Myasthenia gravis: diagnosis delayed or missed

Frequently mistaken for more common conditions, myasthenia gravis is an autoimmune disease of neuromuscular function. Exhausting the muscular system, it fluctuates in severity while not disturbing the sensory processes or leading to atrophy. Drs Jonathan P Sherlock, John C McGourty and Jeremy Brown furnish two case reports charting the difficulty in diagnosis and the treatment options.

Swallowing problems are common in older people. Thirty percent of elderly people acutely admitted to hospital suffer from dysphagia. There are many causes ranging from the simple ageing process (presbyphagia) to malignancy and neurological disease. As dysphagia is debilitating, can be life threatening and is often amenable to treatment, accurate diagnosis is essential. We report two cases recently presenting to our department with dysphagia and in whom there had been a significant delay in arriving at the correct diagnosis of myasthenia gravis.

**Case 1 (used with permission)**
An 88-year-old man with a known oesophageal pouch and hiatus hernia was referred for endoscopy in 2003 with several months of increasing dysphagia and weight loss. The endoscopy was normal, but a subsequent barium swallow study showed a large pouch — which had not changed from 2000 — a sliding hiatus hernia, a tortuous oesophagus and tertiary contractions. His symptoms of dysphagia and regurgitation continued, and were thought to be related to the oesophageal pouch. He was assessed by an ENT (ear, nose and throat) consultant and subsequently underwent a stapling procedure in May 2004. There was initially some improvement in his symptoms, although he continued to have difficulty swallowing fluids. On ENT review in October 2004 he complained that his speech had now also become slurred and monotonous. He was referred to the medical department where his main complaints were coughing and choking after food and drink, nasal regurgitation and at times unintelligible speech. On examination he had a Parkinsonian tremor, slight right ptosis without fatigue, weak eye closure, dysarthric speech, weak cough and mild truncal weakness. Investigations undertaken included acetylcholine receptor antibodies, which were raised at 86 (NR 0–5). Electromyogram (EMG) showed increased ‘jitter’, in keeping with a diagnosis of myasthenia gravis. A CT scan of his chest was normal. He was treated with pyridostigmine and increasing doses of prednisolone with resolution of his symptoms. He has recently started therapy for his coincidental Parkinson’s disease.

**Case 2 (used with permission)**
An 83-year-old lady was referred under the ‘two week rule’ in October 2004 for dysphagia. Endoscopy was normal, and a barium swallow study in November noted reduced peristalsis. Her symptoms continued to worsen, but when re-referred to the gastroenterology clinic no diagnosis was made. In January 2005, she also began to complain of diplopia; ophthalmological assessment in May was inconclusive. In June her swallowing difficulty had become so extreme that she was referred by her GP for emergency admission. At presentation she was found to have bilateral ptosis,
reduced down gaze, shortness of breath, slurred speech, reduced cough reflex, inability to swallow, weak neck flexion and proximal muscle weakness. Swallowing assessment showed reduced oral transit time, and decrease in strength of swallow with time. Myasthenia gravis was diagnosed, and pyridostigmine was administered via nasogastric tube. She regained normal function within 10 days on pyridostigmine 30mg three times a day and prednisolone was started. Subsequently anti-acetylcholine receptor antibody assay came back raised at 91, and CT chest showed no thymoma. On follow-up her myasthenic symptoms remain well controlled.

Discussion
Myasthenia gravis (MG) is a readily treatable cause of dysphagia, and untreated has a high morbidity and mortality. Traditionally, it has been considered a disease of the young and middle-aged, but it is increasingly recognised to be undiagnosed in later life\(^3\)\(^-\)\(^6\). Recent UK population studies have reported an overall annual incidence of between 1.1 and 1.8/100,000, but rising to 9.9/100,000 in elderly males\(^5\). The other factor making diagnosis more difficult in the elderly is the frequent co-existence of other conditions. The first case clearly illustrates this, as this patient had both a pharyngeal pouch and early Parkinson’s disease, with the less common diagnosis of MG not being initially considered.

With the ageing of the population and, given that MG can be treated satisfactorily albeit with a risk of side effects, it would seem timely to heighten clinicians’ awareness of the occurrence of MG in later life. Delay in diagnosis unnecessarily places patients at risk of complications, and results in significant morbidity and cost. As these cases demonstrate, even in the very old, treatment is effective and allows patients to maintain a normal lifestyle.

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