

Treatment of Ramsay–Hunt’s syndrome with multiple cranial nerve involvement and severe dysphagia

A case report

Jong Min Kim, MD^a, Zeeihn Lee, MD, PhD^a, Seungwoo Han, MD, PhD^b, Donghwi Park, MD^{a,*}

Abstract

Rationale: Ramsay–Hunt’s syndrome (RHS) is a disorder characterized by facial paralysis, herpetic eruptions on the auricle, and otic pain due to the reactivation of latent varicella zoster virus in the geniculate ganglion. A few cases of multiple cranial nerve invasion including the vestibulocochlear nerve, glossopharyngeal nerve and vagus nerve have been reported. However, there has been no report about RHS with delayed onset multiple cranial nerve involvement causing severe aspiration, and a clinical course that improved after more than one year of dysphagia rehabilitation and percutaneous endoscopic gastrostomy (PEG). Here, we report on a 67-year old male with delayed onset swallowing difficulty after 16 days of RHS development.

Patient concern: Severe aspiration during swallowing.

Diagnosis: Severe dysphagia caused by RHS with multiple cranial nerve involvement.

Intervention: Application of percutaneous endoscopic gastrostomy (PEG) and rehabilitation therapy of dysphagia.

Outcomes: After 13 months from symptom onset, his PAS improved from 7 to 2 in follow-up video-fluoroscopic swallowing study (VFSS). Then, he was re-admitted, and the PEG tube was removed and oral feeding was started.

Lessons: This case gives us the lesson that optimal doses of acyclovir and corticosteroids are important to prevent progression of multiple cranial involvement in RHS, and swallowing difficulty in RHS patients with multiple cranial nerve involvement can be improved through long-term rehabilitation even if there is no improvement for more than one year.

Abbreviations: PAS = penetration-aspiration scale, PEG = percutaneous endoscopic gastrostomy, RHS = Ramsay–Hunt’s syndrome, VFSS = video-fluoroscopic swallowing study, VZV = varicella zoster virus.

Keywords: acyclovir, corticosteroid, dexamethasone, dysphagia, prednisolone, Ramsay–Hunt syndrome, rehabilitation

1. Introduction

Ramsay–Hunt’s syndrome (RHS) is characterized by herpetic eruption in the external auditory canal, facial paralysis and otalgia.^[1–4] The classical concept of RHS is the reactivation of the varicella zoster virus (VZV) of the alpha herpes family,

involving the facial nerve and the vestibulocochlear nerve.^[1–4] RHS has been reported to have an incidence of five per 100,000, and incidence and severity increases in immune-compromised persons.^[5] Once VZV infection occurs, the virus is never cleared from the body, and keeps itself hidden from the host’s immune system. RHS develops due to reactivation of the virus in the geniculate ganglion. Pathognomonic for the syndrome are vesicular rash on the ear, external auditory canal and peripheral facial nerve palsy accompanied by otalgia. Additionally, symptoms such as dizziness and hearing loss may occur due to involvement of the vestibulocochlear nerve. Multiple cranial nerve involvement is rare in RHS, but it has been reported by several researchers and its prognosis is also known to be worse than other forms of RHS without multiple cranial nerve involvement.^[6–8] However, there has been no report about RHS with delayed onset multiple cranial nerve involvement causing severe aspiration, and a long-term clinical course that improved after more than one year of dysphagia rehabilitation and percutaneous endoscopic gastrostomy (PEG). Here, therefore, we report on a 67-year old male with delayed onset swallowing difficulty after 16 days of RHS development and his clinical course of swallowing difficulty which improved after 13 months of rehabilitation therapy for dysphagia. The patient was informed that data concerning the case would be submitted for publication, and he provided consent. This case report was approved by the ethics committee of our hospital.

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^a Department of Rehabilitation Medicine, Daegu Fatima Hospital, ^b Division of Rheumatology, Department of Internal Medicine, Kyungpook National University School of Medicine, Daegu, Korea.

* Correspondence: Donghwi Park, Department of Rehabilitation Medicine, Department of Rehabilitation Medicine, Daegu Fatima Hospital, Ayangro 99, Dong gu, Daegu 41199, Republic of Korea (e-mail: bdome@hanmail.net).

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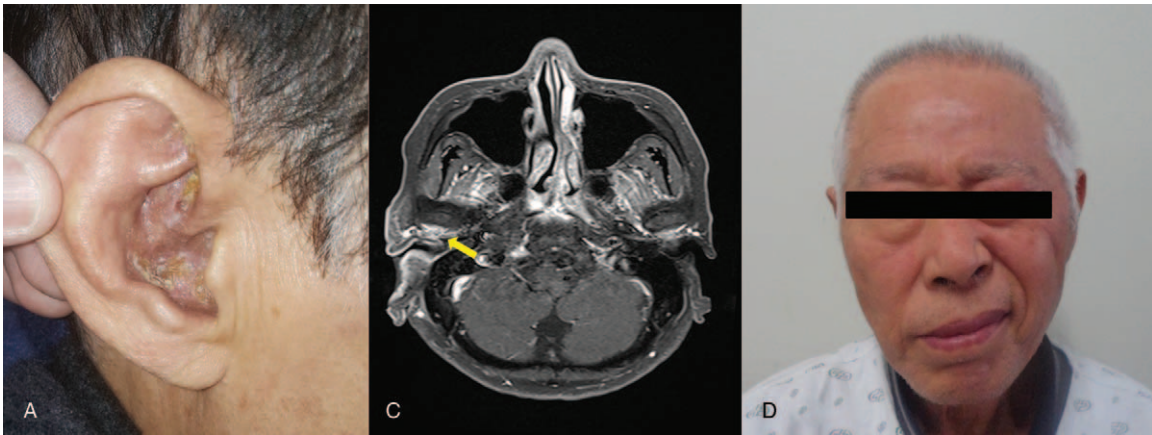


Figure 1. Vesicular eruption on the auricle and peripheral facial nerve palsy. (A) Vesicular eruptions of the auricle in the remission phase. (B) Enhancement of the external ear and auditory canal in a Gadolinium-enhanced MRI of the brain. (C) Peripheral facial nerve palsy including nasolabial fold flattening, drop of oral angle, and decreased movement of forehead wrinkles.

2. Case report

A 67-year old male visited our hospital with a 7-day history of right-sided otalgia. The patient was admitted to the department of otorhinolaryngology. The family and social history of the patient did not reveal any remarkable findings. He had no chronic diseases except diabetes mellitus type 2 for 5 years, and no history of operation. At admission, a physical examination revealed vesicular eruptions and pustules in the right ear auricle and external auditory canal (Fig. 1A), but the left ear was normal. The patient did not have any difficulties in facial expression, swallowing, and other activities of daily life, although he had vertigo and decreased hearing in the ear. Brain magnetic resonance image (MRI) evaluation was performed, and the MRI findings showed no acute cerebral infarction and no definite abnormal findings (Fig. 1B). In serum analysis, an immunoglobulin G (IgG)-specific antibody titer for VZV was 8.12, a highly positive result, coupled with an immunoglobulin M (IgM)-specific antibody titer for varicella zoster of 0.14, a negative result. Then, the patient was given intravenous acyclovir 1500 mg (25 mg/kg/day) and intravenous dexamethasone disodium phosphate 5 mg daily for nine days. After nine days, oral prednisolone 15 mg (0.25 mg/kg/day) was prescribed for a further 5 days. After 16 days from symptom onset, however, he started to complain of additional symptoms of difficulty in swallowing food, hoarseness, sputum, mild dyspnea, and right-sided facial expression muscle weakness (Fig. 1C). In a second physical examination, his gag reflex of the right side was absent and the uvular was deviated to the left. However, tongue movement showed no abnormalities. From a clinical point of view the patient had involvement of the V, VII, VIII, IX, and X right cranial nerves, consistent with multicranial neuritis. In a video-fluoroscopic swallowing study (VFSS), the patient had severe aspiration in all materials, and his penetration-aspiration scale (PAS) was 7.^[9] Additionally, intravenous acyclovir 1500 mg without corticosteroids was administered daily for 6 days. In addition, enteral tube feeding (Levin tube) was used to help prevent further aspiration. Thereafter, rehabilitation therapy for dysphagia including education of adequate positioning and occupational therapy was started.

After 8 weeks from symptom onset, his hearing impairment, dizziness, and facial palsy symptom had improved mildly, but the

patient did not have definite improvement in dysphagia and continued to show severe tracheal aspiration and his PAS was 7 in VFSS. After being informed of the long-term adverse effects of the Levin tube and the need for a percutaneous endoscopic gastrostomy (PEG) tube, he agreed with the application of PEG. Until 13 months from symptom onset, he continued to have rehabilitation therapy of dysphagia in our hospital. After 13 months from symptom onset, his PAS improved from 7 to 2 in follow-up VFSS. Then, he was readmitted, and the PEG tube was removed and oral feeding was started.

3. Discussion

In most other cases of RHS with multiple cranial nerve involvement, symptoms of cranial nerve involvement developed within a few days.^[6–8] However, we report on a patient with delayed onset swallowing difficulty after more than two weeks of RHS development. In our case, acyclovir and corticosteroid were administered in an early phase of disease, but the outcomes were not satisfactory. We thought that the delayed symptoms might be due to insufficient treatment. In the treatment of RHS, the combination of acyclovir and prednisone is the most recommended therapy.^[10] Although the optimal dosage of acyclovir and prednisone has not been established and has varied in previous studies (Table 1), a relatively small dosage of corticosteroid (intravenous dexamethasone disodium phosphate 5 mg daily for nine days and thereafter oral prednisolone 15 mg (0.25 mg/kg/day) daily for 5 days) was administered compared with the other previous studies (1 mg/kg prednisone daily for 7 days).^[10–13] Considering 5 mg of prednisone has an equivalent dose of 0.75 mg of dexamethasone,^[14] 5 mg of dexamethasone is approximately equivalent to 33 mg of prednisone, which is half the dosage of corticosteroid compared with previous studies. Moreover, unlike Bell's palsy, late degeneration of the facial nerve function up to 21 days from the onset of symptoms has been described in RHS.^[11,15] Thus, in some previous studies, pharmacologic treatment of antiviral agents was recommended to be continued until 21 days after symptom onset to cover the concept of late neural degeneration after RHS.^[11,15] Therefore, use of antiviral agents for an insufficient period time and/or

Table 1**Recent combination treatment of Ramsay–Hunt syndrome.**

Study/year	Antiviral treatment	Corticosteroid (prednisolone)
Coulson et al/2011 ^[11]	Famciclovir 250 mg × 3 orally, 21 days	1 mg/kg/ day, 14 days, decreased by 10mg/d until 0 mg
Uri et al/2003 ^[12]	ACY 800 mg × 5/day orally, or 15mg/kg/day IV for 7 days	100 mg × 3/day for 7 days
Kinishi et al/2001 ^[13]	ACY 4000 mg/day orally for 7 days	500→100 mg/day decreased over 7 days
Murakami et al/1997 ^[10]	ACY either 250 mg × 3 IV or 800 mg × 5 orally for 7 days	1mg/kg/ day for 5 days, decreased 10 mg/day until 0, over 10 days

ACY = acyclovir.

insufficient anti-inflammatory effect of corticosteroids on neuroinflammation of the cranial nerves may lead to late involvement of other cranial nerves.

Although the patient's other RHS symptoms, such as facial palsy, improved greatly there was little improvement in dysphagia until 13 months from the initial diagnosis. Previous studies reported that RHS with multiple cranial nerve involvement had a poor recovery rate compared to RHS without multiple cranial nerve involvement. Shim et al^[16] reported that RHS patients with multiple cranial nerve involvement achieved only 27.3% of full recovery compared with 67.7% for RHS patients without multiple cranial nerve involvement. However, considering that there is no significant recovery until 13 months, our case suggests that long-term rehabilitation and clinical follow-up of more than one year may be important in RHS patients with severe dysphagia.

4. Conclusion

Our case gives us the lesson that optimal doses of acyclovir and corticosteroids for a sufficient period time are important to prevent progression of multiple cranial involvements in RHS, and swallowing difficulty in RHS patients with multiple cranial nerve involvement can be improved through long-term rehabilitation even if there is no improvement for more than one year.

Author contributions

Conceptualization: Jong Min Kim, Zeeihn Lee, Seungwoo Han.

Writing – original draft: Donghwi Park.

Writing – review & editing: Donghwi Park.

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