

Dysphonia as the main symptom in Myasthenia Gravis, a case report

Rafael Albertino, MD; Larissa Brasil, MD; Augusto Vabo, MD; Fernanda Magela, MD;
Oswaldo Júnior, MD; Flávio da Silva, MD; Marcelo Vieira, MD;
Department of Otorhinolaryngology, Hospital das Forças Armadas

ABSTRACT

Myasthenia Gravis (MG) is a rare autoimmune neuromuscular disorder that affects the postsynaptic membrane at the neuromuscular junction, leading to fluctuating, fatigable muscle weakness. Although classically associated with ocular symptoms, bulbar involvement particularly in elderly patients may be the initial or predominant presentation, complicating diagnosis.

The case presents a 77-year-old male with multiple comorbidities, admitted with progressive dysphagia, dysphonia, and lower limb weakness. Videolaryngoscopy revealed initial unilateral and subsequent bilateral vocal cord paralysis. Given the fluctuating pattern of symptoms and worsening fatigue by the end of the day, MG was suspected. Empirical treatment with pyridostigmine resulted in marked clinical improvement, supporting the diagnosis.

This case emphasizes the importance of considering MG in elderly patients with isolated or atypical otorhinolaryngological symptoms. Early recognition and treatment can significantly improve outcomes and prevent serious complications, such as aspiration and respiratory failure.



CONTACT

Rafael Saba Albertino
Hospital das Forças Armadas
St. Sudoeste – Cruzeiro
Brasília – Federal District - Brazil
otorrinolaringologiahfa@gmail.com

INTRODUCTION

Myasthenia gravis is a rare and underdiagnosed autoimmune neurological disease. It affects the postsynaptic portion of the neuromuscular junction, interfering with the musculoskeletal system. It primarily manifests in women between 20 and 30 years of age and men between 60 and 80 years of age.

The main symptom of the condition is fatigable weakness, which can affect any striated muscle. Involvement of the extraocular, facial, and oropharyngeal muscles is most often observed. Therefore, it can lead to significant morbidity or even mortality, which is generally preventable with timely diagnosis and appropriate treatment, considering the significant decrease in current mortality rates.

CASE REPORT

GOM, a 77-year-old male, with systemic arterial hypertension, heart failure with preserved ejection fraction, dyslipidemia, atrophic gastritis, and myocardial revascularization in 2024. Admitted on October 1, 2024, to the emergency, presenting with dyspnea on moderate exertion associated with a secretory cough, dysphonia, and progressive dysphagia that began 6 months earlier, initially with solids and later with liquids.

During his stay in the Intensive Care Unit, a consult from the Otorhinolaryngology department was requested on October 3, 2024, and videolaryngoscopy was performed. A diagnosis of unilateral vocal cord paralysis (left vocal cord in adduction and right vocal cord in a paramedian position) was made. The patient presented with decreased glottic clearance, fatigue, and difficulty walking due to progressive loss of strength in the lower limbs, worsening mainly at the end of the day. He also had dyspnea with minimal exertion. Videolaryngoscopy on October 14, 2024, revealed bilateral paramedian vocal cord paralysis, with tracheal lumen involvement.

Diagnostic hypotheses included myasthenia gravis, degenerative neuromuscular diseases, and motor neuron lesions. Empirical treatment with pyridostigmine was initiated, with significant clinical improvement, including removal of the nasogastric tube and initiation of exclusive oral nutrition, as well as significant improvement in phonation. Videolaryngoscopy on November 1, 2024, revealed laryngeal anatomical and functional improvement, with present mobility and vocal folds with mild hypotonia.

IMAGE

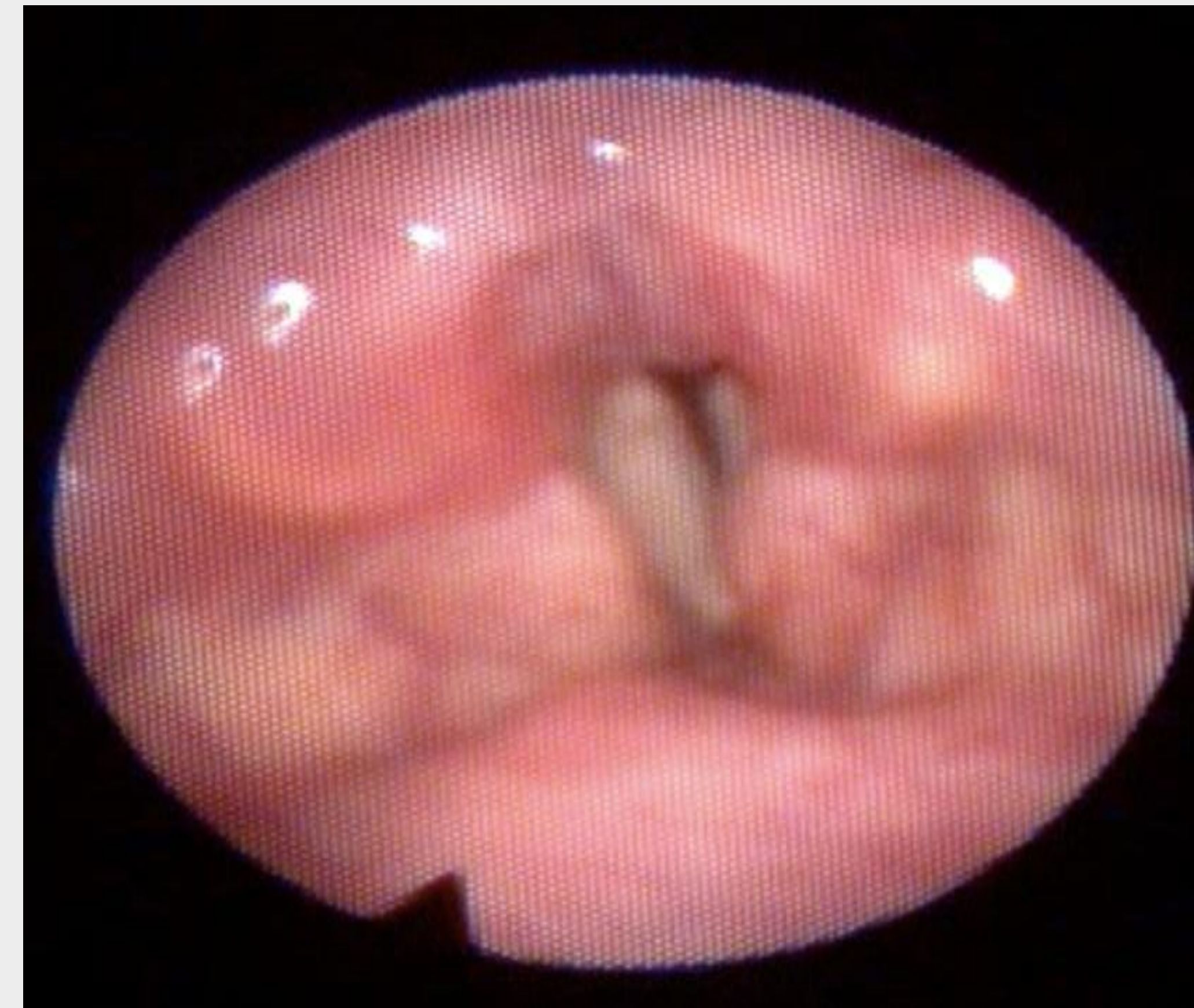


Image 1. Left vocal cord in adduction and right vocal cord in a paramedian position.

DISCUSSION

Despite its low incidence, Myasthenia Gravis is the main pathology affecting the neuromuscular junction. Precipitating factors are not identified early in the disease in most patients. However, there are limited reports of a link with viral infectious diseases, as well as a causal association with emotional stress, surgery, physical exercise, trauma, antibiotic use, menstrual cycle, and pregnancy.

Skeletal muscles innervated by the cranial nerves are especially vulnerable, and their involvement results in extraocular and facial muscle weakness or bulbar and oropharyngeal weakness. Speech and chewing may be affected, as well as pharyngeal function and swallowing coordination, placing the patient at greater risk of pulmonary aspiration of oral or gastric contents.

Otorhinolaryngological symptoms such as bulbar weakness, dysphagia, dysarthria, and dysphonia account for 15% of initial symptoms, often without prominent ocular symptoms. This type of presentation is more common in the elderly. Regarding pharyngolaryngeal manifestations, dysarthria was the most common primary symptom, followed by dysphagia and dysphonia.

Symptoms fluctuate, worsening as the day progresses, especially after prolonged use of the affected muscles. Furthermore, the clinical course may be marked by periods of exacerbation and remission.

Based on a clinical picture compatible with myasthenia gravis, AChR-Ab and MuSK-Ab tests are recommended, with sufficient positive results; if negative, electrodiagnostic testing is the next step. When the clinical picture is highly suggestive but none of the tests confirms it, the response to pyridostigmine may play a role in supporting the diagnosis. This is especially true in ocular myasthenia gravis or when more accurate antibody tests are not available.

The condition is treated symptomatically with pyridostigmine, an acetylcholinesterase inhibitor at the neuromuscular junction, increasing the availability of acetylcholine in the synaptic cleft. It therefore functions as a palliative treatment, aiming to compensate for the reduced number of functional acetylcholine receptors with increased availability of acetylcholine in the synaptic cleft. The drug has few serious side effects and, if effective, acts quickly.

An imminent myasthenic crisis requires hospitalization and careful monitoring of respiratory and bulbar functions. Admission should be to a facility with the possibility of transfer to the intensive care unit if the patient progresses to an acute crisis.

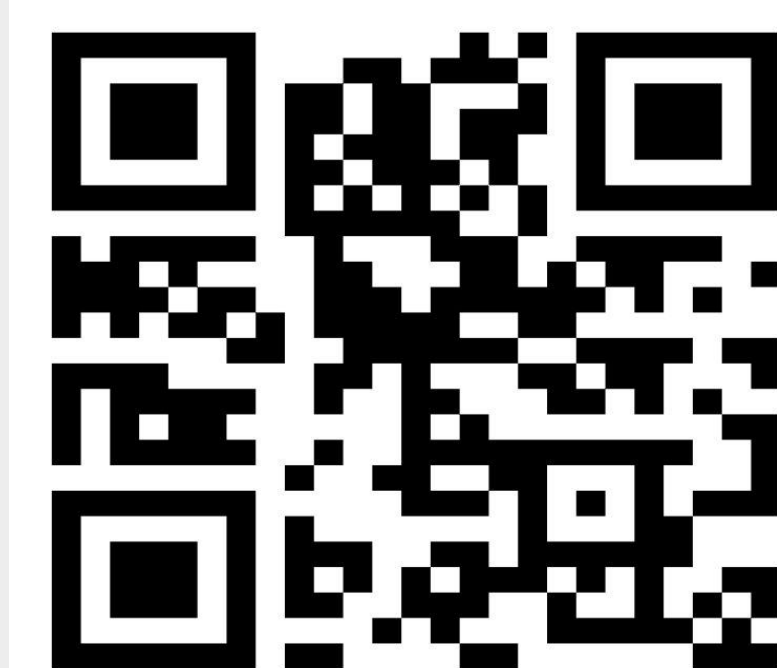
CONCLUSION

Given the negative impact of this condition on patients' quality of life, it is crucial to suspect it during clinical investigations of patients with otorhinolaryngological signs and symptoms.

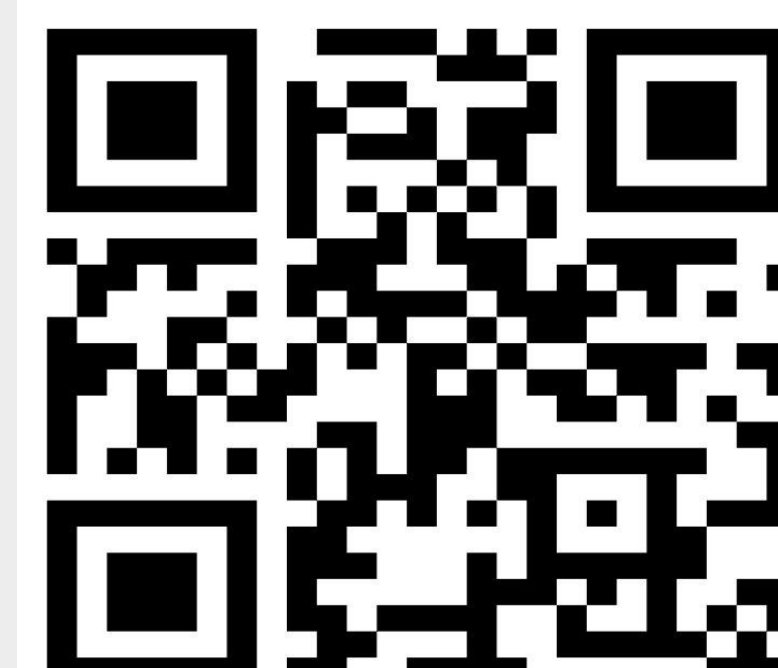
The pathophysiology of Myasthenia Gravis has gradually been clarified, and therapies have been developed to alleviate clinical manifestations and slow disease progression. Early diagnosis of otorhinolaryngological complaints is essential for the appropriate management of Myasthenia Gravis, preventing complications inherent to the natural progression of the disease and reducing systemic involvement, which has high complication rates.

REFERENCES

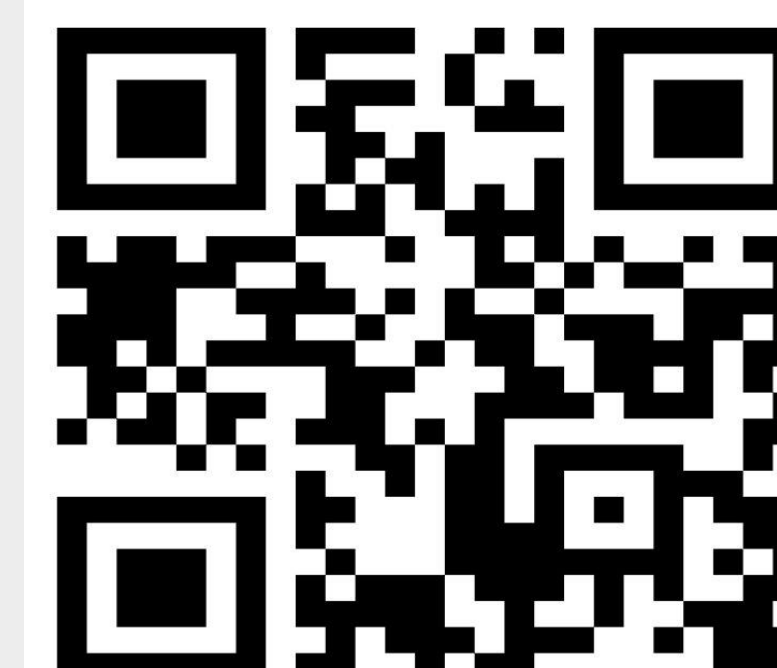
- Andrade N. "Manifestações Otorrinolaringológicas da Miastenia Gravis: Relato de Caso." *Revista Científica Hospital Santa Izabel*. 2021; v.5, n.1:31-34
- Sharp H. R., Degrip A., Mitchell D. B., & Heller A. "Bulbar presentations of myasthenia gravis in the elderly patient." *The Journal of Laryngology & Otology*. 2001;115(1):1-3.
- Inoue M., Kojima Y., Kinboshi M., Nakagawa T., Kanda M., Shibasaki H. "A case of myasthenia gravis presenting solely with bulbar palsy unassociated with easy fatigability." *Clinical Neurology*. 2013;53(3):229-234.
- Santos C, Areias C, Neto M, Paupério A, André C, Antunes L. *Paralisia bilateral das cordas vocais: causas e sua abordagem – análise dos últimos 12 anos*. *Revista Portuguesa de Otorrinolaringologia-Cirurgia de Cabeça e Pescoço*. 2024;62(3):253-261
- Nemr NK, Simões-Zenari M, Ferreira T, Fernandes H, Mansur LL. *Disfonia como principal queixa num quadro de miastenia grave: diagnóstico e fonoterapia*. *CoDAS*. 2013;25(3)



Video 1. 30/07/2024



Video 2. 14/10/2024



Video 3. 01/11/2024