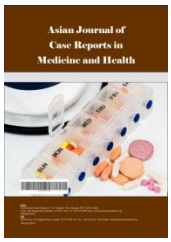

*Asian Journal of Case Reports in Medicine and Health*

*7(3): 14-18, 2022; Article no.AJCRMH.88662*

# Ramsay Hunt Syndrome with Cranial Polyneuropathy and Aseptic Meningoencephalitis: A Case Report

**W. L. M. Soysa [a*], Francisca Sivagnanam [a] and Nipuna Ranaweera [a]**

[a] *National Hospital of Sri Lanka, Sri Lanka.*

***Authors' contributions***

*This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.*

***Article Information***

**Open Peer Review History:**
This journal follows the Advanced Open Peer Review policy. Identity of the Reviewers, Editor(s) and additional Reviewers, peer review comments, different versions of the manuscript, comments of the editors, etc are available here: https://www.sdiarticle5.com/review-history/88662

***Case Study***

***Received 26 April 2022***
***Accepted 02 July 2022***
***Published 06 July 2022***

## ABSTRACT

**Introduction:** Ramsay Hunt Syndrome is characterized by paroxysmal ear pain, vesicular rash around the auricle and ear canal with ipsilateral peripheral fascial nerve palsy. It is due to reactivation of Varicella Zoster Virus (VZV) in the fascial nerve ganglion. It is considered to be the second most common cause of peripheral fascial nerve palsy. Involvement of other cranial nerves is uncommon. Rarely, dissemination of infection can lead to meningoencephalitis.

**Case Presentation:** A 14-year-old immunocompetent boy presented with fever associated with vertigo, fascial asymmetry and difficulty in swallowing. He also complained of a rash on his left ear associated with ear pain, one week prior to this presentation. He had a few crusted lesions on his left ear pinna and ear canal on examination. Cranial nerve examination revealed a left sided lower motor neuron type fascial and palatal palsy. Cerebrospinal fluid (CSF) analysis was suggestive of viral meningoencephalitis. He was treated for Ramsay Hunt Syndrome with cranial polyneuropathy and meningoencephalitis with Acyclovir and steroids. He had a uncomplicated recovery.

**Conclusion:** This case highlights the importance of clinicians diagnosing Ramsay Hunt Syndrome amongst patients presenting with multiple cranial nerve palsies and meningoencephalitis. It is a reminder that careful history and meticulous examination can lead to prompt recognition of a reversible cause of cranial polyneuropathy.

**Corresponding author: E-mail: lumalms101192@gmail.com, lumalms@gmail.com;*

*Keywords: Ramsay hunt syndrome; cranial nerves; acyclovir; varicella zoster; meningoencephalitis.*

## 1. INTRODUCTION

Ramsay Hunt syndrome (RHS), which was first described by J. Ramsay Hunt in 1907, refers to the association of unilateral peripheral facial nerve palsy and reactivation of Varicella Zoster Virus (VZV) along the sensory nerves innervating the ear [1]. RHS has an incidence of around 5 per 100,000 and is known to account for around 12% of fascial nerve palsies [2] Although the fascial neve is most commonly involved, other cranial nerve involvement is possible. They include VII, VIII, IX, X, V and III/XI cranial nerves, in descending order of involvement. The disease occurs due to reactivation of VZVin the geniculate ganglion of the fascial nerve. It can be confirmed by demonstration of VZV DNA in the geniculate ganglion [3] Hunt explained the mechanism of multiple cranial nerve palsy as an involvement of adjacent ganglia by contiguous anatomical contact from the original source of inflammation [4] Another theory suggested the spread of the virus through a common blood supply to cranial nerves. Other associations of RHS are Horner's syndrome, herpes zoster uveitis, meningoencephalitis, and the syndrome of inappropriate secretion of antidiuretic hormone [5-7].

This case is unique as the boy had both cranial polyneuropathy as well as meningoencephalitis, both being uncommon associations of RHS. Such complications usually occur amongst immunocompromised patients. But this case report highlights the need to consider the possibility of RHS amongst immunocompetent patients presenting with multiple cranial nerve involvement and meningoencephalitis.

## 2. CASE REPORT

A 14-year-old previously healthy boy was admitted with a complaint of vertigo and fever for one week. One week prior to onset of fever, he complained of a painful rash on his left ear which had resolved spontaneously. He described the rash to be clusters of small drops like lesions. He also complained of pain in his left ear and tinnitus. He had fever during that week which was low grade in nature, not associated with chills and rigors. He also complained of dysphagia, nasal regurgitation and imbalance for one week. He denied any weakness of his limbs. He had no headache, photophobia, phonophobia or neck stiffness. He had no fits or bladder/bowel incontinence. He had chicken pox when he was 7 years old which was uncomplicated. He had no significant past medical, surgical or allergic history. He is a school going child with average school performance. He denies smoking, alcohol or substance abuse.

On examination he was thin built and not in any respiratory distress. He had a left sided lower motor neuron type fascial nerve palsy (Fig. 1) and a left sided palatal palsy (Fig. 2). Eighth cranial nerve examination revealed normal hearing with no sensorineural or conductive hearing losses. He had an ataxic gait with swaying to the left. Romberg's test was positive. Rest of the cranial nerve examination was normal. He had no other cerebellar signs. Upper and lower limb examination was normal. He had no neck stiffness and Kernig's sign was negative. Ear examination revealed crusted lesions on the pinna and external auditory canal suggestive of a recent Herpes Zoster infection (Fig. 3). There were few vesicles on the left side of palate as well. Rest of the systemic examination was normal.

**Fig. 1. Left fascial nerve palsy**

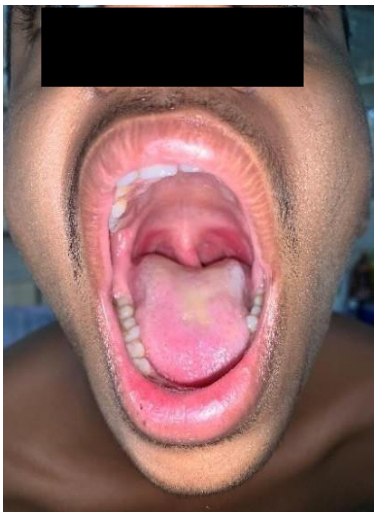

**Fig. 2. Left sided palatal palsy**

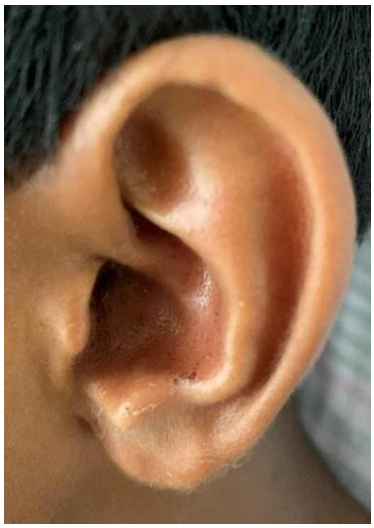

**Fig. 3. Herpes Zoster infection**

Routine blood investigations were normal. Pure Tone Audiometry showed no hearing loss. Cerebrospinal Fluid (CSF) analysis results are shown in Table 1.

MRI brain was normal with no evidence of tumor, infection or demyelinating disease.

A clinical diagnosis of Ramsay Hunt Syndrome was made, with involvement of seventh, eighth and tenth cranial nerves associated with VZV meningoencephalitis. He was started on IV Acyclovir 10 mg/kg three times a day for 14 days along with prednisolone 1 mg/kg/day for ten days, which was then tapered off.

Fascial weakness on House Brackmann scale was grade 3 (Moderate) at presentation and grade 2 (mild) on discharge. He was review ed after one month. It was found that the fascial and palatal weakness had completely resolved. He did not have vertigo or other features of vestibular component of eighth cranial nerve involvement.

**Table 1. CSF results**

| | |
|---|---|
| Protein | 73 mg/dl |
| Cell Count | 86/$mm^3$ |
| Neutrophils | 1/$mm^3$ |
| Lymphocytes | 85/$mm^3$ |
| CSF Sugar | 68 mg/dl |
| Random Blood Sugar | 96 mg/dl |
| Varicella Zoster Virus PCR | Positive |
| Herpes Simplex Virus 1 and 2 PCR | Negative |
| CytomegaloVirus PCR | Negative |
| Tuberculosis Gene Xpert | Negative |
| CSF culture | Negative |

## 3. DISCUSSION

Reactivation of the VZV in the geniculate ganglion causing lower motor neuron fascial nerve paly is a well described phenomenon and a common presentation in clinical practice. Involving of other cranial nerves, although less common has been reported in literature. This patient had involvement of the ipsilateral tenth cranial nerve suggested by the left sided palatal palsy. Involvement of the eight cranial nerve was the likely cause for the vertigo and positive Romberg's test. Normal hearing suggested involvement of only the vestibular component of the eight cranial nerve. It has been reported that the incidence of vestibular nerve involvement is three to four fold higher than that of cochlear nerve involvement [8]. Patient also had CSF fluid analysis suggestive of meningoencephalitis, with CSF being positive for VZV DNA. The patient's clinical presentation was possibly due to dissemination of reactivated VZV, causing local meningoencephalitis and multiple cranial nerve involvement. This is a rare combination of symptoms with a few cases reported previously [9] Further literature review showed that such extensive spread of the virus was rarely seen in immunocompetent patients [10].

Due to the widespread involvement of unilateral cranial nerves, the possibility of malignant infiltration and sarcoidosis was considered, although unusual at his age. [11] Screening for these entities were negative.

The beneficial evidence of antiviral therapy in RHS has been documented in literature [12]. This patient was started on early antiviral treatment as his CSF analysis was suggestive of a viral meningoencephalitis, which is associated with high mortality and morbidity. Treatment with Acyclovir reduces the duration of symptoms [13].

MRI and other neuroimaging being normal in such patients have been reported before [14] This highlights the fact that Ramsay Hunt Syndrome with cranial polyneuropathies should be diagnosed clinically.

## 4. CONCLUSION

Peripheral fascial nerve palsy is a relatively common presentation in clinical practice and physicians should always consider the possibility of Ramsay Hunt syndrome amongst such patients. Involvement of multiple cranial nerves should not diverge the diagnosis from RHS but should rather be supportive of it. Proper neurological assessment of other cranial nerves in patients with obvious fascial nerve palsies remain crucial. This case report highlights the possibility of coexisting meningoencephalitis in patients with RHS and the need for performing CSF analysis in them. This will allow early initiation of anti- viral drugs that will significantly improve the outcome of such patients.

## INFORMED CONSENT

Informed written consent was obtained from the patient's father to publish details regarding the patient's condition including photographic evidence.

## ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

## COMPETING INTERESTS

Authors have declared that no competing interests exist.

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*The peer review history for this paper can be accessed here:*
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