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Case Report

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A Case Report of Ramsay Hunt Syndrome with Concurrent Vestibulocochlear Nerve Involvement

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Abstract

Background: *Ramsay-Hunt syndrome (RSH) is a rare clinical condition characterized by herpes zoster oticus and ipslilateral facial nerve paralysis caused by reactivation of latent varicella-zoster virus (VZV) in the sensory root of geniculate ganglia. This syndrome must be distinguished from other causes of facial palsy such as Bell's palsy, stroke, Lyme disease and malignancies. Accurate dignosis and treatment in the first 72 hours of are crucial to improve the prognosis and to prevent later complications including postherpetic neuralgia and corneal damage.*

Case Report: *A 72-year-old female without any past medical history was admitted to emergency center of Vali Asr Hospital in Zanjan with hearing loss, ererythematous papules on the right external ear, otalgia, vertigo and dysgeusia. One day after hospitalization, the patient developed unilateral right-sided facial palsy and crusted erythematous papules on the right external ear.*

Conclusion: *Ramsay-Hunt syndrome is an uncommon peripheral neuropathy of the facial nerve caused by reactivation of VZV in the geniculate ganglion. Most cases occur in immunocompromised and elderly. Pharmacological interventions can reduce recovery time and prevent complications. Specialists are not unanimous on the choice of medication, dosage and the duration of therapy, thus more research is warranted.*

Keywords: Ramsay Hunt Syndrome, Herpes Zoster Oticus

Background

Ramsay-Hunt syndrome (RSH) is a rare clinical condition characterized by herpes zoster oticus and ipslilateral facial nerve paralysis caused by reactivation of latent varicella-zoster virus (VZV) in the sensory root of geniculate ganglia. This syndrome was firstly Described by James Ramsay Hunt in 1907. The elderly and immunocompromised individuals are at the highest risk and Children are rarely affected [1, 2].

Involvement of other Cranial nerves V, IX, X, XI, and XII might occur in this syndrome [2]. Clinical manifestations of RHS are various. Vesicular rashes iusually occur on the external ear and occasionally on oral cavit. In severe cases of HZO, involvement of the vestibular nerve causes patients to develop neurological hearing loss and vestibular symptoms [1].

This syndrome must be distinguished from other causes of facial palsy such as Bell's palsy, stroke, Lyme disease and malignancies. The diagnosis of RHS is primarily based on clinical presentation and physical examination. It is self limited, and the aim of pharmacological intervention is to alleviate symptoms rapidly and to prevent complications [1].

The treatment of choice includes a combination of antivirals and ccorticosterois. Accurate dignosis and treatment in the first 72 hours of are crucial to improve the prognosis and to prevent

later complications including postherpetic neuralgia and corneal damage [1, 3].

Case Report

Herein We present a case of Ramsay-hunt syndrome (RHS) in an immunocompetent adult female in iran. A 72-year-old female without any past medical history was admitted to emergency center of Vali Asr Hospital in Zanjan with hearing loss, erery-thematous papules on the right external ear, otalgia, vertigo and dysgeusia. One day after hospitalization, the patient developed facial paresis and crusted erythematous papules on the right external ear. [figure 1] physical examination of the oral cavity and pharynx was normal. The Otoscopy was normal. The pupils are of normal size and responsive to light. The patient was alert and hemodynamically stable.

Neurology consultation was requested, and Acute cerebrovascular disease (CVD) was ruled out. Brain computational tomography(CT) and magnet resonance imaging (MRI) were reported normal. Laboratory tests' results were in the normal range. The diagnosed of RHS syndrome was established based on clincical manifestations and physical examination.

Oral valacyclovir(1000mg three times a day) and intravenous methylprednisolone(1g/day) were administered for 10 days and 5 days respectively. The dose of methylprednisolone was tapered for the next 10 days. Artificial tear and eyeshield were given for eye protection. Intravenous acetaminophen and oral gabapentin were ordered for pain alleviation. During the first week of hospitalization, erythamatous rashes disappeared and vertigo relieved.

The patient was discharged with a course of oral acyclovir (400 mg five times a day) and oral prednisolone. Furthermore, she was advised to inoculate a dose of VZV vaccine.

The patient was followed up for one month. After 6 weeks the facial paresis improved considerably and regained sense of taste and hearing.

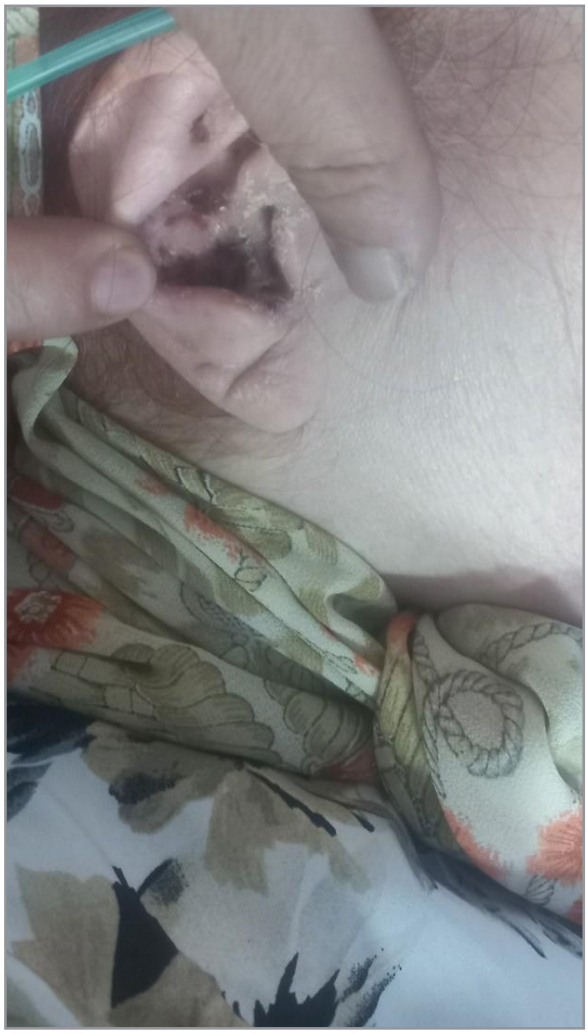

Figure1: Crusted erythematous rashes on the right external ear

Discussion

Herpes zoster oticus occurs when latent infection of varicella zoster virus reactivates in the geniculate ganglion. This condition is called Ramsay hunt syndrome if accompanied with facial palsy on the same side and other related symptoms [1]. In this study we described a rare case of Ramsay Hunt syndrome with involvement of the 7th and 8th cranial nerves who was successfully treated with significant relief of symptomes and signs.

Gondivkar et described a case of RHS presenting facial palsy on the left side and vesicular eruptions on the external ear. A combination of intravenous acyclovir and steroid was administered [4]. In a case study, a 16-year-old adolescent developed facial palsy a few days after developing herpes zoster oticus. The patient was treated with acyclovir(2400mg daily) without systemic corticosteroid [5]. In another study, penciclovir (250 mg twice daily) and methylprednisolone (40 mg) were used for treatment of RHS [6].

However, we started oral valacyclovir (1000 mg three times a day) for 10 days and pulsed dose of intravenous methylprednisolone for 5 days. Ghezta et reported a case of RHS in a 37 year old man without involvement of vetibulococochlear nerve. The patient had left sided facial palsy and erythematous rashes in the oral cavity with secondary candidiasis. The patient was treated with oral acyclovir(800mg qid) and oral presnisolone(60 mg) for seven days [3].

The development of RHS after vaccination has been reported in other case studies, although our patient had no recent history of vaccination [7, 8]. Postdischarge care is important for complete recovery and prevention of complications. Vaccination against the varicella-zoster virus can prevent the development of Ramsay-Hunt syndrome and is recommended for children and the adults over 50.

Conclusion

Ramsay-Hunt syndrome is an uncommon peripheral neuropathy of the facial nerve caused by reactivation of VZV in the geniculate ganglion. Most cases occur in immunocompromised and elderly. Pharmacological interventions can reduce recovery time and prevent complications. Specialists are not unanimous on the choice of medication, dosage and the duration of therapy, thus more research is warranted.

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