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Pediatric Ramsay Hunt Syndrome: An uncommon cause of facial paralysis in children

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Abstract
Ramsay Hunt Syndrome (RHS) arises due to the reactivation of the varicella-zoster virus (VZV) manifesting as peripheral facial paralysis (PFP), periauricular pain, vesicular eruptions in the external auditory canal and ear, and vestibulocochlear dysfunction. Although RHS is uncommon in children, it ranks as the second most prevalent cause of non-traumatic facial paralysis in childhood. Diagnosis hinges on clinical evaluation, and the recommended treatment involves a combination of high-dose corticosteroids and acyclovir. This case study focuses on a 13-year-old adolescent with right ear pain, without fever. Within four hours of admission to the emergency room (ER), the patient developed a vesiculobullous rash in the right auditory canal and exhibited signs of lower motor neuron-type facial paralysis. RHS was considered, prompting the initiation of treatment with acyclovir and prednisolone, resulting in progressive clinical improvement. This underscores the importance of thorough examination and prompt initiation of therapy in suspected cases of RHS. This approach enhances the recovery rate of facial nerve paralysis and positively influences the disease prognosis.

Keywords: Ramsay hunt syndrome, varicella-zoster virus, periauricular pain

Introduction
Ramsay Hunt syndrome (RHS) is a relatively uncommon occurrence in pediatric cases of facial paralysis, representing only 10% of instances in this age group [1, 4]. Despite its lower frequency, RHS stands as the second most prevalent cause of nontraumatic facial palsy in childhood, following Bell's palsy. The documented incidence of RHS in children under 10 years old is reported to be 2.7 cases per 100,000 [1, 4]. The reactivation of the Varicella zoster virus (VZV) plays a significant role in 25 to 50% of peripheral facial paralysis (PFP) cases in the pediatric population aged between 6 and 15 [2-5]. In younger children under 6 years old with PFP, VZV reactivation is implicated in 9-10% of cases [2-5]. Notably, the risk of VZV reactivation seems to increase with a younger age at the time of initial Varicella zoster virus infection [2-5]. RHS manifests with symptoms such as PFP, inner ear dysfunction, periauricular pain, and herpetiform vesicles in the auricular region or mouth, resulting from the reactivation of latent VZV in the geniculate ganglion of the facial or vestibulocochlear nerves. This typically occurs a few years after the primary infection [3, 4]. The diagnosis primarily relies on clinical evaluation. There is ongoing debate about the use of corticosteroids and antivirals in pediatric patients, including uncertainties about their duration and route of administration, mainly due to a lack of dedicated studies. Nevertheless, early intervention in RHS is crucial to prevent disease progression, improve prognosis, and mitigate potential complications [2, 5].

Case Report
A 13-year-old adolescent was hospitalized after experiencing worsening right ear pain for five days, without fever. He had previously been diagnosed with acute media otitis and was undergoing treatment with amoxicillin and clavulanate. The patient, who had a history of type 1 diabetes since the age of 10 and had a past varicella infection at 8, showed signs of right auricular edema, pain on mastoid palpation, and redness of the ipsilateral eardrum during examination. A vesiculobullous rash appeared on the right ear canal and half of the palate (Figure 1), along with right facial nerve palsy (House-Brackmann scale II),

vestibulocochlear symptoms, hearing loss, and vertigo. Routine tests and a cranial CT scan yielded normal results. A positive PCR test confirmed a VZV infection on the vesicular lesion. A pure tone audiogram revealed mild sensorineural hearing loss on the right side. The patient received intravenous acyclovir (30 mg/Kg/day), oral prednisolone (60 mg/day), and continued antibiotics for acute otitis media with ceftriaxone (50 mg/Kg/day). A rehabilitation program involving massage, stretching exercises, and neuromuscular facial re-education was initiated during hospitalization. Following a seven-day course of medication and a gradual tapering of prednisolone over two weeks, the patient was discharged with a normal audiogram. However, reduced occlusion strength and slight asymmetry of mouth movements persisted (Figure 2). The rehabilitation program continued with thrice-weekly sessions for about a month, leading to complete recovery of facial nerve function within 20 days post-discharge.

Discussion

The diagnosis of RHS relies on a comprehensive examination of the patient's clinical history and physical condition. Typically, individuals may experience ear pain lasting 1-3 days as the initial symptom [1, 3]. RHS is characterized by a classic triad of symptoms, including ipsilateral facial paralysis, ear pain, and the presence of vesicles in the ear canal or on the auricle [1-3]. Other associated symptoms may include changes in taste, dry eyes, tearing, increased sensitivity to sound, nasal obstruction, and dysarthria. Vestibulocochlear nerve involvement can lead to hearing loss (In 24.4% of affected children), tinnitus, and vertigo, while vagus nerve involvement may manifest as hoarseness or difficulty in swallowing

Vesicles may appear subsequent to neurological symptoms, with pain often being the initial indicator. While vesicles typically emerge on the auricle, they can also be found on the affected side of the face, scalp, palate, and tongue [2-4]. In some cases, patients may not exhibit vesicles, and their primary complaints may revolve around severe pain and facial paralysis, resembling a variant known as zoster sine herpete, making it challenging to distinguish from Bell's palsy [5].

The prevalence of VZV reactivation is higher among children aged 6–15 compared to younger counterparts [2]. After contracting varicella, the virus becomes latent in the geniculate ganglion of the facial nerve, leading to RHS upon reactivation [1, 4]. Alongside clinical assessment, diagnostic measures such as enzyme-linked immunosorbent assay (ELISA) for serum anti-VZV IgG and IgM antibody titers, viral culture, and identification of viral DNA in samples of tears, saliva, or exudate from the external ear canal may be necessary alongside clinical assessment to rule out alternative causes [3]. Audiological studies may also be imperative, especially during follow-up.

Several therapeutic options are available for RHS, but the literature recommends early initiation of antiviral treatment within the initial 72 hours, coupled with high-dose steroid therapy, to hinder VZV replication and prevent complete facial nerve degeneration [2-5]. Simultaneous administration of prednisolone and acyclovir upon vesicle presentation resulted in notable improvement in facial nerve function by day 20. The concurrent use of antivirals and steroids proves more efficacious than steroids alone [2-5]. The concurrent use of antivirals and steroids proves more efficacious than steroids alone [2-5].

The prognosis for facial paralysis in RHS is less favorable than that in Bell's palsy, with only 10% of RHS patients achieving complete recovery [1, 4]. However, the outlook for RHS in children is more promising than in adults, and factors contributing to a poorer prognosis include audiovestibular findings, advanced facial paralysis at presentation, and delayed initiation of treatment [1-3].

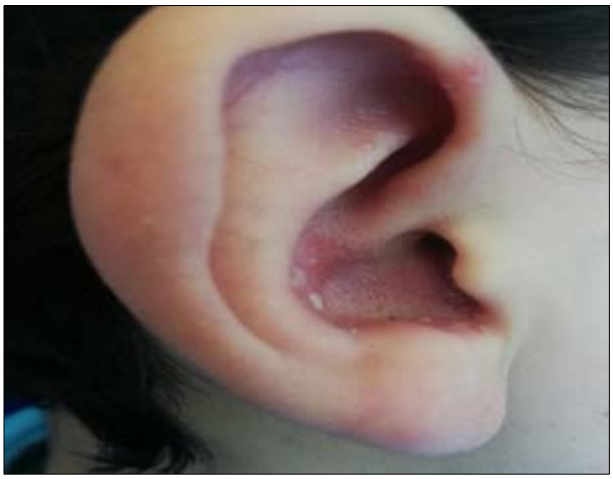

Fig 1: Skin lesions on the right ear

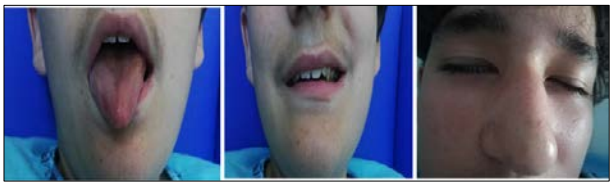

Fig 2: Right facial palsy, with decreased occlusion strength of the right eye and deviation of the labial commissure

Conclusion

The presentation of RHS can vary depending on the cranial nerves impacted and the degree of their involvement. Timely commencement of treatment, a high index of clinical suspicion, and laboratory verification of the infection are crucial for a favorable outcome, mitigating the risk of consequential motor nerve impairments.

Conflict of Interest

Not available

Financial Support

Not available

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