Ramsay Hunt syndrome presenting as simple otitis externa

Daniel Kim, MD, BSc;* Munsif Bhimani, MD†

ABSTRACT

Ramsay Hunt syndrome is a rare complication of herpes zoster in which reactivation of latent varicella zoster virus infection occurs in the geniculate ganglion, causing otalgia, auricular vesicles and peripheral facial paralysis. Because these symptoms do not always present at the onset, this syndrome can be misdiagnosed. We report the case of a patient who was diagnosed with simple otitis externa after presenting to the emergency department (ED) with a 3-day history of right-sided otalgia. Her condition subsequently evolved to include right-sided auricular vesicles and right-sided facial weakness. She presented to the ED again after 2 days and was correctly diagnosed with Ramsay Hunt syndrome. We describe the clinical presentation, diagnostic findings and management of this uncommon but important entity.

Keywords: Ramsay Hunt syndrome, varicella zoster virus, herpes zoster

RÉSUMÉ

Le syndrome de Ramsay Hunt est une complication rare du zona (herpès zoster). La réactivation de l'infection latente du virus zona-varicelle survient dans le ganglion géniculé, causant une otalgie, des éruptions vésiculeuses sur l'oreille ainsi qu'une paralysie faciale périphérique. Comme ces symptômes sont parfois absents au début de cette affection, un mauvais diagnostic est occasionnellement posé. Nous présentons le cas d'une patiente ayant reçu un simple diagnostic d'otite externe après s'être présentée à l'urgence avec des antécédents d'otalgie du côté droit au cours des trois jours précédents. Son état a évolué, donnant lieu à des éruptions vésiculeuses sur l'oreille du côté droit et à une faiblesse faciale du côté droit. Deux jours plus tard, lors d'une autre visite à l'urgence, on a posé le bon diagnostic de syndrome de Ramsay Hunt. Nous décrivons la présentation clinique, le diagnostic et la prise en charge de cette complication rare, mais grave.

Introduction

In 1907, James Ramsay Hunt described a syndrome of otalgia, auricular vesicles and peripheral facial paralysis.[1] At that time, it was well accepted that infection of ganglia and skin by a herpes virus produced a characteristic dermatomal distribution of pain and vesicular rash.[2] Hunt hypothesized that this syndrome was a result of herpetic infection of the geniculate (seventh nerve) ganglion. He also described other accompanying signs and symptoms

From the *Division of Emergency Medicine, Department of Medicine, University of Toronto, Toronto, Ont.; at the time of writing: Schulich School of Medicine and Dentistry, University of Western Ontario, London, Ont., and the †Department of Emergency Medicine, London Health Sciences Centre and Division of Emergency Medicine, Schulich School of Medicine and Dentistry, University of Western Ontario, London, Ont.

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including tinnitus, hearing loss, nausea, vomiting, vertigo and nystagmus.[1] It is now known that reactivation of varicella zoster virus (VZV) after primary varicella infection precipitates Ramsay Hunt syndrome (Box 1).[3] We report a case of Ramsay Hunt syndrome that presented initially as simple otitis externa.

Case report

A 51-year-old woman presented to the emergency department (ED) with a 3-day history of gradual increasing right-sided head pain overlying her right ear, exacerbated by traction on the pinna. She had no fever, neck stiffness, vomiting or neurologic symptoms. The head pain was different from her past migraines, especially in that it radiated to the right mastoid. Her past medical history was significant for hypertension and a childhood history of varicella infection.

On examination, her temperature was 37.0°C, heart rate was 77 beats/minute, blood pressure was 144/88 mm Hg, respiratory rate was 18 breaths/minute and oxygen saturation was 99% on room air. Cranial nerves II–XII were intact, pupils were equal and reactive to light, strength and sensation were normal, cerebellar testing was normal and gait was normal. Examination of the head and neck revealed a reddened, swollen right external auditory canal with a normal tympanic membrane. The rest of the physical examination was unremarkable.

Complete blood count and serum biochemistry, including creatinine, urea, glucose and electrolytes, were normal. A noncontrast CT scan of the head, ordered to rule out malignant otitis externa and mastoiditis, was interpreted as normal, so the patient was diagnosed as having simple otitis externa and discharged from the ED with ciprofloxacin ototopical drops.

Box 1. Clinical features of Ramsay Hunt syndrome

Key clinical features

- Acute peripheral facial paralysis
- Vesicles occurring anywhere along the sensory distribution of the facial nerve, including the anterior two-thirds of the tongue, the pinna or the external auditory canal
- Otalgia

Additional clinical features

- Tinnitus
- Hearing loss
- Vertigo
- Nausea
- Vomiting
- Nystagmus
- Change in taste perception

Two days later, she returned to the ED with worsening head pain and several new symptoms, including right-sided pulsatile tinnitus, right-sided aural fullness, vertigo, reduced ability to close the right eye and decreased taste sensation. She had no hearing loss. On examination, her temperature was 37.1°C, heart rate was 74 beats/minute, blood pressure was 138/72 mm Hg, respiratory rate was 20 breaths/minute and oxygen saturation was 98% on room air. Facial sensation was intact, but there was mild right-sided facial weakness with sluggish right eye closure. In addition, there was right auricular swelling and redness with 3 small vesicles on the concha. The right tympanic membrane was not visualized owing to meatal swelling, but the left tympanic membrane was normal. The oral cavity and oropharynx were normal, with no vesicles. Ocular examination demonstrated a spontaneous, lateral, left-beating gaze nystagmus but normal conjunctiva and sclera.

The patient was seen by an ear, nose and throat (ENT) consultant while in the ED, and the diagnosis of Ramsay Hunt syndrome was confirmed. The ENT consultant administered an initial dose of intravenous acyclovir and steroids, and discharged the patient with a 2-week course of oral acyclovir and steroids. She was also referred to an ophthalmology clinic for corneal assessment as well as education to prevent corneal irritation and injury. At 3-week follow-up, the patient's symptoms, including pain, vesicles and facial weakness, had all resolved.

Discussion

Background

After primary infection with VZV, latent infection is established in the sensory dorsal root ganglia. Reactivation of endogenous VZV infection within these sensory ganglia causes herpes zoster, a syndrome characterized by a painful, unilateral vesicular eruption in a restricted dermatomal distribution.[4] Dermatomal pain may precede lesions by 48–72 hours and total disease duration is 7–10 days. Immunocompromised and elderly patients may have a more prolonged and severe course.[5] Ramsay Hunt syndrome can be precipitated by reactivation of VZV in the geniculate ganglion, resulting in peripheral facial paralysis, otalgia and auricular vesicles.[3]

Epidemiology

Herpes zoster has been described in all age groups, and lifetime risk is estimated to be 10%–20%. The incidence of herpes zoster is about 150–300 cases per 100 000, with the

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incidence dramatically increased in patients older than 60 years.[6] Ramsay Hunt syndrome is much less common, approximately 5 cases per 100 000 population; nevertheless, it is the second most common cause of atraumatic facial paralysis.[7] The incidence of herpes zoster in patients with peripheral facial palsy is 4.5%–8.9%. Compared with Bell palsy, Ramsay Hunt syndrome generally has more severe paralysis at onset, and patients are less likely to recover completely.[8]

Diagnosis

The strict definition of Ramsay Hunt syndrome is peripheral facial nerve palsy accompanied by erythematous vesicular rash on the ear or in the mouth. The diagnosis is based on history and physical examination.[9] Gadolinium-enhanced magnetic resonance imaging and cerebrospinal fluid examination have no diagnostic or prognostic value.[10] Polymerase chain reaction (PCR) can detect VZV in saliva, tears, middle ear fluid and blood mononuclear cells, but it is not necessary to establish the diagnosis.[11] Ramsay Hunt syndrome can be misdiagnosed at initial presentation, particularly in the absence of skin lesions. In approximately 10% of cases, there is no vesicular rash with the facial paralysis, but there is either a 4-fold rise in antibody to VZV or the detection of VZV DNA in skin, blood mononuclear cells or middle ear fluid. This condition is known as Ramsay Hunt syndrome sine herpete.[9]

James Ramsay Hunt postulated that the combination of herpes zoster oticus and facial paralysis results from the reactivation of latent VZV in the geniculate ganglion. Unfortunately, he never examined the geniculate ganglion in autopsies of patients with past history of Ramsay Hunt syndrome.[1] Since then, PCR has demonstrated the presence of VZV DNA in the geniculate ganglion[3] and in the facial sheath[12] of patients with Ramsay Hunt syndrome, strengthening Hunt's original hypothesis. Hunt also described other signs and symptoms, including tinnitus, hearing loss, nausea, vomiting, vertigo and nystagmus. He explained these vestibulocochlear features by the close proximity of the geniculate ganglion to cranial nerve VIII within the bony facial canal.[1] In addition to cranial nerve VIII, nerves V, IX and X can also be affected.[7]

Differential diagnosis

The differential diagnosis for Ramsay Hunt syndrome includes Bell palsy, otitis externa and trigeminal neuralgia. Bell palsy is defined as an acute peripheral facial nerve palsy of unknown cause, although recent research links it to various viral infections, especially herpes simplex virus type 1. Diagnosis is based on the sudden onset of unilateral peripheral facial paralysis, usually over hours, but sometimes more gradually. Additional symptoms such as decreased tearing, hyperacusis, loss of taste sensation over the anterior two-thirds of the tongue and ear pain are variable. Bell palsy does not involve the presence of vesicles in the external meatus.[13]

Otitis externa, inflammation of the external auditory canal or auricle, commonly presents with otalgia, pruritus, discharge and hearing loss. Examination usually reveals pain with tragal pressure and a red edematous ear canal. Facial palsy is not associated with otitis externa.[14] Trigeminal neuralgia is manifest by sudden, usually unilateral, severe, brief, stabbing, recurrent episodes of pain in the distribution of one or more branches of the trigeminal nerve. Most importantly, trigeminal neuralgia does not cause neurologic deficit, and the pain cannot be attributed to another disorder.[15]

Natural history

The natural history of Ramsay Hunt syndrome was detailed in a retrospective study of 102 untreated patients.[8] Maximal loss of facial nerve function occurred within 1 week, measured with a subjective muscle strength scale and response to facial nerve stimulation. Complete paralysis was twice as frequent as incomplete paralysis and occurred more often in patients older than 50 years. Of the patients with partial facial nerve function at disease onset, 66% recovered completely, whereas only 10% of those who presented with complete loss of function recovered completely.[8] In another retrospective study of 26 patients treated with acyclovir and steroids, age greater than 60 years, diabetes mellitus, essential hypertension and associated vertigo were identified as prognostic factors for worse outcome and decreased chance of recovery.[16]

Management

The largest retrospective treatment study showed a statistically significant improvement in patients treated with acyclovir and prednisone within 3 days of onset. Complete recovery occurred in 75% of patients treated within the first 3 days, but in only 30% of those treated after 7 days. This suggests that prompt diagnosis and management improves outcome in Ramsay Hunt syndrome. Importantly, no statistically significant outcome differences were noted between patients treated with intravenous or oral acyclovir.[17] A large prospective study demonstrated that combination therapy with acyclovir and steroids led to better recovery of facial nerve function than steroids alone.[18] These findings were confirmed by responses to nerve excitability testing. Although there are no evidence-based dosing recommendations,

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published trials typically administered acyclovir at 800 mg by mouth 5 times/day for 7–10 days and prednisone at 1 mg/kg/day by mouth for 5 days followed by a taper.[17] One study evaluated famciclovir 500 mg by mouth 3 times daily for 7 days after an initial course of acyclovir,[16] but there are no existing data describing outcomes with other antiviral agents. The collective published literature suggests that the combination of acyclovir and corticosteroids improve overall prognosis, especially when given early.

Ramsay Hunt patients should have ophthalmology assessments to evaluate them for ocular complications of facial nerve palsy, since incomplete eye closure exposes the cornea to drying and foreign body irritation. Mild facial paresis usually does not require therapy, but moderate to severe deficits require a corneal moistening regimen consisting of artificial tears during daytime and lubricating ointment at night. Taping of the lower eyelid to the lateral canthus can also be used to improve lid closure.[19]

Conclusion

Ramsay Hunt syndrome is a rare complication of herpes zoster infection affecting the geniculate ganglion and facial nerve. Physicians need to be aware that the peripheral facial paralysis, vesicular rashes and otalgia do not always present at the same time and may take time to evolve before declaring themselves as Ramsay Hunt syndrome. Early diagnosis is important so that acyclovir and steroid therapy can be initiated as soon as possible to maximize the recovery rate of facial nerve function.

Competing interests: None declared.

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Correspondence to: Dr. Daniel Kim, 1912-633 Bay St., Toronto ON M5G 2G4; dkim000@gmail.com