

Ramsay Hunt syndrome

Steven J. Montague MD, A. Ross Morton MD

■ Cite as: *CMAJ* 2017 February 27;189:E320. doi: 10.1503/cmaj.160483

An 83-year-old woman with dementia was admitted to hospital after her caregivers noted a one-week history of increasing confusion, episodic vertigo and difficulty speaking. The day before the patient's admission, she had complained of pain in her left ear, and caretakers noted drooping on the left side of her face.

Because of the patient's dementia, taking a reliable history of tinnitus and hearing loss was not possible, and undertaking magnetic resonance imaging was difficult. Left facial droop and mild dysphasia were observed on physical examination, consistent with palsy of the left cranial nerve VII (Figure 1A; Appendix 1, video available at www.cmaj.ca/lookup/suppl/doi:10.1503/cmaj.160483/-/DC1). Laboratory investigations and computed tomography imaging were unremarkable. Four days after the patient's admission to hospital, the patient's left ear — unremarkable on admission — had clustered vesicles in the external auditory meatus, coinciding with the distribution of the somatosensory chorda tympani branch of cranial nerve VII (Figure 1B). We diagnosed herpes zoster oticus (Ramsay Hunt syndrome, type 2).

Herpes zoster has an annual incidence of 3.2–4.2 cases per 1000.¹ Incidence rises in the fifth decade of life and peaks in the eighth decade.² Lifetime incidence is estimated at 32%.¹ Facial nerve involvement occurs in only 1% of patients, and predominantly affects women in their fifth and sixth decades.²

Treatment consists of analgesics, antivirals and corticosteroids. Suggested antivirals include acyclovir (800 mg, 5 times daily), valacyclovir (1000 mg, 3 times daily) or famcyclovir (500 mg, 3 times daily) for 7–10 days.^{2,3} Acyclovir administered intravenously (10 mg/kg every 8 hours for 7 days) is recommended for immunocompromised patients or patients with visceral, disseminated or ocular involvement.⁴ Suggested corticosteroid dosing is prednisone, 60 mg daily for 14 days, then tapered over 7 days.³ Efficacy of treatment is time sensitive. If treatment with antivirals and corticosteroids is begun within 3 days, 4–7 days, or more than 8 days, paresis resolves 75%, 48% and 30%, respectively.²

To prevent herpes zoster, the herpes zoster vaccine is recommended for patients 60 years of age and older. Within the first five years of administration, the vaccine is 51% effective, dropping to 40% effectiveness four to seven years after administration. Currently, data regarding efficacy beyond seven years is lacking.⁵

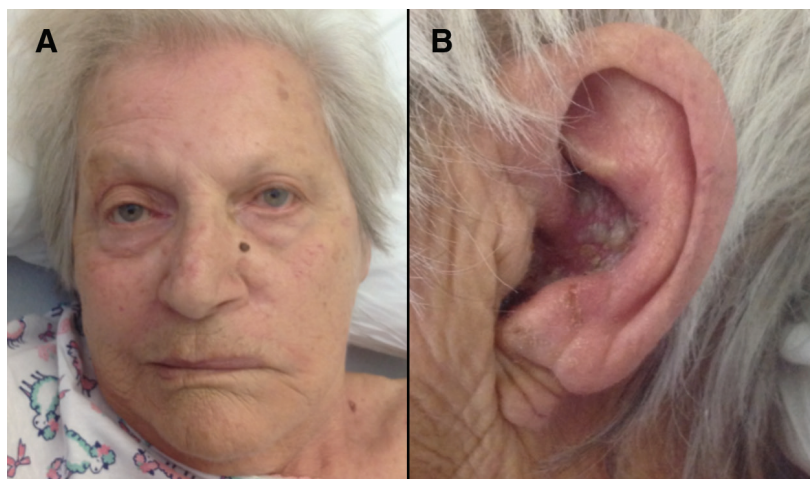


Figure 1: An 83-year-old woman with herpes zoster oticus, also known as Ramsay Hunt syndrome, type 2. On admission to hospital, the patient presented with left facial nerve paresis (A). Four days later, vesicular lesions and redness could be seen in the patient's left external auditory meatus (B).

Our patient's family decided to focus on her comfort while in hospital and not pursue additional tests and treatment. The non-resolution of the patient's symptoms on follow-up reflected the syndrome's natural history rather than treatment failure.

References

1. Harpaz R, Ortega-Sanchez IR, Seward JF. Prevention of herpes zoster: recommendations of the Advisory Committee on Immunization Practices (ACIP). *MMWR Recomm Rep* 2008;57:1-30.
2. Wagner G, Klinge H, Sachse MM. Ramsay Hunt syndrome. *J Dtsch Dermatol Ges* 2012;10:238-44.
3. Dorsch JN. Neurologic syndromes of the head and neck. *Prim Care* 2014;41:133-49.
4. Ahmed AM, Brantley JS, Madkan V, et al. Managing herpes zoster in immunocompromised patients. *Herpes* 2007;14:32-6.
5. Hales CM, Harpaz R, Ortega-Sanchez I, et al. Update on recommendations for use of herpes zoster vaccine. *MMWR Morb Mortal Wkly Rep* 2014;63:729-31.

Competing interests: None declared.

This article has been peer reviewed.

The authors have obtained patient consent.

Affiliation: Department of Medicine, Queen's University, Kingston, Ont.

Correspondence to: Steven Montague, montaug@kgh.kari.net



A video showing a patient with left facial droop and mild dysphasia is available in Appendix 1, at www.cmaj.ca/lookup/suppl/doi:10.1503/cmaj.160483/-/DC1