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Selective or predominant triceps muscle weakness in African–American patients with myasthenia gravis

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Abstract

Myasthenia gravis (MG) can lead to weakness in different patterns of muscle groups. Limb muscle weakness is most typically seen in a limb girdle pattern, although variants exist. In the current study, we aimed to describe a unique MG phenotype consisting of selective or predominant triceps muscle weakness. We performed a retrospective review of MG patients who developed focal or predominant triceps muscle weakness between 2006 and 2016. The clinical, electrophysiological and serological characteristics of these patients were examined. 8 MG patients were identified, including 7 males, all of whom were African–American. Two patients underwent muscle biopsy, and one patient underwent cervical spine decompression surgery. All showed significant improvement following immunosuppressive treatment, although one patient experienced a relapse of muscle weakness. This case series highlights a relatively uncommon MG clinical phenotype of selective triceps muscle weakness, mainly in African–American males, in line with previous literature. Familiarity with this phenotype is important in order to facilitate diagnosis and appropriate treatment for this group, and avoid unnecessary investigations or treatments.

Keywords: Myasthenia gravis; Focal weakness; Triceps; Single-fiber EMG; African–American; Muscle atrophy

1. Introduction

Myasthenia gravis (MG) is an autoimmune disorder of neuromuscular transmission characterized by fluctuating muscle weakness, with a predilection for muscles innervated by cranial nerves. As a result, ptosis, diplopia, dysarthria, dysphagia, and difficulty chewing are amongst the most common symptoms. Limb muscle weakness occurs in 20–30% of affected individuals. In the majority of patients with limb muscle weakness, proximal muscles are preferentially involved, resulting in a limb girdle pattern [1]. However, the clinical pattern of weakness in MG can be highly variable, and there are reports of focal, multifocal, or diffuse muscle weakness in the medical literature [1,2]. Although extensor muscles in the upper extremities, such as the triceps muscle, are known to be involved in MG [3,4], focal or predominant triceps muscle weakness is rare and has only been reported in individual cases [2,5]. Nonetheless, it has been suggested that this phenotype is more common than previously thought, especially in African–American population [6]. In the current study we describe our own series of 8 MG patients with focal or predominant triceps muscle weakness. Recognition of this disease phenotype is important in order to avoid misdiagnosis of other conditions, such as focal radial neuropathy, cervical radiculopathy, or superimposed myopathy.

2. Methods

This study protocol was approved by the Research Ethics Board of the University Health Network at the University of Toronto. We performed a retrospective chart review of patients diagnosed with myasthenia gravis (MG) who developed focal or predominant triceps muscle weakness during the course of their illness, and who attended the Prosserman Family Neuromuscular clinic (University of Toronto) between January 2006 and July 2016. MG diagnosis was based on a compatible clinical presentation, supported by electrophysiological and/or serological evidence in all patients.

For each patient, we extracted the clinical history, serial neurological examinations, electrophysiological studies and any additional tests (serology, diagnostic imaging studies, and muscle

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biopsy) or procedures that were performed for investigation of patients with this clinical phenotype. Clinical examination findings were extracted from the chart, looking at 4 pre-determined time points. These time points included the *initial visit*, the *reference visit* (clinic visit showing maximal triceps muscle weakness), the *follow-up visit* (showing maximal improvement in weakness), and the *most recent follow-up visit*.

3. Results

The clinical, electrophysiological, and serological findings, and ancillary tests performed in 8 MG patients, have been presented in Table 1. The mean age of patients was 48 ± 10 years at the reference visit, with mean duration disease of 7 years (range 0.5–33 years). 7 patients were male, comprised entirely of African–Americans (7/7), and 1 patient was female of Asian background. The follow-up visit showing improvement in triceps muscle weakness occurred a mean of 1.5 years (range: 0.2–6 years) following the initial visit. Within the identified patients, 4/8 had *focal* triceps muscle weakness, and 4/8 had *predominant* triceps muscle weakness at the reference visit, indicating that milder weakness was demonstrated in additional limb muscles (Table 2). Ocular symptoms were reported by 8/8 patients at disease presentation (although not necessarily at the reference visit). All patients were treated with immunosuppressive therapy (prednisone, intravenous immunoglobulins and/or azathioprine) and improved, as demonstrated in Table 2.

Long term follow-up (>5 years after the reference visit) occurred in 3/7 patients. At long-term follow-up, 2 patients (#3 and #5) achieved pharmacological remission with normal strength in all muscle groups, and one patient (#2) experienced significant triceps muscle weakness recurrence, graded 2/5 on MRC (Medical Research Council) scale.

In addition to routine chest CT, imaging studies performed included brain MRI in patients #2, #6 and #7, cervical MRI in patients #2, #3, #7 and #8, brachial plexus/limb MRI in patient #8, and muscle ultrasound in patient #4. Upper limb MRI in patient #8 confirmed the presence of fatty infiltration and volume loss within the right triceps muscle.

Two patients (#4 and #5) underwent biceps muscle biopsy, on the clinical suspicion of a superimposed myopathy. Patient #4 developed isolated triceps muscle weakness while being treated with mycophenolate mofetil and pyridostigmine. Needle electromyography showed myopathic features, and muscle ultrasound demonstrated increased echogenicity and atrophy in bilateral triceps muscles. Patient #5 failed to respond to pyridostigmine, intravenous immunoglobulins, and mycophenolate. Steroid treatment was initially refused, but was started after muscle biopsy, resulting in complete resolution of the triceps weakness. In both patients #4 and #5, the muscle biopsies showed mild and non-specific abnormalities, including a mild variation in fiber size. In another patient (#2), a clinical suspicion of C7 radiculopathy was suspected as the cause of triceps muscle weakness (prior to evaluation in the neuromuscular clinic), and therefore cervical spine decompression and fusion (from C3-C6 levels) was performed. The surgery did not lead to any improvement in symptoms. However, there was notable improvement 1.5 years after the reference visit, subsequent

Table 1
Clinical, electrophysiological and serological characteristics at presentation, and additional investigations.

Age	Presenting symptoms	SFEMG	RNS	AChRAb	Additional investigations
31M	Ptosis	+	−	+	None
34M	Diplopia	+	−	−	Brain and cervical MRI
40M	Diplopia, 4-limb weakness	+	−	+	Cervical MRI
52F	Diplopia, 4-limb weakness	+	+	+	Muscle ultrasound and muscle biopsy
53M	Diplopia,4-limb weakness	+	−	−	Muscle biopsy
55M	Diplopia and ptosis	+	+	+	Brain MRI
57M	Diplopia and ptosis	+	+	−	Brain and cervical MRI
58M	Ptosis	+	+	+	Cervical and brachial plexus

Age – At reference visit. M, male; F, female; SFEMG, single-fiber EMG; RNS, repetitive nerve stimulation; AchRAb, acetylcholine receptor antibodies; NA, not available.

Table 2
Limb muscle weakness at reference and follow up visits, graded by MRC scores.

Reference visit				Follow up visit		
Disease duration (years)	Triceps MRC grade (/5)		Addition limb muscle weakness (/5)	Time since reference visit (years)	Triceps	
	Right	Left			Right	Left
2	2	2	None	0.2	5−	5−
2	2	5	None	1	4	5
3	4	4−	None	2	5	5
33	2	2	None	1	4	4
11	4−	4−	Right ankle dorsiflexion (4)	6	5	5
3	2−	2−	Deltoids (4), neck extension, biceps (4)	0.3	4	4
0.5	3	4−	Distal UL (4)	0.2	4−	4
2	4−	0	Right biceps (4+), finger flexion, extension, and abduction (4)	0.3	5	4+

Reference visit – Presenting or follow up visit showing maximal triceps muscle weakness; Disease duration = time elapsed between presentation and reference visit; Time since ref visit = time elapsed between reference and follow up visits; UL, upper limbs.