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Ramsay Hunt Syndrome: A unique clinical presentation in young patient- Case report and literature review

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Abstract

The Ramsay Hunt Syndrome (RHS), also called as Herpes Zoster Oticus, as well as, Shingles of the geniculate ganglion, is caused by reactivation of Varicella Zoster Virus (VZV) in the geniculate ganglia. It has a constellation of symptoms including vesicular rash of the external ear and ipsilateral two thirds of the tongue with associated paralysis of the seventh cranial nerve. Besides classic symptoms of the disease, the clinician must carefully examine several further presentations like, other cranial nerve involvement. RHS is a rare disease that presents with vague symptoms, so a high index of suspicion and close follow-up are essential. Rapid diagnosis and treatment of this rare neurodermatologic condition is crucial to avoid permanent complications. This case was presented due to its rarity and classical clinical presentation.

Keywords

facial paralysis; zoster oticus; VZV; Ramsay Hunt syndrome

Introduction

The strict definition of the Ramsay Hunt syndrome (RHS) is peripheral facial nerve palsy accompanied by an erythematous vesicular rash on the ear (zoster oticus) or in the mouth. Vesicular rash may manifest prior to or following facial paralysis or may not be observed at any time. J Ramsay Hunt, who described various clinical presentations of facial paralysis and rash, also recognised other frequent symptoms and signs such as tinnitus, hearing loss, nausea, vomiting, vertigo, and nystagmus [1]. He explained that these eighth nerve features are due to the close proximity of the geniculate ganglion to the vestibule-cochlear nerve within the bony facial canal. The incidence of RHS is 5 cases/100,000 people [2]. It affects mostly patients above fifth decade, with no gender predilection. Without treatment, full recovery of the facial paralysis occurs in as little as 20% of cases; this is much improved if treatment is started within 72 hours [3]. The aim of this article is to inform physicians about the presentation, including atypical, and management of RHS in order to facilitate prompt diagnosis, treatment and appropriate specialist referral.

Case Report

A 30-year old male reported to our department (Department of Oral Medicine and Radiology,

Subharti Dental College) with the chief complaint of painful ulcers in the mouth since 6 days following removal of carious right mandibular third molar. The patient presented with the history of pain which was mild, continuous, and radiating in nature, and was associated with fever of low grade since 9 days. After 3 days patient developed fluid-filled blisters distributed over the right half of the face. The patient gave history of chickenpox infection in childhood. History of prior vaccination for herpes zoster infection was not reported. On general physical examination, the patient was of normal built and no abnormality was detected in the nails, gait, upper, and lower limbs. Clinical signs of icterus, pallor, clubbing, oedema, cyanosis, and lymphadenopathy were absent. On evaluation of vital signs, temperature was noted to be 100°F and blood pressure 140/80 mm of Hg.

On extra-oral examination, no abnormality was detected in the eyes, nose, and temporomandibular joint. Clusters of vesicles were present on the right half of face involving cheek, upper lip and tragus of the ear. (Fig.1a,b) The lesions were limited to one half of the face without crossing the midline. On intraoral examination, multiple ulcers were seen on the buccal mucosa, hard palate and tongue on right side. (Fig.1c,d) Ulcers were irregular and measured approximately 4 mm × 5 mm in size. Margins of the ulcer were erythematous and edges were sloped. Correlating the case history and clinical findings, a provisional diagnosis of Herpes Zoster involving both maxillary (palate) and mandibular dermatome was given.

Antiviral drug therapy was started with acyclovir 800 mg five times a day for 5 days along with benzydamine mouthwash as local analgesic. On 5th day, regression in number of extraoral and intraoral lesions were noted with the formation of scar tissue and hypopigmented areas, but patient presented to us with the complaint of slight deviation of mouth towards left side since 3 days. Patient was unable to close his right eyelid with positive bell's phenomenon, decreased forehead wrinkling; slight obliteration of right nasolabial fold and inability to puff his cheek was observed. House Brackmann scoring system was used to grade the severity of facial nerve paralysis. Facial nerve paralysis was found to be of Grade III. Fig 2(a,c) Patient also gave history of itching sensation and watery discharge from the ear 3- 4 days back. No sensory loss on face or any lymphadenopathy was noted. The diagnosis was revised as RHS.

Patient was prescribed with systemic steroids; Tab. Prednisolone was administered in loading dose of 100 mg once daily in the morning for 1 week with local application of topical antiviral agent. After 1 week when the patient was reported, facial paralysis was improved to Grade I of House Brackmann scale. Fig. 2(b,d) Patient was prescribed with the same medication on alternate day basis for next 6 days and further follow up showed good recovery.

Discussion

RHS is an infectious disease and is a rare complication of the VZV infection [4]. 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 [5]. A positive history of varicella zoster infection had been reported in our case in his childhood. This reactivation of dormant virus results in a painful erythematous vesicular rash involving the ear as well as the anterior two/thirds of the ipsilateral tongue and anterior palate.

RHS is the second most common cause of atraumatic peripheral facial paralysis. Before 1986, the

frequency of zoster in patients with peripheral facial paralysis was estimated to be 4.5%-8.9% [6]. About 12 % of all facial palsies are caused by varicella zoster virus [7]. The mean age at onset of zoster among adults (age 22 years and older) is 59.4 years, with 68% of cases occurring in those 50 years and older. Age-adjusted rates are higher in women than in men (3.9 vs 3.2 per 1,000 person-years, respectively) [8,9]. The occurrence of herpes zoster in the head and neck region is reported as 13–35 % [10]. Considering the cranial nerves, theophthalmic nerve accounts for 10–20 % and the facial nerve 1 % of all herpes zoster manifestations [10,11]. RHS is the eponym given to a constellation of symptoms including a vesicular rash of the external ear and the ipsilateral two-third of the tongue associated with paralysis of the seventh cranial nerve.

Most commonly, acute herpes zoster is seen in thoracic dermatomes after re-activation of dormant virus within the thoracic sympathetic ganglia. The second most common site of reactivation is first division of the trigeminal nerve [12] (Fig 3). In less than 0.8% of the general population, dormant VZV may reactivate within the geniculate ganglion and its associated nerves resulting in facial pain, hearing loss, vertigo, vesicles in the ear, and pain [13].

Although in most patients the diagnosis of RHS can be made on clinical grounds, in some patients, especially those with other dermatologic conditions that confuse the clinical picture (e.g. Kaposi's sarcoma), laboratory testing may be beneficial. The individual symptoms of RHS are non-specific and can be seen in other diseases [Table 1]. Some 2-23% of unilateral facial palsies without vesicles are actually herpes zoster sine herpete [14]. The use of CSF analysis or MRI adds no additional diagnostic value [15]. Finally, the gold standard for diagnosing VZV reactivation is polymerase chain reaction of skin, saliva or middle ear fluid samples but this is rarely done clinically [16].

There are two primary goals when treating a patient suffering from RHS: (1) to provide immediate relief of acute pain and symptoms, (2) to prevent complications including exposure keratopathy of the ipsilateral cornea due to the patient's inability to properly close the eye, and (3) to prevent the late complication of postherpetic neuralgia. Clinical experience suggests that earlier treatment is associated with less development of postherpetic neuralgia. Pharmacologic treatment of HZ complicated by RHS is controversial and requires ongoing research. (Table 2) Antivirals and corticosteroids are the current mainstay of treatment. Acyclovir, valacyclovir and famcyclovir have been shown to reduce the duration of acute HZ symptoms and associated long term nerve damage.

Physical modalities- The application of ice packs to the lesions of acute herpes zoster may provide symptomatic relief in some patients, while the application of heat will often increase pain as a result of increased conduction of small fibres. Transcutaneous electrical nerve stimulation and vibration may also provide symptomatic relief. The favourable risk-to-benefit ratio of these physical modalities makes them reasonable alternatives for patients who cannot or will not undergo sympathetic neural blockade and do not tolerate the aforementioned pharmacologic interventions.

Conclusion

RHS is a rare but severe condition defined by herpetic rash of the ear or the mucosa of the mouth and peripheral facial nerve palsy. The incidence of the disease demonstrated an increase rate of occurrence after fifth decade and peaked at eighth decade, parallel to the decreasing cellular immunity

with the aging process. The present case is rare in its presentation as the patient belongs to younger age group. The pain associated with acute VZV infection usually precedes the appearance of the classic vesicular skin lesions by 5 to 7 days. Prompt diagnosis and treatment (ideally within 72 hours of the onset of symptoms) are crucial to secure the best outcomes. The ideal approach for treatment is still controversial and a multidisciplinary approach is essential for the follow up and recovery of these patients.

Tables

Table 1: Differential diagnosis of Ramsay hunt syndrome

Differential diagnosis	Features
Bell's palsy	Bell's palsy is a diagnosis of exclusion for unilateral facial weakness.[17] Erythematous vesicular rash and otalgia suggest Ramsay Hunt.
Otitis externa	The ear pain in RHS can be mistaken for otitis.[18] Onset of rash and development of facial palsy differentiate Ramsay Hunt from otitis.
Trigeminal neuralgia	Trigeminal neuralgia pain is similar to Ramsay Hunt but tends to be paroxysmal and stimulated by triggers. It is not associated with any skin manifestations or neurological losses.[19]

Table 2: Pharmacologic treatment of HZ complicated by RHS

Antiviral agents-	1. Famcyclovir, 500mg q8 hrs 2. Valacyclovir 1gm tid for 10 days and 3. Acyclovir 800mg/day 5 times a day · Shorten the course of the acute cutaneous manifestations of the VZV infection. · prevent the development of post-herpetic neuralgia(PHN) and attenuate the severity and duration of disease [13].
Steroids	Oral prednisolone- 1 mg/kg/day for 5 days followed by a 10 day taper • Large RCTs showed that adjunct corticosteroids resulted in quicker healing of the rash and decreased incidence and severity of pain [20].
Steroids and antivirals	The largest RHS treatment study was a retrospective analysis of 80 cases. Patients treated with acyclovir and prednisone within 72 hours of symptom onset had a complete recovery rate of 75% v/s those treated after 7 days, who had a complete recovery rate of 30% [21,22].
Sympathetic nerve block-	Combination of local anaesthetic, epinephrine and steroid
Opioid analgesics -	Controlled-release oxycodone (Oxycontin) in variable dose Daily dietary fibre supplementation and milk of magnesia prevent constipation.
Tricyclic antidepressants	Amitriptyline† • Up to 150 mg per day • Sedation, dry mouth, blurred vision, constipation, urinary retentionDesipramine (Norpramin) • Up to 150 mg per day
Anticonvulsants	• Gabapentin (Neurontin) 1,800 to 3,600 mg per day • Somnolence, dizziness, edema, dry mouth • Pregabalin (Lyrica) 150 to 600 mg per day

Topical agents	Tepid solution of aluminium sulphate[23] • Promote drying of crustings Capsaicin 0.075% cream (Zostrix)[24] • Applied three or four times per day • Burning skin Lidocaine 5% patch (Lidoderm) Maximum three patches per day

Figures

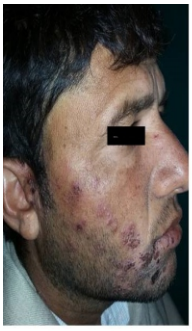


Fig.1(a)

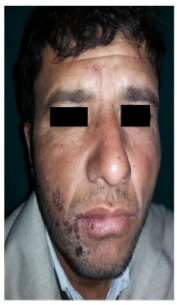


Fig.1(b)

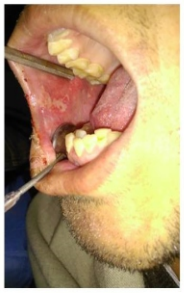


Fig.1(c)

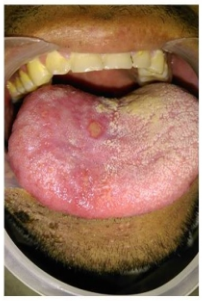


Fig. 1(d)

Fig.1(a,b) Extraoral images showing crustings of vesicular lesions on the right side of face involving tragus of ear, cheek area and upper and lower lip Fig.1(c,d) Intraoral images showing ulcerations on the right buccal mucosa and dorsal surface of tongue.

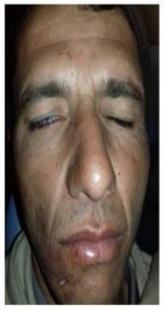


Fig. 2 a

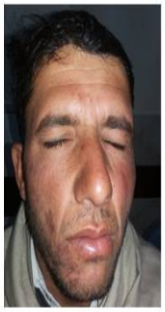


Fig 2 b

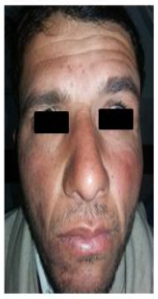


Fig. 2 c

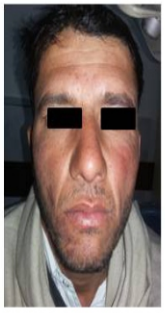
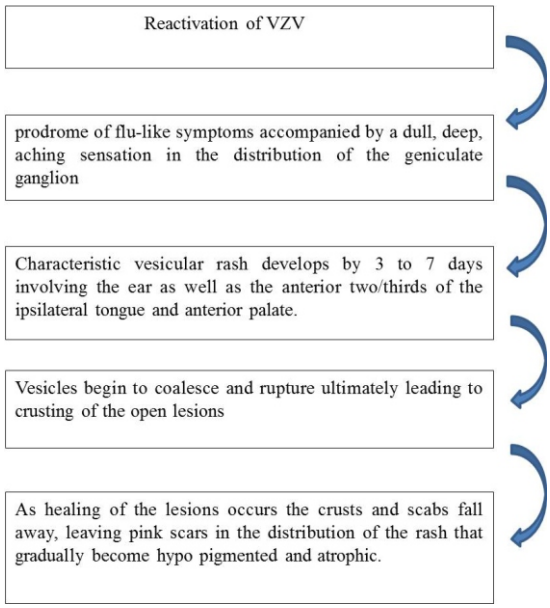


Fig. 2 d

Fig. 2(a,b) Depicts ability to close the eyes before and after treatment.
Fig. 2 (c,d) Depicts decreased forehead wrinkling on right side before treatment and improved condition after treatment.



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