

A Case of Ramsay Hunt Syndrome with Atypical Presentation

Atipik Prezantasyonlu Bir Ramsay Hunt Sendromu Olgusu

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SUMMARY

Ramsay Hunt syndrome is a rare complication of herpes zoster which results from the reactivation of the latent varicella-zoster virus in the geniculate ganglion. Although facial nerve is the most common affected nerve in Ramsay Hunt syndrome, other cranial and cervical nerves can also be affected. We present an atypical case of Ramsay Hunt syndrome in a 42-year-old male, with cervical nerve involvement. As spontaneous recovery rate in Ramsay Hunt syndrome is low, early diagnosis and treatment plays a key role in full recovery of paralysis.

Key words: Facial palsy; Ramsay Hunt syndrome; varicella-zoster virus.

ÖZET

Ramsay Hunt sendromu, varisella-zoster virüsün latent olarak kaldığı genikulat ganglionda aktifleşmesiyle oluşan herpes zosterin nadir bir komplikasyonudur. Ramsay Hunt sendromunda fasiyal sinir en sık etkilenen sinir olmasına rağmen diğer kranial sinirler ve servikal sinirler de tutulabilir. Bu yazıda, 42 yaşındaki erkek hastada servikal tutulumun da eşlik ettiği atipik bir Ramsay Hunt sendromu olgusu sunuldu. Ramsay Hunt sendromunda spontan iyileşme oranları düşük olduğundan bu hastaların tanılarının erken dönemde konması ve tedavilerinin hemen başlanması paralizinin tam olarak iyileşmesinde kilit role sahiptir.

Anahtar sözcükler: Fasiyal paralizi; Ramsay Hunt sendromu; varisella-zoster virüsü.

Introduction

Ramsay Hunt Syndrome (RHS), also known as Herpes Zoster Oticus, was first defined by James Ramsay Hunt in 1907. [1] The disease is a rare complication of the primary varicella zoster virus infection (VZV). Activated many years after inoculation, VZV is followed by a latency period in the geniculate ganglion and spreads along the sensory tract of the facial nerve. Ipsilateral facial paralysis, otalgia, and vesicular rash of the skin of external auditory canal compose the classical triad of the disease. In contrary to Bell's palsy, the patients who are untreated or delayed in their treatment have poor prognosis and the full recovery rate is as low as 10-30% in these patients. [2,3] RHS is also known as cranial polyneuropathy and besides facial nerve, cranial nerves 5, 8, 9, 10, 11, 12, and C2-C4 cervical nerves may also be affected. The eighth cranial nerve is the most common involved nerve together with facial nerve, while the others are rarely involved. [4]

Here, we report a case of RHS, in which C2-C4 cervical nerves are affected together with the facial nerve.

Case Report

A 42-year-old man presented to our emergency department with the complaints of redness, numbness, and pain around left eye and face following an episode of otalgia, otorrhea, decreased hearing, and swelling of the left ear lasting three days. He did not have any chronic disease and medication history. His vital signs were as follows: blood pressure, 140/90 mmHg; heart rate, 76 bpm; respiratory rate, 16/minute; and temperature, 36.7 °C. Physical examination revealed edema of the external auditory canal and auricula of the left ear with a vesicular rash and serous discharge, periorbital and facial erythema and edema, stage 2 peripheric facial paralysis on the left side and vesicular rash of the left anterior cervical region (Figure 1a-c). Ophthalmic examination revealed no

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Figure 1. (a) Left facial palsy and periorbital edema. (b) Vesicular rash on the left face and periauricular region with serous auricular discharge. (c) Vesicular rash on the left anterior cervical region.

pathological findings. Hematological or biochemical findings were not substantial except a white blood cell count of 14.6 K/mL. The patient was hospitalized with the diagnosis of RHS and acyclovir 10 mg/kg/day and prednisolone 1 mg/kg/day were initiated. Following the regression of his clinical symptoms and physical findings, he was discharged at the third day of his admittance for ambulatory treatment and follow-up.

Discussion

In United States of America, it is estimated that one out of every three people are infected by VZV.^[5] However, clinical findings of herpes zoster do not develop in all infected subjects. The population based herpes zoster incidence was reported as 22.4 / 10,000 in a prospective study conducted by Paul and Thiel.^[6] RHS develops in 0.2% of primary herpes zoster infections.^[5] The incidence of the disease demonstrated an increase after fifth decade and peaked at eighth decade, parallel to the decreasing cellular immunity with the aging process. It is 20% more common in females compared to males.^[7]

The present case developed facial paralysis and vesicles around the left ear and neck following an episode of otalgia, otorrhea, decreased hearing, and swelling of the left ear, which lasted three days. Vesicular rash may manifest prior to or following facial paralysis or may not be observed at any time. When present, these vesicles may be observed at the external auditory canal, auricle, skin of the cheeks, the anterior two thirds of the tongue, hard palate, or cervical region.^[4] Our patient had vesicles at the external auditory

canal, auricle, and anterior cervical region with no presentation in other regions. The key clinical finding is facial paralysis and RHS, which covers 12% of cases with non-traumatic facial paralysis.^[7] Otolgia is the second most common symptom and 73% of the patients manifest otalgia as a complaint.^[2] Involvement of the vestibulocochlear nerve together with facial nerve is observed in 50% of the cases, causing hearing loss, vertigo, nausea, vomiting and nystagmus.^[4] Dysphagia, disturbances of gaze and taste, double vision suggest involvement of other cranial nerves. Furthermore, cardiac arrhythmias may develop when vagal nerve is involved.^[2,4] Our patient did not have any cranial nerve involvement other than facial nerve.

The cervical involvement observed in our case is not a typical finding in RHS.^[5] Three different theories are proposed for this clinical situation. The first theory is spreading of VZV through cerebrospinal fluid (CSF) or by hematogenous way. In 38% of the cases with cutaneous herpes zoster, pleocytosis is detected in CSF.^[8] Another study reported presence of 21% of the VZV in CSF of the patients manifesting cutaneous lesions.^[9] The second proposed theory is the presence of anastomoses between cervical nerves and branches of the facial nerve as an anatomical variation, leading to the spread of inflammation to more than one dermatome along these anastomoses.^[5,10] The last theory is the simultaneous activation of the virus in more than one ganglion, causing clinical symptoms.^[5]

The case presented here was diagnosed clinically. There was no need for ancillary laboratory or imaging studies. RHS is a clinical diagnosis with the unilateral facial paresis, otalgia

and vesicular rash in ipsilateral ear, hard palate, and the anterior two third of the tongue being sufficient for diagnosis. It is hard to differentiate between RHS and Bell's palsy when vesicles are not present in patients. Complement fixation tests and increased antibody levels may support the diagnosis. The detection of the virus by polymerase chain reaction in mononuclear cells of external auditory canal fluid, tear, CSF, and blood is accepted as the gold-standard in diagnosis of VZV.^[11] Jonsson et al. evaluated the cases with facial paralysis and reported that there were no extra benefits of contrast enhanced magnetic resonance imaging for either diagnosis or prognosis of the patients.^[12]

There are not so many randomized, prospective controlled studies for RHS in the medical literature. The data is mostly based on the case reports and retrospective case series. The actual management is starting with a combination of an antiviral agent and steroids.^[13] The most commonly used antiviral agent is acyclovir, however; a new generation antiviral drugs such as valacyclovir, famciclovir, penciclovir, and brivudine are being preferred due to increasing resistance.^[14] Prednisolone is suggested as the drug of choice for steroid therapy. In a retrospective study, Murakami et al. compared the effectiveness of the combination therapy of acyclovir and prednisolone with prednisolone therapy alone. They concluded that the full neurological recovery rate of the combination therapy group was significantly higher than the group treated merely with prednisolone. Moreover, when patients were grouped according to the time of initiation of drug therapy, the group in which therapy was initiated in the first three days showed a full neurological recovery rate of 75%. The rate decreased to 48% in the second group in which therapy was initiated in 4 to 7 days similar to the decrease of 30% in the third group in which therapy was initiated in later than 7 days. There was no clinically significant difference between therapeutic effects of parenteral and oral administration of acyclovir.^[3] Kinishi et al. compared the groups of patients who started acyclovir alone and prednisolone alone therapy in the first seven days. They reported significantly better nerve stimulation test results in the acyclovir alone group.^[15]

Since patients suffer from severe pain, pain management must also be taken into consideration in addition to the treatments mentioned above. Cellulitis may develop following infection of the vesicles, necessitating antibiotic therapy. Following discharge from the hospital, most of the patients need rehabilitation and long term follow-up.^[5] Our patient was administered a combination therapy of acyclovir and prednisolone after the clinical diagnosis was made. He was discharged at the third day for ambulatory treatment and follow-up.

The most important aspect of RHS when compared to Bell's palsy is the significantly worse prognosis when left untreated. In non-treated patients or patients in whom treatment was started later than seven days, the full neurological recovery rate is significantly as low as 10 to 30%. Advanced age, involvement of more than one cranial nerve, and a higher stage of facial paralysis during first admittance are well-defined prognostic indicators of RHS.^[2,3] The absence of the prognostic factors as well as the early initiation of therapy yielded full neurological recovery in the present case, consistent with the literature. The most common complication of RHS is postherpetic neuralgia that increases with age. Meningoencephalitis, myelitis, cerebrovascular problems associated with vasculitis, ventriculitis, and cerebral venous thrombosis may also develop.^[4] Examination of our patient during follow-up visits indicated no development of any of these listed complications.

Conclusion

RHS is a medical emergent condition necessitating early diagnosis and treatment in order to have a good prognosis. In patients with facial paralysis but no external vesicular findings, otoscopic and audiometric examinations must be performed. The combination therapy with antivirals and steroids must be initiated as soon as possible in order to minimize the risk of permanent neuronal damage.

Conflict of Interest

The authors declare that there is no potential conflicts of interest.

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