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Early diagnosis and treatment of Ramsay Hunt syndrome: a case report

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Abstract

Background Ramsay Hunt syndrome (RHS), a rare complication of varicella-zoster virus (VZV) reactivation, presents with ipsilateral facial paralysis, ear pain, and vesicular rash. Early recognition is crucial for prompt treatment and optimal outcomes.

Case presentation We report a case of a 67-year-old woman with RHS who presented with right-sided facial palsy, severe ear pain, and fluid-filled blisters. Prompt diagnosis and initiation of antiviral and glucocorticoid therapy led to significant symptom improvement.

Conclusions Healthcare providers should consider RHS in patients with facial palsy, especially when accompanied by ear pain and vesicular rash, to facilitate early intervention and prevent long-term complications.

Keywords Ramsay Hunt syndrome, Peripheral facial palsy, Herpes zoster oticus, Geniculate ganglion reactivation

Background

Peripheral facial palsy, indicative of a lower motor neuron lesion, is most frequently caused by idiopathic Bell's palsy but can also result from infections, cholesteatoma, trauma, malignancy, autoimmune disorders, and pregnancy [1]. While the prognosis for Bell's palsy is generally favorable, the outcome for other causes depends on the underlying condition. Identifying the etiology is essential for effective treatment and improved outcomes.

Herpes zoster infection, caused by the reactivation of the VZV, typically presents with a painful, self-limiting dermatomal rash. After initial exposure resulting in varicella, the virus remains dormant in the dorsal root ganglia. Upon reactivation, it manifests as herpes zoster, starting with tingling pain along the affected dermatome, followed by a vesicular eruption within 2–3 days, characterized by painful grouped herpetiform vesicles on an erythematous base. Although usually self-limiting, herpes zoster can cause neurological complications such as aseptic meningitis, encephalitis, postherpetic neuralgia, herpes zoster ophthalmicus (HZO), acute retinal necrosis, and RHS.

RHS is a significant otologic complication of VZV reactivation. The latent virus reactivates in the geniculate ganglion and may spread to the eighth cranial nerve, and in rare cases, it can involve multiple cranial nerves, particularly cranial nerves V, IX, and X [2]. RHS typically presents with a triad of ipsilateral facial paralysis, ear pain, and vesicles in the auditory canal or on the auricle [3]. Other symptoms, including altered taste perception, tongue lesions, hearing abnormalities (decreased hearing, tinnitus, hyperacusis), lacrimation, vertigo, and nystagmus, are frequently reported due to the involvement of the affected nerves. This case aims to present a woman's clinical course and imaging findings of HZO.

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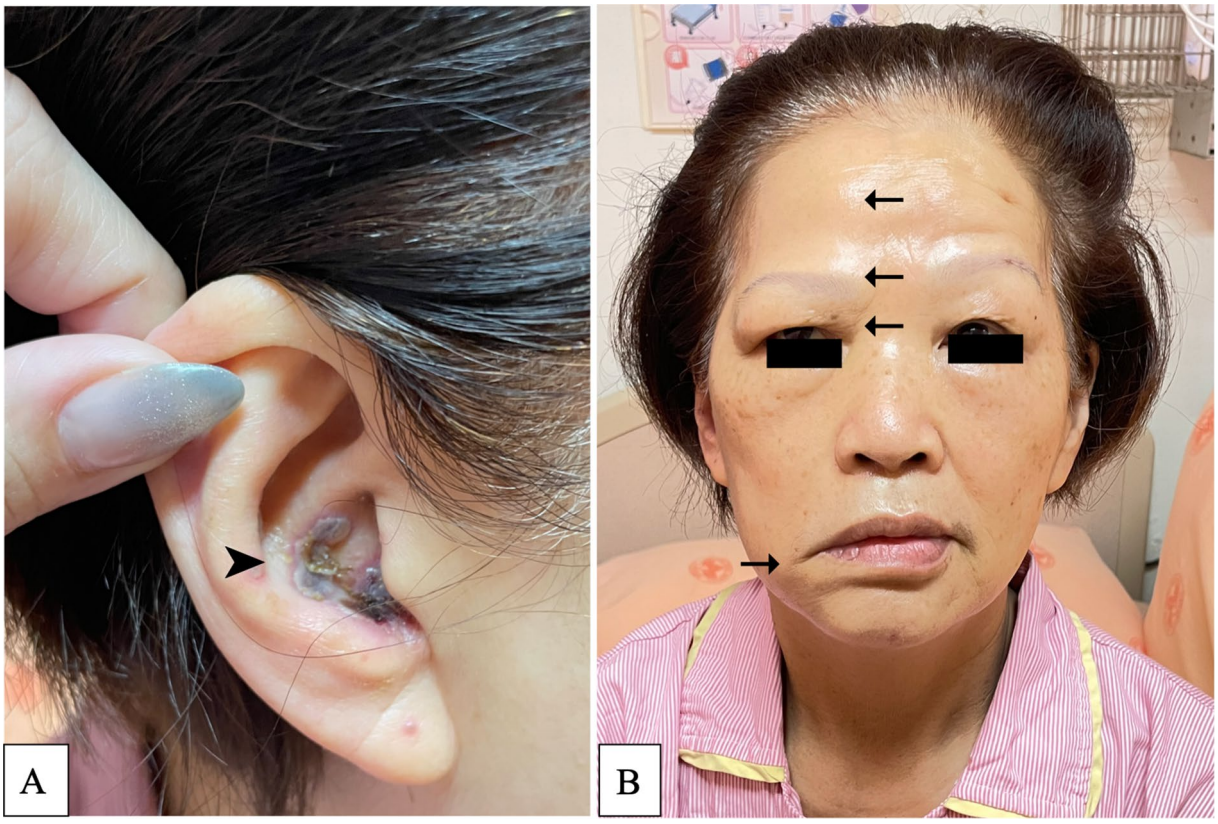

Fig. 1 Clinical manifestations of Ramsay Hunt Syndrome. **A**. Grouped vesicles (arrowhead) on the patient's right ear. **B**: Right peripheral facial palsy (arrows), characterized by the absence of forehead wrinkling, eyebrow ptosis, facial drooping, and a flattened nasolabial fold

Case presentation

A 67-year-old woman with no known history of systemic disease presented to the emergency department with a 5-day history of progressive right ear discomfort. She initially reported tingling pain in her right ear, which progressed to facial asymmetry, characterized by drooping on the right side of her face and an inability to fully close her right eye four days after symptom onset. On the day prior to admission, she experienced worsening ear pain and noticed fluid-filled blisters on and around her right ear.

She denied any recent trauma, fever, or upper respiratory symptoms. Her medical history was unremarkable, with no prior episodes of facial paralysis or significant infections. She reported no history of immunocompromised conditions, recent vaccinations, or travel.

Upon admission, her vital signs were stable, and systemic examination was unremarkable except grouped vesicles on her right auricle, extending to the external auditory canal and tympanic membrane (Fig. 1A). She exhibited right peripheral facial palsy (Fig. 1B), characterized by facial drooping, incomplete closure of the right eye, and a lack of forehead movement on the affected side, indicating moderate to severe dysfunction. The House-Brackmann system score was grade IV.

She was diagnosed with RHS and received intravenous (IV) acyclovir 500 mg every 8 h and methylprednisolone 40 mg daily for 9 days during her hospital stay. An electromyography (EMG) test was performed prior to discharge to evaluate the extent of nerve dysfunction. The EMG findings indicated a right facial nerve lesion in the hyperacute stage, showing reduced motor unit recruitment and decreased amplitude in the right facial muscles. Spontaneous activity was absent, and voluntary effort revealed partial but incomplete motor unit activation, suggesting ongoing but improving nerve function. Her facial palsy showed significant clinical improvement but had not fully resolved by discharge. Upon discharge, she was prescribed a tapered course of oral methylprednisolone: 12 mg twice daily for 2 days, 8 mg twice daily for 2 days, and 4 mg daily for 2 days with follow-up instructions.

Discussion

Facial palsy is a common cranial nerve disorder that necessitates prompt and accurate assessment to distinguish between central and peripheral etiologies, as this distinction significantly impacts treatment and prognosis. Evaluating the forehead musculature is crucial in this process. The frontalis, corrugator, and orbicularis

muscles are innervated centrally; thus, weakness in these muscles typically indicates a peripheral facial nerve issue, while sparing of these muscles suggests a central lesion. Clinicians can identify the problem's source by assessing the patient's ability to tightly close their eyes or wrinkle their forehead. Normal movement on the unaffected side further highlights the involvement of the forehead musculature.

Idiopathic causes account for more than half of peripheral facial palsy cases, with other etiologies including viral infections, diabetes, Lyme disease, and sarcoidosis. Among viral causes, Herpes simplex virus is the most common pathogen, followed by Herpes zoster. James Ramsay Hunt, an American neurologist, first described three distinct syndromes, with HZO, also known as type 2 HZO, being the best known. being the most well-known. RHS is relatively uncommon, with less than 1% of zoster cases involving the facial nerve and resulting in RHS. RHS is typically characterized by facial paralysis and a rash affecting the ear, predominantly in Hunt's Zone, which includes the tympanic membrane, middle ear, and cavum conchae. Additional symptoms frequently reported include ipsilateral altered taste perception and tongue lesions, hearing abnormalities (such as decreased hearing, tinnitus, and hyperacusis), vestibular disturbances (such as vertigo), and lacrimation, though these symptoms do not necessarily coincide [3].

Several grading systems are used to evaluate the severity of facial palsy, including the House-Brackmann, Sydney, Sunnybrook [4]. The House-Brackmann system is often preferred due to its quick and standardized evaluation, whereas others offer more detailed and complex assessments. However, no universally accepted standard grading system exists. The diagnosis of RHS is primarily based on clinical presentation, but in uncertain cases, laboratory tests such as polymerase chain reaction (PCR) testing, direct fluorescent antibody (DFA) testing, and viral culture PCR can confirm the diagnosis using cerebrospinal fluid, blood, and other non-cutaneous specimens.

Treatment for RHS includes symptom control, antiviral agents, and glucocorticoids [5]. Elderly and immunocompromised patients are at increased risk for varicella-zoster virus reactivation and its complications. RHS has a high rate of complete recovery, with up to 70.4% of patients regaining facial nerve function, particularly with early medical treatment. Therefore, early identification and treatment of RHS are essential.

Conclusions

Early recognition and treatment of RHS are essential to prevent long-term complications. In this case, a 67-year-old woman received early diagnosis and antiviral therapy, leading to significant symptom improvement. Healthcare providers should consider RHS in patients with facial palsy, especially when accompanied by ear pain and vesicular rash, to ensure prompt and effective treatment.

Abbreviations

RHS	Ramsay Hunt Syndrome
VZV	Varicella-Zoster Virus
HZO	Herpes Zoster Ophthalmicus
IV	Intravenous
EMG	Electromyography
PCR	Polymerase Chain Reaction
DFA	Direct Fluorescent Antibody

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Author contributions

Chen, Yu wrote the manuscript. Yen-Chia, Chen served as the corresponding author and was responsible for overseeing all other aspects of the study. Haw-Yu, Lee contributed to data collection.

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Data availability

No datasets were generated or analysed during the current study.

Declarations

Ethics approval and consent to participate

Not applicable.

Consent for publication

Written informed consent was obtained from the patient's relative for the publication of this case report and accompanying images.

Competing interests

The authors declare no competing interests.

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