Multifaceted rehabilitation strategies for ramsay hunt syndrome: a case report and a review of literature

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CASE REPORT

ABSTRACT

Background: Ramsay Hunt Syndrome (RHS) is a scarce diagnosis involving unilateral facial paralysis resulting from the reactivation of Varicella-Zoster Virus (VZV) infection in the past, which causes pain, asymmetrical facial expressions, and difficulty in speaking, eating and drinking. The most effective treatment is still debated, and limited research is available. Rehabilitation program is considered as the core treatment that gives functional improvement. The aim of this case study is to present clinical findings in RHS and the rehabilitation program to maximize facial expression recovery.

Case Description: A 59-year-old man came to rehabilitation medicine outpatient clinic two weeks after diagnosed as RHS, with complaints of fever; severe pain and rashes on the left side of the face; and ear discomfort on the same side. Physical examination revealed peripheral facial nerve palsy on the left side with drooping of the left eyelid, left end of the mouth, and difficulty in oral communication. Electroneuromyography (ENMG) examination showed axonal facial nerve paralysis on the left side. The patient was referred to the physiatrist and got rehabilitation program with neuromuscular electrical stimulation (NMES), facial massage and biofeedback exercise for facial muscle 3 times a week for 4 weeks.

Discussion: After 4 weeks rehabilitation program, the patient showed improvement as House-Brackmann grades improved from a grade IV to a grade II; improvements in communication, facial symmetry at both rest and motion; significant improvement in experiencing pain.

Conclusions: This study suggested that rehabilitation program with NMES, facial massage and biofeedback exercise are safe, efficacious and provide good outcomes in the treatment of Ramsay Hunt Syndrome.

Keywords: ramsay hunt syndrome, facial nerve palsy, rehabilitation


INTRODUCTION

Ramsay Hunt Syndrome (RHS) is a scarce diagnosis of preceding Varicella-Zoster Virus (VZV) reactivation in the geniculate ganglion.1 It typically involves the seventh cranial nerves (CN VII), causing unilateral facial paralysis, asymmetric facial expression, and eye watering. The VZV infection can also have an impact on multiple lower cranial nerves in consequence of the juxtaposition of their routes in the temporal bone, including trigeminal nerve (CN V), vestibulocochlear nerve (CN VIII), the glossopharyngeal nerve (CN IX), vagus nerve (CN X), and hypoglossal nerve (CN XII) which not always presenting the sign of dysfunction, but may include the symptoms of vertigo, tinnitus, hearing loss, nystagmus, the pain and difficulty of speaking, drinking and eating.2,3

The recovery rate of facial paralysis has been reported to be as high as 75% after early diagnosis and embarking antiviral treatment within 72 hours of the onset of symptoms.4,5 Research showed that acyclovir and high-dose corticosteroids increase the recovery of facial nerve function in RHS.5 Patients with multiple cranial nerve involvement in RHS have been shown to have a full recovery rate of only 27.3%, and with a single cranial nerve involvement the percentage of full recovery can reach 67.7%.6 Compared to Bell’s palsy, patients with RHS have more severe paralysis initially and are less likely to fully recover.6 Study have recommended that a rehabilitation is considered the core treatment as it gives improvement on social and functional limitations. The most effective treatment is still debatable, and limited study discussing physical therapy interventions for RHS patients is available. Rehabilitation programs for facial paralysis include transcutaneous electrical stimulation, massage, neuromuscular re-education, exercises, and tapping. Research has shown different results related to physical therapy for patients diagnosed with facial paralysis.2,5,6,10 The aim of this case study is to present clinical findings in RHS and the rehabilitation program to maximize facial expression recovery.

CASE REPORT

Patient history
A 59-year-old man came to rehabilitation medicine outpatient clinic at Bhayangkara Hospital, Bali, two
weeks after a diagnosis of Ramsay Hunt Syndrome (RHS), with complaints of fever; severe pain and rashes on the left-sided face; and ear discomfort on the same side. Detailed history unveiled that the onset of symptoms starts a week back starting with fever and preauricular pain followed by formation of vesicles on the left side of the face (Figure 1). The patient also showed paralysis of the left facial nerve by drooping from the end of the mouth on the same side. There was no other significant medical history. The patient was suggested to the neurology outpatient clinic, and started analgesic therapy in conjunction with antiviral therapy acyclovir 800mg, 5 times a day for 7 days as well as systemic steroid therapy with methylprednisolone 16 mg 3 times a day for 7 days which was slowly tapered off. Patient also referred to do an electroneuromyography (ENMG) examination and showed axonal facial nerve palsy on the left side (Figure 2). The patient was referred to physiatrist after 2-3 weeks with severe facial muscle weakness on the left side, difficulty in eye closure, dryness of the eye and asymmetry of the facial tone at rest.

**Physical examination**

The initial evaluation was performed on first visit for all systems after signing informed consent. In cardiovascular findings, he was normotensive with blood pressure 120/76 mmHg and pulse 75 beats/minute. Musculoskeletal examination was done by measuring manual muscle testing (MMT) of the muscles of facial expression.\(^8\)\(^,\)\(^14\)\(^,\)\(^15\) Manual muscle testing for facial muscles took approximately 10 minutes. It was performed on sitting position. At first visit, the examination revealed MMT of left frontalis muscle was poor, left corrugator supercilli muscle was grade poor, left orbicularis oculi muscle was grade poor, left zygomaticus mayor muscle was grade poor, left risorius muscle was grade poor, and left orbicularis oris muscle was grade trace, as seen in Figure 3, respectively. Neuromuscular findings revealed taste of the tongue was normal, hyperacusis was not found in patient and Schimer’s test was negative.

Daily activity functions on this patient was also examined. Limitation found in feeding because he was difficult in chewing on the left side due to muscle weakness, in communication because of impaired speech, in face expression because of difficulty in smiling, and difficulty in gargling because some water keep drooling unintentionally due to lopping of the left corner of the mouth. At the psychosocial level and participation restriction, his expressions and verbally communications were moderately affected.

**Outcome measurement**

Cranial nerve testing was demonstrated as to perform facial nerve involvement as unilateral facial nerve paralysis acquired. Facial symmetry was measured by the severity of paralysis using the House-Brackmann Grading System. The House-Brackmann grade measures grade I being normal and grade VI being total paralysis.\(^13\) The House-Brackmann baseline scale was established. It is valid and accepted by the American Academy of Otolaryngology Head and Neck. This is the most commonly used scale and has good inter-rater problems. However, its sensitivity to facial facial changes is low. This scale takes into account overall facial paralysis by assessing tone, symmetry, rest position, forehead, eye and mouth movements.\(^13\) His grade was a grade IV at initial visit.

**Treatment**

Patient was trained in biofeedback exercises for facial muscles for better movement progress, soft tissue mobilization to reduce pain and improve mobility, facial massage and patient education related to Ramsay Hunt Syndrome (RHS).

The patient agreed to have electrical stimulation initially for muscle activation and pain management.\(^8\)\(^,\)\(^14\)\(^,\)\(^15\) Electrical stimulation treatments include direct current at an intensity sufficient to produce a motor response, a pulsatile waveform that is transmitted at a sub-motor threshold, to manage pain. The patient underwent therapy sessions lasting 45 minutes, 3 times a week for 4 weeks. Each session began with a brief re-evaluation of facial motor function three times a week, as seen in Figure 4.\(^8\)\(^,\)\(^14\)\(^,\)\(^15\)

The therapeutic rehabilitation guide applied in this study consisted of sequentially structured exercises to align the two sides of the face, and the task of reintegrating emotions with expression.\(^14\) Our aim is to improve the facial alignment at rest and during movement. The patient is taught: (1) massaging his face every day for 10-15 minutes (avoiding lesions on the face), which consists of kneading and stretching of the affected; (2) Breathing and relaxation exercises to feel and feel the connective and relaxing muscles in the face; (3) Biofeedback exercises using a mirror for 30 minutes, twice every day, such as: raising the forehead; open eyes wide; lift the upper lip; tighten the lips; smile; (4) Exercises of covering eyes and lips with varying speed and strength; (5) Exercises to increase awareness of lip movements - need to eat, drink and gargle; (6) Practice of pronouncing letters and words, and (7) Exercises for emotional expression.
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Outcomes

The outcomes were recorded once a week (Table 1). The photographs were used to record treatment progress, facial tone changes and to visually trace active expression and passive rest.

Self-reported disability was also measured for treatment progress from initial until the last visit. The patient showed a definite improvement within two weeks with only dysesthesia and a tingling sensation in the maxillary branching area of the marginal mandibular nerve. All the symptoms had diminished after one month therapy, and the patient could smile nearly symmetrical. (Figure 5).

DISCUSSION

The prognosis for facial paralysis is worse in RHS than in the idiopathic form. Facial nerve involvement due to inflammation caused by viral neuronitis and secondly due to facial nerve edema. The likelihood of full recovery from facial muscle control in patients diagnosed with RHS is 27.3-67.3%. In facial nerve palsy, the facial muscles degenerate and there are cortical changes relevant to nerve input. With facial reinnervation, motor function will return but cortical mapping will not be the same as the pre-injury muscle. Neurumuscular re-education is essential for recruiting the appropriate muscles and ensuring adequate cortical maps. A multifaceted rehabilitation strategy that combines the goal of selective muscle control with appropriate cortical input is the most successful in maximizing facial recovery. The main objective of this study was to evaluate a multifaceted rehabilitation strategy in early rehabilitation to improve facial symmetry and muscle activation in RHS. All facial muscle strength was measured at each visit: ‘trace’ and ‘poor’ for nearly all facial muscles at the first visit. Electrical stimulation was initiated in the first week of intervention (2 weeks after diagnosis) and up to 12 visits. The rehabilitation program started with the NMES. Electrical stimulation treatment includes direct current at an intensity sufficient to generate a motor response, a pulsatile waveform that is sent to

Table 1. Outcomes of House-Brackmann grade and facial muscles strength findings per week post-diagnosis

<table>
<thead>
<tr>
<th>Week after diagnosis</th>
<th>House-Brackmann grade</th>
<th>Frontalis</th>
<th>Corrugator supercilli</th>
<th>Orbicularis oculi</th>
<th>Zygomaticus mayor</th>
<th>Risorius</th>
<th>Orbicularis oris</th>
</tr>
</thead>
<tbody>
<tr>
<td>2</td>
<td>IV</td>
<td>Poor</td>
<td>Poor</td>
<td>Poor</td>
<td>Poor</td>
<td>Trace to poor</td>
<td>Trace</td>
</tr>
<tr>
<td>3</td>
<td>III</td>
<td>Poor to good</td>
<td>Poor to good</td>
<td>Poor to good</td>
<td>Poor</td>
<td>Poor</td>
<td>Trace to poor</td>
</tr>
<tr>
<td>4</td>
<td>III-II</td>
<td>Good</td>
<td>Good</td>
<td>Normal</td>
<td>Good</td>
<td>Poor to good</td>
<td>Poor to good</td>
</tr>
<tr>
<td>5</td>
<td>II-I</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Good to normal</td>
<td>Good</td>
<td>Good</td>
</tr>
</tbody>
</table>

Figure 1. Lateral and frontal view of patient: presenting with vesicles, crusting, swelling on the left side of the face in acute onset (A) and first diagnosed as RHS at day 4 (B)

Figure 2. Electroneuromyography (ENMG) examination showed axonal facial nerve paralysis on the left side
the threshold of the sub motor, to reduce pain three times a week. Electrical stimulation is given at an early stage because it is supported by a circuit that can have positive outcomes in muscle activation and pain management and no published evidence of harm has been demonstrated.\textsuperscript{2,5,10,13} Facial massage was performed 10-15 minutes per day consisting of effleurage, kneading and stretching exercises on the affected side. Studies show that light massage can help muscle relaxation.\textsuperscript{4} Relaxation training is needed to increase the patient's awareness of facial tones and the small stimuli needed to increase muscle activity.\textsuperscript{4} Biofeedback training can be used to help patients begin voluntary movements and increase facial muscle strength with specific exercises in front of the mirror.\textsuperscript{4} Observation in front of the mirror at the movement of the unaffected side can provide visual clues to help the patient make movements on the affected side of the face.

The severity of facial paralysis is measured by the House-Brackmann Grading System. In the first week's intervention, the House-Brackmann score in this patient was grade IV (moderately severe dysfunction). There are several negative impacts on the functional aspects of a patient’s daily life such as the ability to drink, eat and express. After the fourth week, the score is increased to grade II. This rehabilitation program significantly enhances the House-Brackmann Grading System. This finding is in accordance with recent studies.\textsuperscript{5,15}

In the second week to the fourth week of intervention, the House-Brackmann score increased from grade IV to grade II. The patient showed improvement in resting asymmetry, symmetrical volunteer movements, and mild development in snarl, lip pucker, and closing eyes. The patient became more cooperative in examinations and starts an exercise program at home when he realized the changes in his condition for the better. This is supported by the research of Beurskens et al (2004) which states that facial massage and exercise as physical therapy can increase the growth and production of collagen and connective tissue in facial muscles and restore facial muscle function.\textsuperscript{16} Visual feedback is also said to control muscle activity in the facial muscles. Research by Hu et al (2001) have also shown that exercise and facial massage can improve facial symmetry in patients with long-term facial nerve paresis.\textsuperscript{11} Overall, there were greater improvements in the House-Brackmann scale after physical intervention.

A cochrane review found moderate quality evidence from one trial by Beurskens et al (2004) and suggested that this type of therapy has some benefit in chronic cases of idiopathic facial paralysis.\textsuperscript{3,16}

\textbf{Figure 1.} Lateral and frontal view of patient: presenting with vesicles, crusting, swelling on the left side of the face in acute onset (A) and first diagnosed as RHS at day 4 (B)

\textbf{Figure 3.} Physical examination after one-week post-diagnosis Ramsay Hunt Syndrome, and resolution of vesicles after 2 weeks from initial onset.

\textbf{Figure 4.} A first week of initial electrical stimulation therapy and started along with exercise and massage of the face

\textbf{Figure 5.} Healing of lesions after 3-4 weeks with minimal scarring, and progression of facial nerve palsy
Exercise therapy enhanced the House-Brackmann scale and increases the social subscale of the facial disability index. These results are consistent with what we found in which treatment showed a reduction in physical and social impairment. Thus, rehabilitation therapy can be used in the treatment of RHS to improve facial asymmetry in less time. Initial retraining should start with EMG biofeedback training because patients get accurate, direct feedback to ensure the right muscles are being recruited. With the assurance that the patient is doing the exercises correctly, mirror feedback exercises can be started.

The limitations of this study were that patient was not followed up for a longer period of time, and home exercises were not supervised. Future research in this kind of therapy should aim to assess its long-term effects with larger sample sizes. High quality evidence to assess daily and weekly session evaluations.

CONCLUSION
Proper diagnosis and treatment are essential to achieve the best cure. The patient received neuromuscular electrical stimulation (NMES), facial massage and biofeedback exercise therapy at the first week of intervention. To our knowledge, this is the first study that establishes the rehabilitation therapy of Ramsay Hunt Syndrome (RHS) in Bhayangkara Hospital Denpasar. The patient showed improvements in House-Brackmann scale, and facial muscle strength. Further better quality research is entailed to ponder electrical stimulation, facial massage and biofeedback training as a choice of safe, efficacious and provide good outcomes in the treatment of RHS.

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CONFLICT OF INTEREST
We declare that there were no conflicts of interest in this study.

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AUTHOR CONTRIBUTION
All of authors are equally contributed to the study from the study framework, data gathering, data analysis, until reporting the result of study.

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