Myasthenia gravis—a rare presentation with tongue atrophy and fasciculation

J. Burch1, C. Warren-Gash1, V. Ingham1, M. Patel1, D. Bennett2, K. R. Chaudhuri2

1Elderly Medicine Directorate, University Hospital Lewisham, London, UK
2Department of Neurology, Kings College Hospital, London, UK

Address correspondence to: J. Burch. Email: jessiburch@doctors.org.uk

Abstract

We report the case of an unusual presentation of myasthenia gravis with tongue atrophy and fasciculation. Myasthenia gravis is an autoimmune condition associated with weakness and fatigability of voluntary muscles. In >50%, the initial symptoms and signs are related to extraocular muscle weakness, such as diplopia or ptosis [Tsung K, Seggev JS. An unusual cause of dysphagia. West J Med 1995; 163: 159–60]. Rarely, it is known to affect bulbar muscles and can lead to dysphagia and respiratory compromise.

Keywords: myasthenia, elderly, fasciculations

Case report

A 76-year-old man presented with a 3-week history of progressive dysarthria and dysphagia, describing extended mastication with difficulty in food bolus manipulation. He was dysarthric with evidence of tongue atrophy and fasciculation. In the absence of other clinical features, the differential diagnosis included amyotrophic lateral sclerosis and paraneoplastic disease.

His initial blood tests, chest X-ray and CT brain scan were normal. Subsequently, an MRI brain showed generalised age-related atrophy. His dysphagia worsened and a videofluoroscopy revealed severe global oropharyngeal dysphagia. Nasogastric feeding was commenced. Specific blood tests including an autoimmune screen, antianoglobulin antibodies and tumour markers were negative, but his acetylcholine receptor antibodies were elevated at 23.4 nmol/l (normal range 0–0.2 nmol/l). Electromyographic studies confirmed a diagnosis of bulbar myasthenia gravis, showing classical decremental response, jitter and blocking. Pyridostigmine and prednisolone treatment were commenced.

His disease was unusually aggressive in its course. He developed complications including steroid-induced psychosis, diabetes requiring insulin, pneumonia requiring ventilation and atrial fibrillation. He also sustained a myocardial infarct resulting in pulmonary oedema and a gastrointestinal bleed due to a gastric ulcer.

Discussion

We present an atypical course of a relatively rare condition. This patient had an atypical phenotype resembling bulbar amyotrophic lateral sclerosis. This case illustrates the importance of acetylcholine receptor antibody tests in cases of unexplained bulbar or lower motor neurone pathology, even when the classical fatigability of myasthenia is absent.

The annual incidence of myasthenia gravis is widely accepted as 2–4 per million [1]. It is twice as common in females with peak age of diagnosis in the third decade. More recently, diagnostic trends indicate that male patients present much later between the sixth and seventh decade [1]. In elderly patients, it is frequently misdiagnosed or has a significant delay in the diagnosis [1, 2]. It has previously been suggested that the average time in diagnosing myasthenia was 4.5 months in the >60 age group, compared to 2.5 months in younger patients [1]. For diagnostic purposes, the acetylcholine receptor antibodies are very specific, and up to 90% will have antibodies, especially in elderly male patients [2].

The survival in elderly patients has been attributed to adequate treatment [3] with anticholinesterases and steroids. It is unusual for patients to present with bulbar weakness and it is the presenting complaint in only 6% [4]. However, 28% report some form of dysphagia or dysarthria at some point during the disease [4]. A case series has indicated that bulbar symptoms, as a presentation of the disease, are more common in the elderly [5]. The voice is affected due to facial
nerve involvement and lip incompetence. Tongue and masticatory weakness are due to hypoglossal and trigeminal nerve involvement respectively [6, 7]. As with our patient, weakness of the oropharyngeal muscles produces dysphagia, and this is a major cause of morbidity in myasthenia [8]. Patients are frequently shown to aspirate silently when a videofluoroscopy is performed [8]. Tongue atrophy has been described in a minority of patients with myasthenia. In a study of 752 patients with myasthenia, only 10 exhibited atrophy, and it was described to present later on in the disease [9], which was not the case with our gentleman. Moreover, the dysphagia and dysphonia that patients with myasthenia develop are almost always refractory to treatment [8], often requiring artificial nutrition [4].

**Conclusion**

Bulbar presentations are well recognised in myasthenia gravis, but tongue atrophy is rare and late in the disease. We have found no previous published cases of associated fasciculation. The atrophy and fasciculation in this particular case were certainly misleading and delayed his eventual diagnosis. Myasthenia needs to be considered, even in the absence of classical symptoms and in all age groups, in order to be diagnosed and thus treated appropriately.

**Key points**

- Myasthenia gravis can present with bulbar symptoms alone.
- It is a diagnostic possibility in patients with weakness, even when fatigability is not a prominent feature.
- Elderly patients are frequently misdiagnosed, which may result in further medical complications.

**References**


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