

# Ramsay Hunt Syndrome with Hemiparesis and Hemihypoesthesia: Report of 2 Cases

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**Abstract-** Ramsay Hunt syndrome (RHS) complicated with encephalitis is rare, and the appearance of hemi-motosensory deficit in RHS is even rarer. We reported two such cases. Both patients had a peripheral type facial palsy, facial numbness and zoster in the ipsilateral ear canal. The consciousness was clear and cognitive function was normal. The functional deficits of extremities were ipsilateral to the facial palsy in one case, but contralateral in the other. Magnetic resonance imaging of the brain in both cases showed no abnormality. The manifestations were compared with two more cases from the literature.

**Key Words:** Ramsay Hunt syndrome, Encephalitis, Hemiparesis, Hemi-hypoesthesia

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## INTRODUCTION

Varicellar zoster virus (VZV), a double-strand DNA virus, belongs to the family of *Herpesviridae* and the subfamily of  *$\alpha$ -herpesvirinae*. It is transmitted by respiratory droplets of the host and causes chickenpox and shingles (zoster) in human. In year 1907, Hunt reported the clinical syndrome of herpes zoster oticus, consisting of facial paralysis of lower-motor-neuron type, otalgia and auricular vesicles<sup>(1)</sup>. Although he was not the first one to report this syndrome, it was later named as Ramsay Hunt syndrome (RHS). The syndrome is believed to be caused by reactivation of VZV in the geniculate ganglion and produces skin lesions in corresponding muco-cutaneous areas, including the ear and the ipsilateral side of the tongue. Various vestibule-

cochlear manifestations may occur if cranial nerve VIII is affected because of a bystander effect<sup>(2,3)</sup>. In most cases, RHS usually consists of symptoms related to injury to the ganglions and peripheral nerves. RHS accounted for 4.5% (116/2570) of patients with peripheral facial nerve palsies<sup>(4)</sup>. On the other hand, there were different neurological symptoms accompanying Bell's palsy, including facial tingling (46%), blurred vision (29%), ipsilateral limb paresthesias (17%), ipsilateral limb weakness (14%) and feelings of clumsiness (13%)<sup>(5)</sup>.

According to the study of Kin et al.<sup>(6)</sup>, there were only 8 cases in the English and Japanese literature that documented the coexistence of RHS and VZV encephalitis (7 from previous reports and 1 of their own). They attributed the small number of cases to the

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lack of clearly documented evidence in the previous reports and the rarity of both RHS and VZV encephalitis. Only two cases in their review exhibited weakness of extremities. We report here another two cases of RHS with motosensory impairment in the ipsilateral or contralateral limbs.

## CASE REPORT

### Case 1

The patient was a 47-year-old male Taiwanese. He denied past history of chickenpox in his childhood, major systemic disease or operations. He did not take any regular medications. He did administrative service in the government and have not traveled to foreign countries recently.

Initially (day 1), he suffered from flu-like symptoms, including sneezing, mild tinnitus in the right ear and episodes of involuntary twitching in the right face. He also noticed vague pain in the right post-auricular area. Because these symptoms were subtle and had no effect on his daily activities, he did not take it seriously. However, it became more difficult to control his right face when he smiled or ate on day 3. He thus hurried to our hospital for help. Physical examination revealed right peripheral-type facial palsy and post-auricular pain. There is no fever, vesicles or limbs weakness. Oral prednisolone 40 mg per day was prescribed for 4 days under the impression of Bell's palsy. Neurophysiological tests, including nerve conduction velocity study of the facial nerves and blink reflexes were arranged.

Although feeling more at ease, the patient experienced additional symptoms in the next 4 days, including impaired hearing of the right ear, mild fever and unsteady gait with deviation to the left side. Also, the post-auricular pain exacerbated and spread to the right ear, cheek and orbital areas. He could not work anymore and had to come to our emergency room on day 8 when he found himself feverish (up to 38°C). On examination, his consciousness was clear and he was orientated to time, place and people. In addition to the symptoms mentioned above, tinny vesicles and tenderness in and near the right ear canal were noticed. Hearing acuity of the right ear was decreased and mild weakness of the left

upper and lower extremities (5- on the upper and lower extremities) was detected. Deep tendon reflexes in all four limbs were symmetric and normal. The left side of his body, including limbs and trunk, were less sensitive to pain by pinprick test (7/10). His left hand showed dysmetria at finger-nose-finger test. Barany's past pointing test revealed lateral deviation of the left hand.

He was hospitalized from day 10 to day 20. During admission, his basic biochemistry data were within normal limits. Electroencephalography was normal. Results of nerve conduction velocity study were compatible with a clinical diagnosis of peripheral-type facial palsy on the right side. Brainstem auditory evoked potential was normal. Brain magnetic resonance imaging (MRI) with and without gadolinium on day 9 revealed no abnormality.

Lumbar puncture was performed smoothly on day 11. The open/close pressure was 185/130 mmH<sub>2</sub>O. The appearance of cerebrospinal fluid (CSF) was clear. However, the WBC count was elevated to 225/mm<sup>3</sup> while the RBC count were 0/mm<sup>3</sup>. The leukocytes consisted of lymphocytes (96%) and macrophage (4%). Other CSF data included decreased glucose level (39 mg/dL, 128 mg/dL in simultaneous serum sample), elevated total protein (88 mg/dL) and lactate (2.2 mmol/L). However, viral isolation from CSF and cotton swab of post-auricular skin both reported negative results. VZV antigen was absent and polymerase chain reaction revealed no nucleic acid of herpes simplex virus in the CSF.

His symptoms improved after treatment with intravenous acyclovir (10 mg/kg q8h) for 10 days. The hypoesthesia to pinprick on the right side of face, the left side of body, and in the left limbs improved from 7/10 to 9~10/10. Dysmetria of the left limbs recovered completely. There were still residual deficits, including weakness of the left extremities, right post-auricular pain, and tinnitus on the day of discharge, when the facial palsy was in Brackmann stage VI.

### Case 2

The patient was a 44-year-old healthy woman. The first symptom was tinnitus of the right ear (day 1). Swelling sensation and dull pain of the right ear and reddening of the right eye appeared 10 days later (day 11).

In another 2 weeks (day 25), decreased taste sensation, right facial hypoesthesia and right facial palsy developed with tearing from the right eye, easy choking, numbness and weakness of the right extremities. She came to our emergency room on the next day (day 26). Tiny crust and scarring in the acoustic meatus and swelling of the right auricle were noticed. The right peripheral-type facial palsy was in Brackmann stage IV. Under the tentative diagnosis of Ramsay Hunt syndrome, oral valaciclovir (1000 mg tid) and prednisolone (60 mg qd) were prescribed for 3 days and she was discharged. Because the symptoms persisted, she was referred to the outpatient clinic of the department of neurology on day 33. In addition to the right peripheral-type facial palsy, there were dysphagia and motor-sensory deficits in the right limbs. Under the impression of Ramsay Hunt syndrome complicated with encephalitis, she was hospitalized on the same day, roughly one month after the onset of the first symptom.

Complete neurological examination revealed right peripheral-type facial palsy, right hemiparesis (grade 4 muscle power in both upper and lower limbs) and hypoesthesia to vibration and pinprick (3/10 on the right side of the face, 5/10 in the right upper limb and trunk, and 7/10 in the right lower limb). There was no dysmetria revealed by the finger-nose-finger or heel-knee-shin tests. The taste sensations to bitter, salty and sweet stimulants were intact. Her basic biochemistry data were within normal limits. Lumbar puncture was performed smoothly on day 34. The open/close pressure was 150/100 mmH<sub>2</sub>O. The appearance of CSF was clear, but there were mildly elevated WBC (12/mm<sup>3</sup>, with 10 lymphocytes and 2 polymorphonuclear cells) and RBC counts (41/mm<sup>3</sup>). The remaining CSF was sent for other studies, including sugar (77 mg/dL, 133 mg/dL in simultaneous serum sample), total protein (37 mg/dL) and lactate (1.6 mmol/L). Viral isolation of CSF and cotton swab of post-auricular skin both revealed negative result. VZV IgG and antigen were absent in the CSF. The evoked potential studies, including visual, sensory, motor and brainstem-auditory evoked potentials, were all normal. Facial nerve conduction velocity study and blink reflexes on day 35 confirmed the diagnosis of peripher-

al-type facial palsy on the right side. Mini-mental state examination showed a score of 28/30 and cognitive ability screening instrument 93/100, both being within normal limits. MRI of the brain with and without gadolinium was performed on day 34 and revealed no abnormality. Tc-99m single photon emission computed tomography showed no deficit of cerebral blood perfusion, either. Intravenous acyclovir (500 mg q8h) was given for 7 days from day 33. After treatment, the facial palsy improved. The muscle power of the right upper limb increased to 5- and the right lower limb to 4+. The pain sensation by pinprick improved to 10/10 on the face and 8/10 in the body and limbs.

## DISCUSSION

RHS is a clinical syndrome that may include different combinations of symptoms. The core definition of RHS requires (1) herpes zoster auricularis or herpes zoster in any of the sensory zones corresponding to the trigeminal branches in the face, and (2) ipsilateral facial palsy. The definition of VZV encephalitis is confusing. According to the report of Kin et al.<sup>(6)</sup>, it requires impairment of consciousness and focal signs of the central nervous system. In addition, the symptoms should be associated with VZV infection, which had to be confirmed by clinical (herpes zoster) or laboratory findings (positive VZV DNA in CSF, anti-VZV IgM in CSF or a four-fold or greater increase in the anti-IgG titer in CSF obtained at an interval of at least 2 weeks). In other words, involvement of any structure or system in the central nervous system could be regarded as encephalitis. Kim et al.<sup>(7)</sup> reported a case of RHS complicated by a brainstem lesion (rhombencephalitis). The presentation, however, was regarded as the manifestation of nucleus invasion in cranial polyneuropathy in other reports<sup>(8,9)</sup>. Booss and Esiri<sup>(10)</sup> classified the neurological complications of herpes zoster infection into syndromes and circumstances. The syndromes included stroke, aseptic meningitis, cranial neuropathy/brain stem encephalitis, cerebellitis, encephalitis and myelitis. In this classification, encephalitis was more restricted to the impairment of consciousness and cognition.

The coexistence of RHS and VZV encephalitis is rare and Kin et al.<sup>(6)</sup> found only 8 cases, including one case of their own, in English and Japanese literature<sup>(6)</sup>. When the data were examined more closely, we found that these patients could be divided into two groups according to the presence of disturbed consciousness (encephalitis) and pyramidal sign (stroke-like), similar to the classification of Booss and Esiri<sup>(10)</sup>. Since our cases showed hemiparesis without disturbance of consciousness, we compare our cases with those previously reported cases in the stroke-like group (Table). The motor and sensory deficits of the limbs were on the same side in all the cases, and were on the same side as the deficits of the face in 3 cases. Dysmetria was described in one case. Image studies were performed in 3 cases with lesions reported in one case. Brain MRI performed in case 4 showed a lesion in the middle of the pons, more to the contralateral side of the midline<sup>(11)</sup>. Delayed ipsilateral cerebral infarction leading to contralateral hemiparesis

was a well documented complication of herpes zoster ophthalmicus<sup>(12-15)</sup>. It was suggested that the virus may travel along the ophthalmic branch of the trigeminal nerve to reach the vessel wall of internal carotid artery in the cavernous sinus. Affected vessels showed thrombosis and inflammation. However, the lesion in the pons of case 3 was more likely ascribable to a direct extension from the infected cranial nerve nucleus. Cases of cerebral infarction outside the facial area following zoster were also reported<sup>(16,17)</sup>. These cases indicated that virus could be disseminated to the vessels via blood or CSF. Image studies of the brain in both of our cases showed no lesion. The pathogenesis of ipsilateral weakness and sensory deficit in case 2 was especially intriguing. There are two possibilities. The virus may spread from the facial nerve to the basilar artery and then to the structures in the brain stem, causing brain stem encephalitis or brain stem infarction. On the other hand, there may be generalized vasculopathy of different severity, resulting in focal encephalitis or stroke. In fact, for the neurological manifestations of VZV infection, the border between encephalitis and vasculopathy is getting blurred. Gilden<sup>(18)</sup> suggested to discard the term VZV encephalitis completely and use only VZV vasculopathy, because vasculopathy is the main pathogenetic mechanism for the neurological syndromes of VZV infection. In other words, small vessel vasculopathy may be responsible for symptoms of diffuse encephalitis, whereas large vessel vasculopathy would have manifestations of focal deficits.

In a retrospective study of 265 patients with peripheral-type facial palsy<sup>(19)</sup>, abnormal CSF findings could occur in both Bell's palsy (12.6%) and RHS (64.7%). In this regard, the most frequent abnormalities of CSF were pleocytosis. However, the incidence (41.3% in the cases of Bell's palsy with abnormal CSF findings and 100% in RHS) and cell count (on average 23 WBC/mm<sup>3</sup> in Bell's palsy and 84 in RHS) were significantly different. The CSF of case 1 taken on day 11, showing prominent leukocytosis (225/mm<sup>3</sup>) with lymphocyte predominance, indicating an aseptic infection or inflammation of the central nervous system and thus in favor of the diagnosis of RHS. Leukocytosis in the second case was mild

**Table.** Comparison of the cases in the present study and from review of the literatures

Case no.*	1	2	3	4
Disturbed consciousness	-	-	+	
Facial palsy	R	R	R	L
Facial numbness	R	R	R	L
Hemiparesis	L	R	R	L
Hemihypoesthesia	L	R	R	L
Dysmetria	L	-	ND	-
Image modality	CT, MRI	CT, MRI	ND	MRI
Image finding	-	-	ND	+
CSF				
WBC (/mm <sup>3</sup> )	225	12	184	0
Protein (mg/dL)	88	37 <sup>†</sup>	ND	Normal
Viral detection	- <sup>†</sup>	-	ND	- <sup>§</sup>

R: right; L: left; ND: not described.

\* Cases 1 and 2 are cases 1 and 2 described in this report, case 3 is from Taukayama<sup>(20)</sup> and case 4 is from Mizock et al.<sup>(11)</sup>. <sup>†</sup>CSF VZV antigen, viral culture, and serum VZV IgG were all negative. <sup>‡</sup>CSF VZV IgG and antigen, and viral culture were all negative. <sup>§</sup>CSF VZV PCR was negative and serum VZV IgG was positive.

(12/mm<sup>3</sup>). It could be ascribed to the relatively “late” lumbar puncture which was performed 34 days after the onset of first symptom, and/or to the partial treatment by oral valaciclovir.

According to the recommendations for the management of herpes zoster<sup>(20)</sup>, RHS was a VZV infection of non-truncal involvement and should be treated with systemic anti-viral agents. Moreover, the recommendation also emphasized that severe neurological complications of VZV, including meningitis, encephalitis and myelitis, should be treated by not only by systemically but specifically by intravenous acyclovir. In these 2 cases, if we had missed their weakness, sensory deficits and other focal signs, we would have given them oral but not intravenous acyclovir for RHS. This could be an under-treatment and result in increased risk of further complications. The cases presented in this report underscore the importance of a complete neurological examination of face and limbs in patients with a chief complaint of facial weakness or a tentative diagnosis of peripheral-type facial palsy.

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