Dysphonia as the primary complaint in a case of myasthenia gravis: diagnosis and speech therapy

Disfonia como principal queixa num quadro de miastenia grave: diagnóstico e fonerapia

ABSTRACT

Myasthenia gravis is an autoimmune disease, manifested by progressive muscular weakness and fatigue. There are frequent ocular and bulbar symptoms, among them, dysphonia. This article reports a case of dysphonia that contributed to the diagnosis of myasthenia gravis through a speech evaluation. The patient sought speech therapy with the ENT diagnosis of presbyphonia. The perceptual voice assessment and acoustic analysis pointed out respiration, glottal voice source and resonance affections. Considering that some of the data obtained from anamnesis and vocal assessments were not directly related to presbyphonia, the speech therapist discussed the case with the physician and they both concluded it was necessary to refer the patient to a neurological evaluation. The neurologist then raised the diagnostic hypotheses of myasthenia gravis and requested further examinations. The patient underwent speech therapy and drug treatment. A vocal reassessment, which occurred two months after the initial treatment, showed improvement in voice quality, with great impact on quality of life. This article shows the importance of detailed clinical speech evaluation and participation of a speech therapist in an interdisciplinary team.

RESUMO

Miastenia grave é uma doença autoimune que se manifesta por fraqueza e fadiga muscular progressivas. São frequentes os sintomas oculares e bulbares, dentre eles, a disfonia. Este artigo relata um caso de disfonia, cuja avaliação fonoaudiológica contribuiu com o diagnóstico de miastenia grave e seu tratamento. A paciente em questão procurou o atendimento fonoaudiológico com diagnóstico otorrinolaringológico de presbifonia. A avaliação perceptivo-auditiva e acústica da voz identificou alterações em respiração, fonte glótica e filtro/ressonância. Como alguns dados obtidos com a anamnese e observados nas provas vocais não se relacionavam diretamente com a presença de presbifonia, houve a necessidade de discussão do caso com o médico, quando ambos concluíram a necessidade de encaminhar a paciente para avaliação neurológica. O neurologista consultado levantou a hipótese diagnóstica de miastenia grave e solicitou exames. A paciente seguiu em acompanhamento fonoaudiológico e medicamentoso. Na reavaliação vocal, ocorrida cerca de dois meses após o início do tratamento, foi constatada melhora na qualidade vocal, com grande impacto na qualidade de vida. Este trabalho evidencia a importância da avaliação fonoaudiológica detalhada e da participação do fonoaudiólogo na equipe interdisciplinar.
INTRODUCTION

Myasthenia gravis is an autoimmune disease caused by a decrease of acetylcholine receptor and loss of postsynaptic receptors[1,2]. It is manifested by progressive weakness and fatigue of skeletal muscles[3] and affects between 5 and 15 individuals per 100,000, mostly being 30% elderly[5]. The prevalence is higher in women, with a ratio of approximately 2:1[4,5]. There are few clinical and epidemiological data on the occurrence of this disease in Brazil[5].

Myasthenia gravis can affect one or more muscle groups, and the involvement of the levator of upper eyelid and extraocular muscles is quite common; therefore, symptoms such as ptosis and diplopia may be frequent[3,6]. Bulbar symptoms such as dysphonia, dysphagia, weak mastication, dysarthria, and weakness in facial muscles[1] are also common, especially when the disease is manifested after the age of 65[2].

The difficulties with speech and swallowing appear as initial symptoms in more than 27% of cases[4]. A study of 40 patients with myasthenia gravis found dysphonia as the first symptom in 6% of patients and dysphonia appeared with the progression of the disease in 60% of cases[4]. The vocal affections commonly found in these patients are hypernasality, difficulty in sustaining pitch, vocal fatigue, intermittent aphasis, stridor, roughness, glottal incompetence, and changes in vocal quality[2,3,6]. Articulatory imprecision and verbal fluency alterations can also be observed together with dysphonia[2].

In a recent Brazilian study[5], difficulties in swallowing were observed and voice disorders were more common in the age group between 12 and 50 years; in the group of individuals with more than 50 years, the presence of dysphonia was as frequent as diplopia.

The diagnosis of myasthenia gravis is determined from the clinical signs and can be confirmed by electromyography in 95% of cases[3]. The report of the patient is considered essential for the diagnosis. Dysphonia is often not associated with the disease during the ear, nose, and throat (ENT) diagnosis[6].

If untreated, myasthenia gravis can impair the individual due to the damage that can occur in motor endings[6]. The treatment is usually done through medications[3] or, in some cases, through surgery with significant improvement in symptoms[6] and even complete remission in a few patients[7]. Few data are available regarding the referral to associated treatments such as physiotherapy and speech therapy. In a literature review, physiotherapy proved to be indispensable[7] with improvement in motor performance, fatigue reduction, and prevention of generalized respiratory complications. Speech therapy is indicated due to swallowing/mastication alterations, dysphonia, and speech difficulties, the improvement of which contributes to raising the quality of life of patients[8,9].

The purpose of this paper was to report a case and discuss the treatment results of an elderly individual whose speech evaluation was critical to the referral for neurological assessment and, thereafter, to establish the diagnosis of myasthenia gravis and appropriate conducts.

CLINICAL CASE

This study was approved by the Ethics Committee of the institution (236/10). The patient agreed with the realization and dissemination of this research and its results and signed the consent form.

The 84-year-old patient was referred by an ENT doctor for clinical assessment because the diagnosis was not possible via laryngoscopy.

During anamnesis held in our school clinic, the patient mentioned that her voice felt “horrible and nasty” and that she could not speak loudly and had difficulty being understood by others. The main vocal aspects indicated by the patient were vocal fatigue, hoarseness, voice failure, vocal tremor, and the need to make an effort to speak. She also complained of constant coughing and gagging. The patient reported daily use of medications prescribed by the physician due to hypothyroidism, gastroesophageal reflux episodes, diabetes, hypertension, and peripheral vestibular dysfunction.

Upon completion of anamnesis, perceptual voice assessment and acoustic analysis, qualitative analysis of the spectrum, quality of life related to voice, orofacial motricity, speech, and oral functions of mastication and swallowing were carried out. Alterations on the three vocal production systems were observed: breathing, glottal source, and filter — resonance and articulation of speech sounds. During the assessment tasks, the patient had great difficulty coordinating breathing and speech, her pitch was lower, she presented aspirated vocal attack and loudness, and her fundamental frequency (f0) were decreased. The vocal range was reduced, as well as the maximum phonation duration and vocal resistance. With the implementation of the CAPE-V protocol[10,11], dysphonia was considered moderate and intense, with asthenia and roughness as the most affected aspects, followed by instability and breathiness. The patient presented gagging and vocal fatigue with rapid vocal deterioration in spontaneous speech leading to aphasis. Important articulatory imprecision was observed mainly in isolated phonemes, and difficulty coordinating speech organs and sustaining fricatives were observed. The mean fundamental frequency was 107.52 Hz (Table 1).

The acoustic spectrographic analysis was performed using the Spectrogram software and the fundamental frequency was measured with the Praat software. The tracing showed partially visible harmonics, presence of noise between harmonics, substitution of harmonics by noise, interruptions, bifurcations, graphical irregularity, and reduced series of harmonics (Figure 1).

It was observed that vocal alterations produced significant negative impact on the quality of life of the patient, limiting her participation in social and family events. Her score on the Vocal Health Quality questionnaire (VHQ)[12] was 25 on a scale of 0 to 100, where the maximum value represents better quality of life related to voice.

After speech assessment, the patient was referred to laryngological evaluation at the otolaryngology service of a reference hospital — part of routine procedures. The diagnosis was
presbyphonia. However, as speech assessment had raised issues that were not compatible only with this diagnosis — such as important vocal fatigue, dysarthria, and dysphagia — the multidisciplinary team discussed the case and decided to refer the patient for neurological evaluation at the same hospital. The neurologist who evaluated the patient considered the clinical signs sufficiently suggestive for the diagnosis hypothesis of myasthenia gravis and, therefore, initiated drug treatment in parallel to further examination. Such diagnosis was subsequently confirmed.

The patient underwent weekly speech therapy program with an emphasis on techniques for glottal closure, articulation of speech sounds, and improvement of vocal intensity and resistance. To prevent the patient to have vocal fatigue, the selected exercises were distributed at each session so that they would intermix glottal source techniques with others that would not involve vocal production, such as respiration and articulation of voiceless sounds. Exercises were interrupted at the slightest sign of loss of vocal quality/vocal fatigue. The glottal source exercises were not suitable for domestic practice at the beginning of treatment. As vocal fatigue was one of the symptoms that rapidly improved due to the medication, this control gradually became less necessary.

After 2 months of speech therapy and drug use, a new clinical evaluation of voice and speech was carried out when the improvement on the initially identified affections became apparent. Roughness passed to moderate and asthenia and instability passed to mild and moderate, respectively. Only the presence of breathiness became more apparent, which is compatible with increased vocal intensity presented by the patient in the presence of incomplete glottal closure. Furthermore, maximum phonation time and vocal resistance improved and vocal fatigue decreased; also, the fundamental frequency was higher (Table 1).

In the spectrogram, the bifurcations and interruptions initially observed were no longer present, and better definition and greater regularity of harmonics were also observed. Only the

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<th>Table 1. Speech assessment aspects pre- and postspeech therapy</th>
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<td><strong>Aspects</strong></td>
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<td>Vocal quality, CAPE-V protocol</td>
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<td>Respiratory-speech coordination</td>
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<td>Maximum phonatory duration</td>
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<td>Mean fundamental frequency</td>
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Legends: VHQ, vocal health quality.
presence of noise between the harmonics and replacement of harmonics by noise at frequencies above 2,000 Hz increased, which is consistent with the increased presence of breathiness previously reported (Figure 2).

However, the most significant improvement was regarding quality of life related to voice, expressed through the reports of the patient and also through the VHQ index, which increased from 25 to 60.

It is important to indicate that, during speech therapy, auditory evaluation was performed and the patient was diagnosed with presbycusis. This fact may explain her difficulty in perceiving some aspects of vocal production.

**DISCUSSION**

The presentation of this case evidences how speech assessment was important for the multidisciplinary team to correctly diagnose and quickly start treatment, which would not be possible with the ENT diagnosis alone.

Vocal aspects resulting from myasthenia gravis are related to fatigue and weakness of adductor muscles and tension of the vocal folds. Dysphagia may be associated with disorders of pharynx and esophagus muscles, and dysarthria and articulatory imprecision can be justified by the weakness of the muscles of the tongue and palate(2,3). Valuing both clinical aspects and imprecision can be justified by the weakness of the muscles pharynx and esophagus muscles, and dysarthria and articulatory vocal folds. Dysphagia may be associated with disorders of the tongue and palate(2,3).

Diagnosis and early agile multidisciplinary treatment enable improvement of signs and symptoms of myasthenia gravis and, in many cases, even total remission. For patients who present dysphagia associated with this disease, these factors are essential to prevent aspiration, dehydration, and malnutrition(14).

In this case, drug treatment led to the rapid decrease of vocal symptoms and fatigue during speech and provided full participation of the patient in therapy. Furthermore, vocal fatigue was considered in treatment planning, especially with regard to the distribution of exercises during the session and therapeutic attention for the least sign of its presence.

The results exceeded the expectations of the patient and professionals involved, mainly because of the presence of other associated diseases such as gastrointestinal reflux, presbycusis, and hypothyroidism, the latter being very common in cases of myasthenia gravis(15). Practitioners should give special attention to the described aspects in the analysis for differential diagnosis.

**FINAL COMMENTS**

This case study reiterates the importance of clinical assessment for the diagnosis of patients with voice and speech alterations. The combination of laryngoscopy, clinical signs, aspects reported by the patient, and interdisciplinary dialogue allowed the complete understanding of the framework and helped define the diagnosis with appropriate referral and determination of appropriate care. The results obtained in relation to both medical treatment and speech therapy were highly positive and reinforced the relevance of the latter not only to prevent complications but to provide better quality of life.

* NKN and LLM were in charge of the project and study design, as well as the general orientation as to the stages of execution and elaboration of the manuscript; MSZ supervised data collection and collaborated with data analysis and with the writing of the manuscript; TSF and HRF were responsible for data collection and formatting.

**REFERENCES**


