Ramsay Hunt Syndrome Revisited

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Practice Points

- Systemic treatment with a short course of corticosteroids and an antiviral agent may be effective for Ramsay Hunt syndrome; however, it is reasonable to maintain antiviral therapy until maximum neurologic function is regained.
- Consider neurologic consultation for patients with Ramsay Hunt syndrome to provide additional evaluation, management recommendations, and treatment follow-up, especially if partial or complete facial palsy persists.

Varicella-zoster infections commonly are seen in clinical practice; however, Ramsay Hunt syndrome caused by reactivation of latent varicella-zoster virus in the geniculate ganglion is rare. We report the case of a 30-year-old woman who developed Ramsay Hunt syndrome and also review the characteristic features of this disease.

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Herpes zoster is a reactivation of a latent varicella-zoster infection that usually presents as a cluster of erythematous vesicles in a unilateral dermatomal distribution.[1-3] Ramsay Hunt syndrome, which is caused by reactivation of latent varicella-zoster virus in the geniculate ganglion, is associated with ear pain, erythematous vesicles on the ear, facial nerve paralysis, and vestibulocochlear dysfunction.[4-8] We report the case of a 30-year-old woman who developed Ramsay Hunt syndrome and also review the characteristic features of this disease.

Case Report

A 30-year-old woman who was otherwise healthy presented with an eruption of nonpruritic lesions

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The authors report no conflict of interest.

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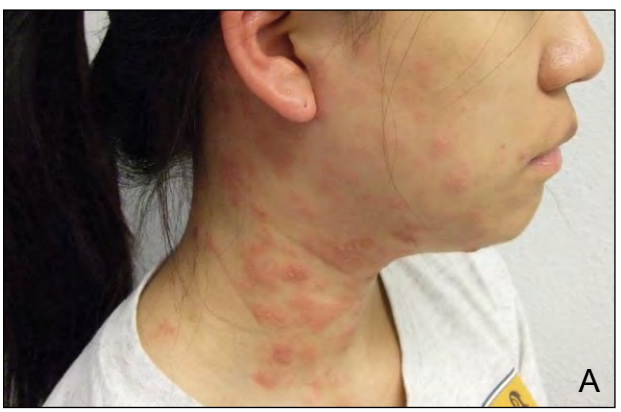


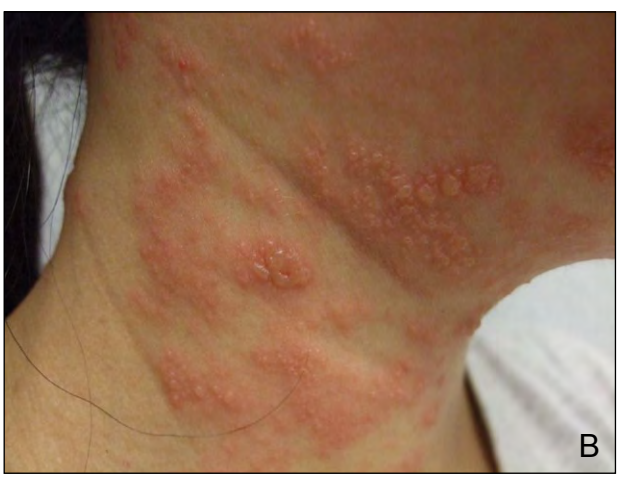
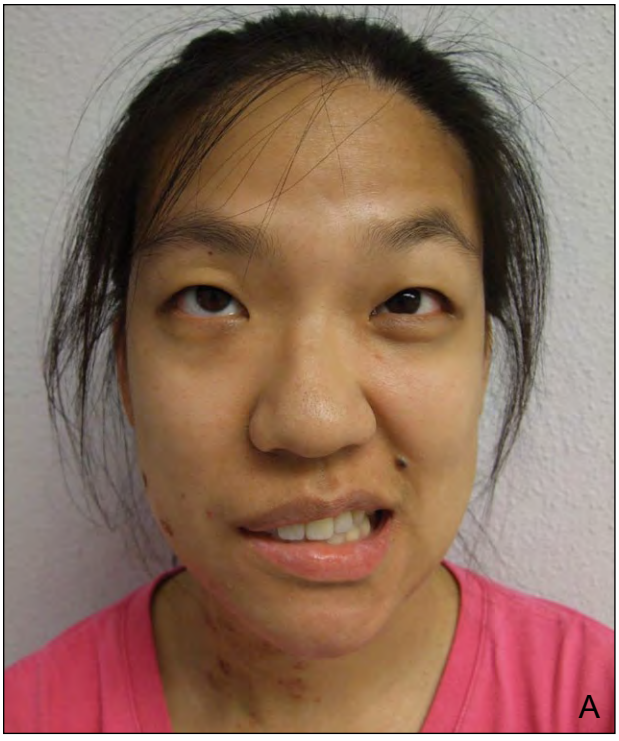


Figure 1. A 30-year-old woman presented with an eruption of nonpruritic lesions on the right side of her face, neck, scalp, and upper chest (A). She later was diagnosed with Ramsay Hunt syndrome. The erythematous vesicles on the neck occurred in multiple dermatomes, indicating varicella-zoster infection (B).



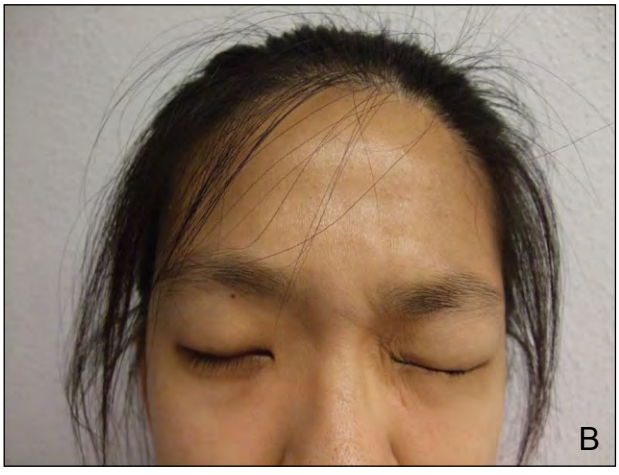
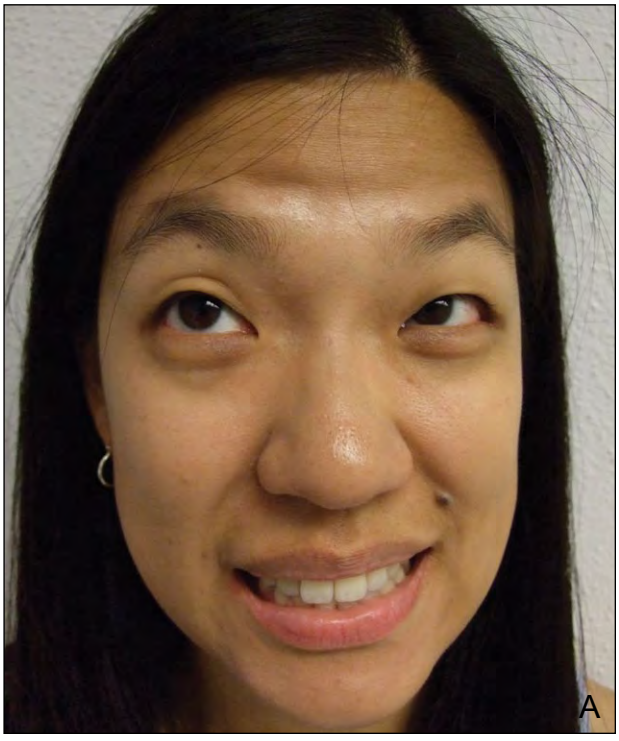


Figure 2. Peripheral seventh cranial nerve paralysis occurred following reactivation of latent varicella-zoster infection affecting the second and third right cervical nerves, the mandibular division of the fifth cranial nerve, and the seventh and eighth cranial nerves. The patient could not raise her right eyebrow or complete a smile on the ipsilateral side of her face (A); she also was not able to tightly close her right eye (B).



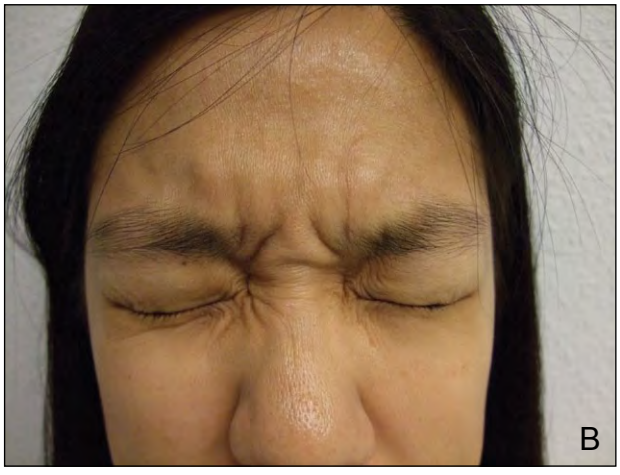


Figure 3. Following treatment, the patient demonstrated complete resolution of facial paralysis associated with Ramsay Hunt syndrome. The patient could raise both eyebrows equally and complete a smile (A); she also was able to tightly close both eyes (B).

on the right side of her face, neck, scalp, and upper chest of 2 days' duration. The patient also reported soreness of the face and a tingling sensation on the right side of her tongue. Her medical history was remarkable for varicella at 5 years of age.

On cutaneous examination, erythematous vesicles involving the right cranial nerve V3, right cervical nerves C2 and C3, anterior and posterior external right ear, and right auditory canal were observed (Figure 1). A regimen of oral valacyclovir hydrochloride (1 g 3 times daily) for treatment of herpes zoster was initiated, and acetaminophen (300 mg) combined with codeine phosphate (30 mg) was prescribed for pain (every 4 to 6 hours as needed).

Topical acetic acid soaks using a solution of white vinegar (1 cup) and water (4 cups) were performed 3 times daily for local care.

The patient returned for follow-up evaluation 1 week later. Near-complete resolution of all skin lesions was observed; however, the patient noticed a sudden onset of right-sided facial paralysis 2 days earlier. The neurologic deficit corresponded to the function of the peripheral seventh cranial nerve. The patient was unable to complete a smile on the right side of the face, elevate the right eyebrow, or tightly close the right eye (Figure 2). The patient's clinical history and examination indicated a diagnosis of Ramsay Hunt syndrome.

Treatment was modified with the initiation of prednisone (50 mg for 5 days with subsequent taper by 10 mg every other day) and replacement of the patient's oral antiviral medication with oral acyclovir (800 mg 5 times daily), a less expensive alternative. The patient also was evaluated by a neurologist who agreed with the diagnosis of Ramsay Hunt syndrome and subsequent therapeutic interventions.

The patient returned for follow-up examination 2 weeks later after completing the 13-day prednisone regimen. Her facial palsy showed substantial improvement, and postinflammatory hyperpigmentation was observed where the lesions had been on the right cheek, neck, scalp, and upper chest.

The patient continued treatment with oral acyclovir for an additional 3 weeks. Follow-up assessment revealed complete resolution of her facial palsy. She regained full neurologic function and was able to complete a smile, raise the right eyebrow, and tightly close both eyes (Figure 3).

Comment

The varicella-zoster virus is the herpesvirus that is responsible for causing both varicella (chickenpox) and herpes zoster (shingles). Varicella is a primary transient infection that establishes latency in the dorsal root ganglia. Herpes zoster is the reactivation of latent varicella-zoster virus that occurs idiopathically or due to immunosuppression. Herpes zoster usually is confined to a single dermatome or contiguous unilateral dermatomes as erythematous clustered vesicles and often is preceded by pain. Systemic antiviral therapy (ie, acyclovir, famciclovir, valacyclovir) is the standard treatment.[1-3]

Bell palsy is the abrupt onset of isolated peripheral facial nerve weakness or paralysis of unknown etiology.[9-12] Clinical presentation includes limited retraction of the angle of the mouth and the inability to raise the involved eyebrow or tightly close the involved eye.[11] In the majority of patients (70%–80%), symptoms completely resolve within 6 weeks.[9,12] The etiologies of Bell palsy typically fall into 3 broad categories: infectious, autoimmune, and oncologic.[10] The leading etiologic factor with the most supporting evidence is herpes simplex virus type 1.[11,12] Treatment of Bell palsy often includes a short course (2–14 days) of systemic corticosteroids as well as an oral antiviral agent, which may or may not give added benefit.[9,11]

Ramsay Hunt syndrome, also known as herpes zoster oticus and facial paralysis, is a reactivation of latent varicella-zoster virus in the geniculate ganglion.[4-8] The disease is named after Dr. James Ramsay Hunt (1874-1937), a preeminent American neurologist whose research on herpetic geniculate ganglion syndrome in 1907[13] established Ramsay Hunt syndrome as a clinicopathologic entity and also revealed that the facial nerve has both sensory and motor components.[14]

Ramsay Hunt syndrome is characterized by facial nerve paralysis, ear pain, and auricular vesicles. Other symptoms may include dry mouth and eyes, hearing loss, hyperacusis, nausea, nystagmus, taste impairment, tinnitus, vertigo, and vomiting.[4-8] The proposed mechanism for involvement of the eighth cranial nerve is secondary to the close proximity of the geniculate ganglion to the vestibulocochlear nerve within the facial canal; symptoms result from both the inflammatory edema, which subsequently applies pressure to the nerve, and direct sequelae of the varicella-zoster virus on the nerve.[4,7] In addition to cranial nerve VIII, cranial nerves V, VII, IX, and X also may be involved in patients with Ramsay Hunt syndrome.[8]

The annual incidence of Ramsay Hunt syndrome in the United States is approximately 5 cases per every 100,000 patients, with the majority occurring in elderly individuals.[4] Ramsay Hunt syndrome is second only to Bell palsy as the most common cause of acute-onset peripheral facial paralysis[6,8]; however, Ramsay Hunt syndrome results in greater neurologic dysfunction, including a more complete facial nerve palsy, and a poorer prognosis than Bell palsy.[4,6,8]

Diagnosis is chiefly based on clinical signs, such as the combination of severe ear pain, vesicles on the pinna, and facial palsy. Vesicles usually appear prior to or concurrent with facial paralysis but may present after in some cases.[4] Use of polymerase chain reaction to detect varicella-zoster virus from the cerebrospinal fluid, ear mucosa, facial nerve sheath, and vesicles can confirm the clinical diagnosis.[4,5,8] Prominent enhancement of the facial and vestibular nerves, as demonstrated using gadolinium-enhanced magnetic resonance imaging, also is a typical finding in Ramsay Hunt syndrome.[5]

Treatment of Ramsay Hunt syndrome, preferably within 3 days of onset, often yields notable

improvement. Similar to Bell palsy management, the first-choice treatment option for Ramsay Hunt syndrome is a short course of systemic corticosteroids and an antiviral agent. However, one study questioned the efficacy of an antiviral agent for treatment of this disease, as the addition of an antiviral drug did not show benefit over corticosteroid use alone.[15] Nonetheless, a randomized clinical trial evaluating both therapies is necessary and adverse effects of antiviral medications must be considered before they are administered.[15]

Our patient's characteristic presentation of Ramsay Hunt syndrome was successfully treated with systemic corticosteroids and antiviral therapy. Ramsay Hunt syndrome must be considered when lesions from a herpes zoster virus infection affect the ear and ipsilateral facial paralysis is observed. Systemic treatment with a short course of corticosteroids and an antiviral agent may be effective. As observed in our case, it may not be unreasonable to maintain antiviral therapy until maximum neurologic function is regained, while ensuring that no adverse events occur.

Conclusion

Ramsay Hunt syndrome is a peripheral seventh cranial nerve palsy presenting with ipsilateral otalgia, erythematous vesicles on the ear, and facial paralysis. The reactivation of latent varicella-zoster virus in the geniculate ganglion as well as the close proximity of the facial and vestibulocochlear nerves leads to a constellation of symptoms that have been associated with this disease. Even with appropriate treatment, not all patients regain complete neurologic function. Therefore, it is important to consider neurologic consultation for additional evaluation, management recommendations, and treatment follow-up, especially if partial or complete facial palsy persists.

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