

## CASE REPORT

## A Rare Cause of Facial Palsy

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### ABSTRACT

#### Abstract:

A 41-year-old female presented with a three-week history of sore throat, cough, sneezing, and a right fronto-occipital headache. Over three days, she developed intractable nausea and vomiting, ataxia, right otalgia, and a worsening headache. Her physical exam was remarkable for a complete right facial palsy, vesicles on her right upper lip, hyperacusis, and an impaired gait. A non-contrasted computed tomography scan demonstrated no acute process within the brain. Magnetic resonance imaging showed nonspecific increased signals in the cerebellar cortical sulci and occipital lobes, an ill-defined leptomeningeal enhancement in the cerebellar vermis, and an enhancement of the right cranial nerve V. At the time, consideration was given to herpes simplex virus (HSV) encephalitis and meningoencephalitis, and the patient was treated empirically with oral acyclovir and methylprednisolone. Lumbar puncture demonstrated lymphocytic pleocytosis and a varicella zoster virus PCR of vesicular fluid from the lip was positive. These findings are consistent with herpes zoster oticus, also known as Ramsay Hunt syndrome (RHS) and associated meningoencephalitis. The patient was continued on corticosteroids and antiviral therapy; she had a recurrence within two months and was given another course of acyclovir.

#### Case Report:

A 41-year-old female with a past medical history of asthma, anxiety, and hypothyroidism, presented with headache, some nausea and vomiting, and right facial weakness. The patient had originally presented to the Emergency Department (ED) with sore throat, coughing, sneezing, and a generalized headache that was more intense over the right fronto-occipital region, and she was initially diagnosed with sinusitis, treated with a course of analgesics and ciprofloxacin, and sent home. However, she presented multiple times to the ED, and she continued to have worsening right-sided facial swelling and facial droop of the same side, and she eventually developed intractable nausea and vomiting. At that juncture, she received a CT of the head demonstrating bilateral maxillary sinus disease, which was treated with clarithromycin, and she was discharged home. The patient again presented to the ED with the same unresolved symptoms along with a new onset of ataxia, worsening right facial weakness, and the new onset of right otalgia. She was admitted to the hospital for management.

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On admission, her physical examination was remarkable for a complete right facial palsy, vesicles on her upper lip (Figure 1), and a positive Romberg's sign. Her facial palsy was assessed as a grade II using the House-Brackmann Score. She was otherwise afebrile with stable vital signs. The patient reported a history of herpes simplex, and she had noted the appearance of the vesicles one to two days prior to admission.



Figure 1. Initial physical exam. Note the right facial paralysis and HSV vesicles on upper lip.

MRI of the brain demonstrated a nonspecific increased signal in the cerebellar cortical sulci and occipital lobes. In addition, there was an ill-defined leptomeningeal enhancement in the cerebellar vermis, an abnormal enhancement in the right auditory internal canal, and an enhancement of the right facial nerve (Figure 2).

A lumbar puncture was performed and cerebrospinal fluid (CSF) analysis showed 215 WBCs with 94% lymphocytes and elevated protein with normal glucose, but specific viral diagnostic testing was not performed on the CSF at that time. The patient was thought to have HSV meningitis or meningoencephalitis with Bell's palsy. A subsequent physical exam on hospital day two demonstrated the new onset of vesicles on the right pinna and a right-sided hyperacusis.

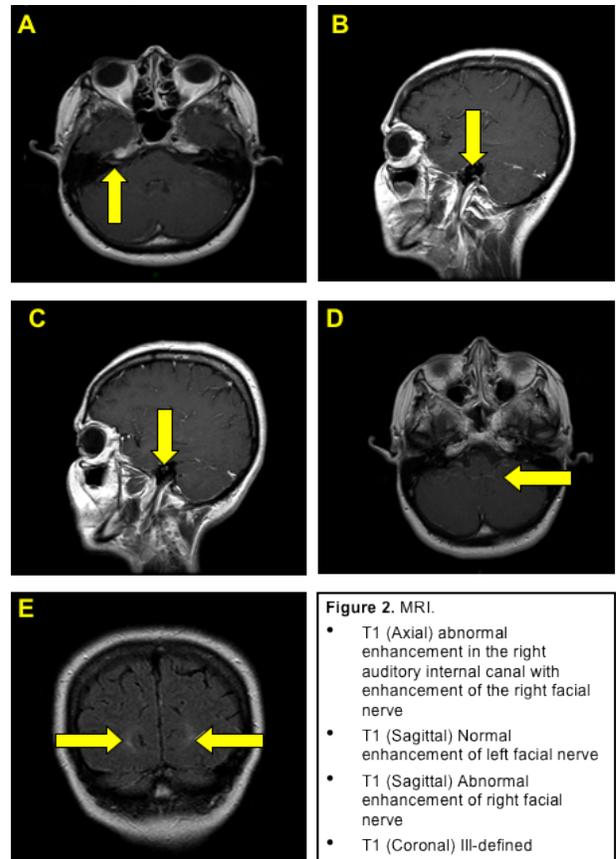


Figure 2: Radiographic findings

A varicella zoster virus (VZV) polymerase chain reaction (PCR) of the fluid from the vesicles on the right pinna returned positive, and the diagnosis of Ramsay Hunt Syndrome with zoster meningoencephalitis was made. The patient was started on a ten-day course of oral acyclovir and a five-day course of oral prednisone. She continued to receive hydrocodone, and promethazine for symptomatic relief in addition to her home medications of levothyroxine and alprazolam. The patient was discharged home on hospital day 12 with two weeks of valacyclovir and an oral prednisone taper with close follow up with internal medicine and neurology.

### Discussion:

Ramsay Hunt Syndrome (RHS) was first described by James Ramsay Hunt in 1907. Hunt described a syndrome which included otalgia, auricular vesicles, and peripheral facial paralysis, and he astutely predicted that the syndrome

was due to a viral infection of the geniculate ganglion. The incidence in the United States is about five cases in 100,000, and there is a significant increase in incidence in those over age 60, much like VZV<sup>1</sup>. However, a literature review reveals a wide array of manifestations, which is due to the varying reactivation patterns of the VZV. The presentations have included a simple otitis externa, a pseudo-Garcin-Guillain syndrome (a unilateral paralysis of nearly all cranial nerves), lesions in the spinal trigeminal nucleus and tract, brainstem lesions, cerebellitis, and syndrome of inappropriate secretion of antidiuretic hormone.<sup>1, 2, 3, 4, 5, 6, 7, 8, 9</sup>

As noted above, RHS is caused by the VZV reactivation in the geniculate ganglion. This leads to symptoms such as tinnitus, hearing loss, nausea, vomiting, vertigo, and nystagmus. In addition to the most common presenting symptoms, other important exam findings are hyperacusis and changes in taste perception. Interestingly, hyperacusis is due to paralysis of the stapedius, which is innervated by the geniculate ganglion. While the CN VII findings are easily explained, the other symptoms such as tinnitus, hearing loss, and vertigo with associated nausea and vomiting are due to the shared blood supplies of the geniculate ganglion and cranial nerves VIII, IX, X, XI, and XII. The ascending pharyngeal artery supplies the glossopharyngeal, vagal, accessory, and hypoglossal nerves. Additionally, the middle meningeal artery supplies the facial nerve along with the maxillary and mandibular branches of the trigeminal nerve. VZV has a known proclivity for axonal spread among the nerves.<sup>1</sup>

The diagnosis of RHS is generally made based on clinical findings, but in the early stages of the disease, it can be difficult to distinguish from the more common Bell's palsy; however, if the patient presents with the classic triad of otalgia, auricular vesicles, and CN VII nerve palsy, treatment for VZV should be considered as early intervention has been shown to improve long term outcomes of facial paralysis.<sup>10</sup> In a clear clinical case, an MRI scan with gadolinium and lumbar puncture should not be required to make the diagnosis of RHS and initiate therapy. However, in atypical presentations, these tests can offer insight into the disease process. A VZV PCR can be performed on vesicular fluid, and this can offer a definitive diagnosis.<sup>2, 11, 12</sup>

The standard of care for treatment is 800 mg acyclovir PO for 7 – 10 days and 60 mg prednisone orally for 3 – 5 days. Also, 500 mg of famciclovir PO can be used in place of acyclovir.<sup>13, 14,</sup>

<sup>15</sup> It should also be noted that there is no statistical difference between using oral and intravenous antiviral or steroid medications.<sup>16</sup> Interestingly, a Cochrane Review article did not find evidence for a beneficial effect of antiviral drugs with regard to the outcome of RHS. However, the article does note that this does not necessarily indicate that antivirals are ineffective.<sup>14</sup> Another Cochrane Review article examines the use of prednisone for RHS, and this study was unable to comment on the efficacy of prednisone in the treatment of RHS; the study goes on to call for further research into the matter.<sup>15</sup> Acyclovir only affects VZV replication, consequently once VZV stops replicating the drug has no effect. Following infection, VZV remains latent within the peripheral sensory ganglia, and relapse is possible. Best outcomes are obtained when treatment is started within 72 hours of initial symptom onset.<sup>16</sup>

The prognosis of RHS is closely correlated with the House-Brackmann scale (Figure 3). This scale is a measure of facial nerve dysfunction. It has been shown that recovery rates with House-Brackmann grade II or better were 84.6% in those with RHS.<sup>17, 18</sup> Prognosis has been shown to be adversely affected by increased age, diabetes mellitus, essential hypertension, and vertigo.<sup>19</sup> When compared to Bell's palsy, RHS has a less favorable prognosis.<sup>20</sup>

Grade	Description	Function %
I	Normal	100
II	Slight	76 – 99
III	Moderate	51 – 75
IV	Moderately Severe	26 – 50
V	Severe	1 – 25
VI	Total	0

Figure 3: House Brachmann Scale (16)

Somewhat unique to our case was the patient's meningoencephalitis due to VZV. A literature review reveals only a handful of cases that present in such a manner.<sup>21, 22, 23</sup> Given our patient's constellation of symptoms of nausea, vomiting, ataxia, and profound headache and the radiological findings,

she seemed to have a fairly rare presentation. Following discharge the patient did have a relapse of her condition.

One final variation of RHS, which should be mentioned, is *zoster sine herpette*. In this form of RHS, a patient will present with a peripheral facial paralysis without the characteristic ear or mouth lesions but will have a four-fold rise in antibody titer to VZV. This particular presentation may be confused with Bell's palsy. The diagnosis of this form of RHS requires PCR testing of saliva.<sup>16</sup>

## Conclusion

RHS is an important cause of unilateral facial paralysis, characterized by the clinical triad of herpetic lesions, otalgia, and facial paralysis. Depending on which group of cranial nerves is affected, a wide array of clinical presentations may occur. The current standard of care for treatment is acyclovir and prednisone; however, large studies to prove efficacy of this therapeutic approach have not been conducted. Given RHS's distinctive clinical presentation, it is important for a practicing clinician to be able to recognize this condition and initiate treatment within 72 hours of symptom onset. With treatment, most patients make a full recovery; however, this condition can lead to permanent facial paralysis.

## Authors

**Sources of Funding:** There was no funding for this project.

**Conflicts of Interest:** The authors have no conflicts of interest to report.

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