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Case Report

Dysphagia as a Main Symptom of Myasthenia Gravis in a Middle-aged Adult Male: A Case Report

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ABSTRACT

Background: Myasthenia gravis (MG) is a rare neuromuscular disorder resulting from the destruction of acetylcholine receptors at the neuromuscular junction by IgG antibodies. Although dysphagia is a common symptom in generalized form of MG, it is rarely reported as a sole manifestation of the disease, specifically in younger patients. Herein, we report a middle-aged adult patient with dysphagia as the sole manifestation of MG.

Case Presentation: A 49-year-old male complaining of severe dysphagia underwent an extensive clinical and paraclinical examination. Oropharyngeal dysphagia was confirmed by an experienced speech-language pathologist using the water swallowing test. Unilateral right paresis of the soft palate and vocal fold was confirmed using laryngeal video stroboscopy. Chest computerized tomography (CT) scan, brain Magnetic resonance imaging (MRI), and routine blood, urine, and thyroid tests were normal. In electromyography, slow repetitive nerve stimulation (RNS) showed a decremental response in the right nasalis muscle. The diagnostic neostigmine test with 1.5 mg of intramuscular neostigmine led to significant recovery of laryngeal and pharyngeal motor dysfunction and dysphagia within 12 hours, so the MG diagnosis was confirmed. The patient was treated with plasmapheresis, pyridostigmine, and prednisolone, which followed an improving course and led to better swallowing of solid and liquid foods.

Conclusion: MG should be considered as a diagnosis in middle-aged adults with complaints of dysphagia.

Introduction

Dysphagia is a condition that leads to problems in swallowing and occurs as an important symptom of various diseases and disorders. The prevalence of dysphagia has been reported to range from 7% to 22% among people over the age of 50 years. Old age, gastroesophageal reflux disorder (GERD), head and neck trauma and malignancies, and cerebrovascular attacks (CVA) as well as neurodegenerative conditions such as Parkinson's disease, multiple sclerosis, and myasthenia gravis (MG) are risk factors that may lead to dysphagia [1].

In myasthenia gravis, an uncommon autoimmune disease, the neuromuscular junction's acetylcholine receptors are attacked by IgG antibodies, which leads to muscle contraction problems and weakness [2]. The weakness is usually ocular, bulbar, or seen in proximal muscle groups and the neck, but respiratory muscles may also be affected in some patients [3]. About 85% of patients with the generalized form of MG have antibodies to the acetylcholine receptor [2]. Thus, a positive result of these antibodies and other antibodies such as anti-striated muscle or anti-muscle

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specific kinase (MuSK antibody) are diagnostic for the disease [1]. The first line of treatment for MG is oral anticholinesterase such as pyridostigmine. Prednisone, azathioprine, or mycophenolate mofetil are used for immunosuppression. Plasmapheresis and intravenous immune globulin therapy are performed for acute exacerbations [2].

Dysphagia is reported in 15%-40% of cases of the generalized form of the disease [1]; however, dysphagia is the presenting symptom in only 6% of patients with MG. Dysphagia as the sole manifestation of the disease is rare, specifically in the younger patients [4, 5], and requires a multidisciplinary approach for examination and management [6]. In the present article, we report a previously reported case of a younger patient with dysphagia as the sole manifestation of myasthenia gravis.

Case Report

Participant

The patient was a 49-year-old man whose chief complaint was a progressive dysphagia for solid, semi-solid, and liquid foods from six days prior to his hospitalization, at which time, the dysphagia became severe. There were no symptoms of odynophagia, diplopia, fatigue, muscle weakness, shortness of breath, or speech problems. Symptoms did not fluctuate during the day. Personal history of the disease and family history of neurological and autoimmune diseases were also negative.

Clinical Findings

The vital signs and general physical examination were normal. The patient counted to 32 in the single breath count test (SBCT). Examination of the cranial nerves showed that his pupils were mid-size and reacted to light. Visual acuity and eye movements were normal. The patient did not have ptosis. His facial muscles were symmetrical and normal, and his tongue movements were normal. Examination of the locomotor system did not show any muscular atrophy of the limbs. The strength of the flexor and extensor muscles of the neck was normal. The strength of the muscles of the upper and lower limbs was 5/5. Deep tendon reflexes were normal. Bilateral plantar reflexes were downward. There was no sensory disturbance, and other neurological examinations were normal.

A moderate hypernasal speech, nasal regurgitation, multiple secondary swallowing efforts, wet voice, and coughing, choking, and frequent throat clearing during and after the water swallowing test were reported in the clinical examination by a speech-language pathologist. The gag reflex was reduced on the right side. The uvula deviated to the left and the surface of the soft palate was lower on the right than on the opposite side. Unilateral paresis of the right palatoglossal and palatopharyngeal folds led to an asymmetrical view (Figure 1). A nasogastric tube was fixed for efficient feeding.

Diagnostic Assessment

In paraclinical examinations, routine blood, urine, and thyroid tests were normal. The rheumatoid factor was negative, and anti-nuclear antibodies (ANA) were in normal range. There was no evidence of hyperplasia or thymoma in the chest CT scan. Magnetic resonance imaging of the brain did not show any pathological lesions. In video laryngeal stroboscopy, no mass or structural anomaly was seen in the hypopharynx or glottis, but in functional analysis, the unilateral abductor's paralysis of the right aryepiglottic fold and true vocal cord was evident (Figure 2).

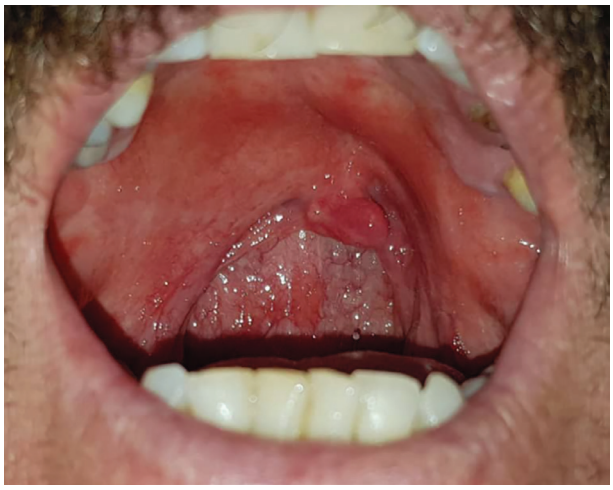

Figure 1: Deviation of uvula to the left side and unilateral right paralysis of the right palatoglossal and palatopharyngeal folds.

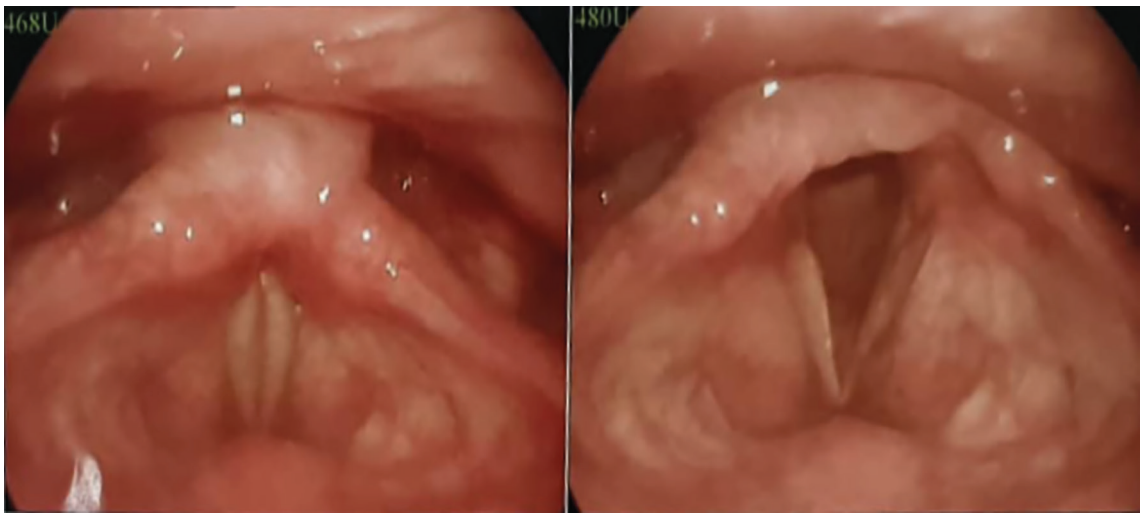

Figure 2: Unilateral abductor's paralysis of the right aryepiglottic and vocal folds.

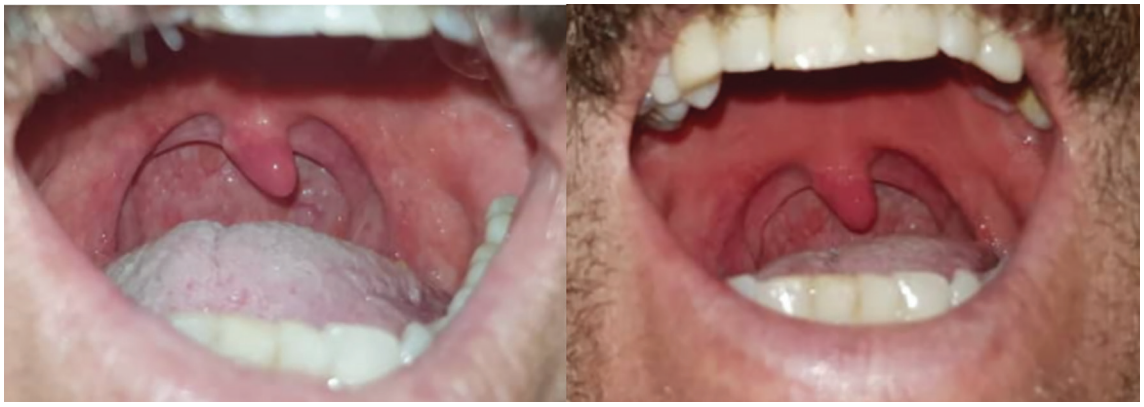

Figure 3: Continuous recovery of unilateral right paralysis after intervention. The uvula gradually returned to the midline. Left photo taken 4 hours and the right 12 hours after intervention.

In electromyography, slow repetitive nerve stimulation (RNS) showed a decremental response in the right nasalis muscle, indicating a post-synaptic neuromuscular junction.

Intervention

Based on the evaluation of the patient's swallowing, first, a nasogastric tube was fixed for efficient feeding. The patient underwent a diagnostic neostigmine test before which 1.5 mg of intramuscular neostigmine and 0.8 mg of subcutaneous atropine were injected five minutes prior to the neostigmine. The palatine folds became significantly symmetrical, and the uvula approached the midline 1 hour after intervention. The patient was monitored every hour for up to 12 hours. The uvula moved to the midline, so the water swallowing test was performed again by a speech-language pathologist (Figure 3). The test results revealed no signs of aspiration, wet voice, any secondary swallowing efforts, or coughing, choking, or frequent throat clearing during or after the water swallowing test. Based on the test results, oral feeding with liquid was started, and the nasogastric tube was discontinued.

Serum levels of acetylcholinesterase antibodies were normal (0.1 nmol/L) and negative for the anti-musk antibody. The patient was first treated with plasmapheresis and subsequently with pyridostigmine and prednisolone, which followed a course of improvement and led to better swallowing of solid and liquid foods. After one month, prednisolone was gradually tapered, and the patient underwent maintenance therapy with 7.5 mg prednisolone every other day and 60 mg pyridostigmine every 6 hours.

Discussion

Patients with dysphagia should be comprehensively evaluated using a multidisciplinary approach for better diagnosis and management. A thorough examination by a speech-language pathologist is necessary to determine the severity of the disorder and to order any compensatory method and behavioral or food modification for safe oral feeding. Additionally, systematical clinical and paraclinical examinations for the etiology and specific underlying diseases or disorders should be considered as soon as possible.

Myasthenia gravis is an uncommon autoimmune disease that usually leads to ocular and bulbar weakness as well a weakness in the proximal muscle groups (most commonly the upper limbs) and neck. In addition, the respiratory, extraocular, pharyngeal, and laryngeal muscles may be involved [3].

Although dysphagia is a common symptom of the generalized form of MG [1], it is rarely seen as a presenting symptom and more rarely as the sole manifestation of the disease, specifically in younger patients [4, 5]. The present case revealed that dysphagia may occur as a sole manifestation symptom of the disease in middle-aged adults. The present report emphasizes that neuromuscular disorders, particularly MG, must be considered among middle-aged adults with complaints of dysphagia and problems swallowing. Early diagnosis and avoidance drugs which potentiate muscular weakness not only reduce unnecessary paraclinical workups, but also reduce morbidity caused by severe malnutrition or aspiration pneumonia. The main focus of the present case is to emphasize MG as a probable diagnosis among patients who refer to speech therapy, neurology, and gastrology clinics with dysphagia as a chief complaint.

Conflict of Interest: None declared.

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