Austin Journal of Clinical Case Reports

Austin Publishing Group

Open Access Full Text Article

Case Report

# Asymmetric Involvement of Abductor Digiti Minimi Found by Repetitive Nerve Stimulation in Myasthenia Gravis

**Qi Wang[1] and Hai-feng Li[2]***

[1]Department of Neurology, Affiliated Hospital of Qingdao University, China

[2]Department of Neurology, Qilu Hospital of Shandong University, China

***Corresponding author:** Hai-feng Li, Department of Neurology, Qilu Hospital (Qingdao) of Shandong University, No. 758, Hefei Road, Qingdao, China

**Received:** March 18, 2014; **Accepted:** April 10, 2014; **Published:** April 15, 2014

## Background

Myasthenia gravis (MG) is an autoimmune disease involving neuromuscular junction, which is characterized as fluctuating weakness and fatigue in various muscle groups. Amplitude decrement is often found in involved muscles and sometimes in clinically normal muscles in low frequency repetitive nerve stimulation (RNS) test. The abnormal response elicited in RNS is mostly seen in facial muscles or proximal limb muscles. Distal involvement was reported as a rare manifestation of MG, alone or in combination with other muscle involvement. We report a MG patient with asymmetric abductor digiti minimi (ADM) involvement.

## Case Report

A seventy-year old male presented with fluctuating blurred vision for two months in Dec 2010. The symptom tended to worsen in the afternoon and relieved after rest. Mild ptosis after sustained up gaze for 60 seconds and mild limited movement in adduction and abduction was found in his left eye. The ptosis resolved with rest, but eye movement did not. The diameters and responses to light of pupils were normal. No involvement of other muscle groups was noticed in a standardized Jolly test. The grip strength of his left hand was 29 kilograms, and that of right hand was 32.5 kilograms. Brain MRI was normal except for small focal ischemia lesions in bilateral corona radiate. Chest CT was normal. T3, T4 and TSH, anti-thyroglobulin antibody, anti-thyroid peroxidase antibody and anti-TSH receptor antibody were in normal ranges. AChR binding antibodies were 1.87nmol/L (positive cutoff: 0.45 nmol/L, RSR Limited, UK). The symptom and signs relieved slightly after intramuscular injection of neostigmine 1mg and disappeared completely 40 minutes after injection in a repeated test with 1.5mg, and reappeared 90 minutes after injection. Amplitude decrement were elicited in the right ulnar nerve (recorded at ADM) at low frequencies (28% at 2Hz, 34% at 3Hz and 36% at 5Hz), while normal responses in the right facial, axillary and median nerves (recorded at orbicularis oculi, deltoid and abductor pollicis brevis) at low frequencies (2, 3 and 5 Hz). Normal response was elicited in the right ulnar nerve at high frequency (20Hz). Motor conduction studies in the right median and ulnar nerves were normal. When asked to abduct his little fingers on both sides, he could not abduct his right little finger to the same angle as the left one. Bilateral abduction and opposition of thumb and flexion of fingers were normal. There was no muscle atrophy and sensory involvement in his both hands. The RNS test was repeated on the next day and found similar responses in right ulnar nerve (18% at 2Hz, 28% and 30% at 3Hz, and 29% and 34% at 5Hz), while normal responses in the left ulnar and axillary nerves. Motor conduction studies in the right and left ulnar nerves were normal.

The patient was given prednisone (1mg/kg/day) after MG was diagnosed. The ptosis disappeared 20 days after treatment and eye movement was in full range, with slightly blurred vision after sustained left gazing. Slight impairment of right little finger abduction was noticed. The steroid was tapered. All symptoms and signs including impaired right little finger abduction disappeared 50 days after treatment. The dosage of prednisone was tapered to 25mg/day. The RNS of right facial, axillary and median nerves, and RNS and motor conduction studies of bilateral ulnar nerve were repeated with normal results sixty days after treatment. Steroid was tapered gradually and decreased to 5mg/day one year later and was continued till now, with one minor fluctuation of ptosis for 10 days, which recovered without increasing the dosage of steroid.

## Discussion

The asymmetric involvement of ADM was found with RNS test, which performed two months after the initial ocular symptoms in our patient. No other muscles except for extra ocular muscles were found to be involved by a standard Jolly test, including grip strength of both hands. Normal RNS responses in other nerves along with normal motor conduction studies in the right ulnar nerve led us to suspect that the ADM involvement was due to MG. Normal motor conduction in ulnar nerve and no atrophy and sensory involvement excluded ulnar nerve injury. The ADM involvement resolved gradually after treatment with steroid. Repeated RNS test of the ulnar nerve was normal, which was consistent with clinical improvement, confirming our hypothesis on the ADM involvement.

The decremental response was obtained mostly in facial nerves and proximal limb nerves in the studies in unclassified MG patients (including both ocular and generalized MG) [1-3], and in the studies with specific data of ocular MG patients [4-8]. The ulnar nerve/ADM involvement in RNS was reported as a less common finding [1-8]. In the 90 ocular patients in the series of Cui et al [4], only one patient (1.1%) showed ulnar nerve involvement in RNS test, which was accompanied with involvement of two other nerves. There was




**Citation:** Wang Q, Li H-f. Asymmetric Involvement of Abductor Digiti Minimi Found by Repetitive Nerve Stimulation in Myasthenia Gravis. Austin J Clin Case Rep. 2014;1(1): 1001.

no isolated ulnar nerve involvement in the series of Chen et al [8]. The decremental response in the right ulnar nerve on the background of normal responses in other nerves, especially in the right median nerve prompted us to suspect MG as the underlying mechanism.

Distal muscle involvement is a rare manifestation in MG. The reported incidence of this in two large series was 3% [9] and 7.1% [10,11]. Distal involvement occurred at any age in the adulthood, with similar proportion between males and females. Distal weakness was more commonly seen in hand muscles, particularly finger extensors, and was reported as an initial symptom or the only presenting symptom, although in some patients distal involvement developed several years after involvement of other muscles. Additional mild proximal muscle involvement, minor ocular and bulbar symptoms were evident in some patients. Although presented with an asymmetric mode, patients subsequently tended to develop bilateral distal weakness if left untreated [9-12]. In the series of Nations et al, all 9 patients had elevated AChR antibodies. RNS of ulnar nerve recording at ADM demonstrated abnormal results in 87% of the patients examined. The one patient with a normal ulnar RNS results had a significant decrement in the accessory nerve. The results of routine nerve conduction studies and needle EMG were normal in the 5 patients examined. None of the patients had thymoma on chest CT [9]. In the series of Werner et al, two-thirds of the patients were AChR antibody positive and 83% showed a pathological decrement in the accessory, facial and/or median nerves. The Tensilon test was positive in all patients examined [10,11]. Distal weakness in MG often improves with immunosuppressive therapy, although mild weakness in the hand muscles or other muscles may persist in the setting of normalized proximal limb strength [9-12]. Our patient was consistent with this scenario. His ADM involvement was noticed two months after the onset of extra ocular symptoms. Chest CT was normal, and AChR antibody was positive. All symptoms and signs disappeared after steroid treatment, with only minor fluctuation of ptosis and no relapse involving other muscles. Early treatment when the patient was still in his early course and persistent low-dose steroid may be the underlying mechanism of the good outcome.

This case suggests that ulnar nerve RNS may be sensitive in detecting distal involvement of MG. The standard Jolly test includes few examinations on distal muscles. Bilateral proximal and distal nerve RNS and more examinations on distal muscle strength should be performed in MG patients with suspected distal involvement.

## Acknowledgement

This study is supported by the National Natural Science Foundation of China (Grant No. 81070963) and Shandong Provincial Natural Science Foundation (Grant No. ZR2010HM019).

## References

1. Misra UK, Kalita J, Srivastava A. A study of diagnostic yield, technical ease and patient discomfort of low rate repetitive nerve stimulation test in patients with myasthenia gravis. Electromyogr Clin Neurophysiol. 2006; 46: 337-341.
2. Witoonpanich R, Dejthevaporn C, Sriphrapradang A, Pulkes T. Electrophysiological and immunological study in myasthenia gravis: diagnostic sensitivity and correlation. Clin Neurophysiol. 2011; 122: 1873-1877.
3. Wang W, Chen YP, Wang ZK, Wei DN, Yin L. A cohort study on myasthenia gravis patients in China. Neurol Sci. 2013; 34: 1759-1764.
4. Cui LY, Guan YZ, Wang H, Tang XF. Single fiber electromyography in the diagnosis of ocular myasthenia gravis: report of 90 cases. Chin Med J (Engl). 2004; 117: 848-851.
5. Costa J, Evangelista T, Conceição I, de Carvalho M. Repetitive nerve stimulation in myasthenia gravis--relative sensitivity of different muscles. Clin Neurophysiol. 2004; 115: 2776-2782.
6. Witoonpanich R, Barakul S, Dejthevaporn C. Relative fatigability of muscles in response to repetitive nerve stimulation in myasthenia gravis. J Med Assoc Thai. 2006; 89: 2047-2049.
7. Liang YX, Chen ZY, Ge H, Liao SJ, Yao XL. The sensitivity analysis of electrophysiological examination for ocular myasthenia gravis. Chin J Clin Neurosci. 2010; 18: 407-409.
8. Chen YP, Wang W, Wei DN. Clinical value of low-frequency repetitive nerve stimulation in myasthenia gravis. Chin Med J. 2011; 91: 1178-1180.
9. Nations SP, Wolfe GI, Amato AA, Jackson CE, Bryan WW, Barohn RJ. Distal myasthenia gravis. Neurology. 1999; 52: 632-634.
10. Werner P, Kiechl S, Löscher W, Poewe W, Willeit J. Distal myasthenia gravis frequency and clinical course in a large prospective series. Acta Neurol Scand. 2003; 108: 209-211.
11. Iwasaki Y, Igarashi O, Kawabe K, Kiyozuka T, Kawase Y, Aoyagi J, et al. MG with distal muscle involvement. Acta Neurol Scand. 2004; 110: 271-272.
12. Karacostas D, Mavromatis I, Georgakoudas G, Artemis N, Milonas I. Isolated distal hand weakness as the only presenting symptom of myasthenia gravis. Eur J Neurol. 2002; 9: 429-430.

Austin J Clin Case Rep - Volume 1 Issue 1 - 2014
**ISSN : 2381-912X** | www.austinpublishinggroup.com


**Citation:** Wang Q, Li H-f. Asymmetric Involvement of Abductor Digiti Minimi Found by Repetitive Nerve Stimulation in Myasthenia Gravis. Austin J Clin Case Rep. 2014;1(1): 1001.