

## CASE REPORT

# Where are the vesicles? A case report of Ramsay Hunt syndrome

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### Abstract

A 1-year-10-month-old child developed left-sided Ramsay Hunt syndrome (RHS) without vesicles 1 month after an episode of varicella zoster infection. No ear symptoms, including hearing loss, tinnitus, or imbalance, were reported. The external ear and otoscopic examinations were unremarkable. He achieved adequate recovery with corticosteroid treatment. This case report discusses the unusual presentation of RHS without vesicles, the diagnostic dilemma in young children, varicella zoster virus hepatitis, treatment modalities, and the role of vaccination in its prevention.

### Introduction

Facial nerve palsy can be of upper motor neuron (UMN) or lower motor neuron (LMN) pathology. UMN facial nerve palsy is caused by a lesion affecting the supranuclear pathway, such as a stroke or brain tumour, and results in weakness of the lower face. In contrast, a lesion that involves the nuclear or infranuclear pathways will lead to LMN facial nerve palsy, resulting in weakness of both the upper and lower face.<sup>1</sup> Sudden-onset LMN facial nerve palsy may have various causes, including complications of chronic otitis media, viral infections, temporal bone trauma, or parotid pathology. In children, it is estimated that the overall incidence of facial nerve palsy is 21.1/100,000 per year. Idiopathic (Bell's palsy) and Lyme borreliosis (*Borrelia* bacterial infection transmitted via tick bite) are the most common etiologies, while Ramsay Hunt syndrome (RHS) is rare and accounts for only 2.8% of all cases of facial nerve palsy.<sup>2</sup>

RHS, also known as herpes zoster oticus, is a variant of herpes zoster (HZ). It is caused by the reactivation of varicella zoster at the geniculate ganglion causing peripheral neuralgia, and it is often associated with a vesicular papular rash around the cutaneous distribution of the 7th cranial nerve; this distribution includes the external ear canal, pinna, and sometimes the tympanic membrane.<sup>3</sup> Occasionally, it may cause adjacent inflammation of the vestibulocochlear nerve within the internal acoustic meatus, resulting in vertigo, tinnitus, and hearing loss.<sup>4</sup> Some cases may also present with lesions over

the distribution of the lingual nerve within the oral cavity, which also carries fibres for taste from the facial nerve.

This case report presents a case of RHS resulting in isolated LMN facial nerve palsy in a child without the key feature of vesicular rash.

### Case Presentation

A 1-year-10-month-old boy developed sudden-onset left-sided facial weakness with the inability to close his left eye. He had no history of trauma, insect bite, or upper respiratory tract infection. He had no otologic symptoms, including ear discharge, hearing loss, or imbalance, but was seen to frequently tugging at his left pinna.

He received a 5-day course of oral prednisolone from a primary care provider, but his condition continued to worsen. He had a history of varicella infection 1 month prior to presentation, with vesicular rashes appearing over his face, trunk, and limbs, which resolved after 10 days and he had been well since. A referral to the ENT outpatient clinic was made in view of his worsening facial nerve palsy.

On examination, the patient had left-sided LMN facial nerve palsy, House–Brackmann grade V, characterised by an absence of forehead movement, inability to fully close his eye, and minimal movement of his mouth (Figure 1). External ear examination was unremarkable. Otoloscopic examination showed bilateral intact tympanic membranes with cone

of light and absence of hyperaemia. There was no observed discharge, vesicles, lesions, or foreign bodies. Oral cavity and oropharyngeal examination were unremarkable. Eye examination revealed superficial punctate keratitis over the left cornea with Bell's phenomenon. There was no nystagmus, and the remainder of the cranial nerve examination was normal. The child was able to walk and run normally without any sign of imbalance or abnormal movement.



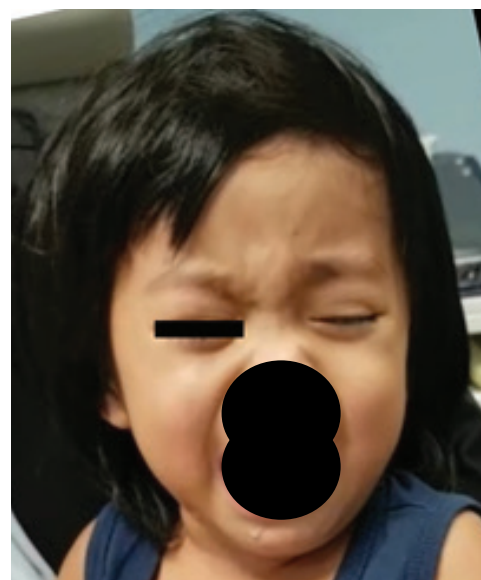
**Figure 1.** Pre-treatment shows left facial nerve palsy House-Brackmann grade V

Laboratory investigation showed an elevated total white cell count of  $21.2 \times 10^9$  cells per litre, with 72.5% lymphocytes. There was also an incidental finding of elevated ALT (62 U/L), which was likely secondary to post-VZV hepatic injury. A COVID-19 polymerase chain reaction (PCR) test was also performed in view of the increasing presentation of paediatric facial nerve palsy during the COVID-19 pandemic, and Malaysia had been experiencing a second wave of the pandemic at the time. The child and his parents were negative for COVID-19 on presentation.

Differential diagnoses included left-sided RHS without vesicles in view of recent VZV infection, and Bell's palsy given that there was no indication of ear or parotid infection. Varicella zoster IgG and IgM antibody titres were available after 1 week and were positive. Therefore, the final diagnosis was a left-sided LMN facial nerve palsy secondary to RHS without vesicles and mild exposure keratopathy.

Intravenous dexamethasone was administered for 5 days with facial rehabilitation. Antiviral therapy was not initiated as it had been more than 72 hours since the onset of the symptoms. The patient's left eye was patched at night,

and he received artificial tears. He completed a 2-week course of corticosteroids and was followed up 2 weeks later at the outpatient clinic. There was significant improvement of the facial nerve palsy from House-Brackmann grade V to II. Moreover, the left eye exposure keratopathy had resolved (**Figure 2**). The liver function tests had also normalised.



**Figure 2.** 2 weeks post-treatment shows left facial nerve palsy House-Brackmann grade II

### Discussion

Zoster sine herpete (ZSH) is an atypical variant of HZ without vesicular skin rash. It produces atypical symptoms, such as neuropathic pain, post herpetic neuralgia, and cranial nerve palsies, such as facial nerve palsy.<sup>5</sup> The infection may involve the cranial nerves, spinal nerves, visceral organs, or autonomic nerves, and may affect either motor neurons or sensory neurons that have no skin innervation.<sup>5</sup> The boy in this case presented with unilateral facial nerve palsy, without the typical auricular rash of RHS. The diagnosis was difficult as there was no dermatomal skin rash, and a high level of suspicion is required when encountering atypical symptoms without any other obvious cause. In this situation, laboratory investigation helped to overcome the diagnostic dilemma with PCR detection of VZV DNA, or anti-VZV IgG and IgM antibody detection via enzyme-linked immunosorbent assay (ELISA).<sup>5</sup> Other viral etiologies include Epstein-Barr virus (EBV), *Cytomegalovirus* (CMV), and herpes simplex.

With no VZV serology performed prior to presentation, the child was quickly diagnosed with Bell's palsy on his initial presentation to

the emergency department. In the strictest of definitions, Bell's palsy is an idiopathic facial nerve palsy and a diagnosis of exclusion after other possible causes of LMN facial nerve injury have been ruled out. Viral infections, including COVID-19, may cause peripheral neuropathy presenting as facial nerve palsies. Increasing numbers of children presenting to the emergency department with sudden-onset facial nerve palsies were reported in Italy during the COVID-19 pandemic; however, all patients were asymptomatic or had a negative COVID-19 diagnostic test. It is unclear whether this trend was observed elsewhere.<sup>6</sup>

Non-cutaneous manifestations of varicella zoster infection, such as hepatitis, are rare and are usually associated with immunocompromised states. In these patients, the condition may rapidly develop into acute fulminant hepatitis, which has a high mortality rate.<sup>7</sup> In contrast, VZV hepatitis in healthy children is a mostly benign and self-limiting condition. Approximately 3% of children in this group may develop hepatitis from varicella zoster infection. Symptomatically, there is no difference between the children with VZV hepatitis and children with varicella zoster infection without hepatitis, and neither group tends to develop hepatomegaly. Most of the children will have their liver function return to normal within 1 month, without any treatment.<sup>8</sup>

The mainstay treatment for RHS is antiviral therapy and corticosteroids. Antiviral therapy is effective in decreasing VZV replication to prevent its proliferation and spread, but it does not eradicate the existing virus. Antivirals help to reduce the duration of viral shedding and new lesion formation while hastening rash healing. However, the therapy has a short therapeutic window and must be initiated within 72 hours of rash onset for it to be effective. Acyclovir, an antiviral agent, has become the first-line treatment for HZ on all body sites, except for the ear. Despite its wide use in RHS, there is a lack of evidence of the benefits of acyclovir being statistically significant randomised-controlled trial (RCT).<sup>9</sup> Corticosteroids have anti-inflammatory properties that help to reduce oedema and minimise facial nerve damage in Bell's palsy<sup>10</sup>; they are also widely used in the treatment of RHS, given that both conditions involve inflammation of the facial nerve. However, unlike Bell's palsy, there is no significant evidence that supports the usage

of corticosteroids in RHS.<sup>11</sup> Although there have been no randomised trials that support both corticosteroids and antiviral therapy in RHS, multiple case series and cohort studies have shown that a combination of both treatments offers benefits and lowers the risk of complications.<sup>12</sup>

The varicella vaccine is a live-attenuated vaccine that contain the Japanese vaccine strain V-Oka.<sup>13</sup> The vaccine was initially developed to prevent varicella infection and has a role in the prevention of HZ and its complications. There are some concerns regarding vaccination, as vaccine-strain VZV establishes a latent infection and can theoretically reactivate into vaccine-strain HZ. Despite the risk of vaccine-strain HZ, the overall incidence of HZ (for both wild-type and vaccine strain) in vaccinated children is 79% lower than in unvaccinated children. Therefore, varicella vaccination is an effective way to prevent HZ and its variant, RHS, in children.<sup>14</sup> Furthermore, the rash is more likely to occur at dermatomes corresponding to the injection site, such as cervical and lumbar, and is likely to preserve other nerves, such as cranial nerves and the brain. From the history in our case, the child had not been vaccinated for VZV, but his RHS symptoms were milder than usual.

### Conclusion

RHS in paediatric patients is rare and has poorer outcomes compared with the more common Bell's palsy. The lack of auricular vesicles and otologic symptoms does not rule out RHS, which presents a diagnostic dilemma for inexperienced practitioners. Despite a lack of evidence, antiviral therapy and corticosteroids remain the treatment of choice for RHS. Varicella vaccination is effective in reducing the overall risk of HZ and preventing its complications.

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### Conflicts of interest

No conflicts of interest were reported.

### Patients' consent for the use of images and content for publication

The patient's parents consented verbally to the use of images and case content for publication.

**What is new in this case report compared to the previous literature?**

- RHS may present without vesicles, which mimics Bell's palsy.
- VZV hepatitis is asymptomatic, benign, and self-limiting in healthy children.
- VZV vaccine reduces the risk of RHS.

**What is the implication to patients?**

Other causes of facial nerve palsy in children need to be considered before being diagnosed as Bell's palsy.

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