Reversible man-in-the-barrel syndrome in myasthenia gravis

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Abstract

Man-in-the-barrel syndrome (MBS) is an uncommon presentation due to bilateral, predominantly proximal muscle weakness that has not been described to be associated with myasthenia gravis. We describe a case of myasthenia gravis presenting as MBS. Additionally, he had significant wasting of the deltoids bilaterally with fibrillations on electromyography (EMG) at rest and brief duration (3-6 ms) bi/triphasic motor unit potentials (MUPs) on submaximal effort apart from a decremental response on repetitive nerve stimulation (RNS) at 2 Hz. While electrophysiology is an important tool in the diagnosis of myasthenia gravis, pathological EMG patterns do not exclude the diagnosis of myasthenia gravis.

Key words

Electromyography (EMG), man-in-the-barrel syndrome (MBS), myasthenia gravis, reversible

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Man-in-the-barrel syndrome (MBS) refers to bilateral, predominantly proximal arm weakness. The etiology of MBS is varied, ranging in localization from central, spinal cord to peripheral including myopathic and neuropathic etiologies.[1-3] Herein we describe a patient who had MBS as a presenting symptom of myasthenia gravis among other atypical features.

Case

A 77-year-old male presented on January 19, 2015 with history of mild left ptosis, intermittent diplopia, and difficulty in raising both arms since December 29, 2014. He had been operated on for bilateral cataracts on December 21 and 28, 2014 under local anesthesia. No neuromuscular blockade was used during the surgeries. The difficulty in raising both arms was fairly severe, symmetrical, and acute in onset. He denied radicular pain, numbness, paresthesiae, or fasciculations. He had no bulbar weakness or complaints in the lower limbs. Examination revealed bilateral fatiguable ptosis and normal extraocular movements. He had wasting of both bilateral shoulder girdle muscles with significant weakness of both deltoids (grade 2/5) [Video 1]. The hip girdle muscles also showed mild weakness (grade 4/5). There were no fasciculations. His gait, sensations, and deep tendon reflexes were normal and the plantars were flexor.

In this case, the key feature was MBS due to a peripheral cause [Table 1]. Given the ptosis and diplopia, myasthenia gravis was considered as a possibility, even though it has not been described to cause MBS. Brachial plexopathy or a bilateral C5-6 radiculopathy could present with MBS and wasting of the deltoids. However, there was no pain, reflexes were

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Table 1: Causes of man-in-the-barrel syndrome

Central causes	Peripheral causes
Cerebrum: Bilateral frontal lobe lesions (precentral gyrus)	Myopathic
Ischemic stroke: Bilateral anterior watershed infarction	Muscular dystrophy: Limb girdle muscular dystrophy, facioscapulohumeral muscular dystrophy
Systemic hypoperfusion or hypovolemia from cardiogenic shock, myocardial infarction	
Cerebral anoxia or hypoxia after pericardial tamponade, cardiac or aortic surgery with extracorporeal circulation	Immune-mediated myopathy: Polymyositis, dermatomyositis
Pontine or extrapontine myelinolysis	
Multiple sclerosis	
Multicentric cerebral glioblastoma multiforme	
Cerebral metastasis	
Spinal cord	Neuropathic
Infarction of the anterior spinal artery and posterior inferior cerebellar artery	Brachial plexopathy
Vertebral artery dissection	Motor neuron disease: Brachial amyotrophic diplegia, monomelic amyotrophy, HIV-associated motor neuron disease
Spinal trauma with cervical cord contusion or hyperextension	Neuropathy: Multifocal motor neuropathy
	Cervical radiculopathy

HIV = Human immunodeficiency virus

preserved with normal sensory examination, and the hip girdle weakness and ptosis could not be explained. The phenotype was atypical for a muscle disease. Because of the short duration of the symptoms it was difficult to explain the muscle atrophy. Involvement of ocular muscles is not seen in metabolic myopathies with short duration of symptoms.

The patient's antiacetylcholine receptor (anti-AChR) antibodies were elevated at 16.73 nMol/L (normal <0.25 nMol/L). Serum creatine phosphokinase (CPK) and thyroid hormones were in the normal range.

Electrophysiological studies showed normal peripheral motor and sensory conduction findings with normal compound muscle action potential amplitudes (CMAP) and decremental response to repetitive nerve stimulation (RNS) study at 2 Hz in the left deltoid (39%) and left orbicularis oculi (38%) [Figure 1]. Needle electromyography (EMG) in both deltoids revealed fibrillation and positive sharp waves at rest with brief duration (3-6 ms) bi/triphasic motor unit potentials (MUPs) on submaximal effort [Figures 2 and 3]. High-amplitude or long-duration potentials were not noted. On strong effort, a mild fallout was noted. Both tibialis anterior, left triceps, left first dorsal interosseus, right abductor digiti minimi, and tongue showed normal EMG findings. A diagnosis of myasthenia gravis was made on the basis of clinical findings, elevated anti-AChR antibodies, and decremental response to low-frequency RNS. He was treated with steroids, the dose increased gradually to reach 1 mg/kg per day. Within 20 days he had significant improvement with an improved range of shoulder movement (right more than left) and no signs or symptoms of extraocular muscle involvement.

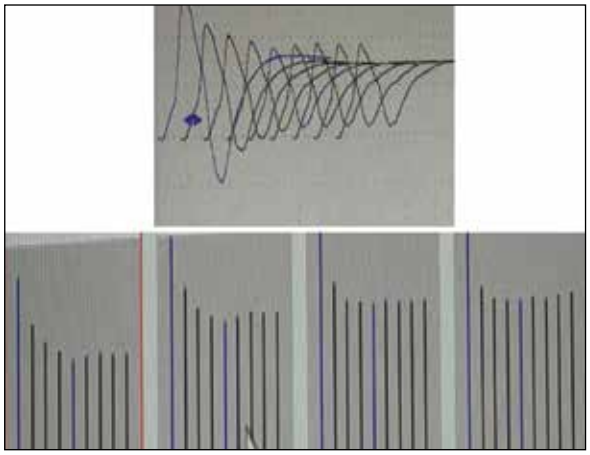

Figure 1: RNS of the axillary nerve, recording from left deltoid, nine stimuli at two per second showing resting and postexercise decremental response

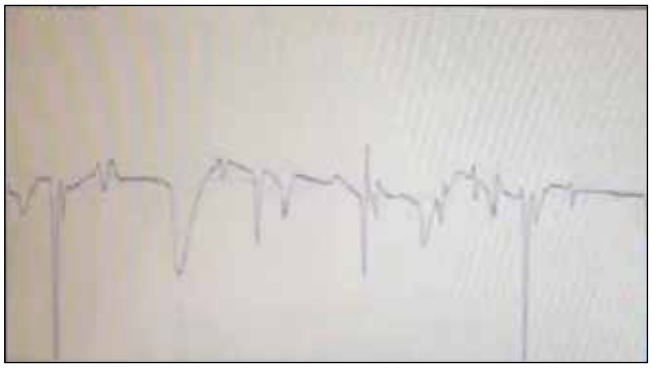

Figure 2: Left deltoid spontaneous activity. Time base 100 ms, amplitude calibration 200 uv/div

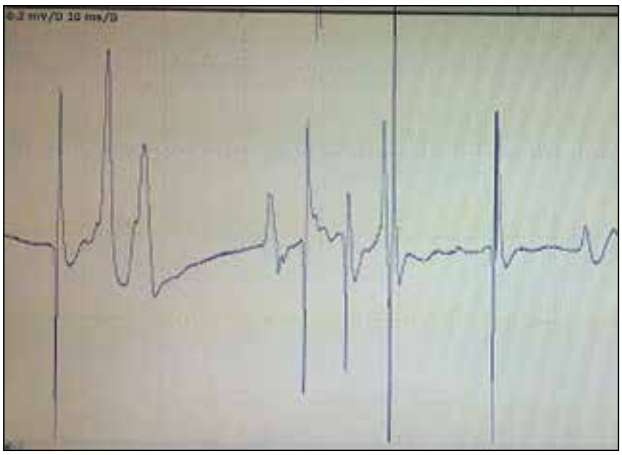

Figure 3: Left deltoid volitional activity showing brief-duration low-amplitude MUPs. Time base 100 ms, amplitude calibration 200 uv/div

Discussion

Predominant proximal arm weakness or MBS syndrome has been initially described with bilateral watershed cerebral infarction and has also been associated with brainstem and cord involvement. Peripheral nervous system involvement is usually insidious in onset and slowly progressive; it is most

commonly associated with motor neuron disease. Myasthenia gravis has not been described to be associated with this rare clinical presentation. Apart from this presentation, the early-onset, severe atrophy of the shoulder girdle muscles, presence of fibrillation potentials, and positive sharp waves at rest and myopathic potentials on activation in the deltoid were unusual for myasthenia gravis.[4]

It is known that acetylcholine exerts a trophic influence on the muscle. Oosterhuis and Bethlem found muscular atrophy, confirmed histologically, in at least one of several groups of muscles in 14 out of 148 patients with generalized myasthenia gravis. As in this case, the proximal muscles were more involved. The mean duration of disease was 11 years compared to 6-8 years for patients without atrophy. No statistical correlation was found between the duration of disease and muscular atrophy, and atrophy was not the presenting symptom, contrary to our case. Six out of 14 patients showed EMG evidence of myopathy.[5] Neurogenic atrophy and pseudohypertrophy of the tongue have also been described in two cases of bulbar myasthenia where EMG showed evidence of neurogenic changes and the histology showed severe neurogenic atrophy with fatty pseudohypertrophy.[6] Axonal neuropathy and myositis as a cause of fibrillation potentials was ruled out by the normal nerve conduction study and the normal CPK levels.[4]

Teaching point

Muscular atrophy and atypical changes on EMG can occur in the setting of myasthenia gravis. This is the first account of a reversible MBS secondary to myasthenia gravis.

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Conflicts of interest

There are no conflicts of interest.

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