

ACUTE HEMIPARESIS WITH HEMICHOREA AND CROSSED HEMIPARESIS; UNUSUAL PRESENTATION FOR MYASTHENIA GRAVIS

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ABSTRACT

Myasthenia gravis is an autoimmune disease mediated by antibodies directed against the acetylcholine receptor. Patients typically present with weakness and fatigability involving predominantly ocular and bulbar muscles. We describe 2 patients with acute weakness, one with a left hemiparetic pattern and chorea, and the other with crossed left hemiparesis and right facial weakness. More typical features of generalised myasthenia developed with time and both had thymic follicular hyperplasia on thymic histopathology. Clinical improvement occurred in both following thymectomy and immunosuppression. These cases exemplify the fact that markedly asymmetric limb weakness can be an early feature of generalised myasthenia gravis.

Keywords: Myasthenia gravis, hemiparesis, hemichorea.

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INTRODUCTION

Myasthenia gravis is an organ specific antibody mediated disease^(1,2) which typically affects ocular and/or bulbar musculature initially⁽³⁾. The disease becomes generalised, in the majority of patients, within thirteen months of these first symptoms with maximal weakness usually occurring within 36 months of onset^(4,5). Limb involvement is relatively uncommon as the sole presentation, and weakness is usually more marked in proximal muscles⁽³⁾. We present 2 cases where the early pattern of weakness, predominant limb involvement and acute history suggested a central lesion, provoking initial investigations aimed at excluding either an ischaemic or demyelinative etiology.

Case 1

LG, a 24-year-old Chinese male, first presented with a one-week history of acute left arm and leg weakness, associated with involuntary movements. Examination confirmed a left hemiparesis with grade 3 power in the arm and grade 4 power in the leg. There was a mild left facial weakness but no sensory deficits, ptosis, dysphagia or dysarthria. The plantar responses were flexor and reflexes symmetric and brisk. Choreiform movements of the left hand and feet, with dystonic posturing, were evident but could be volitionally suppressed.

Both computed tomograms (CT) and a magnetic resonance (MR) scan (1.5 Tesla) of the brain were normal. Surface nerve conduction, concentric needle electromyography (EMG) and visual and auditory evoked potentials were normal. The cerebrospinal fluid (CSF) at lumbar puncture was sterile and acellular, with normal protein levels and electrophoretic pattern.

Progress

The weakness resolved spontaneously over 3 weeks, but he developed pre-syncopal faintness and occasional diplopia. Review showed bilateral ptosis, more prominent on the right, with fatiguability of both deltoids. Repetitive supramaximal stimulation (5Hz) of his nasalis and abductor digiti minimi muscles confirmed 20-35% decremental responses, and 10 mg of intravenous edrophonium reversed ptosis dramatically. The serum acetylcholine receptor antibody titre (AChR) was 20.4 nmol/L (controls 0-0.04 nmol/L).

A thymectomy was performed and an enlarged thymus gland was removed. Histology showed thymic follicular hyperplasia. The patient was started on prednisolone 60 mg/day with pyridostigmine, the former being gradually tailed to 10 mg EOD. He is currently in remission one year post thymectomy.

Case 2

HN, a 44-year-old Malay lady, was admitted for evaluation of left knee pain. Whilst in the ward, she developed acute left arm and leg weakness associated with non vertiginous giddiness. She had a history of surgery to the right eyelid to alleviate unilateral ptosis 10 years previously. Clinical evaluation showed grade 2 power in the left arm with poor finger movements. Left leg power was grade 3, more pronounced proximally. There were no tone changes and the tendon reflexes were symmetrically brisk with flexor plantar responses. There was a lower motor neuron right facial weakness and mild right ptosis.

A CT scan of the brain was normal as was a MR scan (with and without Gadolinium-DTPA) done 2 days later. A 15% facial decremental response was evident on repetitive supramaximal stimulation but this was equivocal (6-8%) in the abductor digiti minimi. Intravenous edrophonium (10 mg) improved arm power but did not reverse the ptosis. The anti AChR antibody titre was 0.04 nmol/L. She was started on limb physiotherapy and pyridostigmine 30 mg QID and improved enough to walk independently.

Progress

A week later, weakness in the left arm and leg returned. A CT scan of the mediastinum showed a thymic mass and she was given a course of plasmapheresis (5 litres exchanged in 2 sessions) with dramatic improvement in left arm power (grade 2 to 4+) and left leg strength (grade 2+ to 4). She was commenced on prednisolone 60 mg/day and at thymectomy, a

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large hyperplastic thymus was removed. Six weeks after surgery, apart from pain at the sternotomy site, she had near normal power in the left arm and leg.

DISCUSSION

Weakness and fatigue in myasthenia gravis are mediated by heterogenous antibodies directed against the nicotinic post synaptic acetylcholine receptor in skeletal muscle^(1,2). Assays for these antibodies form the basis of the most specific investigation for this disorder⁽⁶⁾. Clinical expression is varied but the majority of patients present with involvement of ocular and bulbar musculature. Limb muscles can be involved, usually affecting proximal more than distal musculature⁽³⁾.

It is a well recognised feature of the disease that weakness is often asymmetric and differential, ie it is seldom the case that both levator palpebrae are equally involved and dysphagia may be prominent whilst there is minimal objective limb weakness⁽³⁾. This is also evident in the treatment in that sufficient pyridostigmine to control ptosis and diplopia may result in cholinergic weakness of muscles of deglutition.

However, what is unusual in these 2 patients, is the rapid onset of weakness with initially no evident fluctuation of symptoms. The pattern of weakness was also strikingly asymmetric and in Case #1 produced a left hemiparesis with choreiform movements of the arm. It was only the development of ptosis and variable diplopia that led to the authors investigating him for a neuromuscular transmission disorder. Following thymectomy, his left hemiparesis improved and then resolved. We cannot satisfactorily explain his chorea and did not investigate his CSF for antibodies against AChR, but it is intriguing to speculate on whether IgG class immunoglobulins might be implicated as was described by Husby et al in Sydenham's chorea⁽⁷⁾.

Case 2 already had a history of left ptosis before admission and had been treated surgically for this without confirmation of the cause. However, like the other patient, she acutely developed a left hemiplegia with right facial weakness that was interpreted correctly as being of the lower motor neuron

variety. Nevertheless, the crossed hemiplegia led to the initial investigations directed at excluding a brainstem cerebrovascular ischaemic event. Again, neuroimaging was unremarkable and her weakness persisted but fluctuated in intensity. As in the first case, plasma exchange followed by thymectomy resulted in rapid improvement of weakness.

Clinically, an important feature was the lack of true signs of pyramidal tract release ie spasticity, enhanced deep tendon reflexes or upgoing plantar responses. Also, within the period described in Grob's review⁽⁴⁾, more typical ocular weakness and fluctuating signs became apparent. Additionally, neuroimaging was unremarkable despite prominent motor weakness.

CONCLUSION

These two patients exemplify the protean manner in which myasthenic weakness may initially manifest. In both, no definite corticospinal tract release signs occurred although the pattern of weakness suggested a lesion that was central. Moreover, the acute presentation led to initial consideration of either an ischaemic or demyelinating etiology. Over time, however, both developed more typical features of generalised myasthenia and rapidly responded to appropriate therapy.

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