

A 55-Year-Old Woman with Unilateral Facial Paralysis and Pain

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Figure 1. Clinical photograph of the case patient showing left-sided facial palsy.



Figure 2. Photograph of the case patient's left ear.

CASE PRESENTATION

A 55-year-old woman presented with a complaint of pain and numbness on the left side of her face. Her past medical history included a left hemicolectomy 3 years earlier for ischemic bowel, hypertension, hiatal hernia, and osteoarthritis. Her regular medications included lisinopril 10 mg daily, aspirin 81 mg daily, calcium and vitamin supplements, and ibuprofen as needed. She had 2 teeth extracted 4 days prior to presentation and was placed on clindamycin by her dentist. Over the next 2 days, she developed pain, numbness, and swelling of her left ear and left side of her face. The pain was described as sharp and stabbing in nature. She denied fever, chills, or rash. She admitted to tinnitus of the left ear but denied hearing loss.

On physical examination her vital signs were: temperature, 97.1°F; pulse, 80 bpm; and blood pressure, 120/70 mm Hg. Her mouth was not swollen, and there was no dental abscess. She had a left facial palsy that involved the forehead as well as the lower face; however, she was not flaccid. Her left ear was swollen, and a rash was present in the external auditory canal. Seventh cranial nerve and external ear examinations were performed (**Figures 1 and 2**).

Radiographic evaluation consisting of computed tomography (CT) scanning of the brain and face was notable for bilateral maxillary sinus mucous retention and mild bilateral ethmoid sinus mucosal thickening. Her laboratory examination was notable for a leukocyte count of $7.5 \times 10^3/\text{mm}^3$ with 70% neutrophils. Her electrolyte levels were normal; her blood glucose was elevated at 248 mg/dL.

WHAT IS YOUR DIAGNOSIS?

- (A) Idiopathic Bell's palsy, most likely from herpes simplex virus reactivation
- (B) Post-traumatic seventh cranial nerve injury related to tooth extraction
- (C) Left facial abscess with resultant left-sided facial paralysis
- (D) Ramsay Hunt syndrome
- (E) Uncontrolled diabetes is the most likely cause of the patient's facial nerve paralysis
- (F) Acute bacterial sinusitis with associated facial nerve palsy

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ANSWER

The correct answer is (D) Ramsay Hunt syndrome.

DISCUSSION

Although idiopathic Bell's palsy is the most common cause of facial nerve (cranial nerve VII) palsy, it is not associated with vesicles of the auditory canal. Trauma—usually from a motor vehicle accident or iatrogenic nerve injury during ear surgery—is the second leading cause of facial nerve paralysis; however, trauma to the facial nerve is typically not associated with tooth extraction. A facial abscess could potentially lead to compression of the facial nerve, but this is unlikely and ruled out by a normal CT scan of the face. Hyperglycemia is the most common metabolic cause of idiopathic Bell's palsy; however, it is not associated with the specific findings in this case, that is, auditory canal vesicles, tinnitus, and lancinating facial pain. Although the patient's CT scan demonstrated sinus mucosal thickening, this is not specific for bacterial sinusitis, and without extension of the infection to areas surrounding the facial nerve, it is not likely to cause paralysis.¹

RAMSAY HUNT SYNDROME**Etiology**

The classic definition of Ramsay Hunt syndrome is facial nerve weakness or paralysis together with *zoster oticus*—vesicles of the pinna or outer auditory canal of the ipsilateral ear.² The vesicles can also appear on the hard palate and tongue. This disorder was first described by the American neurologist James Ramsay Hunt in early part of the twentieth century.³ The etiology of this condition is the reactivation of the varicella zoster virus. Varicella zoster virus is the human herpesvirus that leads to chicken pox, typically in childhood. The virus may lie dormant in the cranial nerves and dorsal root ganglia. When the virus reactivates, possibly as a result of advanced age or immunosuppression, it leads to shingles (*zoster*) and sometimes post-herpetic neuralgia (a chronic neuropathic pain syndrome). The exact cause for the reactivation of the virus is unknown.⁴

Clinical Presentation

The first sign that the varicella virus is reactivating may be the characteristic syndrome of severe sharp, lancinating pain. The two most common places for the virus to reactivate are the thorax and the face.⁴ The typical rash associated with *zoster* is a dermatomal vesicular rash. Similar to the original infection, these vesicles will be at many different stages of development. The

form of *zoster* called Ramsay Hunt syndrome involves the re-emergence of the activated virus within the seventh cranial nerve or geniculate ganglion, which lies in close proximity to the vestibulocochlear nerve (eighth cranial nerve).^{5,6}

The proximity of the vestibulocochlear nerve to the geniculate ganglion explains the other features of Ramsay Hunt syndrome, including tinnitus, hearing loss, and vertigo.^{7,8} These symptoms occur only on the affected side.⁸ In comparison to idiopathic Bell's palsy, patients with Ramsay Hunt syndrome typically have more extensive paralysis of the facial nerve and are less likely to fully recover.⁹

The cause of idiopathic Bell's palsy has not been fully elucidated; however, evidence has suggested a role of the herpes simplex virus.¹⁰ Many patients with Ramsay Hunt syndrome present with facial nerve paralysis without vesicles, and because the vesicles are often overlooked, the diagnosis may be missed in a considerable number of patients.¹¹ It has been estimated that 4.5% to 9% of patients diagnosed with idiopathic Bell's palsy have facial paralysis associated with Ramsay Hunt syndrome.¹² There is no simple diagnostic test to determine whether Ramsay Hunt syndrome is the cause of facial weakness in the absence of vesicles. Recently, techniques utilizing the polymerase chain reaction have been used to detect the varicella zoster virus in the geniculate zone of the ear as well as in saliva, but this remains a research tool at the present time.¹³

Treatment

The treatment of Ramsay Hunt syndrome is not entirely agreed upon. In a large retrospective review of 80 patients with Ramsay Hunt syndrome, Murakami et al¹⁴ discovered that patients who were treated with prednisone and acyclovir within 3 days of symptom onset had a statistically significant improvement in facial nerve symptoms (75% recovered) and hearing compared with patients whose treatment started 7 days after symptom onset (30% recovery for facial paralysis). All patients included in the analysis were treated with prednisone (1 mg/kg body weight daily for 5 days followed by a 10 day taper) and with either intravenous acyclovir (250 mg 3 times daily) or oral acyclovir (800 mg 5 times daily). Of note, there was no difference between oral or intravenous acyclovir in recovery from symptoms.

A small trial performed by Uri et al¹⁵ reported that treatment with intravenous acyclovir for 7 days resulted in a recovery rate of 82.6% in 31 patients with Ramsay Hunt syndrome. In a slightly larger trial in which

patients were randomized to either intravenous acyclovir in combination with steroids or steroids alone, the recovery rate for facial nerve paralysis for patients given the combination treatment was 90% compared with a 64% recovery rate in the steroid-alone group.¹⁶

Despite the lack of a large randomized controlled trial, many authorities recommend that Ramsay Hunt syndrome be treated with either oral acyclovir (800 mg 5 times daily) or oral famciclovir (500 mg 3 times daily) for 7 to 10 days, together with oral prednisone at 60 mg daily for 3 to 5 days, usually followed by a 10-day taper.^{4,11} Consideration should be given to the initial use of intravenous acyclovir and an otolaryngology consultation, especially if hearing loss or tinnitus is present.

Long-term complications of Ramsay Hunt include incomplete recovery of facial nerve paralysis, hearing loss, post-herpetic neuralgia, and tinnitus.¹¹ The patient presented was treated aggressively to help prevent the complications of Ramsay Hunt syndrome, initially with intravenous acyclovir, followed by oral famciclovir and prednisone. She was also started on an oral sulfonylurea for the treatment of her elevated blood sugars and educated on eye care to avoid corneal trauma.

CLINICAL COURSE OF CASE PATIENT

Less than 2 months after her initial presentation, the patient had almost full recovery of her facial weakness. Unfortunately, she continued to have left-sided neuropathic pain. She was started on gabapentin and titrated to a dose of 300 mg 3 times daily, but this was later discontinued secondary to intolerable side effects. She has no notable hearing loss; a formal audiology examination was deferred by the patient, but she continues to have left-sided tinnitus.

CONCLUSION

Ramsay Hunt syndrome is an important cause of facial weakness with pain. The natural course has a worse prognosis than Bell's palsy, and physicians should be diligent not to miss the presence of zoster oticus. If treatment is begun early in the course of the disease, the long-term complications can hopefully be avoided. Areas of uncertainty include the ideal treatment for adult patients and diagnostic strategies to determine whether the patient with idiopathic Bell's palsy does, in fact, have Ramsay Hunt syndrome. In the future, large randomized trials may help to guide our diagnostic and treatment decisions. **HP**

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