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Case report

# Neurotrophic corneal ulcer and iridocyclitis directly preceding Ramsay-Hunt Syndrome

Farzan M. Gorgani, MS, DO. [a,b,c,*], Todd L. Beyer, DO, FAAO, FASOPRS. [a,b]

[a] Novus Total Eye Care, 518 West Ave, Tallmadge, OH, USA
[b] Cleveland Clinic Akron General, 1 Akron General Ave, Akron, OH, USA
[c] New Jersey Eye Center- Dello Russo Laser Vision, 1 N Washington Ave, Bergenfield, NJ, USA

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ABSTRACT

*Purpose:* To present to and inform the practitioner of an unusual presentation of Varicella zoster virus and Ramsay-Hunt Syndrome.

*Observations:* A 69-year-old bedbound male with vascular dementia presented to the emergency room with a red right eye with associated tearing and mucus production. The patient could not express if he was in pain. The initial diagnosis from the emergency room was bacterial keratitis, confirmed with a positive pseudomonas culture. However, upon examination by the ophthalmologist it was noted that there was not only a large, infected epithelial defect, but also an intraocular pressure of 35 and a candy-cane hypopyon. The diagnosis of herpes neurotrophic keratitis and iridocyclitis was made and the patient was started on intravenous acyclovir along with the appropriate topical medications. A day later, it was noted that the patient developed a right sided facial palsy and vesicular lesions inside the right ear canal, as confirmed by otolaryngology.

*Conclusion and Importance:* Ramsay-Hunt Syndrome is usually known to the ophthalmologist due to the exposure keratopathy caused by facial palsy. This case demonstrates varicella-zoster virus (VZV) neurotrophic keratitis preceding the development of facial palsy, which can further exacerbate an already neurotrophic cornea. The practitioner should be aware of these signs and symptoms and adjust their treatment with systemic acyclovir-prednisone.

## 1. Introduction

Varicella-zoster virus (VZV) reactivation in the geniculate ganglion, and involvement of the sensory nerves innervating the ear, defines herpes zoster oticus.[1] When associated with facial paralysis, it is known as Ramsay-Hunt Syndrome (RHS), which includes ear pain and vesicular lesions in the ear.[1] Due to the proximity of the facial nerve fascicles to the VIII$^{th}$ nerve in the bony facial canal, these 2 cranial nerves are commonly co-involved characterized by not only facial paralysis and painful vesicular lesions in the ear canal, but also hearing loss.[1] However, poly-cranial-neuropathy has been reported in many cases of RHS due to VZV causing ascending inflammation along the brainstem between several cranial nerve nuclei and dorsal root ganglia. It is also hypothesized that VZV can travel along brainstem reflexes which would allow it to travel to multiple cranial nerves during reactivation.[2–4] RHS is usually known to the ophthalmologist due to the paralytic ectropion and exposure keratopathy caused by facial nerve palsy; it is the second most common cause of facial nerve paralysis behind Bell's palsy.[4]

Cranial nerve V is one of the more commonly co-affected cranial nerves in RHS.[2–4] Herpes zoster ophthalmicus (HZO) with necrotizing retinitis, neurotrophic keratitis, and exposure keratitis have all been reported following the RHS presentation, usually along with vesicular lesions in the V1–V3 dermatomes.[5,6] Our case is the first known case of neurotrophic keratitis and iridocyclitis, which are common sequela of herpes zoster reactivation in the eye, with facial palsy and vesicular lesions in the ear canal occurring subsequently. This would suggest that VZV reactivation in the trigeminal ganglion can also descend to the geniculate ganglion and cause RHS.

## 2. Case report

A 69-year-old bedbound male with type 2 diabetes mellitus (HbA1c 7.3%), end-stage renal disease on dialysis, vascular dementia, and chronic pressure ulcers presented to the emergency room for a red right

* Corresponding author. DO. 518 West Ave Tallmadge, OH, 44278, USA.
*E-mail address:* fgorgani@dellorusso.com (F.M. Gorgani).

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eye along with tearing and mucous production. The patient was unable to provide a history or express if he was in pain due to his mental status. The initial diagnosis in the emergency room was bacterial keratitis and a culture was taken by the emergency room physician, followed by admission for around-the-clock topical antibiotics. The patient was started on moxifloxacin and tobramycin drops alternating every 2 hours. The patient was seen by ophthalmology the next day and the right eye was found to have an intraocular pressure of 35, a large central neurotrophic epithelial defect with a stromal infiltrate, and a 1mm hypopyon and hyphema, also known as a candy-cane hypopyon Fig. 1. There was good eyelid blink and closure. *Pseudomonas aeruginosa* was grown in culture susceptible to the current antibiotics. The diagnosis of herpes keratitis and iridocyclitis was made and the patient was promptly started on intravenous acyclovir along with topical IOP lowering drops and cycloplegics. Given the patient's physical condition, it was decided not to bring him to the operating room for an anterior chamber paracentesis for viral cultures and PCR. Moreover, the clinical signs highly suggested a viral pathology and that the bacterial infiltrate was a super-infection. The next day, the stromal infiltrate was noted to be coalesced and more superficial, so topical prednisolone was added, antibiotic drop frequency was reduced and tobramycin drops was switched to tobramycin ointment. At this time, it was noted that the patient developed a right-sided lower facial palsy and vesicular lesions inside the right ear canal, as confirmed by otolaryngology examination Figs. 2 and 3. The diagnosis of VZV Ramsay-Hunt Syndrome was made and 60mg intravenous prednisone was added, dosed as a weekly taper of 10mg. The patient developed a right lateral lower lid paralytic ectropion, but still had good lid closure and ointment coverage, so a tarsorrhaphy was not performed. The facial palsy improved within 2 days of initiation of intravenous prednisone, along with improvement of the right intraocular pressure, corneal infiltrate, epithelial defect and hypopyon. The prompt response to the anti-viral and steroid medication further suggested that the patient's condition was viral in origin. The patient was discharged on tapering oral prednisone, topical prednisolone and tobramycin ointment.

## 3. Discussion

RHS is known to involve multiple cranial nerves at one time. Cranial nerves VII and VIII are most commonly co-involved due to close proximity of the geniculate ganglion to the VIII$^{th}$ nerve within the bony facial canal.[1] There are several hypotheses describing the ability of VZV to affect multiple cranial nerves simultaneously: several cranial and dorsal root ganglia comprise a chain in which inflammation of a single ganglion could extend to nearby ganglia in the brainstem (ganglionitis), namely cranial nerves V, IX, X, XI, XII. It is also hypothesized that all cranial nerves involved in RHS are connected by pathways of important brainstem reflexes, which would allow VZV to spread along these pathways causing multiple cranial nerves to be involved.[2–4] This case is the first of our knowledge to have a polyneuropathy involving cranial nerve V first, presenting as neurotrophic ulcer and iridocyclitis, followed by the more classic cranial nerve VII involvement. Acyclovir-prednisone administration is generally accepted as the standard of care in improving facial nerve palsy and reducing cranial nerve inflammation, however some studies with acyclovir alone are equivocal.[1,7,8]

The facial palsy invariably can cause lower lid paralytic ectropion and exposure keratopathy. Previous case reports have been presented with CN V and VII co-involvement presenting as neurotrophic keratitis and necrotizing retinitis, with RHS as the primary presentation. The patient with neurotrophic keratitis initially presented to the otolaryngologist and was diagnosed with RHS. The patient subsequently developed vesicular lesions along V1 and V2, followed by a painful, red eye and was diagnosed with neurotrophic keratitis. The authors hypothesized that VZV laid dormant in both the trigeminal and geniculate ganglia and reactivated simultaneously.[5] Another patient was described to have acquired immunodeficiency syndrome and developed left-sided hearing loss, vesicular lesions in the left V3 distribution and necrotizing herpetic retinopathy of the left eye. Several days later, the patient developed contralateral signs of VZV; right-sided hearing loss and necrotizing retinitis of the right eye. MRI confirmed neuritis in bilateral CN VIII. The authors concluded that the ophthalmologist should monitor the fundus for retinitis in patients with RHS.[6] Lastly, a patient was reported to have sequential RHS, HZO and disseminated-cutaneous herpes zoster, which more commonly happens in immunocompromised

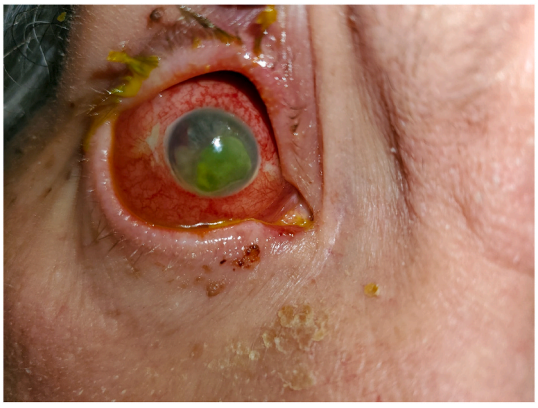

**Fig. 1.** External photograph of the right eye. A large central epithelial defect is seen along with a small hypopyon mixed with hyphema, known as a candy-cane hypopyon. This is classically associated with herpes iritis.

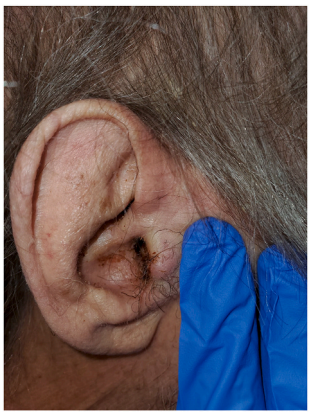

**Fig. 2.** External photograph of the right ear. New vesicular eruptions in the right ear canal as confirmed by otolaryngology.

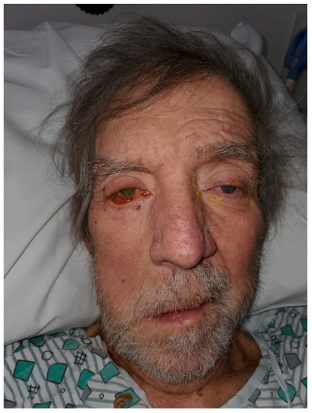

**Fig. 3.** External photograph of the patients face. A new onset right facial palsy causing a paralytic lower lid ectropion.

patients, but can rarely occur in immunocompetent. The authors suggested a similar mechanism as with our patient, that the affected sensory nerves can affect adjacent cranial nerves through anastomoses in the brainstem and even the spinal ganglia.[9]

Our case differs from the others from our literature search as RHS is typically the presenting manifestation of VZV. Our patient is the first known case where CN V was involved initially and the patient developed neurotrophic keratitis and iridocyclitis, which is a typical presentation of CN V VZV, with a subsequent development of RHS. Our hypothesis of this presentation coincides with what was described previously: reactivation of VZV in one ganglion can either cause ganglionitis among neighboring ganglia or that the virus traveled to the geniculate ganglion via brainstem pathways. The practitioner should be aware that facial palsy can develop with VZV reactivation and can exacerbate an already neurotrophic cornea.

## Patient consent

Permission for photographs was obtained by the patient's next-of-kin.

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## Intellectual property

We confirm that we have given due consideration to the protection of intellectual property associated with this work and that there are no impediments to publication, including the timing of publication, with respect to intellectual property. In so doing we confirm that we have followed the regulations of our institutions concerning intellectual property.

## Research ethics

We further confirm that any aspect of the work covered in this manuscript that has involved human patients has been conducted with the ethical approval of all relevant bodies and that such approvals are acknowledged within the manuscript.

Written consent to publish potentially identifying information, such as details or the case and photographs, was obtained from the patient(s) or their legal guardian(s).

## Authorship

All listed authors meet the ICMJE criteria.
We attest that all authors contributed significantly to the creation of this manuscript, each having fulfilled criteria as established by the ICMJE.

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## Authors' contributions

Farzan Gorgani, MS, DO.: Conceptualization, Methodology, Investigation, Writing - Original Draft, Visualization.

Todd L. Beyer, DO, FAAO, FASOPRS, FAACS.: Writing - Review & Editing, Supervision, Project administration.

## Declaration of competing interest

No conflict of interest exists.

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