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Review Article

Bulbar presentations of myasthenia gravis in the elderly patient

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Abstract
We report on three cases of patients whose primary symptoms of myasthenia gravis were related to the upper aerodigestive tract. Symptoms had been present unrecognized in all patients for up to three years, and one patient subsequently developed a myasthenic crisis. We highlight the clinical features of myasthenia gravis to allow its prompt recognition in patients presenting to the ENT surgeon or physician.

Key words: Myasthenia Gravis; Dysarthria; Dysphagia; Aged

Introduction

Myasthenia gravis is an acquired condition with a reported annual incidence of three to four per million and a prevelance of about four in 100 000, although recent studies suggest its annual incidence may be as high as nine to 10 per million.[1] It is twice as common in women as in men, and its peak incidence is said to be around the age of 30,[2] but Schon *et al.*[1] found the peak age in both sexes was 70 to 80 years and that numbers were greatest in elderly men. Kleiner-Fisman and Kott[3] also state that 20 per cent of patients with the disease are in the seventh decade or older.

The condition is characterized by muscle weakness and fatigueability, particularly of ocular, proximal limb and bulbar muscles. Fifty-three per cent of patients present with ocular muscle weakness causing diplopia and ptosis.[4] In younger patients, up to 70 per cent may have thymic hyperplasia and in the older age group, up to 10 per cent have an associated thymic tumour. A link is also recognized between myasthenia gravis and thyroid disease, pernicious anaemia and rheumatological complaints such as rheumatoid arthritis and systemic lupus erythema tosus.[2]

With myasthenia gravis unlikely to present with predominant bulbar weakness, the ENT surgeon will be presented very rarely with an undiagnosed case. The diagnosis of myasthenia gravis is an important one, as the disease may progress to a myasthenic crisis, where the patient has such severe generalized weakness that they may be unable to maintain ventilation and require intensive care support.

Case report

Case 1

A 68-year-old man presented to the ENT out-patient department with a five-week history of dysphagia predominantly to liquids. This was combined with symptoms of palatal insufficiency and weakness of his tongue, such that he had dysarthria and difficulty in manipulation of a food bolus in his mouth. Direct questioning revealed that over the past 18 months he had had intermittent speech difficulty. He had no visual symptoms. On examination, a right ptosis was evident but ocular movements were normal.

A Tensilon test was positive, and acetylcholine receptor antibody titres high. A computerized tomography (CT) scan of his thorax showed no evident of a thymoma. His symptoms were controlled on 90 mg of pyridostigmine four times daily and 40 mg of prednisolone daily, the doses of which have since been reduced without relapse.

Case 2

A 77-year-old man was referred to the ENT out-patient department with a three-week history of severe dysphagia, dysarthria and nasal regurgitation. On further inquiry, the patient admitted to a three-year history of fluctuating ptosis and diplopia, worse towards the end of the day.

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Examination revealed a left ptosis and weakness of the cervical musculature such that the patient found it difficult at times to maintain his head in an upright posture. Ocular movement was normal.

A Tensilon test proved positive and high serum acetylcholine receptor antibody levels noted. Despite initial treatment with oral anticholinesterases, the patient was admitted to hospital seven days later due to worsening tiredness, swallowing difficulty and respiratory difficulty. He continued to deteriorate despite medical treatment, and 48 hours later required intubation and positive pressure ventilation due to progressive decrease in his respiratory effort. He underwent plasmaphoresis, and after three periods of plasma exchange over a three-week period he was weaned off the ventilator. He is now maintained as an out-patient on pyridostigmine and prednisolone.

Case 3

A 72-year-old man presented to the Elderly Care Medical out-patient department complaining of difficulty in chewing solid food and occasional choking episodes. This worsened through the day and was associated with dysarthria, diplopia, dyspnoea and generalized tiredness and lassitude. On examination, a left ptosis was observed but his ocular movements were normal. Examination by an ophthalmologist at this stage was unremarkable. He was a known hypertensive with a history of ischaemic heart disease, and in 1994 had been diagnosed with a cerebrovascular accident (CVA) with left hemiparesis and numbness, combined with short-term memory loss.

A CT scan of his brain at this time showed generalized atrophy and he recovered fully. A repeat CT scan of his head carried out on this later presentation was unchanged, although a magnetic resonance imaging (MRI) scan was not performed.

A Tensilon test was positive and acetylcholine receptor antibody levels high. A CT scan of his chest was negative for thymoma. Control of the disease was achieved over the ensuing three months, during which time he required three admissions for dosage adjustment including once for a neostigmine infusion when he had aphagia and was severely weak.

Discussion

Myasthenia gravis is an acquired condition caused by circulating IgG antibodies binding to the postsynaptic acetylcholine receptor at the neuromuscular junction. This causes depletion and subsequent destruction of the receptor. Characteristic of the disease is of global fluctuating muscle fatigueability. This usually begins with the ocular and extra-ocular muscles, and up to 10 per cent may have only visual symptoms. The disease may progress to cause more generalized problems, but deep tendon reflexes remain intact.

It is unusual for patients to present with symptoms of dysphagia due to bulbar weakness as a result of myasthenia gravis.[5,6] It is the main complaint of only six per cent of patients with the condition, although 28 per cent report some symptoms of dysphagia or dysarthria at outset.[4] Patients with dysphagia secondary to myasthenia gravis usually have dysfunction of the oropharyngeal phase of swallowing due to the bulbar effects, but may have abnormality of the oesophageal phase as a result of damage at the level of the parasympathetic oesophageal plexus. They may also have weakness of the musculature of the tongue and soft palate, causing difficulty in manipulation of the food bolus, hypernasal speech and nasal regurgitation. Myasthenia gravis may also cause stridor due to upper airway obstruction,[7] and vocal fold palsy due to the condition has been described.[8]

In our case series the predominant symtoms were related to the bulbar effects of the disease. In such circumstances, the differential diagnosis is between conditions such as cerebrovascular accident, multiple sclerosis, progressive bulbar palsy, muscular dystrophy, poly- or dermatomyositis and particularly in the elderly, motor neurone disease (MND). MND tends to cause a gradually progressive weakness with a mixture of upper and lower motor neurone deficits and peripheral muscular weakness and fasciculation. The bulbar signs are also often accompanied by wasting and fasciculation of the tongue, not a feature of myasthenia gravis. Eaton-Lambert syndrome is a rare systemic manifestation of small cell bronchial carcinoma (and rarely other malignancies) caused by decreased release of acetylcholine at the neuromuscular junction. It is distinguishable from true myasthenia gravis as deep tendon reflexes are absent and weakness tends to improve with muscle contraction or repetitive nerve stimulation, whereas in myasthenia gravis it worsens.

Myasthenia-like symptoms may be induced by procainamide, high doses of aminoglycoside antibiotics or penicillamine.

It is important to note that approximately half of patients presenting with a thymic tumour have myasthenia gravis.

The diagnosis of myasthenia gravis is made initially by a Tensilon test. This anticholinesterase increases the amount of acetylcholine at the neuromuscular junction on systemic administration. In a positive test, improvement in weakness is immediate and lasts two to three minutes. False positive results may be seen as a placebo or in rapidly progressive MND, and serum acetylcholine receptor antibody assay is thus mandatory. These are present in approximately 90 per cent of cases of the generalized form of the disease and are not present in any other condition. In addition, antibodies to intrinsic factor, skeletal muscle or thyroid may be found. A further investigation that can be valuable although not specific is repetitive nerve stimulation, where the characteristic muscle fatigueability and decrease in electrical response may be demonstrated.

Treatment is with oral anticholinesterases such as pyridostigmine; this may be supplemented by immunosuppressants such as prednisolone and azathioprine if required, although these second line

agents may take months to start working. Plasmaphoresis may be indicated as an emergency treatment in severe cases. Thymectomy may improve long-term prospects in patients without a thymic tumour, particularly in the younger patient. In this group, up to 60 per cent may respond to such surgical intervention.

Our case series illustrates the variability of the presentation of myasthenia gravis and demonstrates that symptoms related to speech and swallowing may predominate, especially in the older patient. In all our patients diagnosis had been delayed. In *Case 1* symptoms had been present for 18 months and in *Case 2* for three years; he went on to develop a myasthenic crisis and require intensive care support. In *Case 3* the correct diagnosis had been obscured by the presence of cerebrovascular disease, and concurrent ischaemic changes on neuroimaging should not exclude the diagnosis of myasthenia gravis.[3]

All patients demonstrated the characteristic muscle fatigueability, but unusually the weakness was predominantly bulbar. They illustrate the high degree of suspicion required in such patients presenting with bulbar weakness associated with other symptoms of neurological dysfunction, both local and generalized.

Prompt diagnosis allows early collaboration between ENT surgeon, physician and neurologist to ensure optimal treatment and avoidance of complications.

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