ORIGINAL ARTICLE

Vestibulocochlear Function in Patients with Ramsay Hunt Syndrome

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Objective/Hypothesis: To investigate the manifestations of the inner ear symptoms in patients with Ramsay Hunt syndrome (RHS), and the relationship between vestibulocochlear symptoms and caloric test and vestibular evoked myogenic potential (VEMP) test in RHS patients.

Materials and Methods: The clinical courses of 18 patients with RHS were investigated. A detailed history of each patient was recorded, and each patient underwent a battery of tests. Ten of the patients also underwent the caloric test and VEMP test.

Results: With respect to the clinical symptoms of the 18 patients with RHS, 8 had facial palsy (44.4%), 5 had auricular vesicles (27.8%), and 5 had simultaneously occurring cases (27.8%). Of the 18 patients, 11 had vertigo (61.1%), 11 had hearing impairment (61.1%), and 9 patients had both vertigo and hearing impairment (50%). Of the 10 patients who underwent the vestibulocochlear function tests, 7 had abnormal results on the caloric or VEMP test. Of the 5 patients with vertigo, an abnormal caloric response was noted in 5 patients and an abnormal VEMP test was noted in 4 patients. Of the other 5 patients without vertigo, an abnormal caloric response was noted in 1 patient and abnormal VEMP test was noted in another. Of the 6 patients with hearing impairment, abnormal results of the vestibular function tests were noted in 5 patients, and 2 of the other 4 patients without hearing impairment had abnormal vestibular function tests.

Conclusion: The sequence of the appearance of facial palsy and auricular vesicles may be different. The pathophysiology of vertigo may be associated with invasion of superior or inferior vestibular nerves. Patients with abnormal vestibulocochlear test results may not have inner ear symptoms.

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Introduction

Ramsay Hunt syndrome (RHS) was first hypothesized by James Ramsay Hunt in 1907 as geniculate ganglionitis induced by reactivation of varicella-zoster virus (VZV) infection [1]. The clinical triad of RHS includes otalgia, vesicles of the auricle or external ear canal, and facial nerve palsy [2, 3]. The lesions responsible for RHS usually affect the cochlear and/or vestibular systems [4- 6]. Although the involvement of acute vestibulopathologies caused by RHS has been reported, the relationship between clinical manifestations and vestibulocochlear function tests has not. This study aimed to investigate the manifestations of the inner ear symptoms of patients with RHS, and the relationship between vestibulocochlear symptoms and caloric test and vestibular evoked myogenic potential (VEMP) test in RHS patients.

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Materials and Methods

Patients

From November 2008 to April 2010, eighteen patients with RHS visited the Otorhinolaryngology Department of Kaohsiung Medial University Hospital. The inclusion criteria were symptoms of otalgia, vesicles of the auricle or external ear canal, and facial nerve palsy. A detailed history was obtained from all patients. Physical examinations including local findings of the ears, nose and throat, neurological examinations, and subjective pure tone audiometry were administered. In addition, ten of these patients also received the air caloric test, VEMP test.

Air caloric test [7]

The caloric test was performed using a bithermal method (and air) with electronystagmographic recordings (SYNAPSYS, VNG VLMER). The unilateral weakness represents the percentage of difference of the maximum slow-phase velocity between lesioned and healthy labyrinthine response divided by the sum of the maximum slow-phase velocity in both ears. In this study the normal limit of the caloric response is within 20% of the unilateral weakness.

VEMP test [7]

The VEMP test was conducted by way of the unilateral sternocleidomastoid muscle as the target for recording. The surface electromyographic activity was recorded ipsilaterally (Grason-StadIer, Audera Version 2.1), with the patient instructed to maintain lateral head-turning in a sitting position. The other reference and ground electrodes were separately set on the suprasternal notch and forehead. Short tone bursts (95 dBHL, 500 Hz, 5 times/second, 200 times/stimuli) were applied to the unilateral ear through an insertion-type earphone. Following the VEMP test by tone burst stimulation, each patient underwent the tapping method [8]. The forehead of the patient was tapped with a tendon hammer (5 times/second, 200 times/stimuli) to elicit a response through bone-conduction stimulation, as with tone burst stimulation during the process of the recording, the instructor observed the monitor and the patient.

Thirty healthy persons without any ear problems underwent the VEMP test by tone burst method as controls. Comparisons between the VEMP test results of the patients with RHS and those of the controls were carried out to establish the baseline information.

When the interaural amplitude difference (IAD) ratio exceeded 0.36, the condition was labeled as either an augmented VEMP or a depressed VEMP depending on whether the amplitude of the affected side was greater or less than that of the opposite side. The VEMP test was considered abnormal if the IAD ratio was abnormal or if p13 or n23 latency was prolonged compared with that of the 30 normal volunteers.

Results

Of the 18 RHS patients, 6 were men and 12 were women, with an age ranging from 29 to 76 years (mean, 52.3 years). The right side was affected in 11 patients, and the left side was affected in 7 patients. All of the patients had auricular vesicles, otalgia, and facial palsy (House-Brackmann [9] grade II in 1 case, grade III in 4 cases, grade IV in 8 cases, grade V in 2 cases, and grade VI in 3 cases), as shown in Table-1.

With regards to the clinical course of the 18 patients with RHS, 8 had facial palsy (44.4%, 2-6 days), 5 had auricular vesicles (27.8%, 2-12 days), and 5 had simultaneously occurring cases (27.8%, within one day). Of the 18 patients, 11 had subjective vertigo (61.1%), 11 had subjective and objective hearing impairment (61.1%), and 9 patients had both vertigo and hearing impairment (50%).

In the 10 patients who underwent vestibulocochlear function tests, 7 had abnormal results on the caloric or VEMP test. Of the 5 patients with vertigo, an absent caloric response was noted in 5 patients, and an abnormal VEMP test was noted in 4 patients (Table-2). Of the other 5 patients without vertigo, an absent caloric response was noted in one patient and an abnormal VEMP test was noted in another (Table-3). Of the 6 patients with hearing impairment, abnormal vestibular function test results were noted in 5, and 2 of the other 4 patients without hearing impairment had abnormal vestibular function test results.

Discussion

Many investigators have postulated that reactivation of latent VZV in the geniculate ganglion causes auricular vesicles and facial paralysis [1, 2, 10, 11]. The inner ear symptoms of RHS include facial paralysis, tinnitus, hearing loss, hyperacusis (dysacousis), vertigo, and dysgeusia [12]. In 1983, Etholm and Schuknecht [13]

Table 1. Summary of clinical manifestations in patients with Ramsay Hunt syndrome

No.	Age	Sex	Lesion Side	Facial Palsy	Hearing loss	Vertigo	Tinnitus	Clinical Course
1	60	F	R	III	-	-	-	V→F, 1day
2	29	F	R	IV	-	+	+	V→F, 2day
3	39	M	L	IV	-	-	-	F→V, 3day
4	39	M	L	III	-	-	+	V→F, 1day
5	47	F	R	IV	-	+	+	V→F, 3day
6	56	F	L	IV	+	+	-	V→F, 1day
7	71	F	L	IV	+	+	+	F→V, 3day
8	56	M	R	II	-	-	+	V→F, 10day
9	76	F	R	IV	+	+	-	F→V, 4day
10	39	F	R	V	+	+	+	F→V, 5day
11	68	M	R	VI	+	-	+	F→V, 6day
12	63	F	R	IV	+	+	-	V→F, 8day
13	31	M	R	IV	+	+	+	V→F, 1day
14	69	F	L	VI	+	+	+	F→V, 2day
15	43	F	L	III	-	-	-	F→V, 2day
16	57	F	R	V	+	+	+	F→V, 3day
17	42	M	R	III	+	+	+	V→F, 12day
18	56	F	L	VI	+	-	-	F→V, 1day

Facial palsy grading: House-Brackman system, V: vesicles, F: facial palsy
+: positive, -: negative, →: contiguous course

Table 2. Summary of clinical manifestations, Caloric test and VEMP test in 5 patients with vertigo

No.	Age	Sex	Lesion Side	Facial Palsy	Hearing Loss	Tinnitus	Clinical course	VEMP test	Caloric test
6	56	F	L	IV	+	-	V→F, 1D	Abnormal	Abnormal
9	76	F	R	IV	+	-	F→V, 4D	Abnormal	Abnormal
10	39	F	R	V	+	+	F→V, 5D	Normal	Abnormal
12	63	F	R	IV	+	-	V→F, 8D	Abnormal	Abnormal
17	42	M	R	III	+	+	V→F, 12D	Abnormal	Abnormal

Facial palsy grading: House-Brackman system, V: vesicles, F: facial palsy
+: positive, -: negative, →: contiguous course
VEMP: vestibular evoked myogenic potential

Table 3. Summary of clinical manifestations, Caloric test and VEMP test in 5 patients without vertigo

No.	Age	Sex	Lesion Side	Facial Palsy	Hearing Loss	Tinnitus	Clinical course	VEMP test	Caloric test
1	60	F	R	III	-	-	V→F, 1D	Abnormal	Normal
3	39	M	L	IV	-	-	F→V, 3D	Normal	Abnormal
4	39	M	L	III	-	+	V→F, 1D	Normal	Normal
15	43	F	L	III	-	-	F→V, 2D	Normal	Normal
18	56	F	L	VI	+	-	F→V, 1D	Normal	Normal

Facial palsy grading: House-Brackman system, V: vesicles, F: facial palsy
+: positive, -: negative, →: contiguous course
VEMP: vestibular evoked myogenic potential

reported that when facial palsy was associated with inner ear symptoms, the inflammatory lesion was located in the nerve trunk within the internal auditory canal. VZV DNA has been identified in the ipsilateral geniculate ganglion [10] and in the facial nerve sheath [14] of patients with facial paralysis associated with herpes zoster oticus. Recent studies on the pathophysiology of herpes zoster oticus suggest that latent VZV is located in the geniculate ganglia and is either primarily present in the auditory and vestibular afferent ganglia or secondarily by inflammatory edema in the seventh and eighth cranial nerves in the temporal bones or more proximally [4].

Increased pressure induced by nerve edema in the confined space within the facial canal or internal auditory canal may lead to further nerve degeneration[3,10]. In the internal auditory canal there are facial nerves, cochlear nerves, and superior and inferior divisions of vestibular nerves. The cochleovestibular symptoms seem more likely to be explained by viral transmission across the perineural tissue inside the internal auditory canal, rather than spread through the vestibulofacial anastomosis or via the middle ear mucosa [4]. Although some studies have reported that the facial nerve is the most commonly involved cranial nerve, the lesions responsible for RHS usually affect the cochlear and/or vestibular systems, with the incidence of cochlear system involvement ranging from 7% [15] to 85% [5], and the incidence of vestibular system involvement ranging from 72% [6] to 85% [5]. Kaberos et al. [16] suggested that these differences resulted from the criteria applied to diagnose a case as RHS.

In 1977, Nakao et al. [17] reported a study of 85 RHS patients, and less (46%) vestibulocochlear symptoms were noted in the patients with facial palsy. In contrast, most of the RHS patients with vestibulocochlear symptoms had auricular vesicles or facial palsy (67%). Compared to our cases, 8 out of 18 RHS patients had facial palsy and only 1 had no vestibulocochlear symptoms. However, all of the other 5 cases with auricular vesicles had vestibulocochlear symptoms such as vertigo, tinnitus, or hearing loss. We tentatively conclude that the clinical course of the appearance of facial palsy and auricular vesicles may be different. Auricular vesicles may appear 6 days after facial palsy, and facial palsy may present 12 days after auricular vesicles.

We also investigated the relationship between the subjective inner ear symptoms and the objective vestibulocochlear test findings in the RHS patients. In 2003, Lu et al.[18] reported that both superior and inferior divisions of the vestibular nerve attribute to the vertiginous attacks in patients with herpes zoster oticus. We conducted both the air caloric test and VEMP test to evaluate the integrity of vestibulocochlear function in our RHS patients. Only 10 patients underwent the vestibulocochlear function tests, and 7 had abnormal caloric or VEMP test results. Of the 5 patients with vertigo, all of the caloric and VEMP test results were abnormal except for 1 patient in whom the VEMP test was normal. However, in the other 5 patients without vertigo, a weakness of caloric response was noted in one patient and abnormal VEMP test was noted in another one. Thus we conclude that in RHS patients with vertigo, the caloric or VEMP test usually reveals abnormal results, and in RHS patients without vertigo, the caloric or VEMP test usually reveals normal results. However, if the caloric test or VEMP test reveals abnormal results, the patient may not have any inner ear symptoms.

Conclusion

RHS is caused by reactivation of the varicella-zoster virus, which may affect all of the nerve trunk within the internal auditory canal and cause a variety of inner ear symptoms. Furthermore, the sequence of the appearance of facial palsy and auricular vesicles may be different. We tentatively conclude that the vestibulocochlear test in RHS patients is not consistent with inner ear symptoms.

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