



## Ramsay hunt syndrome: a case report

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DOI: <https://doi.org/10.54448/mdnt23303>

Received: 06-12-2023; Revised: 08-27-2023; Accepted: 09-10-2023; Published: 09-13-2023; MedNEXT-id: e23303

### Abstract

Ramsay Hunt syndrome, known as Herpes zoster oticus, is a rare neurological condition due to the reactivation of the varicella-zoster virus (VZV) in the geniculate ganglion of the seventh cranial nerve. The triad is composed of vesicula-pustule-crust lesions in the ear and external auditory canal, lagophthalmos, and ipsilateral paralysis. Diagnosis is clinical and drug treatment is with antivirals and corticosteroids. The objective of this work is to report a case of Ramsay Hunt Syndrome and reinforce the importance of early diagnosis to minimize neurological damage. Final considerations: It is important to highlight that although the triad findings appeared late, this did not affect the diagnosis and treatment of the patient, who today presents a significant improvement in symptoms and quality of life.

**Keywords:** Ramsay Hunt Syndrome. Bell's Palsy. Corticosteroids.

### Introduction

Acute peripheral facial paralysis occurs due to an injury to the lower motor neuron of the facial nerve and in more than 70% of cases idiopathic (Bell's Palsy), however, it can also be caused by infections (varicella-zoster virus, cytomegalovirus), diseases autoimmune diseases (Guillain-Barré syndrome, systemic lupus

erythematosus, Sjögren's syndrome, Kawasaki disease), sarcoidosis, neurological diseases (acute disseminated encephalomyelitis, among others), trauma, iatrogenesis, pregnancy, neoplasms (glomus tumor, vestibular schwannoma, parotid) [1].

Ramsay Hunt syndrome, also known as Herpes zoster oticus, is a rare neurological condition due to the reactivation of the varicella-zoster virus (VZV) in the geniculate ganglion of the seventh cranial nerve [2]. Although the primary infection is incident, mainly in children, less than 1% of cases result in the syndrome. Furthermore, Ramsay Hunt syndrome is responsible for 16% of unilateral facial paralysis in children and 18% in adults [3].

The classic triad of Ramsay Hunt syndrome is composed of vesicula-pustule-crust lesions in the ear and external auditory canal, lagophthalmos, and ipsilateral paralysis [2]. Other findings, such as hearing loss, neuralgia, and tinnitus, may occur and, less frequently, dysgeusia may be observed, dry eye, nasal obstruction, and dysarthria. Advanced age, pregnancy, lack of vaccination for the varicella-zoster virus, newborns, immunosuppression, and physical and psychological stress are factors that predispose to the reactivation of the virus [3].

Diagnosis is clinically based on clinical history and physical neurological examination. Complementation with tomography or magnetic resonance imaging is important due to possible differential diagnoses [4].

Drug treatment is carried out with corticosteroids and antivirals.

Therefore, the objective of this study is to report an atypical case of Ramsay Hunt Syndrome, addressing the diagnosis and the need for early treatment.

## Methods

### Study Design

The present study was prepared according to the rules of the CARE case report Available at: <https://www.care-statement.org/>. Accessed in: 06/11/2023. This case study was carried out after the patient's authorization through the Informed Consent Form. The patient's medical records were analyzed, correlating the findings with the literature. The medical records were collected and analyzed at Hospital Escola Emílio Carlos, Catanduva, São Paulo, Brazil.

### Ethical Approval

This study was approved by the Research Ethics Committee of Centro Universitário Padre Albino (UNIFIPA) with approval number 6.273.833.

## Case Report

### Patient Information and Clinical Findings, Timeline, Diagnostic Assessment, Therapeutic Intervention, and Follow-up

The patient, a female, 53 years old, born and living in Catanduva, São Paulo, Brazil, smoker, and hypertensive, sought emergency medical care at Hospital Padre Albino due to otalgia on the left, ipsilateral headache without improvement with simple analgesia for 8 days that progressed to hemiface paresthesia left.

On physical examination, the patient presented left lagophthalmos, reduced sensation in the hemiface and left hemiparesis and dysarthria. During otoscopy, she gave mild pain upon manipulation of the left external auditory canal, intact tympanic membrane, bilaterally translucent. The non-contrast head tomography showed no changes. Then, the patient was discharged with prednisone for seven days, due to the diagnostic hypothesis of Bell's Palsy.

The patient returned to care after 4 days of treatment, with the clinical condition remaining and the appearance of vesicular lesions that evolved into crusted lesions in the left external auditory canal. Associated with the condition was a loss of strength in the left lower limb and gait ataxia, at the time, while using acyclovir prescribed in another service. In the neurological examination, paresis was observed in the lower limbs, mainly on the left, and gait ataxia, with lagophthalmos and hemiparesis remaining on the left. With the new

clinical findings, the diagnostic hypothesis of Ramsay Hunt syndrome was suggested.

During hospitalization for the investigation, asymmetric enhancement was observed on brain Magnetic resonance imaging (MRI) in the intracanalicular path of the facial nerve on the left after contrast administration (Figures 1 and 2).

Figure 1. Contrast-enhanced skull tomography in the coronal section showing asymmetric enhancement in the intracanalicular path of the facial nerve.

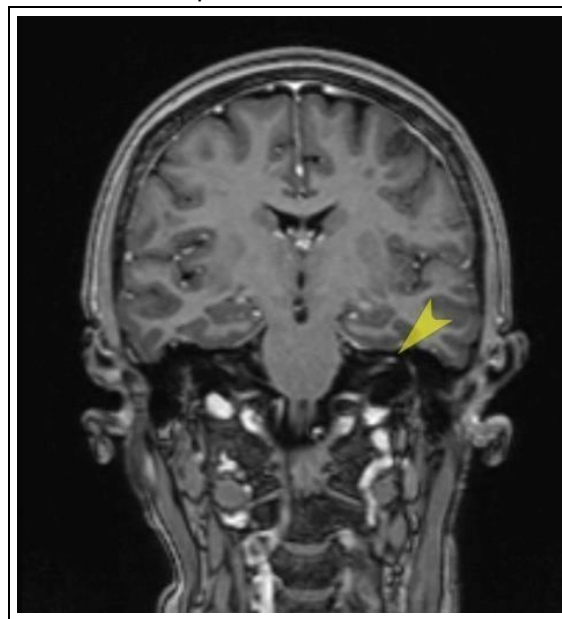
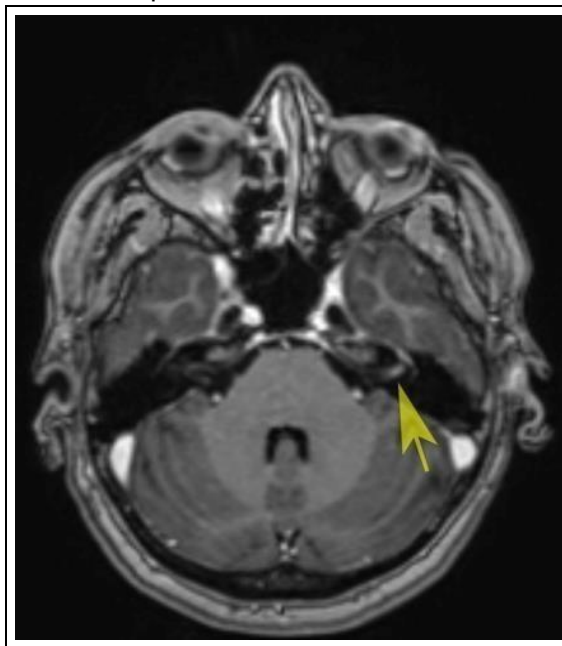


Figure 2. Contrast-enhanced cranial tomography in the axial section showing asymmetric enhancement in the intracanalicular path of the facial nerve.



The patient's condition improved partially after treatment with corticosteroid therapy and intravenous acyclovir and was discharged from the hospital after four days to complete home treatment and return to the

outpatient clinic. After 1 month, she presented remission of lesions in the left ear canal and partial improvement of deficits.

Two months after hospital discharge, the patient showed partial improvement in gait, however, he reported daily biparietal and occipital headaches, preceded by tinnitus in the left ear, which improved with simple analgesia associated with dysphagia for solids. Due to pain, the patient was hospitalized again for investigation of herpetic encephalitis. On magnetic resonance imaging with contrast, no changes were observed.

The examination of the cerebrospinal fluid showed a clear and colorless appearance, with nucleated cells of  $4/\text{mm}^3$  (reference value = up to  $3/\text{mm}^3$ ) - without differential, red blood cells ( $3/\text{mm}^3$ ), chloride (126 mg/dL); glucose (55 mg/dL), proteins (41 mg/dL), negative culture and non-reactive FtaBS. HbsAg, anti-HIV, anti-HCV, and serology for brucellosis were non-reactive. Thus, empirical treatment for herpetic meningoencephalitis was performed with intravenous acyclovir and corticosteroid therapy.

One year after the onset of the condition, the patient maintains dysarthric speech, gait ataxia, tinnitus, vertigo, and dysphagia despite therapy with motor physiotherapy and speech therapy.

## Discussion

Ramsay Hunt Syndrome can present as a cranial polyneuropathy due to the anatomical proximity of the nerves [4,5]. It is considered the second cause of non-traumatic facial paralysis [6]. It usually presents with a typical pattern of development, with emphasis on the triad vesicula-pustule-crust eruption in the ear pinna and around the external auditory canal, ipsilateral peripheral facial paralysis, and ipsilateral lagophthalmos. In the case described here, attention is drawn to the late appearance of the characteristic dermatological lesions, as well as the late appearance of facial paralysis. It should be a differential diagnosis of Bell's palsy.

Cutaneous findings are found in Hunt's zone, which encompasses the eardrum, external auditory canal, and the central portion of the ear [5,7]. Also noteworthy is the change in gait, not commonly associated with the syndrome. In all cases, it is extremely important to start treatment with corticosteroids and acyclovir early to reduce the duration of active disease and reduce neural damage and post-herpetic neuralgia [8]. It is important to combine motor physiotherapy to treat motor sequelae, which tend to be more serious and less likely to improve when compared to Bell's palsy.

## Final Considerations

It is important to highlight that although the triad findings appeared late, this did not affect the diagnosis and treatment of the patient, who today presents a significant improvement in symptoms and quality of life.

## Acknowledgement

Not applicable.

## Ethical Approval

Not applicable.

## Informed consent

Not applicable.

## Funding

Not applicable.

## Data sharing statement

No additional data are available.

## Conflict of interest

The authors declare no conflict of interest.

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Was performed.

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