharyngeal contraction[8], changes of esophageal peristalsis, weakness of the tongue[9,10], alteration of the motility of the lower esophagus and delay in the gastric emptying[11].

When associated with neoplasia, the DM can begin more acute, with early and severe skin manifestations. Muscle changes may be late, but with rapid and progressive evolution. The relationship with neoplasia is more common in the DM (15%) than in polymyositis (9%), and it is more frequent in adults over 60 years[12].

The description of Speech Language Pathology and Audiology findings is found to be important due to the scarcity of data in the literature and because it is a rare condition. From the foregoing, the aim of this study was to describe the Speech Language Pathology and Audiology findings in a case diagnosed with dermatomyositis.[3]

Description of the clinical history and procedures

White, female, 68 years old patient, accompanied at the Rheumatology Service of the University Hospital (UH) of the Universidade Federal de Sergipe [Federal University of Sergipe (UFS)] since 2006. The first symptoms revealed edema, eczema on the neck region, upper torso and abdomen and was medicated, guided and often accompanied by a rheumatologist. The patient presented no satisfactory response to the treatments instituted.

In mid-2012, the symptoms got worse and the patient reported hair loss, muscle weakness and headache. In late 2013, she reported intense sporadic subjective dizziness in orthostasis, as well as the abuse of choking and weight loss (14 kg) due to the difficulty in feeding function.

In March 2014, she presented a diagnostic hypothesis of pulmonary nodules in diagnostic elucidation phase, introducing, in this occasion, dyspnea complaint, cough, chest pain, intolerance to physical exercises and respiratory failure. The diagnosis of DM was performed according to clinical and laboratory criteria adopted by the UH Rheumatology clinic at UFS.

For the performance of an interview and clinical evaluation in orofacial motricity, we used the adapted MBGR Protocol13. The evaluation of swallowing is based on the Speech Language Pathology and Audiology protocol of risk assessment for dysphagia14 (PARD), consisting of liquid, pasty and solid food swallowing test.

During the Speech Language Pathology and Audiology initial interview, the patient showed difficulty in swallowing solid consistency, with episodes of choking and the feeling of "food stuck in the throat", with complaints of breathlessness and the need for multiple swallows, as well as burning sensation in the mouth, especially in the tongue, difficulty of opening the mouth and generalized muscle weakness.

The Speech Language Pathology and Audiology clinical examination revealed the face and scalp with papular erythema and eyelid edema. Straight facial type, with normally closed lips position with the presence of saliva accumulation; intraoral mucosa wound; strength and mobility of lips and reduced tongue; insufficient elevation of the soft palate; tongue with white spots, habitual position on the floor of the mouth, hypofunctioning and decreased sensitivity.

She presented a restriction in mouth opening of 24,6 mm - dimension measured by digital caliper (6 "WesternR PRO - with resolution and reproducibility of 0.01 mm); asymmetrical face with reddish spots and the possibility of route of oral feeding.

During the functional evaluation with dietary supply of pasty (yoghurt), liquid (water) and solid (filled wafer) consistencies, it was observed inefficient chewing, with reduced speed, elevation of reduced larynx, inappropriate tongue movement for all consistencies, as well as inadequate bolus propulsion for pasty and solid consistencies. The presence of cough reflex was observed after swallowing, also to solid consistency, and choking for liquid and change in voice quality after swallowing solid and liquid. It was also noted the presence of noisy cervical auscultation after swallowing liquid and solid.

As a speech therapy conduct, the patient was referred to Otolaryngology and Speech Language Pathology and Audiology evaluation.

This work was approved by the Ethics Committee in Research of the University Hospital of UFS, under the number CAAE-278668014.0.0000 (5546). Because the study involved a research with human beings, it has been implemented in accordance and compliance with the ethical principles of autonomy, non-maleficence, beneficence and justice of the Resolution 466/12 of the National Health Council.

Discussion

The DM is a rare disease that presents a manifestation such as proximal muscle weakness [5,6], symptom quoted by the patient at the beginning of the disease.

This condition can affect anyone at any age, but the peak of incidence occurs between the fifth and the sixth decades of life. Women are affected twice as often as men[3,4], corroborating the results of this study, given that the patient has currently 68 years old and is accompanied for eight years in the rheumatology clinic of the University Hospital of UFS.

The relationship with neoplasia is more common in the cases of DM, being substantially higher than in other inflammatory myositis[12]. It is a more frequent case in adults, especially among elders. There is a higher probability of patients over 45 years old with DM to present some kind of neoplasia, being even more common in adults over sixty

years old[12,14]. More malignancies associated are ovarian, lung, pancreas, stomach and colon cancer and non-Hodgkin lymphoma[12].

Speech Language Pathology and Audiology manifestations are scarce in the literature. Studies related to rheumatic autoimmune diseases [16,17,18] revealed restriction in mouth opening, inefficient chewing and dysphagia, but in cases of individuals with systemic sclerosis.

In our case, the patient presented a dimension in the range of mouth opening, with less than 45 millimeters (mm). Values between 45 and 60 mm of amplitude are cited in the literature[19] as ideal for the adult, and the opening less than 40 mm is considered a warning of possible muscle and/or joint problems. This result justifies a speech therapist professional in the staff working with these individuals, because the restriction on opening the mouth can lead to changes in oral functions[19] in oropharyngeal inspection, speech, nutrition and oral hygiene[20].

The DM can be related to gastrointestinal motility dysfunction. In this situation, dysphagia can be found in 10-15% of the patients[12,21].

Regarding the complaint and the findings related to the disorder of swallowing, it may occur due to the myopathy involvement of striated portion of the proximal esophagus and the hypopharynx[8-10]. Changes in the pharyngeal stage of swallowing may be due to the strength loss of pharyngeal contraction, insufficient elevation of the soft palate, as well as the weakness of the tongue[21].

Dysphagia, dysphonia and dyspnea are manifestations cited in the literature [22], also observed in the clinical case described.

In individuals with DM, the esophageal dysphagia may involve both solid (complaint ratified in our study) and liquid consistencies[21]. The impairments of skeletal muscles of the posterior pharyngeal wall and proximal third of the esophagus can lead to high dysphagia, with reflux of food for the upper airway, heartburn, odynophagia and dysphonia[23]. Dysphagia may be present at any time during the disease process, presenting as most common symptoms the sensation of food stuck in the throat, coughing, and difficulty with dry and solid foods[24].

It is known that the presence of dysphagia may predispose to aspiration pneumonia, being considered one of the factors that lead to the death of the affected subjects[25], and may promote weight loss, as quoted by the patient, with loss of fourteen kilos in the course of the disease. Because of the foregoing and the generalized muscle weakness resulting from the dermatomyositis, it is necessary to work together with Nutrition, in order to promote nutritional intake as soon as possible.

Study[26] revealed that, often, rheumatic patients are exposed to several risk factors associated with the significant increase in the occurrence of fungal infection. This hypothesizes that the whitish plaques in the tongue of the studied individual, as well as the aforementioned complaint of burning in such a structure, can be justified in virtue of this possibility.

In addition, the imposition of Speech Therapy is odd for the improvement of living and health conditions, particularly on the evidence of dysphagia conditions. The literature reported benefits in Speech Language Pathology and Audiology to adolescents diagnosed with dermatomyositis[25].

It is noteworthy that facing the Speech Language Pathology and Audiology implications found, the rarity of the condition and the lack of updated references, especially in the Speech Language Pathology and Audiology area, it was deemed appropriate to disclose the results of the clinical evaluation of patients with DM, aiming at awakening interdisciplinarity with the Rheumatology.

The establishment of interdisciplinary teams (rheumatologist, Otolaryngology specialist, pulmonologist and neurologist, speech therapist, nutritionist, psychologist, physiotherapist, etc.) and early intervention could minimize the consequences arising from the disease, which currently is severely compromised because of widespread muscle weakness, as ratified by literature[6].

Final Considerations

This case report allows it to evidence Speech Language Pathology and Audiology demonstrations in the present case, as limitation in mouth opening, mobility of the speech organs, difficulty in swallowing for solid and liquid consistency with episodes of choking, dyspnea, need for multiple swallowing, burning (lips and tongue), presence of wet voice after swallowing and general muscle weakness. It is necessary to consider conducting further research with the description of cases or larger sample groups, so that it can demonstrate

the occurrence of Speech Language Pathology and Audiology disorders, justifying the intervention of Speech Language Pathology and Audiology in the context of dermatomyositis. This study may contribute to the development of systematic Speech Language Pathology and Audiology approach for the recovery of the normal dynamics of oral functions and interdisciplinary approach in this condition.

In addition, the Speech Language Pathology and Audiology intervention in patients with dermatomyositis should be established as soon as possible, given that changes in mobility and execution of oral functions, in particular, swallowing, can interfere significantly in the nutritional status and well-being of individuals affected by this condition.

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