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Case Report

CASE REPORT

Misdiagnosis of myasthenia gravis presenting with tongue and palatal weakness

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Abstract

We discuss the case of an elderly male who presented with a history of dysphagia, dysphonia, palatal weakness and a sensation of tongue swelling, each symptom of varying time duration. Myasthenia gravis may have a variety of presentations that include ocular fatigability, respiratory muscle weakness and bulbar symptoms. The variety of these myasthenic syndromes can serve as a barrier to diagnosis and can often result in delayed or incorrect diagnosis. In this report, we present an atypical presentation of a relatively rare condition.

INTRODUCTION

Myasthenia gravis (MG) is an autoimmune condition in which a lack of immune self-tolerance results in autoantibodies formed against the acetylcholine receptor at the neuromuscular junction of striated muscle. Disease presentation is characterized by fatigability and variability. Typically, the disease results in fluctuating weakness of striated muscle primarily affecting ocular and respiratory muscles. The symptoms of disease are often amplified by the presence of physiological stress such as infection. MG can occasionally present with bulbar symptoms. However, bulbar symptoms as the presenting feature is understood to be a rare presentation of the disease, occurring in just 6% of patients [1].

CASE REPORT

An elderly man presented to the Acute Medical Assessment Unit with dysphagia for both solids and liquids of 6 weeks duration. He also complained of a subjective sensation of tongue swelling for the previous four days, for which he consulted his general practitioner who made the diagnosis of anaphylaxis and treated with epinephrine, without improvement. On assessment in the hospital, dysphonia was noted with a significant nasal quality to his voice on prolonged speech. There was no objective tongue or lip swelling noted by the attending physician. Furthermore, the patient had tongue weakness and fatigability which resulted in the patient being unable to elevate his tongue. There were no objective means by which the patient's tongue fatigability could have been directly measured, this was reported symptomatically as the patient described being less able to physically move his tongue by the end of his meals. The patient's background history was significant for renal cell carcinoma and therapeutic nephrectomy. Other co-morbid conditions included insulin dependent type two Diabetes Mellitus and uncomplicated hypertension. His vital signs were stable and he had no evidence of other focal neurological signs. In the absence of other clinical features, differential diagnoses at this point included Amyotrophic Lateral Sclerosis, Cerebrovascular Accident, CNS Metastasis or Paraneoplastic disease and Cranial Polyneuropathy. His initial serology demonstrated neutrophilia

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and his Chest X-ray illustrated a left sided basal pneumonia. CT Brain showed no evidence of acute infarction or metastasis, only changes of global cortical atrophy with an increased ventricle size, indicative of age related change. Following review by a Consultant Neurologist, a diagnosis of bulbar MG was suspected and anti-Ach receptor antibodies were ordered, which were subsequently positive. CT thorax was clear, with no evidence of thymoma. If clinical suspicion remained high and the patient's anti-Ach antibodies were negative, the use of repetitive nerve stimulation would have been used. The patient was started on Neostigmine therapy with initial dosing of 60 mg twice daily, with a steady increase to 60 mg three times daily was initiated, along with oral Prednisolone 30 mg once daily. Following this, the patient reported gradual symptomatic relief of both dysphonia and dysphagia and was almost symptom free on discharge 1 week after diagnosis. Considering the patient's response to treatment for MG, further testing including MRI was not performed. This would have been of most relevance in further evaluating the patient for the presence of brain metastases.

DISCUSSION

MG continues to be one of the most difficult conditions to diagnose due to the variability of its presentation. The symptoms of which may beguile the assessing physician as the fluctuant weakness of the disease process may be mistaken both for age related change and/or pathology of central neurological origin. Symptom onset peaks in women typically in the third decade, although peak onset in men demonstrates a bimodal distribution in both the third and sixth decades of life [2]. The pathognomonic presenting symptom of ocular weakness in MG is present in only 60% of cases [3] and as such, the diagnosis of MG in the elderly patient is often delayed as this symptom may eventually become present but is often lacking at the time of presentation. The entity of late onset MG has a particularly evident male predominance in patients over 60 years, which often presents with bulbar symptoms [4]. However, bulbar weakness as the sole presenting complaint is present in only 6% of patients. Approximately 28% develop dysphagia or dysarthria at some point throughout their disease [1]. The involvement of the neuromuscular junction can result in a fatiguable flaccid dysarthria with both voice alteration and tongue weakness occuring. Weakness of palatal muscles subsequently results in the development of a nasal quality of speech, which becomes more evident in direct relation to the duration of speech. Tongue weakness may result in dysphagia due to inability to manipulate a food bolus along with incoordination of swallow. In the case of our patient, a barium swallow revealed an incoordinated swallow, resulting in leakage of contrast media into the oesophagus. Atrophy of the tongue may occur if tongue weakness becomes chronic, with the development of accentuated median and lateral furrows [5]. In our case presented, the dysarthria and subjective sensation of a swollen tongue was initially misdiagnosed as anaphylaxis. The delay in a diagnosis of MG can have severe implications for any patient but particularly the elderly population with an increased likelihood of deterioration and subsequent risk of myasthenia crisis [6]. Furthermore, in cases such as this where there is significant dysphagia, the issue arises of whether a gastrostomy tube should be placed. Often there is patient objection to this. In the case of isolated bulbar symptoms a clinician should not rush to implement gastrostomy feeding prior to the consideration of a local and reversible neurological cause for dysphagia such as MG. The avoidance of this invasive and distressing procedure highlights the importance of an expedient clinical diagnosis.

CONCLUSION

A high index of clinical suspicion is required to make the diagnosis of MG. Although bulbar symptoms such as dysphonia and dysphagia are a well-recognized component of the disease, in isolation they may direct the physician down various routes of neurological investigation. The sensation of tongue swelling without objective evidence of any anatomical abnormality, combined with palatal weakness and dysphonia should warrant investigation of a local neurological cause such as MG. Suspicion of MG should prompt neurology referral and subsequent investigations including anti-acetylcholine receptor antibodies and/or anti-muscle specific kinase (anti-MuSK) antibodies and single fibre electromyography (SFEMG) studies for definitive diagnosis.

EDUCATIONAL POINTS

- Myasthenia gravis can rarely present with symptoms of tongue weakness
- Bulbar symptoms such as dysphonia and dysphagia should prompt the consideration of myasthenia gravis
- Isolated bulbar symptoms are more common in an elderly patient population and these symptoms are usually reversible with treatment. As such implementation of gastrostomy feeding should not be the initial action taken by the attending physician.

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CONFLICT OF INTEREST STATEMENT

No conflicts of interest.

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ETHICAL APPROVAL

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CONSENT

Patient consent was received.

GUARANTOR

Megan Marshal.

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