대한임상신경생리학회지 7(2):117~120, 2005 ISSN 1229-6414

# 급성 벨마비와 동시에 발생한 반대측 반얼굴연축

대구가톨릭대학교 의과대학 신경과학교실

**이 동 국**

# Contralateral Hemifacial Spasm Occurred Simultaneously in Acute Bell's Palsy

**Dong Kuck Lee, M.D.**

*Department of Neurology School of Medicine Catholic University of Daegu*

Hemifacial spasm (HFS) may develop after Bell's palsy (BP). But it was not reported that contralateral HFS occurred simultaneously in acute BP. A 25-year-old woman admitted with left HFS occurred simultaneously in acute right BP for 6 days. Past, family, and social history were unremarkable. Nerve conduction studies (NCS) and blink reflex (BR) test showed bilateral facial neuropathies. Brain MRI and cerebral angiography were normal. The symptoms and signs of HFS and BP were improved slowly after acyclovir and prednisolone therapy. Follow-up serial NCS and BR also showed a rapid improvement.

**Key Words**: Bell's palsy, Contralateral hemifacial spasm

Peripheral facial nerve paresis or paralysis is the most common manifestation of a seventh cranial nerve lesion because of its innervation of the muscles of facial expression.[1] So the appearance of a severe, unilateral, infranuclear facial palsy is one of the most distinctive disorder in clinical medicine. The most common cause of an acute, acquired, nontraumatic lesion is Bells' palsy (BP), in which there is a flaccid paresis of all mimetic muscles on the involved side.[2] But the etiology and pathophysiology of BP remain unknown. The diagnosis is one of exclusion, so most cases of facial paralysis reflect idiopathic BP.[3] BP is generally a benign condition but may complicates various disorders as hemifacial spasm (HFS), postparalytic movements and synkinesis, and crocodile tears.[4]

Address for correspondence
**Dong Kuck Lee, M.D.**
Department of Neurology School of Medicine
Catholic University of Daegu
3056-6 Daemyung 4 dong Namgu Daegu 705-718 Korea
Tel: +82-53-650-4267 Fax: +82-53-654-9786
E-mail : dklee@cu.ac.kr

## Case presentation

A 25-year-old woman admitted with left HFS occurred simultaneously with acute right BP for 6 days. She was a healthy clerk and past, family, and social history were unremarkable. Six days before admission, she noticed a left eyelid twitching. But she observed it without treatment. Four days before admission, she noted a mouth angle deviation to left at mornig. And left eyelid twitching was spread slowly to left all face. So she treated at local neurosurgical and oriental medical clinic but effect was absent.

Vital signs were stable and general physical examinations were normal. And neurologic examination showed a left HFS with a right BP. But other neurologic examinations including deep tendon reflexes were normal. CBC, ESR, EKG, admission pannel, chest X-ray, CSF studies and urinalysis test were normal. Nerve conduction studies (NCS) and blink reflex (BR) showed bilateral facial neuropathies. Brain MRI and MRA studies were normal. The symptoms and signs of HFS and BP were improved slowly after acyclovir and prednisolone therapy. Follow-up serial NCS

 

and BR also showed a rapid improvement in a month of disease onset.

## Discussion

BP is one of the commonest conditions seen in neurologic practice, accounting for approximately 50% of the cases of peripheral facial paralysis. In about 60 to 75% of patients with facial palsy, no cause is found, and the syndrome is then termed idiopathic facial palsy, also known as BP.[4-6]

But bilateral facial paralysis (facial diplegia) is uncommon, occurring less than 1% as frequently as unilateral paralysis. Bilateral involvement may be due to congenital developmental anomalies, or associated with infectious, traumatic, postinfectious, granulomatous, or neoplastic processes. Facial diplegia is most often a manifestation of the Guillain-Barré syndrome and may also occur in Lyme disease. And bilateral facial paralysis may also occur with collagen vascular diseases or osteopetrosis and may rarely be idiopathic (bilateral BP).[5,7]

The cause of BP is still unknown. Most probably, it is a mononeuritis cranialis caused either by viral infection or an autoimmune disease. Also ischemia may be involved in the etiology.[8] My patient was an idiopathic peripheral facial palsy.

Typically in BP, the R1 and R2 responses of BR test may be preserved within the first day or two after the onset of symptoms and then become progressivly delayed or unobtainable in the following days, with a peak in the conduction delay at approximately 5 days. Because of this evolving process and the possibility of late degeneration, it is generally agreed that prognostic assessment cannot be made until after the end of the first week and that serial studies are more helpful. The prognosis for recovery of function in BP is considered favorable if the R1 component persists or reappears after the first week and unfavorable if the R1 and R2 remain absent after the first 3-4 weeks. It is important to understand that the BR is complementary to the direct response and that both should be examined for prognostic purposes. The BR can also be of value in documenting and quantifying the presence of aberrant regeneration with synkinesis after BP.[9]

Clinically my patient showed a right BP but electrophysiologically she revealed bilateral facial neuropathy by NCS and BR test. And electrophysiologic findings showed a progressive improvement as clinical recovery in a month.

In late phase of facial palsy without degeneration, the latency of R1 is usually delayed by a few milliseconds and returns to normal in 2 to 4 months. In patients with substantial degeneration, R1 is absent and the direct motor response is markedly reduced in amplitude for months up to a few years. In patients with severe facial nerve axonal damage, axonal sprouting and regeneration will occur. Facial synkinesis occurs in most patients who have experienced significant axonal degeneration. The most probable cause is abnormal regeneration due to excessive branching of axons at the lesion or ephaptic transmission. However, some evidence indicates changes in the facial nucleus as well, because lesions involving just one facial branch may give rise to synkinesis in all hemifacial muscles. Eekhof et al[10] recorded ephaptic transmission in 50% of their patients, who developed facial synkinesis after BP, and suggested that there may be some alteration in the facial nucleus exitability.[6]

In HFS, there are recurring episodes of involuntary unilateral spasms of the facial musculature invariably beginning about the lateral canthus of the eye on the involved side and gradually spreading downward, over many months to several years to involve the remainder of the facial musculature[3].

HFS may be develop as a sequela to BP but may also be due to an irritative lesion of the facial nerve. However, in the most common form of HFS, the cause and pathology are unknown.[11] But the cause of the HFS is presumed to be a compression of the facial nerve at the root exit zone by a blood vessel. Theories propose ephaptic transmission between nerve fibers or a hyperactivity of the facial motor nucleus as a result of the nerve irritation. When performing microvascular decompression as a therapy, surgeons have identified PICA or AICA in varying proportions and less often the vertebral artery as responsible for about 90% of the cases. Occasionally, a vein is identified, while up to 25% of patients exhibit more than one vessel as possible cause. Only rarely is a tumor or a vascular malformation discovered. While HFS can be diagnosed clinically, tumors and other nonvascular causes should be

excluded by neuroimaging. EMG showes synchronous arrhythmic discharge in different ipsilateral facial muscles.[8] Therefore pathophysiology of the HFS is believed to be nerve root compression and demyelination. The demyelinated axon is capable of activating adjacent nerve fibers by ephaptic transmission. Another possible source of the spasm is ectopic exitation arising in injured fibers.[4,7]

Electrophysiologic techniques have been used by various investigators to try to support either of the two main theories as to the etiology of HFS : ephaptic transmission between adjacent facial nerve fibers at a site of localized compression, or alteration of the excitability threshold of the facial motor nucleus secondary to peripheral injury of the nerve leading to increased firing of the facial motor nerve fibers.[1]

The loss of contralateral synkinesis after unilateral facial nerve microvascular decompression surgery in a patient with bilateral HFS also suggests a role of the facial motor nucleus, with loss of facilitatory effect from the side ipsilateral to the surgical procedure.[12] My case also can be understood by this concept.

Kimura et al.[13] suggests that clinical and electrophysiologic evidence of synkinesis occurring both sporadically or after ipsilateral facial nerve paralysis makes it less likely that it is due to aberrant regeneration of facial nerve fibers.

And the phenomenon of synkinesis has also been argued to be due to a peripheral facial nerve lesion. Moller and Jannetta[14] proposed that synkinesis was generated by a different process than HFS, most likely by aberrant reverberation or a kindling phenomenon in the facial motor nucleus, with antidromic nerve impulses arising from a peripheral trigger zone such as the root entry zone as a result of vascular compression and spreading to the facial motor nucleus, leading to orthodromic stimulation of motor nerve branches and therefore synkinesis.

Ferguson[15] proposed that HFS occurred as a result of reorganization of the facial motor nucleus after peripheral injury or compression, thereby unmasking normal and new reflexive and interconnecting pathways leading to multiple combinations of facial motor movements during HFS, and that ephaptic transmission is too limited in its potential scope to explain the generation of stereotyped facial spasm. Because the facial motor nucleus has multiple unique afferent inputs (spinal cord, red nucleus, superior colliculus), such a central reorganization is possible.

BP is generally a benign condition but may complicates various disorders as HFS, postparalytic movements and synkinesis, and crocodile tears.

Differential diagnoses of HFS include facial synkinesis after facial nerve palsy (increased latencies of R1 and R2 of the BR on the affected side), facial myokymia (constant rapid undulation of facial muscles, such as multiple sclerosis), and facial tic (brief stereotypic complex movement).[8] And HFS also must be differentiated clinically from other facial movement disorders such as blepharospasm, oromandibular dystonia, facial focal motor seizures, craniofacial myoclonus, hemimasticatory spasm, cephalic tetanus, and craniofacial tremor.[1]

The postparalytic HFS, though infrequent, should be differentiated from the primary HFS, as well as from synkinesis due to aberrant regeneration after BP (post-Bells' palsy synkinesis). In all three conditions, synkinesis between upper and lower facial muscles is clinically evident. Similarly, synkinetic responses are often evoked by selective stimulation of facial nerve branches or the supraorbital nerve. Simultaneous recording of the ongoing activity of orbicularis oculi and orbicularis oris may show that post-Bells' palsy synkinesis every burst of activity induced in the orbicularis oculi-that is, in spontaneous or reflex blinks-spreads to the orbicularis oris. On the contrary, in HFS, spread occurs on many, but not all, occasions.[6]

And postparalytic facial spasms generally reflect either fixed contraction of the facial muscles or synkinesis or both. There is always a history of preceding facial weakness, and spontaneous spasms are generally absent. Spastic paretic facial contracture may be confused with HFS. The involved side is weak and contracted, unlike the situation in HFS, in which the facial muscles are relaxed between twitches. Facial myokymia is characterized clinically by undulating movements of the facial muscles. Facial tics are rapid, stereotyped, and relatively well coordinated movements that may also occur in regions distant from the face. Hemimasticatory spasm

involves muscles innervated by both the facial and trigeminal nerves.[4]

Montagna et al.[16] examined the changes in idiopathic versus postparalytic HFS during sleep and found that, although HFS may persist during sleep, it was reduced significantly in both types of patients during deeper stages of sleep. They proposed that this partial diminution of HFS during sleep stages is suggestive of central inhibition of a central facial nuclear component involved in the generation of the spasm. My patient also showed reduced HFS during deep sleep.

HFS may develop after BP. But it was not reported that contralateral HFS occurred simultaneously in acute BP. Clinically the patient showed a right BP but electrophysiologically she revealed bilateral facial neuropathies by NCS and BR test. So left HFS may related with ipsilateral peripheral facial neuropathy.

HFS may be develop as a sequela to BP but may also be due to an irritative lesion of the facial nerve. However, in the most common form of HFS, the cause and pathology are unknown. But the cause of the HFS is presumed to be a compression of the facial nerve at the root exit zone by a blood vessel. But my case did not show any abnormalities in MRI and MRA studies. I can guess the HFS evoked due to ephaptic transmission between adjacent facial nerve fibers at a site of localized compression or alteration of the excitability threshold of the facial motor nucleus secondary to peripheral injury of the nerve leading to increased firing of the facial motor nerve fibers.

## REFERENCES


1. Klein CM. Diseases of the seventh cranial nerve. In: Dyck PJ, Thomas PK. Peripheral neuropathy. 4th ed. Vol. 2. *New York: Elsevier Saunders*, 2005;1219-1252.
2. Sweeney PJ, Hanson MR. Cranial neuropathies. In: Bradley WG, Daroff RB, Fenichel GM, Jankovic J. Neurology in clinical practice. 4th ed. Vol. 2. *New York: Butterworth Heinemann*, 2004;2107-2117.
3. Griffin JW. Peripheral neuropathies. In: Goldman L, Ausiello D. Cecil's textbook of medicine. 22nd ed. Vol. 2. *New York: Saunders*, 2004;2379-2387.
4. Brackmann DE, Fetterman BL. Cranial nerve VII: facial nerve. In: Goetz CG. Textbook of clinical neurology. 2th ed. *New York: Saunders*, 2003;181-194.
5. Brazis PW, Masdeu JC, Biller J. Localization in clinical neurology. 4th ed. *New York: Lippincott williams & Wilkins*, 2001;289-307.
6. Aramideh M, Valls-Sole J, Cruccu G, Ongerboer de Visser BW. Assessment of disorders of the cranial nerves. In: Brown WF, Bolton CF, Aminoff MJ. Neuromuscular function and disease. 1st ed. *New York: Saunders*, 2002;757-780.
7. Ropper AH, Brown RH. Adams and Victor's principles of neurology. 8th ed. *New York: McGraw-Hill*, 2005;1178-1190.
8. Schrader V, Sumner A. Idiopathic(Bell's) facial palsy. In: Brandt T, Caplan LR, Dichgans J, Christoph Diener H, Kennard C. Neurological Disorders course and treatment. 1st ed. *New York: Academic Press*, 1996;113-116.
9. Lucuano CA. Other useful electrodiagnostic techniques: blink reflex, masseter reflex, and silent periods. In: Bertorini TE. Clinical evaluation and diagnostic tests for neuromuscular disorders. 1st ed. *New York: Butterworth Heinema*n, 2002;239-245.
10. Eekhof JLA, Aramideh M, Speelman JD, et al: Blink reflex and lateral spreading in patients with synkinesia after Bell's palsy and in hemifacial spasm. *Eur J Neurol* 2000;43:141-146.
11. Flint Beal M, Hauser SL. Trigeminal neuralgia, Bell's palsy, and other cranial nerve disorders. In: Kasper DL, Braunwald E, Fauci AS, Hauser SL, Longo DL, Larry Jameson J. Harrison's principles of internal medicine. 16th ed. Vol. 2. *New York: McGraw-Hill*, 2005;2434-2437.
12. Schulze-Bonhage A, Ferbert A. Electrophysiological recordings in bilateral hemifacial spasm. *J Neurol Neurosurg Psychiatry* 65:408-410;1998.
13. Kimura, J, Rodnitzky, RL, Okawara SH. Electrophysiologic analysis of aberrant regeneration after facial nerve paralysis. *Neurology* 25:989-993;1975.
14. Moller AR, Jannetta PJ. On the origin of synkinesis in hemifacial spasm: results of intracranial recordings. *J Neurosurg* 61:569-576;1984.
15. Ferguson JH. Hemifacial spasm and the facial nucleus. *Ann Neurol* 4:97-103;1978.
16. Montagna P, Imbriaco A, Zucconi M, et al. Hemifacial spasm in sleep. *Neurology* 36:270-273;1986.