Ramsay Hunt Syndrome with Oral Findings: A Rare Case

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CASE REPORT

Ramsay Hunt Syndrome with Oral Findings: A Rare Case

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ABSTRACT

Ramsay Hunt syndrome (RHS) is a disease that is caused by the varicella-zoster virus and is characterized by severe ear pain, auricular vesicular eruptions, and peripheral facial paralysis. Objective: The aim of this case report is to provide information about the clinical findings and treatment process of RHS, which is a rare case and may have oral findings and stress the importance of early diagnosis. Case Report: A 60-year-old male patient had previously consulted an otolaryngologist and a family physician with complaints of vesicular eruptions in the right ear auricle and on the mandible. The patient in whom a diagnosis could not be established presented to the Department of Oral and Maxillofacial Radiology after exacerbated lesions. White plaque-like and ruptured vesicular lesions were observed in the intraoral examination. All vesicular lesions were on one side of the face, and the patient was referred to the dermatology clinic with the diagnosis of RHS. Facial paralysis fully recovered in a short time after early diagnosis and treatment. It should be kept in mind that there may also be oral findings in RHS, and a patient’s intraoral and extraoral examination findings should be evaluated together. Conclusion: Early diagnosis and treatment are highly important in preventing complications such as permanent facial paralysis, vestibulocochlear dysfunction, and hearing loss. Key words: facial paralysis, Ramsay Hunt syndrome, varicella-zoster virus, vesicular eruptions

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INTRODUCTION

Ramsay Hunt syndrome is a disease, which was defined by James Ramsay Hunt in 1907 and is characterized by severe otalgia, vesicular eruptions around the ear, peripheral facial paralysis, and frequent vestibulocochlear dysfunction. This disease is also known as herpes zoster oticus, and the varicella-zoster virus, a member of the herpes virus family, is responsible for it. Ramsay Hunt syndrome (RHS) emerges with facial nerve involvement of the varicella-zoster virus by 1%, and only 8% of these cases have been stated to have vesicles both around the ears and inside the mouth. Pain and vesicular eruptions occur along the nerve that is affected by the reactivation of the virus. Chronic diseases that reduce immune resistance, diabetes mellitus, hypertension, and the use of immunosuppressive drugs cause the reactivation of the virus. It can be seen at any age, but its incidence increases over 60 years of age. This increase has been reported to be proportional to the increase in the incidence of the deficiency associated with the cellular immune system over 60 years of age. Patients with RHS have weakened functions, such as wrinkling their forehead, raising their eyebrows, and closing their eyes, because of the weakening of the facial motor muscles on the affected side. When patients are requested to laugh or show their teeth, these movements are impossible on the affected side, and the face moves toward the healthy side. Oral lesions are in the regions of trigeminal nerve involvement and mobile areas such as the buccal mucosa. The lesions progress toward the midline, and the overlying skin is also involved. Sometimes it can lead to the devitalization of teeth and related bone necrosis.

With this case report, it is aimed to provide information about the clinical findings and treatment process of RHS, which may also produce oral findings, and stress the importance of early diagnosis and treatment in preventing permanent damage.
CASE REPORT

A 60-year-old male patient with diabetes mellitus presented to the Oral and Maxillofacial Radiology Department due to lesions in his mouth and vesicular eruptions on the skin tissue of his lower jaw. It was learned from his anamnesis that the patient had presented to the Department of Ear-Nose-Throat (ENT) with a complaint of ear pain 4 days before and that an ENT specialist had prescribed ciprofloxacin-derived antibiotics (ciprofloxacin hydrochloride 1 g – 3 times a day 2 tablets) and corticosteroid ear drops (dexamethasone 1 g – 3 times a day 1 tablet) upon diagnosing an external auditory canal infection. After using drugs for two days, lesions appeared in the patient’s mouth and on his face. Then, the patient was referred to the dentist by the family physician, and the general dental practitioner prescribed oral antiseptic mouthwash (chlorhexidine gluconate) and analgesic. The patient, whose complaints did not disappear and whose lesions gradually intensified, presented to our department as a last resort.

From the patient’s detailed anamnesis, it was learned that he had pain in the ear and eye. In the patient’s extraoral examination, vesicular eruptions and local crusts were seen in the right ear auricle, preauricular region, external auditory canal and zygomatic region in the right half of his head and on the right side of his lower jaw (Figure 1).

In the intraoral examination of the patient with poor oral hygiene, white plaque-like lesions on the right lateral of the tongue and edema in the tongue papillae were observed. There were ruptured vesicular lesions in the right sublingual region, alveolar mucosa of the lower jaw and the lower lip mucosa, and significant edema on the right side of the lower lip. Lesions of the entire mucosa and skin of the examined patient were unilateral (Figure 2). Apart from the ear, all lesions covering the right side of the mandible and the intraoral region were asymptomatic.

The patient, who could perform eye movements and mouth opening and closing functions completely, had limitations in his tongue movements. The patient did not have any complaints such as nausea, vomiting, and dizziness and did not use diabetes drugs. The patient had no such family history, but he was a smoker.

The patient was referred to the Dermatology Clinic with the preliminary diagnosis of RHS and then was hospitalized in the Dermatology Clinic on the same day. In the laboratory findings, the patient’s fasting blood sugar (161 mg/dL) and white blood cell (WBC - 11.76 10⁹/L) and C-Reactive protein (CRP- 111.36 mg/L) values from hemogram tests were high, whereas his sedimentation rate was normal (2 mm/h). His HIV, HBV, and HCV serology tests were negative.

Oral valacyclovir (1g - 3x1) therapy was started and administered for 7 days. First, his previous corticosteroid dose (dexamethasone 1 g - 3 times a day 1 tablet) was increased, and then vitamins B1, B6, B12 (1g – 1 time a day 1 tablet) and non-steroidal anti-inflammatory drugs were added to his treatment. Moreover, vesicular eruptions decreased after four days, and crusts started to occur in the patient to whom insulin therapy started to be administered (Figure 3).

The patient was discharged after one week, and facial paralysis emerged on the 12th day. The patient, who had mild asymmetry at the lower lip corner on the right side and limited tongue movements, also had difficulty and pain in moving the right eyelid. The patient could fully close his eyes. He was assessed as grade 2 according to the House-Brackmann facial paralysis grading system. In his examinations, HSV
IgG and VZV (varicella-zoster virus) IgM antibodies were positive (Figure 4).

The patient was again hospitalized in the Department of ENT, and the IV administration of corticosteroid (methylprednisolone 40 mg - 1 time a day 2 tablets) and antibiotic (sulbactam 1gr - 4 times a day 1 tablet), vitamins B1, B6, B12, and eye drops (0.5%/15 ml) with a moisturizing effect were started. The patient was discharged again after seven days, and his crusts and peripheral facial paralysis disappeared completely at his next-month follow-up.

DISCUSSION

In RHS, the time of vesicular eruptions is important in prognostic terms. According to the retrospective study conducted by Coulson et al. on 101 patients, the first symptom was ear pain in 55% of patients, whereas vesicles and facial paralysis occurred 2-3 days later. In 2% of patients, vesicles appeared first. Twenty-three percent first had facial paralysis. Furthermore, vesicles were observed on the auricle in 86% of patients, inside the mouth in 7%, and both around the ear and inside the mouth in 8%.

Another study stated that 46.5% of the vesicles in the ear occurred together with facial paralysis, 19.3% before paralysis, and 34.2% after paralysis. The eruption of vesicles before paralysis was reported to affect the prognosis of facial paralysis positively. In the same study, the sense of taste and lacrimation were reported to be impaired.

In this case, who first had ear pain, vesicular lesions inside the mouth emerged a few days after the eruptions around the ear. Twelve days after the onset of eruptions around the ear, facial paralysis occurred on the same side, although valacyclovir was used. The occurrence of paralysis after vesicular eruptions is a good prognostic sign, and the antiviral drug use probably delayed facial paralysis and reduced the House-Brackmann grade of facial paralysis.

In patients with RHS, erythematous eruptions are rarely observed in the ipsilateral anterior two-thirds of the tongue or palate. The chorda tympani is a sensory branch of the facial nerve related to taste in the anterior two-thirds of the tongue.

This nerve anastomoses with the lingual and maxillary nerves that enable the distribution of Ramsay Hunt lesions. A damaged chorda tympani leads to taste impairment in the anterior two-thirds of the affected half of the tongue and reduction in tear, nose, and salivary secretions. Patients with oral lesions report a burning sense or numbness in the relevant mucosal areas. In this case, in which intraoral findings were rarely observed, the patient did not have taste impairment and numbness. There were edemas in the tongue papillae on the right tongue lateral, and the patient’s tongue and alveolar mucosal surface were covered with a plaque-like white layer. Ruptured vesicular lesions were observed in the sublingual region, the lower lip mucosa, and the alveolar mucosa. The patient had a significant edema on the right side of the lower lip. These oral findings show that dentists should also have information about patients with RHS.

In addition to vesicular lesions observed in RHS, which are helpful for establishing a diagnosis, various neurological disorders such as tinnitus, hearing loss, nausea and vomiting, vertigo, and nystagmus may also emerge. The factors that worsen the prognosis of the disease include stress, chemotherapy, immunodeficiency, malnutrition, infection, diabetes, hypertension, and advanced age. In this case, it is thought that the reduced immunity of the patient who did not use diabetes drugs prepared the ground for secondary infection, with the effect of advanced age. In addition to the antiviral therapy administered to the patient, the patient’s blood sugar was also taken under control during the treatment. Moreover, except for vesicular lesions, the patient had pain in the eye only when he had otalgia and facial paralysis. However, the patient did not experience hearing loss and vertigo. This situation can be explained by the fact that the 8th cranial nerve and vestibulocochlear system were not affected owing to early treatment.

Bell’s paralysis is the most frequent cause of facial paralysis and is an acute-onset, idiopathic and short-term paralysis. RHS is the second most frequent cause of acute peripheral facial paralysis following Bell’s paralysis. Facial paralysis in RHS is more severe compared to Bell’s paralysis, and the recovery rate is
lower. Of patients, 71% recover from Bell's paralysis fully. In RHS, on the other hand, only 21% of patients with complete loss of facial functions recover fully. In patients with RHS, this rate was 25% for mild sequelae and 26% for moderate sequelae. It is crucial to start antiviral agents such as acyclovir, valacyclovir, and famciclovir, particularly in the first 72 hours, in terms of the efficiency of the treatment. Along with these drugs, non-steroidal anti-inflammatory drugs, corticosteroids or tricyclic antidepressants can be used to relieve herpetic neuralgia. In the patient diagnosed with RHS, the aim of the treatment was to enable quick recovery, ensure the least tissue sequestration, and stop the pain.

In the literature, there are studies on the efficiency of acyclovir and steroid treatment, and the combination of antiviral and steroid treatments has been proven to be significantly superior to steroid use alone. Murakami et al. observed complete recovery in 80 patients with RHS who were administered acyclovir and steroid (prednisone) in combination. Facial paralysis disappeared in 21 (75%) patients who started treatment in the first three days, 14 (48%) patients who started treatment in 4–7 days, and 7 (30%) patients who started treatment after the seventh day. Steroids have a strong anti-inflammatory effect that reduces inflammation and edema in the nerves linked with herpes zoster infection. Kanerva et al. achieved good recovery (complete recovery or recovery with only mild sequelae) in more than 80% of patients receiving antiviral therapy within 72 hours of the onset of RHS. Half of these patients received corticosteroid therapy concurrently with antiviral therapy.

In this case, steroid therapy was started in addition to valacyclovir antiviral therapy before facial paralysis, and nonsteroidal anti-inflammatory, vitamin B, ciprofloxacin group antibiotic therapy was administered in combination. The above studies show that the complete recovery rate is considerably lower in cases with delayed treatment. In this case, which started late and had a low degree of House-Brackmann facial paralysis, the complete recovery of the paralysis within a month is thought to result from early diagnosis and, thus, the early initiation of antiviral treatment.

CONCLUSION

The presented case had oral involvement, which is rare, unlike the classic effect of RHS. As seen in this case, it should not be forgotten that oral findings may be observed in RHS, and intraoral and extraoral examinations of the patient should be carried out together. Early diagnosis is important for the recovery of damaged nerves. Accordingly, permanent complications can be prevented. Moreover, starting antiviral and steroid therapies as soon as possible in elderly patients and patients with a chronic disease such as diabetes will prevent permanent facial paralysis, vestibulocochlear dysfunction, and hearing loss.

CONFLICT OF INTEREST

None declared.

REFERENCES


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