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CASE REPORT

MYASTHENIA GRAVIS MASQUERADING AS POST-POLIOMYELITIS SYNDROME

Rajiv Singh and Brian Pentland

From the Rehabilitation Medicine Unit, Astley Ainslie Hospital, Edinburgh, UK

A 79-year-old man with previous bulbar poliomyelitis developed dysphagia and was diagnosed as having post-polio syndrome. Over 2 years, his swallowing deteriorated and he suffered an aspiration pneumonia. Only after the subsequent development of fatigue and facial weakness was myasthenia gravis diagnosed. Diagnostic criteria for post-polio syndrome include the exclusion of all other neurological conditions such as myasthenia gravis. Moreover, in any instance where a patient develops new symptoms, it is advisable to reconsider the original diagnosis.

Key words: myasthenia gravis, post-polio syndrome, dysphagia.

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Correspondence address: Rajiv Singh, Rehabilitation Medicine Unit, Astley Ainslie Hospital, 133 Grange Road, Edinburgh EH9 2HL, UK. E-mail: rajiv.singh@lpct.scot.nhs.uk

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INTRODUCTION

Muscular weakness arising many years, often decades, after recovery from acute poliomyelitis is sometimes referred to as post-polio syndrome (PPS) (1). While limb weakness is often prominent, dysphagia is described in up to half of those affected (2). Other features include fatigue, muscle or joint pain, sleep apnoea and respiratory insufficiency (3). In the USA, there are some 300,000 survivors of polio and up to 50% have features of PPS (4). There is therefore a high potential for attributing signs and symptoms to PPS that may arise from other pathology. We describe here a patient in whom the primary diagnosis of PPS masked the presence of myasthenia gravis.

CASE REPORT

A 79-year-old retired clergyman, who had suffered acute poliomyelitis with bulbar and limb involvement in 1947, presented to physicians with a 6-month history of dysphagia and reduced voice volume. A diagnosis of PPS was made and he was referred for speech and language therapy. Videofluoroscopy indicated poor pharyngeal contraction and clearance and he was given advice about swallowing strategies and speech intelligibility. His poor speech had been of particular concern given his reputation as a media personality. Little improvement occurred and 14 months later he developed a severe aspiration pneumonia, which required intravenous antibiotics. He was felt to present a high risk of aspiration and a percutaneous endoscopic gastrostomy tube was inserted. One year later, he developed fatigue and was seen in our neurorehabilitation clinic. On examination there was marked dysarthria, soft palate wasting, facial and jaw weakness but no demonstrable fatigability. There was no ptosis or diplopia and limb motor power was normal.

At this stage, the alternative diagnosis of myasthenia gravis was considered. An assay for anti-acetylcholine receptor antibodies was strongly positive at 311×10^{-10} mmol/l (normal 0–5) and an edrophonium test was positive. Electromyography showed significant reduction in muscle contraction on repetitive stimulation and increased "jitter" further supporting the diagnosis. A computerized tomography scan of the thorax showed no evidence of thymoma. Within a day of starting pyridostigmine he noticed increased muscle strength and he continues to improve on a combination of steroids and pyridostigmine.

DISCUSSION

The presenting features of dysphagia and reduced speech volume in this patient were consistent with a diagnosis of PPS as was the later development of fatigue. There were no features such as ptosis, diplopia or limb weakness to suggest myasthenia gravis. Dysphagia and dysarthria are, however, recognized presenting features of this disease (5). The classical, though not universal characteristic of myasthenic muscle weakness is that motor activity fatigues. Thus the difficulty with eating and swallowing may be absent at the beginning of a meal but becomes evident as it progresses. This pattern was not present. The fatigue described by the patient was a general lethargy and malaise rather than a progressive weakness with activity. The development of facial weakness was the trigger to investigation for myasthenia gravis. High anti-acetylcholine receptor antibody titres are common in young female patients or those with thymoma but has also been reported in elderly patients as in this case (6).

A search of the principal medical databases (Medline, Embase, CINAHL) has failed to find a single report of the

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co-existence of PPS and myasthenia gravis. We would recommend that clinicians retain a high index of suspicion as to the possibility of myasthenia gravis in patients with dysphagia occurring many years after acute poliomyelitis. This seems particularly pertinent as there are sensitive diagnostic tests available for myasthenia gravis, whereas PPS remains a clinical diagnosis lacking strict criteria. Furthermore, treatment is simple and can have a marked effect on strength and fatigue. In this instance early diagnosis and treatment may have prevented a life-threatening aspiration pneumonia. As a principle, it is worthwhile reconsidering an existing diagnosis when new features develop and to exclude the possibility of a new condition.

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