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Ramsay Hunt Syndrome Affecting Cranial Nerves VII and VIII: A Case Report.

Zafirah Hani Mohamad[1], Khamisah Awang Kechik[2]

[1]Oral Pathology and Oral Medicine Unit, Hospital Raja Permaisuri Bainun

[2]Oral Pathology and Oral Medicine Specialist, Oral Pathology and Oral Medicine Unit, Hospital Raja Permaisuri Bainun

ABSTRACT

Ramsay Hunt syndrome (RHS) is a rare complication of herpes zoster which happens after reactivation of Varicella zoster virus (VZV) from the geniculate ganglion. Practitioners should be aware of the presentations and it should be differentiated from Bell's palsy which is a commoner cause of peripheral facial palsy. We present a case of a 57-year-old male who attended our clinic with unilateral oral and skin lesions including his right external auditory meatus, tinnitus and fever. Based on the clear signs and symptoms, he was diagnosed as having herpes zoster. After 1 week of taking acyclovir orally, he developed paralysis of right facial nerve motor function with severe otalgia involving his right ear. The patient was then diagnosed as having postherpetic neuralgia (PHN) with RHS and was prescribed with oral prednisolone, amitriptyline and vitamin B12. After 1-month review, the pain has diminished and his facial nerve function has fully regained. He just experienced some discomfort at the previously affected region. We would like to report an occurrence of a rare complication of herpes zoster to raise awareness among practitioners. The importance of follow up for early detection and timely management of RHS is highlighted. The management of herpes zoster with RHS are also discussed.

KEY WORDS

Herpes; Herpes zoster; Shingles; Facial paralysis; Ramsay Hunt syndrome

INTRODUCTION

Herpes zoster is a secondary viral infection which happens after reactivation of varicella zoster virus (VZV) which remained latent in the sensory dorsal ganglia. The virus migrated along the affected dermatome and this is clinically presented as rashes[1]. A lot of patients with herpes zoster experience prodromal symptoms before the lesions become visible for 2-3 days to more than 1 week in duration[2]. Some individuals with herpes zoster heal completely while others may develop complications such as postherpetic neuralgia (PHN) and Ramsay Hunt syndrome (RHS). A patient with herpes zoster in our clinic developed both complications. In this paper, emphasis is given on RHS.

CASE PRESENTATION

A 57-year-old gentleman without any known co-morbid was brought to our clinic with lesions that have erupted on the right side of his face for the past 4 days with itchiness, tingling sensation, fever and tinnitus of his right ear. It started with painful ulcers inside of his mouth 2 days prior to the skin eruptions.

Treatment had been sought twice before presenting to our clinic. Initially, he visited a clinic a day following the eruption of skin lesions and was prescribed with a systemic steroid, antibiotic, painkiller and an unlabelled topical cream. The patient claimed that he feels his condition worsened after taking the medications. Hence, he stopped the medications and visited another practitioner after 2 days and started taking acyclovir tablet 400 mg for five times daily and acyclovir topical cream which decreased his symptoms.

Clinical examination revealed crops of vesicles on his right maxillary and mandibular dermatome (Figure 1), clearly without crossing the midline (Figure 2), in addition to his right external auditory meatus opening. Intraorally, multiple ulcers and erosions were present on his right buccal mucosa (Figure 3), labial mucosa and lateral border of tongue (Figure 4).

Figure 1. Vesicles involving right maxillary and mandibular branch region.

Figure 2. Vesicles involving right mandibular branch region, not crossing the midline.

Figure 3. Ulcers involving right buccal mucosa.

Figure 4. Ulcers involving right lateral border of tongue.

Based on the clear clinical presentation, he was diagnosed as having herpes zoster involving the maxillary and mandibular branches of trigeminal nerve.

Treatment was commenced with acyclovir tablet 800 mg, five times daily for 1-week duration. Advice on supportive care and contact precautions were also given.

After one week, he developed paralysis of his right facial nerve motor function (Figure 5(A) & 6(A)) and severe throbbing pain on the right side of face including his ear. Previous lesions have dried up. No new lesions were noted both extra and intra-orally (Figure 7-8). The patient was thus diagnosed as having postherpetic neuralgia (PHN) with RHS and was prescribed with a cocktail of prednisolone, amitriptyline and vitamin B12 tablets. Prednisolone was given in a tapering dosage from 60 mg to 10 mg throughout 13 days. Amitriptyline tablet was advised to be taken 25 mg each night and 500mcg of vitamin B12 daily for 2 weeks duration. The patient was further referred to the Ophthalmology department where eye involvement was ruled out.

Figure 5. (A) Loss of right facial nerve function shown during wrinkling forehead and (B) right facial nerve function has regained after 1-month review.

Figure 6. (A) Loss of right facial nerve function shown during blowing cheeks and (B) right facial nerve function has regained after 1-month review.

Figure 7. Ulcers on right buccal mucosa has healed after 1-week review.

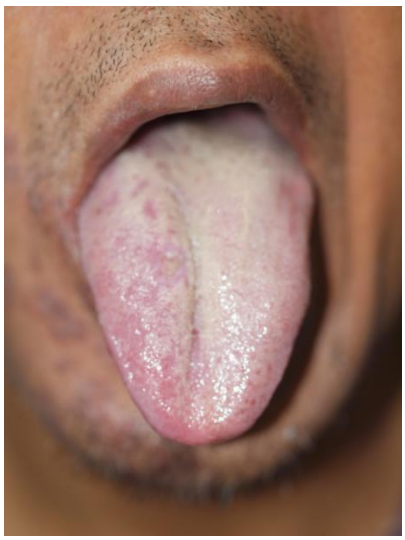

Figure 8. Ulcers on right side of tongue has healed after 1-week review.

After 2 weeks, his pain has decreased except at his right mandibular and tongue area and his facial nerve function improved partially. The patient was thus started with gabapentin tablet 300 mg at night once daily, amitriptyline tablet 25 mg at night once daily and mecobalamin tablet 500 mcg once daily for 1-month duration.

After 1 month, the pain has diminished and his right facial nerve function has fully regained (Figure 5(B)-6(B)). He just experienced some discomfort at the previously affected region. Both amitriptyline and mecobalamin dosage were continued for another 2 months. Gabapentin was gradually tapered down throughout the following 2 months before total discontinuation.

DISCUSSION

A person first infected with VZV will initially develop chickenpox. Then, the virus becomes latent in the sensory ganglia. It can be reactivated later in life, causing herpes zoster which is clinically presented as rashes. The most common complication of herpes zoster is postherpetic neuralgia (PHN)[1]. Besides that, herpes zoster can give rise to other rare complications such as RHS.

RHS happens when VZV is reactivated from the geniculate ganglion. It consists of three pathognomonic sign and symptoms of facial paralysis, earache and herpetic rashes[3]. The close proximity of the geniculate ganglion to the vestibulocochlear nerve is responsible for other frequent signs and symptoms such as hearing loss, nausea, vomiting, vertigo, and nystagmus[3].

Multiple cranial nerve involvement is possible including cranial nerves III, IV, V, VI VII, VIII, IX, X, XI although not as common as cranial nerves VII and VIII involvement. This might give rise to other possible symptoms such as otalgia, hyperacusis, taste disturbances and reduction of tear secretion, hoarseness, weakness of masseter and temporal muscles, asymmetrical rise of the soft palate and/or uvula deviation, dys-sensitivity of the face, tongue deviation, dysphagia, ocular deviation and dilated pupil, in order of descending incidence[4]. Cranial nerves VII and VIII were affected in our patient in the form of facial paralysis and otalgia.

Diagnosis is usually straight forward based on the patient's history, signs and symptoms alone[3]. In our case, the clinical pattern of the lesions and symptoms has shown sufficient evidence in reaching the diagnosis of varicella zoster virus infection.

RHS shouldn't be confused with Bell's palsy where the latter is associated with herpes simplex virus type I and is without visible rashes. The palsy in RHS is also more severe at onset in comparison to Bell's palsy and lesser patients recover their full facial nerve function[3].

It is important to note that the absence of rash doesn't exclude the diagnosis of RHS since there's a condition named zoster sine herpete where peripheral facial palsy happens without rash. Diagnosis is confirmed by a fourfold rise in antibody to

VZV or the detection of VZV DNA[3]. The detection of VZV versus HSV is even helpful for distinguishing RHS from Bell's palsy.

Early treatment of patients with RHS need to be considered with a 7 to 10 day course of antiviral such as acyclovir (800 mg, five times daily), and oral steroid such as prednisone (60 mg daily for 3–5 days)[3]. Combination therapy with steroids and acyclovir increases the recovery rate of RHS compared to steroids alone[5]. A study done by Kinishi et al has compared that statistically significant difference in recovery rate is observed when patients are treated with both steroids and acyclovir (90%) compared to steroids alone (64%)[6].

The earlier the treatment is initiated, the better the prognosis is[7]. It is recommended to be initiated within 72 hours[4]. Acyclovir treatment given by either intravenous or oral route of administration doesn't provide any statistical difference in facial nerve recovery[7]. Complete recovery is possible if not all the time. Poorer prognosis of facial palsy is associated with age, metabolic disease and multiple cranial nerve involvement[5]. In our case, the patient has regained his full facial nerve function even though combination therapy wasn't initiated within the first 72 hours. The good prognosis may be accounted to the relatively young age of the patient, absence of medical illness and involvement of only 2 cranial nerves.

Vaccination against VZV is present worldwide. The Malaysian Society of Geriatric Medicine Position Statement on Vaccination against the Herpes Zoster Virus in Older Adults (updated June 2014) has stated that vaccination is most cost effective to be given in patients aged between 70 to 79 years old[8]. Hence, vaccination was not offered to our patient.

CONCLUSION

RHS is a rare complication of herpes zoster. All practitioners should be aware of the possibility of such a rare complication happening. Early diagnosis by a specialist and combination therapy must be initiated as soon as possible to improve the prognosis of the disease. Hence, it is important to plan a follow up regimen after remission of the acute phase of varicella zoster virus infection. Patient's compliance with the advised oral medication dosage is also mandatory to ensure the success of treatment.

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CONSENT

Patient written consent has been obtained.

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COMPETING INTERESTS

The authors declare that no competing interests exist.

ABBREVIATIONS

PHN: Postherpetic Neuralgia

RHS: Ramsay Hunt syndrome

VZV: Varicella Zoster Virus

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