Austin Journal of Clinical Neurology

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Austin Publishing Group

Case Report

A Unique Case of Unilateral Distal Arm Weakness as a Presentation of Myasthenia Gravis

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Received: August 23, 2021; **Accepted:** September 27, 2021; **Published:** October 04, 2021

Abstract

Background: Myasthenia Gravis (MG) is an autoimmune neuromuscular disorder which typically presents with fluctuating ocular, bulbar or proximal limbs weakness.

Case Presentation: A 71-year-old woman was admitted to our Neurology department with sudden onset of right distal hand weakness which exacerbated within days and subsequently involved also the proximal limbs and ocular muscles following treatment with high-dose steroids (admitted due to suspicion of myelitis). After a broad neurological work up, she was diagnosed with MG, based on the findings of a typical decremental pattern on Repetitive Nerve Stimulation (RNS) test and the presence of Acetylcholine Receptor Antibodies (AChRAbs). Treatment with 5 cycles of plasma exchange induced a dramatic clinical improvement.

Conclusion: This case represents an atypical presentation of MG, simulating Cerebrovascular Accident (CVA) or myelitis and unmasking of signs by treatment with high dose of steroids.

Keywords: Myasthenia Gravis; Distal hand weakness; Ptosis; Anti-acetylcholine receptor antibodies

Abbreviations

MG: Myasthenia Gravis; RNS: Repetitive Nerve Stimulation; AChRAbs: Acetylcholine Receptor Antibodies; CVA: Cerebrovascular Accident; CT: Computed Tomography; MRI: Magnetic Resonance Imaging; CSF: Cerebrospinal Fluid; HHV: Human Herpesvirus; ICA: Iodinated Contrast Agent

Case Presentation

A 71-year-old woman with history of hyperlipidemia and hypothyroidism, was admitted in the emergency room of our hospital, due to sudden onset of distal right hand weakness, which started two days earlier, and subsequent right leg weakness on the day of her admission. She denied any other neurological signs. Neurological examination revealed normal mentation and intact cranial nerves. There was weakness in wrist extension (2/5), wrist flexion (4/5) and interosseous muscles (3/5) in the right arm. There was also weakness of the right iliopsoas muscle (4/5). No other focal or long tract signs were detected, including normal deep tendon reflexes.

A brain and neck Computed Tomography (CT) scan and angiography showed a hypodense area on the right frontal lobe, with no contrast enhancement. A brain Magnetic Resonance Imaging (MRI) was performed and showed chronic ischemic changes with no acute or other pathological findings. The patient was admitted with tentative diagnosis of a possible CVA. Two days later, the patient started complaining of left proximal arm weakness. On her examination there was a new finding of weakness of the proximal muscles of the upper and lower extremities (4+/5). A cervical MRI was unremarkable. A lumbar puncture revealed increased protein (1095 mg/l), normal glucose concentrations (3.3mmcl/l) and pleocytosis (10 cells/ml). Cytopathology of the Cerebrospinal Fluid (CSF) showed normal reactive lymphocytes. CSF immune-electrophoresis did not reveal intrathecal oligoclonal antibodies. Due to a suspicion of myelitis or myeloradiculitis, treatment with high dose intravenous methylprednisolone was initiated. One day after the administration of steroids, the patient complained of binocular diplopia at the lateral gazes, bilaterally and worsening of the limb weakness, that involved the muscles of all four limbs. Neurophthalmologic evaluation revealed bilateral IV cranial nerve palsy, without papilledema. Further evaluation was performed with the suspicion of a central nervous system inflammatory condition. Routine blood tests and an extensive panel of autoantibodies (including ANA, ANCA, APLA, anti-DNA, ENA, LAC, anti-VGCC and anti-gangliosides) were within normal ranges. Tumor markers and a paraneoplastic antibodies panel (in serum and CSF), were negative. Protein immunofixation and electrophoresis did not detect paraprotein. Polymerase chain reaction for Herpes simplex virus (1 and 2), Human Herpesvirus (HHV) 3, HHV-6, and Enterovirus were negative. Nasopharyngeal swab for COVID19 and serology for Human immunodeficiency virus and syphilis, were also negative. A total body CT ruled out systemic neoplasia. The titers of anti-COVID19 spike protein antibodies were high (IgG: 11090.7 AU/ML; pos. >50AU/ML), due to earlier vaccination.

The diplopia continued to deteriorate and was present in all gaze directions. Her repeated neurological examination revealed bilateral ptosis, weakness of neck extensor and flexor muscles (4-/5), weakness (3+/5) of the proximal and distal muscles of the upper limbs and proximal leg weakness (3+/5). The diagnosis of MG was raised and a treatment with 5 cycles of plasma exchange initiated, causing a dramatic improvement within days. A blood test for AChRAbs was

Citation: Shalabi F and Karussis D. A Unique Case of Unilateral Distal Arm Weakness as a Presentation of Myasthenia Gravis. Austin J Clin Neurol 2021; 8(3): 1153.

positive and electromyography revealed a typical decremental pattern on RNS with no signs of peripheral neuropathy of radiculopathy, confirming thus the diagnosis of MG. Treatment with pyridostigmine and low dose of oral prednisone, were initiated.

Discussion

Prominent distal limbs weakness as an initial presentations of MG, is rare, but was been previously reported. In the reported cases, there was no difference from typical MG regarding age, gender or disease-prognosis [1]. Pure asymmetric distal hand weakness is even rarer as an initial presentation of MG [2]. Moreover, in the presented case, the unilateral hand weakness appeared acutely, raising the suspicion of a cerebrovascular event. The high protein and cellularity in the CSF, introduced additional diagnostic thoughts to the direction of myelitis or other immune-mediated CNS disease. We do not have a good explanation for this finding, but high CSF-protein in previously reported MG -cases, was attributed to intercurrent or associated disorders [3].

The additional uniqueness of our case, relates to the unmasking of the symptoms of MG (with multi-focal involvement) following the administration of high dose steroids and/or the use of repeated doses of Iodinated Contrast Agent (ICA) for the purpose the CT scans. Both high dose steroids and iodine have been associated with deterioration or unmasking of MG [4,5]. Concerning ICA, the deterioration of MG signs has been reported to occur with a delay [6], and not so close to the ICA-administration, as in our patient. A possible explanation for the fast effects in our patient elated to the repeated administration of ICA (twice in a short period of time) and the additional treatment with high dose steroids. Those two factors may have acted synergistically, leading to the unmasking of MG.

In summary, the presented case highlights the diversity of MG that occasionally appears with highly atypical presentations which may challenge the physician and cause diagnostic dilemmas or misdiagnosis of the disease, even to a point of false diagnosis of CVA or myelitis. Such cases should raise the alertness of neurologists for rare presentations of MG, even in patients with sudden onset and totally asymmetrical distal limb weakness.

Declaration

Consent for publication: Written informed consent was obtained from the patient for publication of this Case report and any accompanying images. A copy of the written consent is available for review by the Editor of this journal.

References

1. Werner P, Kiechl S, Lo¨scher W, Poewe W, Willeit J. Distal myasthenia gravis-frequency and clinical course in a large prospective series. Acta Neurol Scand. 2003; 108: 209-210.

2. D Karacostas, I Mavromatis, G Georgakoudas, N Artemis, I Milonas. Isolated distal hand weakness as the only presenting symptom of myasthenia gravis. European Journal of Neurology. 2002; 9: 429-432.

3. Sigurdur Thorlacius and Johan A Aarli. The cerebrospinal fluid in myasthenia gravis. Acta Neurol Scand. 1985; 72: 432-443.

4. Deepak K Somashekar, Matthew S Davenport, Richard H Cohan, Jonathan R Dillman, James H Ellis. Effect of Intravenous Low-Osmolality Iodinated Contrast Media on Patients with Myasthenia Gravis. Radiology. 2013; 267: 727-734.

5. Jong Seok Bae, Seok Min Go, Byoung Joon Kim. Clinical predictors of steroid-induced exacerbation in myasthenia gravis. Clinical study. Journal of Clinical Neuroscience. 2006; 13: 1006-1010.

6. Jakob Rath, Matthias Mauritz, Gudrun Zulehner, Eva Hilger, Hakan Cetin, Gregor Kasprian, et al. Iodinated contrast agents in patients with myasthenia gravis: a retrospective cohort study. J Neurol. 2017; 264: 1209-1217.