



Case Study for treatment considerations for patients with Ramsay Hunt Syndrome

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Abstract

“Ramsay Hunt syndrome (RHS) is a rare neurological disorder characterized by paralysis of the facial nerve caused by varicella zoster virus [3].” RHS is not a diagnosis typically seen in physical therapy. Manifesting similar to Bell’s palsy on inception, RHS affects the cranial nerves (CN) with a variety of symptoms that may include: tinnitus, hearing loss, difficulty with speech or swallowing, loss of balance, inner ear involvement, extremity weakness or involvement, and a myriad of other possible symptoms. The purpose of this case study is to enlighten therapists and readers to the possible treatment considerations of the side effects caused by RHS based on the patient’s symptoms and presentation. Patient SC initially presented with shingles to cranial nerves (CN) V, VII, and VIII following a week long headache. Physical therapy (PT) was prescribed in hopes of facilitating and/or alleviating his symptoms of decreased balance and facial control. He responded very well to balance and vestibular training to regain control of his lower extremities to ambulate without his single point cane (SPC). He regained minimal control of his facial musculature with continued tinnitus. It is interesting to point out that he did respond well to electrical stimulation of his facial and cervical musculature while in the clinic with poor to fair carryover after and between treatment sessions.

Keywords: Ramsay Hunt Syndrome, physical therapy, exercise, treatment, therapeutic intervention

Background

Ramsay Hunt syndrome (RHS) is a rare diagnosis and is almost never seen or treated in physical therapy. Because RHS is seldom heard of in the physical therapy clinical setting knowing the signs and symptoms, and in this case the side effects, will help guide treatment. RHS presents similarly to Bell’s palsy (BP) with one exception: RHS is a geniculate herpes zoster infection of the external ear [2]. It is more difficult to differentiate between RHS and BP, unless done so by a physician or neurologist, if the vesicles from herpes zoster infection are missed by the patient. This is why it is important to know the possible symptoms and treatment considerations as patients’ afflictions are often more than external damage from shingles. A systematic review has been conducted resulting in 26 (twenty-six) studies that have documented patients with symptoms similar to SC; of the studies read only two (2) confirm the appropriate use of electrical stimulation for his symptoms [1,4]. With the lack of patient referral to physical therapy (PT) in the research, it’s important to consider treatment options based on how patients present

when diagnosed with RHS versus BP. The purpose of this case study is to bring to light the lesser known treatment options for patients with similar side effects in regards to RHS.

Case Description

At the time of the initial evaluation, SC was a 50 year old male who worked in a highly stressful position with the postal service mostly doing computer work for 10+ years. He reported having overall good general health and provided a list of comorbidities (Table 1). SC was initially treated for a viral sinus infection and diagnosed with BP prior to seeing his neurologist and being diagnosed with RHS. SC’s syndrome development was as follows: a week long headache and nausea, then the observation of blisters and blackened skin in and around his right ear and ear lobe. The blackened skin and blisters are indicative of the shingles infection or herpes zoster oticus. When his headache subsided, he noticed right side facial drooping and severe tinnitus in his right ear. His neurologist determined that cranial nerves (CN) V, VII, and VIII were affected.

Table 1. Co-morbidity List.

- Barrett's esophagus
- Kidney stones
- Acid reflux disease
- Gout
- Testicular disorder
- Sleep apnea
- Postherpetic neuralgia
- Arthritis with psoriasis
- Geniculate herpes zoster

He also developed decreased balance, vertigo, intermittent decreased use of his right lower extremity, right ear hearing loss, fatigue, difficulty with eating, swallowing and talking as well as cervical muscle hypertonicity and right side facial muscle atrophy. He underwent steroid treatments consistent with RHS protocol once the diagnosis was confirmed via his neurologist based on his symptoms. He was referred to PT for loss of balance and facial weakness with a diagnosis of RHS. SC's goals were to "get my hearing back, stop the vertigo, and stop the Bell's palsy."

Clinical Impression/Examination

SC came to PT from his neurologist with a diagnosis of RHS (geniculate herpes zoster). Instructions were to evaluate and treat; facial stimulation status post Bell's palsy. Prior to starting his initial evaluation SC was required to complete a FOTO (Focus on Therapeutic Outcomes) form. According to www.fotoinc.com, FOTO "is rehabilitation software measuring functional orthopedic patient outcomes." It allows for focus on plan of care for improved patient's responses to perceived function, dysfunction and pain summary. At his initial evaluation his scores were 40; computed predicted scores on discharge were 59, while his actual discharge scores were 44. Scores will vary and are individualized based on each patient's response to questions for clinicians to improve care.

Upon entering the clinic and the exam room, SC presented ambulating with a single point cane (SPC) and appeared hesitant in his quality of ambulation. His gait was observed to be slow in nature: taking a larger stride with his left foot versus his right, decreased stance time on his right lower extremity, SPC in his left hand, and mild gluteus medius gait. He reported after the infection he developed increased anxiety in dealing with his symptoms and navigating his surroundings confidently. He also developed gastro-intestinal and type II diabetes mellitus co-morbidities.

During the initial evaluation SC was thoroughly assessed for vestibular involvement, balance deficits, facial muscle control, and cervical and lower extremity mobility, range of motion (ROM), and manual muscle testing (MMT). The aforementioned testing is considered valid and reliable in assessing the various systems in the clinical setting as usual, customary, and best practice. Visual inspection revealed presentation of unilateral facial drooping and cervical posturing of right side bending and

left rotation. In assessing vestibular involvement and reports of vertigo symptoms the following testing was performed specific to the vestibular system to determine deficits: gaze stabilization, head thrust, Dix-Hallpike and Eply's maneuvers: testing revealed positive right ear involvement based on patient's report of symptoms, dizziness and nausea, and right horizontal nystagmus beat. Due to severity of reported symptoms and possibility of increased fall risk, components of the Berg Balance Scale were completed: focusing on static standing in tandem, parallel and single leg stance with eyes open and closed; also performed sit to stand and stand to sit. He demonstrated good sitting tolerance. However, he demonstrated poor standing and activity tolerance during static and dynamic testing. These results were confirmed by observed loss of balance, inability to tolerate standing testing greater than five (5) seconds and reports of increased dizziness and nausea. Facial muscle control was performed in sitting asking SC to perform various facial movements: eyebrow elevation, eye occlusion and opening, smile, frown and lip pursing. ROM of the cervical spine and lower extremities were performed in sitting and supine; assessing all planes focusing on the cervical spine, hips and ankles. He was able to demonstrate ROM within functional limits without gross deviations or limitations. MMT was also assessed in sitting and supine: cervical, upper and lower extremity strength grossly assessed in all directions and planes were functional and within normal limits. While he demonstrated functional strength and ROM about the extremities, SC reported difficulty controlling his right upper and lower extremities due to symptoms of vertigo.

SC's symptoms were as follows in no particular order self reported and observed: right vestibular involvement, severe right ear tinnitus, moderate loss of hearing to the right ear, decreased tongue mobility and use, difficulty swallowing and talking, right sternocleidomastoid (SCM), scalenes, trapezius and suboccipital cervical muscle hypertonicity, right frontalis, orbicularis oculi and oris, zygomaticus, buccinator, masseter, and temporalis facial muscle inhibition and mild atrophy, mild decrease in strength to the right lower extremity, moderate-severe loss of balance and stability, right dry eye, moderate inability to close right eye, and eye fatigue with increased symptoms of eye movement and use. He did report an increase in tinnitus with changes in weather patterns.

Based on his evaluation, diagnosis and research SC presented with deficits consistent with his neurologist diagnosis of RHS. His PT diagnoses were cervicgia, other abnormalities of gait and mobility and dizziness and giddiness. After researching RHS and becoming familiar with the symptoms and side effects it was difficult to determine a definitive prognosis. Careful considerations were taken regarding SC's current state, his goals and desired outcomes and traditional prognoses from the research to determine his prognosis to be poor-fair. A plan of care was developed to focus on his facial control via a home exercise program. Concentration on strength, balance, stability and proprioception were performed in the

clinic under direct therapist supervision. It's important to note, that because of the lack of referrals of patients in the studies to PT, it was difficult to determine reliability and validity of treatments and assign a favorable prognosis. As such, manuals, exercise, and modalities were trialed and completed based on SC's symptoms, side effects and complaints of pain or symptoms based on evidence based and best practice.

Intervention

SC attended 51 of his 52 scheduled appointments. He cancelled only one appointment and was on vacation for two weeks. Of the 51 appointments, ten (10) sessions consisted only of manual intervention and modalities due to performing the initial evaluation and patient reports of increased nausea, pain and vertigo. Also while receiving PT at approximately week 13, SC saw an ENT to receive steroid shots to his right ear to improve his hearing and tinnitus. After the third shot, he reported having no changes in hearing and tinnitus.

Each PT session began with manual intervention focusing on cervical muscle tonicity. Deficits were addressed through soft and deep tissue massage, myofascial release and pressure point muscle release to the suboccipitals, trapezius, scalenes, and SCM. Joint mobilizations to the right cervical spine facets, C1-C7, initially grades one and two, up to grades three and four as SC tolerated, 10-20 repetitions at each level. Manual intervention lasted 20-30 minutes with SC in the supine position. SC preferred head positioning of right rotation and lateral flexion to be able to hear with his left ear. This posture caused significant-severe right cervical muscle hypertonicity. As such,

manual intervention was performed every session to ensure full available cervical mobility to perform dynamic balance and vestibular exercises without limitations. In conjunction with manual intervention, three (3) subsequent visits immediately following the initial evaluation, mechanical traction was performed. SC was supine and received a progression hold cervical distraction program at a 20lb maximum pull. He initially tolerated mechanical traction. After reporting increased pressure and discomfort to his mastoid processes and underlying cervical tissues, traction was stopped and manual intervention resumed.

Sessions proceeded with vestibular exercises for improved balance, stability, and control during ambulation and for proprioceptive awareness for the potential to decrease the use of his SPC. Exercises were performed standing, walking, seated; all performed on even and uneven surfaces. They included dynamic stability with forward propulsion and head movements, dynamic standing with arm movements, and transfer of objects to various heights while standing on even and uneven surfaces. Refer to **Table 2** for a complete list of exercises and progressions. His exercises were gradually progressed to ambulatory and dynamic exercises based on his tolerance and reduction of symptoms. The first follow up session after his initial evaluation, vestibular exercises with eye movements were avoided as the patient reported increased symptoms of dizziness, headaches and severe eye fatigue. At approximately week 13, SC's symptoms and side effects were better managed with habituation becoming more evident, Dix-Hallpike and Epley's maneuvers were added to address symptoms of vertigo.

Table 2. Exercise Progression.

Week 1-2	Seated cervical ROM, progressed to standing, progressed to foam standing VOR 1 & 2 (as tolerated) seated, progressed to standing Standing static normal base of support and tandem stance Rhomberg standing on foam, progressed to eyes closed with arms crossed
Week 3	Standing 180 degree turns with targets, progressed to foam standing Seated trunk flexion holds
Week 4	Ambulation with head turns Standing cone pick-up, progressed to foam standing
Week 5	Sit to stand with head turn targets
Week 6	Tandem ambulation Standing figure 8 arm movements with 2lb medicine ball, progressed to foam standing
Week 7	Foam marching
Week 9	Figure 8 ambulation around cones
Week 11	Side stepping
Week 12	Hurdle ambulation
Week 13	Ambulation with cone pick up
Week 14	Overhead cone moving/activities
Week 15	Grapevine/weaving ambulation
Week 17	Walking with medicine ball rotation Plyotoss standing on foam Retro ambulation
Week 19	Exercise ball sitting with head turns Single leg stance

Modalities were typically performed at the end of treatment sessions. Initially ultrasound was attempted for ten (10) subsequent visits following the initial evaluation. SC complained of swelling and a full sensation about his right ear and surrounding cheek tissue. Ultrasound was initiated to attempt to alleviate any superficial swelling and facilitate and promote healing to the right ear and surrounding tissue: 100%, 1.5w/cm², 1mHz, eight (8) minutes to the right parotid gland. After three (3) visits without any reports of decreased swelling and full sensation, ultrasound was ceased.

Multiple forms of electrical stimulation were attempted following ultrasound for two reasons: H-Wave was used for SC's reports of swelling and full sensation and EMS, Russian and NMES waveforms were used for neuromuscular re-education to the muscles of the right side of his face and neck. H-Wave, which reduces symptoms via cumulative effect, was used in attempt to decrease any potential swelling around the parotid gland, right cervical spine and near the ear was applied using a low setting to influence the lymph system to aid in reducing swelling (13 visits) [4]. Electrode placement varied each session to the right side of SC's face and neck to target different points of the cervical and facial lymph system for 15 minute treatments. With the lack of improvement in using ultrasound and H-Wave to reduce reports of swelling and a full sensation in his right ear, attempts to decrease and/or alleviate this symptom ended. Focus was then directed towards facial and muscle control.

SC performed facial exercises for his home exercise program as he was less self-conscious performing facial expressions in the comfort of his home. In conjunction with facial exercises, EMS 2 C Personal Use Stimulator was initially used (1 visit). The EMS is better able to direct the current via a hand-held probe and applied directly to SC's right facial muscles times ten (10) minutes. The intensity was increased to a therapeutic range as dictated with the device. SC did not tolerate the EMS system as he reported stinging and shocking sensations that were painful. Russian stimulation was then attempted for the next 25 visits: electrode placement to the right facial muscles and the intensity increased until a muscle fasciculation was seen; 20 second contraction with patient muscle contraction, 30 second rest periods for 10 minutes. Russian stimulation was better tolerated as SC was able to dictate the level of stimulation to comfort and have mechanical muscle stimulation. With a poor carry-over between sessions of facial muscle control NMES was attempted for 10 visits to improve neuromuscular facilitation and carry-over: electrodes were placed over the right facial muscles and the intensity was increased to SC's tolerance for 15 minutes. He was unable to gain or maintain any functional control of the right side of his face or reduce reports of swelling with the use of any form of selected modality.

Outcome

After researching RHS and what methods were performed

to regain function, no participants were referred to PT for treatment. Based on this finding, SC was progressed through exercises, treatments and modalities based on his symptoms and how he presented daily using evidence based and best practice. Through vestibular progression, balance exercises and gait training SC was able to ambulate without his SPC and demonstrated improved consistent control of his right upper and lower extremity. This was compared to his initial presentation in which it was difficult for him to ambulate with his SPC. Initially, he presented with head posturing of right lateral flexion and left side bending mostly to be able to hear. With manual intervention, he was able to recover a minimal return of cervical muscle tonicity, flexibility and range of motion demonstrating improved midline cervical posturing. After attempts with an assortment of forms of electrical stimulation [1,4] on SC's face and neck, he was unable to gain facial, eye or swallowing control or speech improvements. Compliance with his home exercise program (HEP) was determined to be good as SC was able to demonstrate the exercises when asked including dictating repetitions and frequency. Additionally, no significant changes or improvements were made regarding his speech, facial muscle control or swallowing abilities with his performance of his home exercise program. Upon discharge from physical therapy, SC's cervical, upper and lower extremity and vestibular symptoms improved enough that he was able to manage them through a HEP he received at discharge. Compared to the initial evaluation, SC made minimal improvements in function confirming his poor-fair prognosis. Unfortunately, to this point, he has only been able to recover full closing of his right eye lid and no significant reportable improvements in swallowing or talking since discharge from PT.

Discussion

According to Sweeny et al. [6] "RHS may initially be indistinguishable from Bell's Palsy...due to delayed development of vesicles after the onset of facial weakness." SC's symptoms were consistent with those of previous patients who have been diagnosed with RHS via research articles [4-6,8,11-19]. The severities and number of symptoms per patient varied, but were overall similar in nature. It is important to note that in all the research performed, no patients were referred to physical therapy. While some patients had a resolution of their symptoms and good prognosis, SC's symptoms improved minimally with poor-fair prognosis. SC made some functional improvements such as the ability to ambulate without his SPC and independent management of his vertigo symptoms and minimal cervical muscle tonicity through his HEP. SC was able to learn the tools to continue with treatment at home and progress as tolerated. PT intervention for this patient was unable to make changes or improvements in his facial control, swallowing, and orating. This insight may be better researched via speech language pathology (SLP) therapy interventions. A combined PT/SLP study regarding this find-

ing would be beneficial in addressing all aspects of patient's deficits and care. Additional research regarding timing of PT intervention compared to onset of RHS would allow for better understanding of the disease progression, reducing side effects and disease severity, and improving prognosis.

Conclusion

In conclusion, due to the onset, severity, system involvement, and side effects RHS is a difficult disease to treat with favorable outcomes for good prognosis. While SC was able to recover gross coordination, balance and stability, only requiring his SPC as needed he continued to have ipsilateral facial, swallowing and orating deficits. For this patient, exercises and vestibular habituation were the best methods to improve his prognosis and overall function. Using clinical judgement in applying modalities is justified when attempting to reduce secondary symptoms or improve muscle facilitation and control. In using modalities for SC, it was based on his reports of swelling and full feeling in his right ear and lack of facial muscle control. It's important to conduct an extensive medical history for co-morbidities, subsequent symptoms leading up to or during disease contraction and length of time between disease contraction and steroid treatments to improve treatment plans, interventions, progression and outcomes. Based on each individual's signs, symptoms and side effects treatment interventions can be tailored to improve deficits for improved quality of life and function. Further research into combined treatments is warranted; i.e. PT and occupational therapy, PT and SLP, etc.

Competing interests

The authors declare that they have no competing interests.

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