

# Herpes zoster oticus: A rare clinical entity

SHAILESH GONDIVKAR, VIREN PARIKH<sup>1</sup>, RIMA PARIKH<sup>1</sup>

## Abstract

Herpes zoster oticus also known as Ramsay Hunt syndrome is a rare complication of herpes zoster in which reactivation of latent varicella zoster virus infection in the geniculate ganglion causes otalgia, auricular vesicles, and peripheral facial paralysis. Ramsay Hunt syndrome is rare in children and affects both sexes equally. Incidence and clinical severity increases when host immunity is compromised. Because these symptoms do not always present at the onset, this syndrome can be misdiagnosed. Although secondary to Bell's palsy in terms of the cause of acute atraumatic peripheral facial paralysis, Ramsay Hunt syndrome, with incidence ranged from 0.3 to 18%, has a worse prognosis. Herpes zoster oticus accounts for about 12% cases of facial palsy, which is usually unilateral and complete and full recovery occurs in only about 20% of untreated patients. The most advisable method to treat Ramsay Hunt syndrome is the combination therapy with acyclovir and prednisone but still not promising, and several prerequisites are required for better results. We present a case of 32-year-old man suffering from Ramsay Hunt syndrome with grade V facial palsy treated effectively with rehabilitation program, after the termination of the combination therapy of acyclovir and prednisone.

**Keywords:** Geniculate ganglion, facial palsy, otalgia, Ramsay Hunt syndrome, unilateral

## Introduction

Ramsay Hunt syndrome (RHS), first described by James Ramsay Hunt in 1907,<sup>[1]</sup> is caused by reactivation of VZV which lies latent in sensory root ganglion for years in a patient who had chickenpox earlier. Involvement of geniculate ganglion of sensory branch of facial nerve leads to herpes zoster oticus (HZO) also known as RHS. Involvement of facial nerve leads to otalgia, lower motor neuron homolateral facial paralysis, and vesicular eruptions in auricle. In severe cases of HZO, involvement of vestibulocochlear nerve leads to sensorineural hearing loss in 10% and vestibular symptoms in 40% patients. Definitive treatment consists of antiviral therapy and steroids. This article describes the case of RHS with grade V facial palsy of House-Brackmann grading system treated effectively with combination therapy of acyclovir and prednisone, supported by rehabilitation program.

## Case Report

A 32-year-old man with previous healthy condition presented to us with pain in left ear for last 2 days. 24 to 36 h after the onset of otalgia, patient developed facial weakness along with vesicular eruptions on conchae and in external auditory meatus of left side. There was history of stressful life events

in past 6 months before the onset of rash. On examination, there was lower motor neuron facial palsy on left side which was complete. Bell's phenomenon was present on left side [Figure 1]. A neurologic examination revealed a weakness in the marginal mandibular branch of the left facial nerve [Figure 2]. Loss of definition in the ipsilateral nasolabial fold [Figure 1] and weakness in the temporal branch of the facial nerve was detected. There were painful adherent crusts and scabs in left conchae and external auditory meatus [Figure 3], associated with unclear hearing of left ear. The oral cavity and oropharynx were normal. Ocular examination demonstrated a spontaneous, lateral, left-beating gaze nystagmus but normal conjunctiva and sclera.

The patient was seen by an ENT consultant and the diagnosis of RHS with grade V facial palsy was confirmed. The ENT consultant administered an initial dose of intravenous acyclovir and steroids, and discharged the patient with a 2-week course of oral acyclovir and steroids. He was also referred to an ophthalmology clinic for corneal assessment. Meanwhile, because of the persistent grade V facial palsy, rehabilitation therapy was requested.

The rehabilitation program that the patient attended included transcutaneous electrical nerve stimulation and facial neuromuscular exercises. The facial exercise program was composed of (1) relaxation of hyperactive muscles, (2) facial massage exercises, (3) biofeedback training using a mirror to let patient know facial movement, and (4) specific facial exercises like smiling, grimacing, and whistling. The patient was instructed by the physiotherapist to follow this exercise program for two times a week, at least 60 min per visit and encouraged him to do methodically the facial exercise himself in front of a mirror at home. It took additional 3 weeks of outpatient rehabilitation program for total remission of his

---

*Department of Oral Diagnosis, Medicine and Radiology, K. M. Shah Dental College and Hospital, Piparia, Vadodara, Gujarat, <sup>1</sup>Parikh Hospital, Veraval, Gujarat, India*

**Correspondence:** Dr. Shailesh Gondivkar,  
# 21, 'Madhuban', Shakuntal colony, VMV Road,  
Amravati - 444 604, Maharashtra, India  
E-mail: shailesh\_gondivkar@yahoo.com



**Figure 1:** Photograph showing the presence of Bell's phenomenon and obliteration of nasolabial fold on left side



**Figure 2:** Photograph while full smiling



**Figure 3:** Photograph showing crusted eruptions on left conchae and external auditory meatus

facial palsy to occur, in addition to the 1-week inpatient rehabilitation.

**Table 1: Clinical features of Ramsay Hunt syndrome**

|                              |  |
|------------------------------|--|
| Key Clinical features        | Acute peripheral facial paralysis<br>Vesicles occurring anywhere along the sensory distribution of the facial nerve, including the anterior two-thirds of the tongue, the pinna or the external auditory canal |
| Additional clinical features | Otalgia<br>Tinnitus<br>Hearing loss<br>Vertigo<br>Nausea<br>Vomiting<br>Nystagmus<br>Change in taste perception  |

## Discussion

Ramsay Hunt suggested that HZO was due to geniculate ganglionitis. However, many contemporary authors agree that this condition represents a polycranial neuronitis. Several observations concluded that the primary etiologic agent of RHS is VZV but Bell's palsy, in contrast, has been attributed to herpes simplex virus type-1.<sup>[2]</sup>

Herpes zoster is seen as a disease of older people (most commonly over 60 years old), and incidence and severity increases with age which may be due to a decline in cellular rather than humoral immunity.<sup>[3]</sup> The VZV reactivation in the geniculate ganglia and subsequent neural inflammation, pressure, and possible destruction of the facial nerve in the temporal bone are suspected to cause facial palsy,<sup>[1]</sup> while VZV migrates from the geniculate ganglia into the skin around the ear or into the oropharynx via the sensory fibers, where it replicates and produces zoster in RHS.<sup>[1]</sup> Frequently, there is involvement of VIII cranial nerve producing hearing impairment and vertigo. Involvement of cranial nerves V, IX, X, XI, and XII occurs less frequently.

Facial palsy and zoster do not always appear simultaneously, and some patients with RHS exhibit facial palsy several days before or after the onset of zoster. VZV also causes acute peripheral facial palsy with the absence of skin lesions; such cases are termed zoster sine herpette and are usually diagnosed using serological assays<sup>[4]</sup> or polymerase chain reaction (PCR).<sup>[5]</sup> The diagnosis of RHS is based on the history and clinical findings mentioned in Table 1. Oral lesions are also present in most cases. Laboratory confirmation of the clinical diagnosis is based on increasing antibody titer in repeated complement fixation tests. PCR can detect VZV in saliva, tears, middle ear fluid, and blood mononuclear cells.<sup>[5]</sup>

The most recommended therapy for RHS is the combination of acyclovir and prednisone.<sup>[6]</sup> Acyclovir is an effective antimicrobial agent against actively replicating herpes zoster viruses. Acyclovir itself is not active. It must first be phosphorylated by viral thymidine kinase to form a

triphosphate. Acyclovir triphosphate inhibits viral DNA polymerase and, thus, DNA replication.<sup>[7]</sup> Importantly, no statistically significant outcome differences were noted between patients treated with intravenous or oral acyclovir.<sup>[7]</sup> Because of increasing viral resistance to acyclovir,<sup>[7]</sup> newer drugs, such as valacyclovir, famciclovir, penciclovir and brivudine, are being more commonly used.

Adjunctive steroid therapy can be helpful in the management of the facial paralysis of RHS.<sup>[8]</sup> A study on 80 RHS patients with different levels of severity treated with acyclovir-prednisone combination showed complete facial recovery, i.e., House grade I, in 52% patients, no matter what their pretreatment gradings were.<sup>[6]</sup>

For a significant improvement of facial recovery and hearing loss, early combination therapy within 3 days of the onset of facial palsy was critical; chances of grade I recovery was less than 30% if therapy started later than 7 days of onset. Previous studies reported better recovery of facial nerve function with combination therapy of acyclovir and steroids than steroids alone.<sup>[8]</sup> However, many authors caution against implementing steroid therapy, especially with periocular lesions, as they fear dissemination of the varicella zoster virus infection.<sup>[9]</sup>

The rehabilitation program for facial palsy includes electrical stimulation, infrared radiation, and facial neuromuscular exercises including automassage, relaxation exercises, inhibition of synkinesis, co-ordination exercises, or emotional expression exercises. The immunization of older persons with a VZV vaccine would boost their cell-mediated immunity to

VZV and thereby provide protection against herpes zoster and postherpetic neuralgia.<sup>[10]</sup>

## References

1. Hunt JR. On herpetic inflammation of the geniculate ganglion. A new syndrome and its complications. *J Nerv Ment Dis* 1907;34: 73-96.
2. Murakami S, Mizobuchi M, Nakashiro Y, Doi T, Hato N, Yanagihara N. Bell palsy and herpes simplex virus: Identification of viral DNA in endoneurial fluid and muscle. *Ann Intern Med* 1996;124:27-30.
3. Burke BL, Steele RW, Beard OW, Wood JS, Cain TD, Marmer DJ. Immune responses to varicella-zoster in the aged. *Arch Intern Med* 1982;142:291-3.
4. Morgan M, Nathwani D. Facial palsy and infection: the unfolding story. *Clin Infect Dis* 1992;14:263-71.
5. Murakami S, Honda N, Mizobuchi M, Nakashiro Y, Hato N, Gyo K. Rapid diagnosis of varicella zoster virus infection in acute facial palsy. *Neurology* 1998;51:1202-5.
6. Murakami S, Hato N, Horiuchi J, Honda N, Gyo K, Yanagihara N. Treatment of Ramsay Hunt syndrome with acyclovir-prednisone: Significance of early diagnosis and treatment. *Ann Neurol* 1997;41:353-7.
7. Dorsky DI, Crumpacker CS. Drugs five years later: Acyclovir. *Ann Intern Med* 1987;107:859-74.
8. Kinishi M, Amatsu M, Mohri M, Saito M, Hasegawa T, Hasegawa S. Acyclovir improves recovery rate of facial nerve palsy in Ramsay Hunt syndrome. *Auris Nasus Larynx* 2001;28:223-6.
9. Hill G, Chauvenet AR, Lovato J, McLean TW. Recent steroid therapy increases severity of varicella infections in children with acute lymphoblastic leukemia. *Pediatrics* 2005;116:523-9.
10. Oxman MN. Immunization to reduce the frequency and severity of herpes zoster and its complications. *Neurology* 1995;45:S41-6.

**Source of Support:** Nil, **Conflict of Interest:** None declared.