Ann.Bull.RILP(1986)No.20, 225-234

IDIOPATHIC ASSOCIATED LARYNGEAL PALSY**

Noriko K. Nishizawa* and Hajime Hirose

1. Introduction

Associated laryngeal palsy is a symptom complex of laryngeal palsy with the involvement of other cranial nerves. Historically, eponymic nomenculature was applied to various combinations of cranial nerve involvement.[1] An enormous number of reports have described the etiology: central and peripheral, tumorous, vascular and degenerative diseases.[2] On the other hand, there is a concept of "idiopathic" associated laryngeal palsy. It is a diagnosis of elimination. As in the case of Bell's palsy and idiopathic recurrent nerve palsy, the origin and etiology of this rather uncommon condition remain unknown.

2. Case Study

Table 1 summarizes the results of a survey on the causes of associated laryngeal palsy in 40 clinical cases observed for the last ten years at the Voice and Speech Clinic, University of Tokyo Hospital. Motor neuron disease was excluded from this study because of its exceptional clinical course and pathogenesis.[2] In 11 cases, intracranial lesions such as tumors, degenerative diseases of the central nervous system and cerebrovascular diseases were found to be the origin of the palsy. In 8 cases, extracranial lesions were suspected. In the remaining 21 cases, the pathogeneses remained unknown even after exhaustive examination, and the diagnosis of idiopathic associated laryngeal palsy was made.

Table 1. Cause of associated laryngeal palsy in 40 cases

intracrninal tumor	3
operation for intracranial tumor	2
extracranial tumor	1
operation for extracranial tumor	5
syringobulbia	2
cerebrovascular disease	3
sarcoidosis	1
trauma	2
idiopathic	21
TOTAL	40

* Department of Otolaryngology, National Rehabilitation Center Hospital, Tokorozawa

** Paper presented at 1986 Neurolaryngology Conference, Kurume, May, 1986

The clinical features of the 21 cases with idiopathic associated laryngeal palsy are shown in Table 2. All cases were over 30 years of age. There was no preponderence of either sex. Some characteristic features could be pointed out about the neurological symptoms.

The palsy was unilateral in 20 cases, while the remaining one case showed crossed paralysis. Symmetrical paralysis was not observed in this study. In 15 cases, the palsies were restricted to the lower cranial nerves: glossopharyngeal, vagal, accesory and hypoglossal nerves. In the other 6 cases, three showed palsies of the lower cranial and facial nerves; one showed palsies of the lower cranial, facial and motor fibers of the trigeminal nerve; and the remaining two developed broad symptoms of upper and lower cranial nerve involvement. Second, dysfunctions of the motor fibers predominated in all the cases. Pure motor palaryses were seen in 18 cases. Minor sensory symptoms, such as hemifacial paresthesia, were observed in three cases. Pyramidal tract signs and cerebellar symptoms were noted in two cases.

As antecedent events, acute upper respiratory infections and irritable pain around the temporal region were noticed in several cases. In one case, an operation under lumbar anesthesia preceded the onset of the paralysis. The onset was acute or subacute in 16 cases in which the palsy became complete within one week. In the other 5 cases, the palsies relapsed or developed gradually over many years. Recovery occurred within several months in 11 cases. In 6 cases, no recovery was seen after a year-long follow-up. The remaining 4 cases are lost to follow-up.

3. Discussion

As seen above, the present series can not be considered homogeneous clinically. Idiopathic associated laryngeal palsy in this study may correspond to those syndromes reported under the name of polineuritis cranialis[3)4)] or multiple cranial neuritis.[5)] As for cranial polineuropathy of unkown etiology, Toyokura[3)] has stated that 1) polineulitis cranialis is not a single disease entity but a syndrome; and 2) polineuritis cranialis is a neuropathy that mainly involves multiple cranial nerves, almost always symmetrically, with or without slight neurological signs involving limbs and trunk. Since the etiology of these syndromes remains unestablised,[6)] we should be careful about subdividing or unifying these cases under hypothetical pathogeneses.[3)4)] Nonetheless, it might be rewarding to analyze the clinical features of these cases from an etiological aspect with special reference to several neurological disorders reported in the literature.

1) Guillain-Barré Syndrome

Guillain-Barré syndrome is an acutely, or subacutely,

Table 2. Clinical manifestations of the 21 cases of idiopathic associated laryngeal palsy

case	III eye	IV	VI	V jaw	VII face	X s.p.	X v.c.	X ct	XI	XII	V sensory	cerebellar	pyramidal
1. 61 F						l	l						
2. 53 F						r	r	?					
3. 36 M						l	l	?					
4. 34 M						l	l	l	l				
5. 62 F						r	r	r	r				
6. 67 M						l	l	l	l				
7. 62 M						r	r	r	r				+
8. 41 F						l	l						+
9. 78 F						l	l	?		l			
10. 33 F						l	l	l		l			
11. 61 M					rl		r	r					
12. 64 F	r	r	r	r	r	r	r	r	r		r		
13. 38 M	l	l	l		l	l	l	?	l		l		
14. 67 F						l	l	l	l				
15. 32 F						r	r	r					
16. 43 M					l	l	l	l					
17. 64 M						l	l	l	l				
18. 54 F						l	l		l	l			
19. 40 F						l	l	?	l				
20. 32 M				l	l		l						
21. 69 M					r	r	r	r	r	r			

s.p.; soft palate, v.c.; vocal cord, c.t.; cricothyroid muscle
r; right, l; left, rl; bilateral, ?; not examined

other manifestations	case no.
1. antecedent event	
cold	3,4,5, 10,11, 20
pain around the ear	3,4, 7, 13, 16
operation	17
2. clinical course	
acute evolution	1,2,3,4,5,6,7,8,9, 14,15,16,17,18,19,20
recovery	1, 3,4,5,6,7,8, 16,17, 19,20
recurrent	10,11,12,13, 21

evolving paralytic disease of unestablished etiology with acellular hyperalbminosis of the cerebrospinal fluid.[7] The palsy may be confined to the cranial nerves without involvement of the other peripheral nerves.[8] In a broad sense, all of the cases in the present study could be included in this syndrome or its subtypes.[4][5] However, the clinical manifestations of the present cases were not strictly compatible with those of the typical Guillain-Barré syndrome. Table 3 is a comparison between the typical type of Guillain-Barré syndrome[9] and the present cases. As can be seen in the table, the typical clinical course of the Guillain-Barré syndrome is rather uniform. In our study, 13 cases, i.e., cases No. 1,3,4,5,6,9,14,15,16,17,18,19 and 20, showed an acute development, benign prognosis and pure motor paralysis without long tract sign. They were thought to be compatible with the criteria of this syndrome. The other 8 cases should be differentiated from this typical type by the chronic or progressive course of their disease and complications of the long tract or cerebellar signs.

Table 3. Comparison between the typical manifestations of the Guillain-Barré syndrome and the present cases

	Guillain-Barré syndrome 26 cases from Sobue[9]	present study 21 cases
under 30 years old	62%	0%
rate for male patients	77%	48%
acute evolution within several weeks	100%	76%
recovery within several months	100%	67%
antecedent event	88%	29%
motor palsy dominant	100%	100%
bilateral	100%	10%
acellular hyperalbminosis of CSF	100%	-
long tract or cerebellar sign	0%	10%

In several of our 13 cases compatible with the Guillain-Barré syndrome, we noticed antecedent events of acute upper respiratory infection or irritable pain around the temporal region, which are known to be common in the Guillain-Barré syndrome. On the other hand, some differences can be pointed out such as age distribution[9)10)] and sex preponderence.[9)11)] In particular, the palsies in these 13 cases were restricted to the unilateral cranial nerves caudal to the trigeminal nerve, while in the Gillain-Barré syndrome symmetrical palsies are common[12)] even in cases of pure cranial palsy.[4)13)8)]

There is an agreement today as to the participation of an aberrant immune response in the pathogenesis of the Guillain-Barré syndrome.[6)15)] On the other hand, Nozoe[14)] has reported three cases of unilateral lower cranial nerve palsy under the diagnosis of polineuritis cranialis while suspecting the participation of a viral infection. Through the existence of polyneuropathy that resembles a Guillain-Barré syndrome restricted to the unilateral lower cranial nerves, it has been suggested that not only systemic reactions, such as allergy, but also local factors, such as viral infection, play an important role in the pathogenesis of this syndrome.

ii) Viral Infection of the Cranial Nerves

Herpes zoster and herpes symplex viruses are supposed to be a causative agent of peripheral nerve palsy.[2)] Accumulating information points to common modes of behavior for these viruses within the peripheral nervous system.[16)]

Hunt[17)] has suggested that herpes zoster invasion in the geniculate ganglion might be responsible for the complication of herpes zoster otics and the involvement of the seventh and eighth cranial nerves. Further, he has pointed out that herpetic lesions of the ganglia of the glossopharyngeal and vagal nerves might be responsible for herpes zoster otics by transmission via Arnold's nerve.[18)] Engström[19)] and Font[20)] have suggested that the region of the auricle and external auditory canal is supplied by the 5th,7th,9th,10th and the upper cervical spinal nerves, and that it is possible for paralysis of the facial nerve with herpetic lesions on or about the auricle to occur from involvement of the ganglia of any of these nerves by zosterian inflammation. Font[20)] has further reported "zoster sine herpete"[21)] and suggested that idiopathic associated laryngeal palsy might be the result of viral infection. These historical discussions are suggestive regarding the possibility of a peripheral pathway for viral invasion into the lower cranial nerves. That is, except for two cases with oculomotor palsies, most of the neurological symptoms of the present cases are explainable by viral transmissions through the peripheral pathways discussed above and through the pathway via the ansa hypoglossi to the hypoglossal nerve.

Recent studies have implicated the herpes simplex virus as a

causative agent in Bell's palsy.[22] Adour[23] and Djupesland[24] have suggested that some reactivating stimulus such as a cold can create an active state of infection of the viruses latent in the ganglion cells and produce cranial polineuritis. Djupesland[24] has suggested not only the peripheral but also the central pathways for the transmission of the virus.

In the present cases, herpetic vesicles diagnostic of herpes infection were not found. In serological study, cases 1, 9 and 21 showed a high titer of the herpes symplex virus. Case 21 showed an especially high titer suggesting a relapsing neuritis caused by herpes symplex virus.

Other viruses such as influenza A2,[25] influenza B[26] and adenovirus[27] have been suggested to be responsible for peripheral neuritis, especially for recurrent nerve palsy. It is still unknown whether these viruses directly invade the periphral nerves, or only act as a trigger for a systemic reaction such as a neuroallergy.

iii) Brain-Stem Encephalitis

In the nucleus ambiguus, the motoneuron pools of the individual muscles have a topographic distribution in relation to one another, rather than being randomly intermingled.[28][29] Consequently, selective palsy of individual muscles could result from localized intranuclear lesions, as in the case of bulbar poliomielitis.[30]

Under the diagnosis of brain-stem encephalitis, Möller[31] has reported 40 cases of unilateral lower cranial nerve palsy with antecedent events of upper respiratory infection or irritable pain in the temporal region, and with an acute development and benign prognosis. He supposes the origin of the palsy to have been a focal lesion in the lower brain-stem for the following reasons: 1) The injury was unilateral, while neuritis and radiculitis often cause bilateral symmetrical paresis. 2) Dissociated paresis, such as paresis of only the vocal cord function of the Xth nerve, indicated nuclear, paranuclear or intranuclear injury rather than injury to the nerve fibers. 3) Mild symptoms of other conduction tracts, especially of the pyramidal tract, occurred to a large extent. Histopathological evidence of degenerative change in the brain-stem with little change in the peripheral nerves has been reported by Arima[32] in the autopsy of a patient with acute cranial nerve palsy. Viral infection[24][32][33] or allergic reaction[32] has been suggested as the etiology of brain-stem encephalitis. It is sometimes difficult to distinguish clinically between central and peripheral injury, especially in cases without signs of central nervous system involvement.[31][32][34] In fact, most recent reports on brain-stem encephalitis have suggested complications of the central nervous system such as ataxia.[32][35][36] In the present cases, acute inflammatory lesions in the brain stem were suspected in cases 7 and 8 with long tract and cerebellar signs.

Further, an EMG study of case 8 established a dissociated injury of the Xth nerve with paresis of the soft palate and vocal cord leaving the cricothyroid muscle intact. It has been suggested that a synchronized voltage in EMG studies is sometimes diagnostic of bulbar palsy.[2)37)] Examinations by methods such as EEG[38)] or CSF might[32)] be necessary.

iv) Chronic Inflammatory Poliradiculoneuropathy

There are rare cases of idiopathic polineuropathy that relapse or progress chronically.[6)8)] They have been reported under the name of relapsing inflammatory polyradiculoneuropathy,[6)] recurrent multiple cranial nerve palsy[39)] or polyneuritis cranialis.[3)] The clinical symptoms of this type of relapse do not essentially differ from those of the acute monophasic form of inflammatory polyradiculoneuropathy.[6)] Etiology is unknown, although an allergic reaction is suspected as in the case of the Guillain-Barré syndrome.[1)] In the present cases, there were five cases of relapsing or progressing palsy. Their clinical course closely resembles those of progressing multiple cranial neuropathy reported by Toyokura.[3)] However, in this rare condition, it is still controversial whether there exists a pure cranial nerve injury without other peripheral nerve palsy.

A differential diagnosis should be made among the degenerative diseases of the central nervous system, multiple scrlosis, sarcoidosis[2)] and chronic nonspecific inflammation of the skull base.[40)]

4. Conclusion

Idiopathic associated laryngeal palsy is a rare condition in neurological symptomatology. Differential diagnoses should be made carefully among tumors cerebrovascular disease, multiple scrlosis, sarcoidosis and degenerative diseases of the nervous system. Even after such differentiation, a final diagnosis of "idiopathic" associated laryngeal palsy is by no means a single disease entity. Etiologically, the intranuclear, internuclear or subnuclear injury resulting from an allergic reaction or viral infection can be supposed. The Guillain-Barré syndrome, a direct invasion of herpes virus into the peripheral nervous system or brain-stem encephlitis was suggested in cases of acutely developing palsy with benign prognosis. In almost one quarter of the present cases, the palsy was chronic or relapsing and resembled the clinical course of relapsing inflammatory polyradiculoneuropathy. However, in this rare condition, it is still controversial whether there exists a pure cranial nerve injury without other peripheral nerve palsy. In order to make a precise diagnosis among the possible conditions above, it is important to describe accurately the clinical picture of individual cases, including the results of electrophisiological, serological and CSF study.

The naming of the classic eponymic syndromes applied to diseases of combinations of cranial nerves does not seem to be practical for the diagnosis of associated laryngeal palsy.[1] They can suggest the location of injury only in cases in which a solitary lesion along the cranial nerve pathway involves a function peripheral to the injury. In cases of "idiopathic" associated laryngeal palsy, combinations of cranial nerve injury can be determined by the topography of the motoneuron pools in the brain stem as in cases of brain-stem encephalitis, by the connection between the individual nerves in the periphery as in cases of viral infection and, only quite randomly in cases of systemic reaction, such as neuroallergy.

References

1) Cody, C.C.: Associated paralyses of the Larynx. Ann. Otol. Rhinol. Laryng. 55:549-561, 1946.
2) Hirose, H: Associated laryngeal palsy. Ishii, H. ed. Recurrent nerve palsy. Bunkodo, Tokyo: pp.184-196, 1984.
3) Toyokura, Y: Cranial polyneuritis: Report of a case. Int. Med. JPN., 3:231-238, 1959.
4) Konodo, K. et.al.: Die idiopathische kranielle Polyneuritis. Clin. Neurol. JPN., 3:167-173, 1963.
5) Kawamura, K. et.al: Clinical observation of patients with unilateral involvement of last 4 cranial nerves. Clin. Neurol. JPN., 13:196-202, 1973.
6) Arnason, B.: Inflammatory poliradiculoneuropathies: Peripheral Neuropathy pp.1110-1148, Dyck ed. Saunders, 1975.
7) Hirayama, K.: Clinical concept of the Guillain-Barré syndrome. Neurolg. Med. JPN., 1:277-284, 1974.
8) Ando, K. et.al.: Idiopathic cranial polyneuritis. Neurol. Med. JPN., 3:387-396, 1975.
9) Sobue, I.: Clinical features and variants of the Guillain-Barré syndrome. Neurol. Med. JPN., 1:321-328, 1974.
10) Araki. S: Neuropathy. Neurological Medicine, Kinpodo JPN, pp.605-617, 1974.
11) Wiederholt, W. C. et.al.: The Landry-Guillain-Barré-Strohl syndrome or polyradiculoneuropathy. Historical review, report on 97 patients and present concept. Mayo Clin. Proc., 39:427, 1964.
12) Osler, L.D. et.al.: The Guillain-Barré syndrome. The need for exact diagnostic criteria. New Engl. J. Med., 262:964, 1960.
13) Shibata, H.et.al.: A case of idiopathic acute cranial polyneuritis. Med. J. Minami-Osaka Hosp., 23:53-56, 1975.
14) Nozoe, I. et.al.: Polyneuritis cranialis -report of three cases-. Otolog. Fukuoka JPN., 20:486-490, 1974.
15) Igata, A. et.al: The immunological aspect of the Guillain-Barré syndrome. Neurol. Med. JPN., 1:297-305, 1974.
16) Baringer, J.R. et.al.: Herpes virus infection of the peripheral nervous system. Peripheral Neuropathy pp.1092-1103, Dyck ed. Saunders, 1975.
17) Hunt,R.: Herpetic inflammation of the geniculate ganglion: A new syndrome and its complications. J. Nerv. & Ment. Dis.

34:73, 1907.
18) Hunt, R.: The symptom-complex of acute posterior poliomyelitis of the geniculate auditory glossopharyngeal and pneumogastric ganglia. Arch. Int. Med. 5:631, 1910.
19) Engström, H.E. et. al.: Herpes zoster of the seventh, eighth, ninth and tenth cranial nerves. Arch. Neuro. Psych., 62:638-652, 1949.
20) Font, J.H.: The juglar foramen syndrome. A.M.A. Arch. Otolaryng. 56:134-141, 1952.
21) Lewis, G.W.: Zoster sine herpete. Brit. Med. J. 2:418-421, 1958.
22) McCormick D.P.: Herpes-simplex virus as cause of Bell's palsy. Lancet 1:937-939, 1972.
23) Adour, K.K.: Cranial Polyneuritis and Bell palsy. Arch. Otolaryng. 102:262-264, 1976.
24) Djupesland, G. et.al. : Acute peripheral facial palsy. part of cranial polyneuropathy? Arch. Otolaryng. 103:641-644, 1977.
25) Ishii, H. et.al.: Inflammatory paralysis of the larynx. J. Otolaryng. JPN., 74:1562-1571, 1971.
26) Sato,I et.al.: Idiopathic laryngeal nerve palsy and prevalence of influenza. J. Otolaryng. JPN., 84:601-607, 1981.
27) Wells, C.E.C.: Neurological complications of so called "influenza"; A winter study in southeast Wales. Brit. Med. J., 13:369-373, 1971.
28) Lawn, A.M.: The localization in the nucleus ambiguus of the rabbit of the cells of origin of motor nerve fibers in the glossopharyngeal nerve and various branches of the vagus nerve by means of retrograde degeneration. J. Comp. Neur. 127:293-306, 1966.
29) Yoshida, Y. et al.: Laryngeal motoneurons of cats. J. Otorhinolaryng. JPN., 85:455-463, 1982.
30) Wyke B.D. et. al. : Neurology of the larynx. Hinchcliffe, R. ed. Scientific Foundations of Otolaryngology. Hinemann, London:pp.546-574, 1976.
31) Möller, F.: Brain-stem encephalitis and epidemic vertigo. Acta Psych. Neurol. Scand. 31:107-115, 1956.
32) Arima, M. et.al.: The origin and the diagnosis of encephalitis in children. Advances in Neurol. Science, JPN. 6:241-265, 1962.
33) Ellison,P.H. et.al.: Herpes simplex: A possible cause of brain-stem encephalitis. Pediatrics 59:240-243, 1977.
34) Vernet,M.: The classification of the syndromes of associated laryngeal palsy. J. Laryngol. Rhinol. Otol., 33: 354-365, 1918.
35) Ogawa, A. et.al.: Clinical course of localized encephalitis in children. J. Pediatr. Practice, JPN. 32:226-233, 1969.
36) Iwasa, H. et.al.: Localized encepharitis, A report of four cases. Otolaryng. Tokyo JPN., 47:247-253, 1975.
37) Sawaki, S. et al.: Disturbances of nucleus ambiguus with resulting manifestation of mixed paralysis of the larynx. Otolaryng. Tokyo JPN., 30:736-738, 1958.
38) Bickerstaff, E.R.: Brain-stem encephalitis. further observations on a grave syndrome with benign prognosis. Brit. Med. J. 1:1384-1387, 1957

39) Symonds, C: Recurrent multiple cranial nerve palsies. J. Neurol. Neurosurg. Psychiat., 21:95, 1958.
40) Kamei, T.: Six cases of associated laryngeal paralysis. J. Otolaryng. JPN. 71:12-18, 1968.