Case Report

Ramsay-Hunt Syndrome in an 8-Year-Old Girl, Case Report and Literature Review

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Keywords

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Abstract

Ramsay-Hunt syndrome results from reactivation of latent varicella-zoster virus and subsequent spread to the seventh (and sometimes eighth) cranial nerve. The syndrome results in unilateral painful vesicular lesions on the external ear with ipsilateral facial weakness or paralysis. Other symptoms may include generalized malaise, altered taste, lacrimation, paroxysmal neuralgic ear pain, and hearing abnormalities. Due to its rare occurrence in young children and the inconsistent co-occurrence of pathognomonic vesicles and peripheral facial paralysis, diagnosis may be delayed or missed. As a result, many affected pediatric patients do not receive appropriate therapy with high-dose corticosteroids in combination with acyclovir.

Introduction

Varicella-zoster virus (VZV) is a pathogenic human herpesvirus responsible for the induction of chickenpox during primary infection. Subsequently, the virus establishes latency in neuronal cells within ganglia, including cranial nerve ganglia such as the geniculate ganglion. Reactivation of the virus primarily affects the facial nerve and often involves the vestibulocochlear nerve, resulting in Ramsay-Hunt syndrome (RHS). The annual incidence of RHS is approximately 5 per 100,000 people, with only 14% of reported cases occurring in children under the age of 16 (1).

The characteristic clinical triad includes facial paralysis, painful erythematous vesicular lesions on the external ear, and vestibulocochlear dysfunction. Other variable features may include nausea, vomiting, altered taste, lacrimation, and paroxysmal neuralgic ear pain. Typically, the disease manifests unilaterally, although rare cases of bilateral facial paralysis have been documented.

The annual incidence of facial nerve palsy in children is estimated to be 21 per 100,000. Common etiologies include idiopathic peripheral facial nerve palsy (Bell's palsy), acute otitis media, and Lyme disease, with RHS accounting for only 2.8% of all cases (2).

James Ramsay Hunt first described the syndrome in 1907, but there is little published large-scale research on its treatment. The recommended approach is a gold-standard combination of systemic acyclovir and high-dose corticosteroids.

Case presentation

An 8-year-old girl presented to the emergency department with drooping of the right corner of her mouth and inability to close her right eye. In addition, she reported pain in her right ear for the previous 3 days, accompanied by excessive tearing in her right eye, for which her general practitioner had previously prescribed an eye ointment. On admission to the emergency department, the patient presented with right-sided lagophthalmos and general malaise, leading to two episodes of vomiting. There were no complaints of tinnitus, hearing loss, dizziness, loss of taste or sensitivity, and no fever. Her medical history revealed no evidence of significant prior varicella infection, tick bite, or erythema migrans. In addition, there was no mention of a possible traumatic cause related to oral, maxillofacial, or nose-throat-ear surgical procedures. Subsequent clinical examination of the ear, nose, and throat revealed acute peripheral facial palsy on the right side. There were no symptoms or clinical findings of nystagmus or meningeal excitation. The severity of facial paralysis was graded according to the House-Brackmann scale (Table 1), with a grade IV status at the time of admission. Laboratory tests, including a complete blood count, C-reactive protein, and erythrocyte sedimentation rate, were within normal limits. Additional serologic testing was performed to identify an underlying viral or bacterial etiology of the facial nerve disorder.

Given the acute vomiting, a lumbar puncture was conducted to exclude central nervous system involvement. Magnetic Resonance Imaging (MRI) and Computed Tomography (CT) scans were not performed due to the patient's young age, the sudden onset of nerve paralysis, and the absence of a previous affliction.

Table 2 provides a chronological overview of the main symptoms and associated treatments. Intravenous corticosteroid therapy (60 mg/day) and ceftriaxone (100 mg/kg/day) were initiated. The anti-inflammatory properties of corticosteroids were expected to reduce the acute facial nerve swelling within the bony canal, thereby reducing the likelihood of residual symptoms. The administration of antibiotics was prompted by the possible occurrence of Lyme borreliosis, given the endemic prevalence of *Borrelia burgdorferi* in the geographical area, the seasonal context of

Table 1: House-Brackmann scale for assessing nerve damage in facial paresis.

Grade	Description	Characteristics
I	Normal function	Normal mimicry in all regions
II	Mild dysfunction	Mild asymmetry of mouth and forehead, complete eye closure with minimal effort
III	Moderate dysfunction	Mild asymmetry of the mouth, full eye closure with effort, slight movement of the forehead
IV	Moderate to severe dysfunction	Asymmetric mouth, incomplete eye closure, no forehead movement
V	Severe dysfunction	Slight mouth movement with effort, incomplete eye closure
VI	No movement	No movement

Table 2: Timeline of symptoms and associated treatments

	Day -3	Day 0	Day 1	Day 2	Day 3	Day 4	Day 5	Day 13
Symptoms	Right-sided earache, lacrimation	Right-sided facial paralysis, vomitus		Right-sided auricular herpetic vesicles			Dismissal	Control
HB grade		IV					II	I
Therapy		CS (60 mg p.o.) + ceftriaxone (100 mg/kg i.v.)	CS (60 mg p.o.) + ceftriaxone (100 mg/kg i.v.)	CS (60 mg p.o.) + aciclovir (100 mg/kg i.v.)	CS (60 mg p.o.) + aciclovir (100 mg/kg i.v.)	CS (60 mg p.o.) + aciclovir (100 mg/kg i.v.)	CS in 4-day tapering schedule + valaciclovir (60 mg/kg p.o.)	Stop treatment

CS = corticosteroids; HB = House-Brackman scale.

spring at the time of admission, and the conceivable pathogenic extent of such an infection (3). To protect the cornea of the affected eye, artificial tears were applied during the day and eye ointment and an eye patch were applied at night. No additional reports of nausea or vomiting were documented during the course of the admission.

On the second day after admission, the patient developed herpetic vesicles on the right auricle. CSF PCR was positive for VZV, confirming RHS. Intravenous aciclovir (100 mg/kg/d) was added to the corticosteroids for five days. Ceftriaxone was discontinued because of negative *Borrelia* serology and CSF PCR. The patient was discharged on day 6 with significant clinical improvement of facial paralysis, graded II on the House-Brackmann scale, and desiccation of the vesicular lesions.

Gradual tapering and discontinuation of corticosteroid therapy was achieved over a four-day period. Subsequently, the patient was started on oral valaciclovir (60 mg/kg/day), which was prescribed until the next follow-up visit 8 days later. During this follow-up, mild and limited asymmetry persisted at the ocular and oral levels. A follow-up visit 4-6 weeks after discharge revealed further resolution of symptoms.

Table 3: Most described medical condition associated with acute peripheral facial paresis in children

Medical condition	Disease or pathogen
Idiopathic	Bell's palsy
Infectious - Bacterial - Viral	 Lyme disease Herpes simplex virus/ Varicella zoster virus/ Coxsackievirus/ Epstein-Barr virus/ Cytomegalovirus/ Mumps virus/ Adenovirus/ Rubella virus/ Influenza B
Neoplastic	Acute leukemia/ Schwannoma/ Cholesteatoma
Traumatic & iatrogenic	
Others	Acute otitis media

Discussion

Acute peripheral facial paralysis in pediatric patients presents with a variety of potential etiologies, including infectious, neoplastic, traumatic, or idiopathic, as outlined in Table 3. Since idiopathic causes account for approximately half of all cases, and given the lack of a clear infectious focus on initial presentation, Bell's palsy initially emerged as the preferred diagnosis in our patient. Despite the negative history and clinical examination, Lyme disease could not be completely excluded, given the endemicity of *Borrelia burgdorferi* in the patient's home area and the temporal context of spring at admission. Notably, VZV, Epstein-Barr virus, and Coxsackievirus are the most commonly documented viral pathogens associated with peripheral facial paralysis.

RHS is a neurological disorder that may result from reactivation of VZV. The diagnosis of RHS is primarily based on clinical manifestations, as serologic testing is limited to the detection of antibodies to VZV, the presence of which does not unequivocally indicate ongoing infection. Confirmatory tests can be performed on cerebrospinal fluid or specimens obtained from herpetic vesicles, and sometimes tear fluid. In addition, MRI scans are used to rule out other conditions and may show swelling of the seventh cranial nerve, which is stained by contrast.

The timing of herpetic vesicles in relation to facial paralysis varies between children and adults. In children, these vesicles typically appear several days after the onset of facial paralysis, while in adults they often occur simultaneously (4). This observation underscores the importance of close and vigilant monitoring of children suspected of having RHS.

While spontaneous remission with complete recovery occurs in only about 20% of VZV infections resulting in facial paralysis, numerous case series and cohort studies have shown that a combination of antiviral therapy and corticosteroids can improve the overall prognosis and reduce the risk of complications such as permanent facial paralysis and deafness (5). In addition, antiviral therapy may have a significant beneficial effect on subjective hearing loss and vestibulocochlear nerve excitability testing (NET). A combination of aciclovir and corticosteroids is recommended to improve the rate of recovery of seventh and eighth cranial nerve function (6). Treatment should be initiated within 72 hours of symptom onset to take advantage of the narrow therapeutic window (7). However, the lack of pathognomonic vesicles and reliable early detection tests for VZV infection often leads to delayed diagnosis, especially in cases of zoster sine herpete, where an initial misdiagnosis as Bell's palsy leads to monotherapy with high-dose corticosteroids (8). While acyclovir is traditionally considered the drug of choice, valaciclovir may be used as a maintenance treatment due to its lower dosing frequency and better adherence.

Varicella vaccination is considered an effective approach to prevent RHS in children, although vaccinated individuals are not completely immune to varicella and its associated complications. Although the incidence of VZV reactivation appears to be lower in vaccinated children than in those who contract natural varicella infection, further studies are needed to prove the effectiveness of vaccination in preventing RHS (9).

Conclusion

RHS is a rare complication of latent VZV infection involving the facial nerve and occasionally the vestibulocochlear nerve. Despite its rarity, RHS can manifest in young children whose history of varicella infection is not always well documented. Clinicians must be aware that the hallmark symptoms of RHS, including peripheral facial palsy, vesicular rash, and otalgia, may not occur simultaneously. While the diagnosis is primarily based on clinical features, confirmatory testing can aid in accurate identification. Prompt initiation of treatment with a combination of antiviral and corticosteroid therapy within 72 hours of symptom onset is essential to improve prognosis and minimize neurologic dysfunction. Vigilant monitoring is essential for children with facial paralysis, with the nerve excitability test and the House-Brackmann scale serving as tools to assess the impact of the disease. Varicella vaccination has the potential to prevent RHS, although further research is needed to determine its efficacy in this regard.

Conflict of interest

The authors mention no conflict of interest.

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