

## An Atypical Ramsay-Hunt Syndrome: Case Report

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### Abstract

### Case Report

After the initial infection, which manifests as varicella, the varicella-zoster virus (VZV) remains dormant in the sensory nerve ganglia. It reactivates in elderly people or in immunocompromised patients. When this reactivation involves the geniculate ganglion, it typically causes the Ramsay-Hunt syndrome manifesting as shingles in the Hunt's zone which includes the eardrum, the external auditory canal and the cavum conchae, as well as a peripheral facial nerve palsy, earache and sensitivity or sensorial disturbances of the nearby cranial and spinal nerves. Our case describes a 40-year-old patient who developed an atypical Ramsay-Hunt syndrome associating only peripheral facial nerve paralysis and a herpetiform vesiculopustular rash in the retroauricular area, with no otologic, vestibular or sensory signs. In fact, there are anastomoses between the different cranial nerve fibers and ganglia with the spinal nerves and ganglia, which may explain the occurrence of herpes zoster in the C2 dermatome with peripheral facial nerve palsy.

**Keywords:** Ramsay-Hunt syndrome – Atypical – Varicella-zoster virus - C2 dermatome.

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## INTRODUCTION

After chickenpox, the primary infection caused by varicella-zoster virus (VZV), this virus seeks refuge in the spinal or cranial sensory nerve ganglia, where it remains latent for years. When VZV reactivates, particularly in the elderly or immunocompromised patients, it causes shingles, which classically manifests as cutaneous and neurological lesions.

When this reactivation involves the geniculate ganglion, it typically causes herpes zoster oticus associated with an acute peripheral facial nerve palsy, which some authors refer to as Ramsay-Hunt syndrome, although the definition of this diagnosis is not unanimous.

In fact, we report here a case of Ramsay-Hunt syndrome manifesting as retroauricular zoster with a peripheral facial nerve palsy, but not meeting the criteria of Malin *et al.*, Robillard *et al.*, or even Sweeney and Gilden, which raises the need to review the diagnostic criteria to include certain atypical forms, of which our case is one.

## CASE REPORT

A 40-year-old patient with no particular medical history, presented to the emergency department with an acute peripheral facial nerve palsy and vesiculopustular lesions grouped in clusters on an erythematous base in the right retroauricular area in the mastoid region (figure 1).



Figure 1: Herpetiform lesions in the right retroauricular area on the C2 dermatome

The patient had no otologic signs nor vertigo. The ear was not painful, there were no lesions in the conchae nor in the external auditory canal and the eardrum was normal in appearance.

Neurological examination revealed a right peripheral facial nerve palsy with facial asymmetry, inability to close the right eye with a positive Bell's phenomenon, and a discrete drooping of the right corner of the mouth (figure 2).



**Figure 2: Right peripheral facial nerve palsy with facial asymmetry, inability to close the right eye with a positive Bell's phenomenon, and a discrete drooping of the right corner of the mouth**

The patient also had difficulty blowing and whistling, and when the cheeks puffed up, air escaped from the affected side (figure 3).



**Figure 3: Air escaping from the affected side when the patient tries to puff his cheeks in a peripheral facial nerve palsy**

Forehead wrinkles were less pronounced on the affected side.

Taste on the anterior two-thirds of the tongue was preserved, and the patient showed no nystagmus.

Examination revealed a tender, mobile, soft right sub-mandibular adenopathy.

Given the acute nature of his symptomatology, the patient was hospitalized for antiviral treatment and IV corticosteroid therapy. Complementary examinations

revealed diabetes. The course was favorable under treatment. The patient received neuromuscular facial re-education sessions.

## DISCUSSION

The Ramsay-Hunt area includes the eardrum, the external auditory canal and the cavum conchae which is the central portion of the ear.

Ramsay-Hunt syndrome is defined by Malin *et al.*, as zoster oticus associated with peripheral facial nerve paresis with taste disturbances and reduction of tear secretion, sensitivity disturbances in the innervation area of the trigeminal nerve, sensitivity disturbances in the cervical dermatomes and lesions of the acoustic nerve and the vestibular nerve.

Robillard *et al.*, define it as a triad of ear pain, peripheral facial nerve paresis and herpes zoster lesions on the external auditory tract, the conchae or other cervical dermatomes.

Sweeney and Gildea on the other hand define it as the association of peripheral facial nerve paresis and zoster lesions on the ear or oral mucosa (Wagner *et al.*, 2012)

Our patient presented with zosteriform lesions outside the Ramsay-Hunt zone, in the territory of the C2 cervical nerve. Indeed, given the anastomosis of facial nerve fibers with other cranial and cervical

nerves (Rivera *et al.*, 2024), skin lesions can occur beyond the Hunt area. These lesions may involve the entire external ear, the hemifacial innervation zone of the trigeminal nerve, and the skin dermatomes from C2 to C4.

## CONCLUSION

Herpes zoster does not have to reach Hunt's area to be associated with peripheral facial nerve paralysis. In fact given the anastomoses between the various nerve fibers and the cervical ganglia, retroauricular herpes zoster may be accompanied by peripheral facial nerve paralysis.

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