

CLINICAL VIGNETTE

Ramsey Hunt Syndrome (herpes zoster oticus) in a 38-Year-Old Woman

Rauz A. Eshraghi, M.D. and Shelese Newmark, M.D.

Case Report

A 38-year-old previously healthy mother of two presented to her primary care physician with complaints of a new rash. Her symptoms began three days prior with pain along her right mandible that subsequently extended over her right buccal mucosa, behind her right ear, and extended to her right scalp. She initially had attributed her symptoms to either dental caries or a temporomandibular joint disorder and had presented to her dentist for evaluation. Her dentist noted a vesicular rash immediately below her right ear, which the patient had not previously appreciated. She denied any fevers or chills. He treated her with amoxicillin and recommended evaluation by her primary care physician for herpes zoster.

Upon examination by her primary care physician, her vital signs were normal. She had several vesicles immediately below the angle of the mandible and a subcentimeter, palpable, mobile lymph node in the right anterior cervical chain. The right tympanic membrane and ear canal were free of any lesions. She noted tenderness to light touch on the right scalp. Otherwise, examination of the eyes and oral mucosa were unremarkable.

She was treated with valacyclovir 1 gram orally every 8 hours for 7 days. Gabapentin was added to treat concomitant neuropathic pain. Given that she had a 4-year-old at home who had previously been vaccinated for Varicella and that she was breastfeeding a 9-month-old, appropriate preventive precautions were reviewed with the patient.

Approximately 5 days later the patient noticed edema over the right face and slight ptosis of the right eyelid. She also noted increasing right otalgia and upon further questioning, hyperacusis. Given these new symptoms, she returned to her primary care physician concerned she may be having a stroke.

Repeat examination now showed decreased tone and flattening of the right nasolabial fold. Her smile was downslowing on the right and when asked to raise her eyebrows, there was minimal movement of the right forehead. Evaluation of the external auditory canal revealed a crusted lesion.

She was started on prednisone and reassured that her symptoms were consistent with complications of varicella zoster reactivation rather than an acute cerebrovascular

accident. Per instructions, she followed-up in 2 weeks at which time there was significant resolution of her symptoms. Movement of forehead had significantly improved. She also noted that her 4-year-old had developed chickenpox in the interim despite precautions and history of vaccination. Her infant remained unaffected.

Discussion

Herpes zoster, a reactivation of the varicella zoster virus (VZV), affects approximately one out of three persons living in the United States during their lifetime.¹ This roughly translates to 1 million cases of shingles annually in the US alone.² Though more commonly affecting adults over the age of 50, VZV reactivation can occur at any age.

A rare known complication of herpes zoster is **Ramsay Hunt syndrome** or herpes zoster oticus. Affected patients often present with facial paralysis, tinnitus, hearing loss, hyperacusis, vertigo, dysgeusia, or decreased lacrimation. Classically it is defined by ipsilateral facial paralysis, otalgia, and lesions in the auditory canal or auricle.³

While postherpetic neuralgia is the most frequent complication of VZV reactivation and can affect between 9-34% of patients, Ramsay Hunt syndrome typically affects less than 1% of patients.^{4,5}

Unlike Bell's Palsy, Ramsay Hunt syndrome is considered a polycranial neuropathy. Neuritis can affect cranial nerves V, VII, VIII, IX, and X. Autopsy studies have shown presence of VZV in the geniculate ganglion, which is responsible for sensory innervation of the anterior tongue, soft palate, skin of the external auditory canal and mastoid process.⁶

Patients who develop Ramsay Hunt syndrome often manifest more severe symptoms than those with Bell's Palsy and are less likely to fully recover.⁷ In addition, because it can also affect the vestibular and cochlear nerves, patients can develop vertigo and tinnitus. Long-term complications include incomplete hearing loss, balance difficulties, and vertigo.

Interestingly, patients with Ramsay Hunt syndrome do not always initially present with skin lesions. Eruptions can occur after the onset of facial paralysis. Therefore, early clinical evaluation for facial paralysis should include evaluation for

hearing loss. In turn, the diagnosis of Ramsey Hunt syndrome should be considered when severe facial or otologic pain accompanies paralysis.

Similar to Bell's Palsy, treatment should be initiated immediately with corticosteroids and often an antiviral medication. Though the use of antiviral therapy has been demonstrated to improve pain and possibly diminish symptoms of postherpetic neuralgia in herpes zoster, there are limited data on their benefit in Ramsay Hunt syndrome.⁸

Treatment with corticosteroids and antivirals is generally based on consensus recommendations as there are few randomized controlled studies to validate their use. However, one study using retrospective analysis of early treatment with acyclovir and prednisone (within 3 days of onset of symptoms) showed almost 75% attained complete recovery. In comparison, the same analysis demonstrated that patients who began treatment more than 7 days after symptom onset had only a 30% likelihood of complete symptom resolution.⁷ Therefore, treatment should ideally be initiated within 72 hours of symptom onset. Recommended pharmacotherapy includes a 7-day course of prednisone 60-80 mg daily or prednisolone 60 mg daily for 5 days with a taper to complete a 10-day course.

In addition, ocular care including application of lubricants to prevent corneal abrasion or drying are advised, particularly if eyelid closure is compromised. Patients should be instructed to use artificial tears hourly and ocular lubricants at night.

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Submitted on December 20, 2013