대한임상신경생리학회지 17(1):35-37, 2015
http://dx.doi.org/10.14253/kjcn.2015.17.1.35
eISSN 2288-1026 pISSN 1229-6414

Brief Communication

안구근육 침범 없이 양쪽얼굴신경마비로 나타난 중증근무력증

제주대학교 의학전문대학원 신경과학교실

강철후 · 김홍전 · 오정환 · 강사윤

Myasthenia Gravis Presenting with Facial Diplegia without Ocular Muscle Involvement

Chul-Hoo Kang, Hong Jun Kim, Jung-Hwan Oh, Sa-Yoon Kang

Department of Neurology, Jeju National University School of Medicine, Jeju, Korea

Key Words: Acetylcholine receptor, Facial palsy, Myasthenia gravis

Received 2 August 2014; received in revised form 6 January 2015; accepted 29 May 2015.

Bilateral simultaneous facial palsy is a rare clinical entity that can result from various etiologies and thus representing a diagnostic challenge.[1] Myasthenia gravis (MG) is an autoimmune disorder usually caused by antibodies directed towards the skeletal muscle nicotinic acetylcholine receptor that lead to a compromise of neuromuscular transmission. Facial palsy is common in MG, but usually occurs together with ocular involvement. The involvement of facial muscles as a presenting feature without ocular weakness is extremely rare.[2-4]

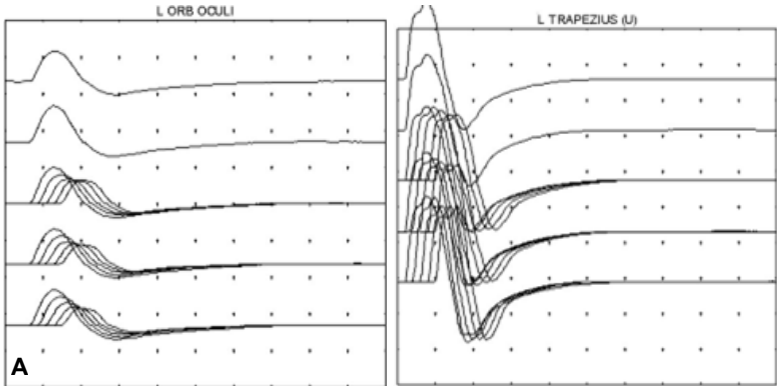
Case Report

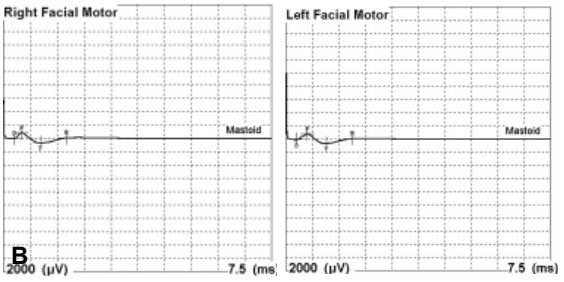
A 36-year-old woman presented with a 1-year history of difficulty in facial expression. Her medical history was unremarkable. Neurological examination showed bilateral facial weakness with lagophthalmos. The patient could not whistle or blow her cheeks. Mild dysarthria was present. Limitation of extraocular muscle and ptosis were not observed. The patient had no problems with tearing, taste, and hearing. Limb muscle strength, sensory examination, and deep tendon reflexes were normal. Pathologic reflexes, cerebellar ataxia, and autonomic dysfunction were not observed. Brain MRI and cerebrospinal fluid examination were normal. There were neither clinical nor laboratory findings of systemic disease. Blink reflex test showed no response, but facial nerve stimulation demonstrated normal distal latency and bilateral reduced amplitude of compound muscle action potential. Repetitive nerve stimulation test revealed a decremental response in the orbicularis oculi and trapezius muscles (Fig. 1). Pharmacologic test with neostigmine showed improvement in facial weakness. Serum acetylcholine receptor antibody titer was significantly increased at 7.905 nmol/L (normal < 0.2). A diagnosis of MG was made based on serological, pharmacological and electrophysiological results. Chest CT was normal. The patient had moderate improvement in bilateral facial weakness and speech on oral pyridostigmine. Diplopia and ptosis were absent on consecutive examination for a year follow-up with pyridostigmine therapy.

Address for correspondence;
Sa-Yoon Kang
Department of Neurology, Jeju National University School of Medicine, 15 Aran 13-gil, Jeju 690-767, Korea
Tel: +82-64-754-8175 Fax: +82-64-717-1630
E-mail: neurokang@jejunu.ac.kr

*This research was supported by the 2014 scientific promotion program funded by Jeju National University.

Copyright 2015 by The Korean Society of Clinical Neurophysiology
This is an Open Access article distributed under the terms of the Creative Commons Attribution Non-Commercial License (http://creativecommons.org/licenses/by-nc/3.0) which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.





Motor Nerves

Site	NR	Onset (ms)	Norm Onset (ms)	P-T Amp (mV)	Norm Amp (mV)	Neg Dur (ms)	Segment Name
Right Facial (Obic Oris)							
Mastoid		2.93	<3.08	1.50	>1.1	4.69	Mastoid-Obic Oris
Left Facial (Obic Oris)							
Mastoid		2.93	<3.08	1.43	>1.1	5.39	Mastoid-Obic Oris

Figure 1. The results of repetitive nerve stimulation test and facial nerve conduction study. The repetitive nerve stimulation at 2 Hz recording over the orbicularis oculi (left) and trapezius (right) muscles showed decremental response (A). Facial nerve conduction study showed normal distal latency and bilateral reduced amplitude of compound muscle action potentials (B).

Discussion

Bilateral simultaneous facial palsy is a rare neurological manifestation of diverse causes such as Lyme disease, Guillain-Barré syndrome, trauma, and sarcoidosis.[5] Diagnostic workup for a patient presenting with bilateral facial palsy depends on the history. The history should include time sequence of onset, prior history of facial palsy, recent viral or upper respiratory infection, otological symptoms, change in taste, or recent immunization. The first priority in the workup is to rule out a life-threatening disease such as leukemia or Guillain-Barré syndrome. However, to our knowledge, there are a few reports of facial diplegia as the only manifestation of MG.

More than 75% of MG patients present with visual complaints of droopy eyelids or double vision. Differences in acetylcholine receptor isoform expression, safety factor, firing frequency and sensitivity to complement deposition have been suggested to explain the increased vulnerability of extraocular muscles in MG compared to other skeletal muscles. The facial and bulbar muscles are frequently affected in generalized MG and their weakness occurs in about 5-10% of patients at the onset of the disease.[6] However, the involvement of facial muscles without ocular weakness is extremely rare at the presentation of MG. Three similar cases have been previously reported in literature.[2-4] Two of the reported patients were not serologically confirmed, and MG was associated with thymoma and clinical remission followed thymectomy.[3,4] Jeannet et al. reported isolated facial and bulbar paresis in a neonatal MG patient.[7] However, this case was different in that the patient neither had acetylcholine receptor antibody, nor responded to any treatment. Prominent facial and bulbar involvement is also encountered in MG patients with anti-MuSK antibody. Although we did not evaluate the anti-MuSK antibody in our patient, it was unlikely that our patient had anti-MuSK antibody considering the favorable response to oral pyridostigmine.

The present case described a patient with MG presenting as bilateral facial weakness and dysarthria without ocular signs. We suggest that the patient with bilateral simultaneous facial palsy may need to be screened for MG even in the absence of ocular muscle involvement.

REFERENCES

1. Keane JR. Bilateral seventh nerve palsy: analysis of 43 cases and review of the literature. *Neurology* 1988;44:1198-1202.
2. Cucurachi L, Cattaneo L, Gemignani F, Pavesi G. Late onset

generalized myasthenia gravis presenting with facial weakness and bulbar signs without extraocular muscle involvement. *Neurol Sci* 2009;30:343-344.
3. Chia LG. Facial weakness without ocular weakness in myasthenia gravis. *Muscle Nerve* 1988;11:185-186.
4. Kini PG. Juvenile myasthenia gravis with predominant facial weakness in a 7-year-old boy. *Int J Pediatr Otorhinolaryngol* 1995;32:167-169.
5. Jain V, Deshmukh A, Gollomp S. Bilateral facial paralysis: case presentation and discussion of differential diagnosis. *J Gen Intern Med* 2006;21:7-10.
6. Keesey JC. Clinical evaluation and management of myasthenia gravis. *Muscle Nerve* 2004;29:484-505.
7. Jeannet PY, Marcoz JP, Kuntzer T, Roulet-Perez E. Isolated facial and bulbar paresis: a persisent manifestation of neonatal myasthenia gravis. *Neurology* 2008;70:237-238.