CASE REPORT

# Ramsay Hunt Syndrome: A Case Report Underlining the Importance of Early Diagnosis to Prevent Complications.

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Submitted: 04/09/2024. Revised edition: 20/12/2024. Accepted: 07/02/2025. Published online: 01/06/2025.

**Abstract**

Ramsay Hunt Syndrome (RHS) is a rare neurological disorder and a sequelae of Herpes zoster, resulting from the reactivation of the Varicella zoster virus (VZV) in the geniculate ganglion. Patients usually present with acute unilateral peripheral facial nerve paralysis, often accompanied by vesicular eruptions in the external auditory canal or oral mucosa, a hallmark of herpes zoster oticus. Due to its overlapping clinical features, RHS is frequently misdiagnosed as Bell's palsy, a more common cause of facial paralysis seen in primary care settings. This case report underscores the potential for RHS misdiagnosis, in which can lead to delayed initiation of antiviral and corticosteroid therapy, resulting in suboptimal patient outcome. This case emphasizes the importance of early recognition of the condition, which are critical to improving prognosis, reducing complications, and enhancing the quality of life for affected patients.

**Keywords**: *Ramsay Hunt, Herpes zoster oticus ,facial nerve palsy.*

## Introduction

Herpes zoster oticus, another name for Ramsay Hunt Syndrome (RHS), is a late manifestation of varicella-zoster virus (VSV) infection. It results from inflammation of the geniculate ganglion of the 7th cranial nerve. The initial presentations are ipsilateral facial paralysis, vesicles at the ear, and otalgia [1]. RHS is a clinical diagnosis that requires early treatment to reduce the risk of complications. This case report describes a patient with RHS who developed prolonged facial nerve palsy due to delayed diagnosis.

## Case presentation

A 39-year-old man presented with intense left ear pain radiating to the left temporal skull region for one week, associated with a skin lesion on the left pinna. He also reported left-sided facial weakness and an inability to close his left eye. The patient denied headaches, dizziness, tinnitus, hearing impairment, recent trauma, insect bite, or upper respiratory tract infections. Prior to visiting our clinic, the patient sought treatment at another health facility, where he was prescribed a 3-day course of prednisolone. However, he noticed no improvement of his symptoms and decided to seek a second opinion. The patient had a history of varicella zoster during childhood, but no other significant past medical history. He also denied engaging in high- risk behaviours. On examination, the patient exhibited left-sided lower motor neuron nerve palsy, categorized as House-Brackmann grade IV. Examination of the left ear revealed yellow crusting lesions over the concha and desquamation of the skin over the antihelix [Figure 1]. Otoscopic examination identified a few vesicular rashes in the external auditory canal, with bilateral intact tympanic membranes. Routine blood tests showed no abnormalities. Serological test for varicella-zoster virus (VZV) IgG and IgM antibodies was not performed due to unavailability.

Based on the history and clinical findings, a diagnosis of RHS was made. The patient started on oral acyclovir 800 mg five times daily for one week and oral prednisolone 60mg once daily, tapered over two weeks. He was also prescribed analgesics, normal saline eye drops, and advised to use an eye patch over the left eye to prevent corneal irritation. Additionally, he was referred to the physiotherapy department for facial exercises and scheduled a follow-up appointment in two weeks to reassess his symptoms.

One week later, the patient returned to the clinic with worsening otalgia and yellowish, foul-smelling discharge from the left ear in the past three days. Examination revealed minimal dark crusting over the cymbal concha and yellow crusting with slough over the concha cavum, extending to the intertragic notch [Figure 2]. Otoscope examination showed an erythematous external auditory canal with minimal slough, but the tympanic membrane remained intact. The patient was diagnosed with acute otitis externa secondary to herpes oticus and started on ofloxacin ear drops, five drops twice daily.

During the third visit, one week later, the patient reported improvement in otalgia and no new symptoms. By this time, he had completed a 7-day course of acyclovir and was on a tapering dose of prednisolone. Physical examination revealed that the previous lesions had turned black and crusted over the concha area [Figure 3]. Despite the improvement in skin lesions, the facial nerve palsy persisted with no improvement. He was given another follow-up appointment in one month to reassess his symptoms, and physiotherapy was continued. Upon review after one month, the skin lesions over the pinna was healed entirely [Figure 4]. However, the facial nerve palsy remained at grade IV [Figure 5]. The patient was advised to continue follow-up with physiotherapy for facial exercises to improve his facial palsy.

## Discussion

RHS is a condition that accounts for approximately 0.3% to 18% of cases of acute non-traumatic facial palsy, making it the second most common cause after Bell's palsy [2]. Once the VZV infects a person, it remains dormant in the spinal and cranial nerve ganglia for life. Reactivation with further replication of the virus can occur during triggering events, leading to the virus spreading into the dermatome associated with the involved ganglion through sensory nerve fibers [3]. All individuals are at risk of getting RHS from early adulthood, regardless of their immune status [4]. Factors that can trigger RHS include stress, infections, malnutrition, cytotoxic drug use, diabetes mellitus, and malignant tumors [4].

RHS is primarily a clinical diagnosis. It is characterized by a vesicular rash involving the auricle, external auditory canal, soft palate, and pharynx. The key clinical features include facial nerve paralysis, otalgia, tinnitus, vertigo, and hearing impairment. In some cases, involvement of additional cranial nerves, such as cranial nerves V, VIII, IX, or X, may also occur [5]. Definitive confirmation of the diagnosis requires serological analysis detecting IgG and IgM antibodies against VZV or polymerase chain reaction (PCR) detection of VZV DNA [4].In this case, the diagnosis was made based on the clinical history and physical examination, as the patient presented with typical features of RHS. The most prominent finding of patients affected by RHS is unilateral hemifacial paralysis. Diagnosing acute facial paralysis demands a high degree of clinical vigilance, as early in its course, it can be challenging to differentiate from Bell's palsy, particularly when facial weakness manifests before the appearance of vesicles [6]. Physicians should perform a thorough facial nerve assessment and grade the severity of facial nerve palsy based on the House-Brackmann grading system. This involves evaluating asymmetry at rest over the face and neck and assessing the major extratemporal branches of the facial nerve. The House-Brackmann grading system helps physicians determine the severity of facial paralysis [1]. Grading of facial nerve palsy, according to House-Brackmann, is further described in Table 1.

Healthcare providers must carefully consider differential diagnoses of facial nerve palsy before initiating treatment for acute facial paralysis to ensure optimal patient outcomes. In this case, the patient was initially treated for Bell's palsy due to symptoms of otalgia and left-side facial nerve palsy without any vesicular rashes , which led to a delay in diagnosis and treatment initiation, eventually worsening the prognosis. It is also essential for healthcare providers to recognize atypical presentations of RHS. In the initial stages of RHS, before the appearance of vesicular rashes and facial palsy, the involvement of multiple cranial nerves can produce diverse early symptoms, increasing the risk of misdiagnosis [4].In the only prospective study on patients with RHS, 14% developed vesicular eruptions after the onset of facial paralysis. The atypical condition known as Zoster Sine Herpete is characterized by peripheral facial paralysis in the absence of ear or oral vesicular rash, with potential involvement of the cervical dermatome [5]. Therefore, a thorough clinical history, physical examination, and recognition of atypical presentation by primary care physicians are essential to avoid misdiagnosis. Another study concluded that the overall possible misdiagnosis of RHS is approximately 41% [7].

Herpes zoster is typically a self-limiting condition, and treatment primarily aims to reduce the risk of long-term complications. High-dose corticosteroids, administered orally or intravenously, should be used in combination with antiviral therapy for optimal management [1]. Oral antiviral treatment is limited to a 7 to 10-day course and should be initiated within 72 hours of rash onset for optimal effectiveness. These medications are well-tolerated at standard doses, such as acyclovir (800 mg five times daily for 7–10 days), famciclovir (500 mg three times daily for 7 days), or valacyclovir (1000 mg three times

daily for 7 days) [8]. The optimal duration of steroid therapy remains uncertain, with recommendations ranging from 4 to 37 days. Treatment should begin with a high dose, typically prednisone at 1 mg/kg/day (up to a maximum of 60 mg) or an equivalent, followed by a gradual tapering schedule [1]. In this case study, the patient was started on oral acyclovir 800mg 5 times daily for one week, oral prednisolone 60 mg once daily, and tapering regimen over two weeks. Symptomatic management is also essential. For pain management, acetaminophen, non-steroidal anti-inflammatory drugs (NSAIDs), and long-acting opioids can be used. Artificial tears used during the day and ocular lubricant ointment at night can help prevent exposure keratopathy. Healthcare providers must advise patients with lagopthalmos to use an eye patch to avoid corneal abrasion [1]. Treatment for RHS is identical regardless of the timing of diagnosis [9]. However, late diagnosis may cause complications such as postherpetic neuralgia, synkinesis, corneal abrasion, and exposure keratopathy [1].

## Conclusion

Healthcare providers must be able to distinguish the typical and non-typical symptoms of RHS and consider differential diagnoses for acute facial nerve palsy. Patients presenting with sudden onset of facial nerve palsy should be thoroughly assessed to establish the correct diagnosis. In this case report, we emphasized the importance of early detection of RHS. Complications arising from late diagnosis have also been highlighted. Permanent facial palsy and other possible sequelae of RHS can significantly impact a patient's social life. Therefore, early diagnosis and treatment are crucial to prevent complications [6].


## Acknowledgment

The authors express their gratitude to Hospital Director, OR Head department OR/ AND clinic staff of the Department of Otorhinolaryngology, Hospital Taiping.


## Conflict of interest

All authors declare no conflicts of interest.

**Ethics:** The patient provided consent for the use of images and the publication of his case.

## Authors' contributions

AHS contributed to the writing, review, and editing of the manuscript. SR contributed to drafting the manuscript, conducting the literature search, and reviewing and editing the manuscript.

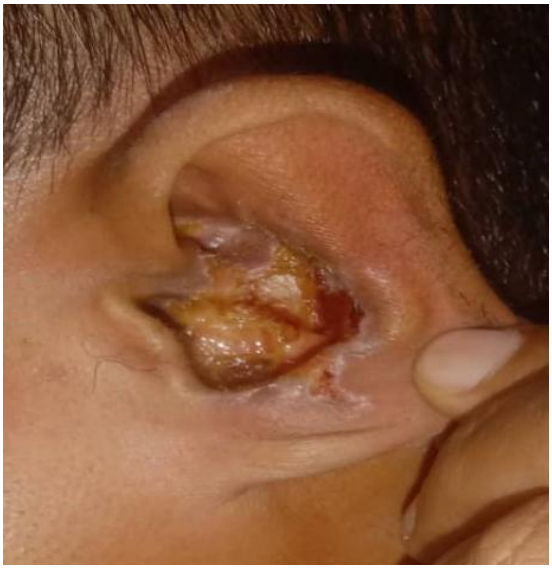

Figure 1. Photograph of the left ear. There was an area of yellow crusting over the concha with desquamation of skin over the antihelix. The pinna was mildly erythematous.

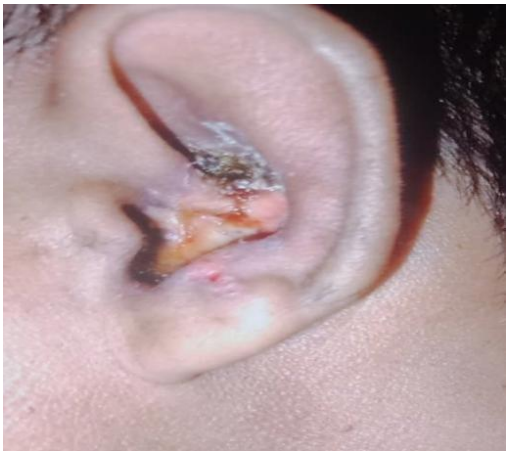

Figure 2. Photograph of the left ear. Minimal dark crusting over the cymba concha. Yellow crusting over the concha cavum extended to the intertragic notch.

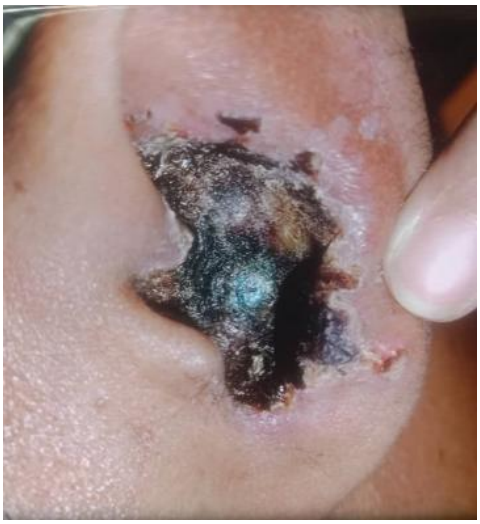

Figure 3. Photograph of the left ear. Noted black crusting over the area of the concha.

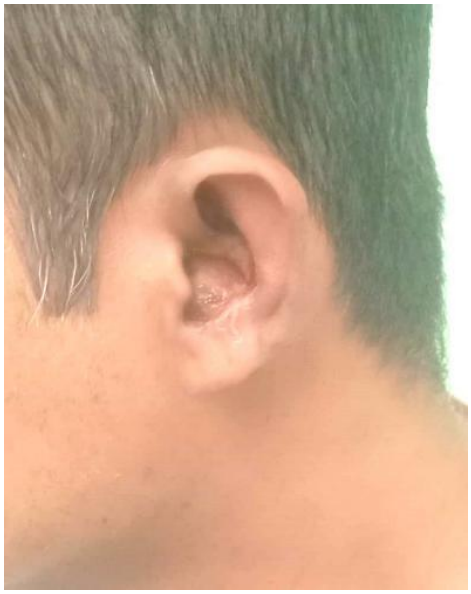

Figure 4. The skin lesion over the left pinna completely healed; no new lesion was seen.

Figure 5. Left facial nerve palsy House-Brackmann grade IV persisted after one month

Table 1. House-Brackmann Classification of Facial Function.[10]

| **Grade** | **Gross** | **Motion** |
|---|---|---|
| I. Normal | Normal facial function | Normal facial function |
| II. Mild dysfunction | On close inspection might notice minimal weakness.<br><br>Slight synkinesis possible<br><br>At rest, no facial asymmetry and good tone | Forehead movement sustains from moderate to good<br><br>Able to close eyes completely<br><br>Minimal asymmetry of the mouth |
| III. Moderate dysfunction | Evident of asymmetry but not much difference compare to the other side<br><br>Synkinesis, contracture, or hemifacial spasm can be seen but not severe<br><br>At rest, bilateral normal symmetry and tone | Forehead movement light to moderate<br><br>Able to close eyes completely with effort<br><br>Slight weakness over the mouth even with maximum effort |
| IV. Moderately severe dysfunction | Prominent weakness with/without disfiguring asymmetry<br><br>At rest, bilateral normal symmetry and tone | No forehead movement<br><br>Incomplete eye closure<br><br>Mouth appears asymmetrical with maximum effort |
| V. Severe dysfunction | Slight movement only<br>Evidence of asymmetry at rest | No forehead movement<br>Incomplete eye closure<br>Slight movement of the mouth |
| VI. Total paralysis | No movement | No movement |

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