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An Unusual Presentation of Ramsay Hunt Syndrome With Secondary Cellulitis: A Case Report And Review of Literature

[1]Dr Ajithkumar, Assistant Professor, Department of OMFS ,Govt. Dental College, Alappuzha, Kerala, India.

[2]Dr Sandeep Rajan O, Assistant Professor, Department of OMFS ,Govt. Dental College, Alappuzha, Kerala, India.

[3]Dr Naveen Nandagopal, Senior Resident, Department of OMFS ,Govt. Dental College, Alappuzha, Kerala, India.

Corresponding Author: Dr Ajithkumar, Assistant Professor, Department of OMFS ,Govt. Dental College, Alappuzha, Kerala, India.

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Abstract

Background: Ramsay Hunt syndrome is a rare and worst complication of varicella zoster virus reactivation, characterized by ipsilateral lower motor neuron facial palsy with ipsilateral paroxysmal auricular pain and erythematous vesicular rash/ herpetic blisters around the auricle and auditory canal. The rapid synaptic spreading capacity of varicella zoster virus, low spontaneous recovery rate and its potential sequelae make its early diagnosis, timely medical intervention and periodic follow up more vital to significantly improve its prognosis.

Method: Here we present a referred case with perioral blisters and diffused facial swelling followed by an attempted extraction of a lower molar carious tooth, which have an unexpected diagnosis of Ramsay Hunt Syndrome.

Result: In this case of Ramsay Hunt Syndrome, antiviral therapy and steroid reamin the mainstay of management. Antibiotic is also administered in view of secondary cellulitis. Famciclovir is the antiviral used in this case instead of acyclovir, as acyclovir reported increasing resistance as per literature reviews.

Conclusion: Even though early administration of antivirals and steroids can provide an excellent recovery of Ramsay Hunt Syndrome patients, early diagnosis and timely medical interventions have a paramount importance in such patients due to its potential sequelae and low spontaneous recovery rate.

Keywords: Ramsay Hunt Syndrome; Vesicular blisters; Cellulitis; Famciclovir; Dexamethasone

Introduction

Ramsay Hunt syndrome is a rare and worst complication of varicella zoster virus reactivation, characterized by ipsilateral lower motor neuron facial palsy with

ipsilateral paroxysmal auricular pain and erythematous vesicular rash/ herpetic blisters around the auricle and auditory canal. It was first described by James Ramsay Hunt in 1907.[1,2] Reactivation of the Varicella Zoster Virus (VZV) involving geniculate ganglion of facial nerve is considered to be the accepted etiology of Ramsay-Hunt Syndrome. It is mainly caused by varicella zoster virus type 3 with predilection to involve sensory nerves.[3] It can also affect cranial nerves VIII, IX, V and VI due to the proximity of their pathway alignments in the temporal bone. In a study by Ramsay Hunt, he described the condition of herpes virus involving cranial nerves as cephalic herpes zoster, in which peripheral facial nerve paralysis were common. Involvement of facial nerve result in facial paralysis where as the involvement of 5,6,8,9 cranial nerves cause vertigo, tinnitus, hearing loss and nystagmus.[1,4]

Even though early administration of oral steroids and acyclovir can provide an excellent recovery of Ramsay Hunt Syndrome patients, early diagnosis and timely medical interventions have a paramount importance in such patients due to its potential sequelae and low spontaneous recovery rate. Here we present a referred case with perioral blisters and diffused facial swelling followed by an attempted extraction of a lower molar carious tooth, which have an unexpected diagnosis of Ramsay Hunt Syndrome.

Case Presentation

A 56 year old male was referred from a private clinic to the department of oral and maxilofacial surgery, Government medical college Alappuzha with complaints of pain and swelling over left side of the lower face and multiple erosive lesions over upper and lower labial mucosa. The detailed history revealed that the patient had fever and few vesicular blisters over upper lip and perioral regions one and half week ago, which get self resolved. Later he had pain and swelling in relation to caries exposed left lower molar tooth, so that he consulted a nearby dental clinic for its extraction. Extraction was attempted from there after a course of antibiotics, which was failed due to severe pain. He was under antibiotics again for 3 more days. During this course he had multiple vesicles over perioral region and severe earache and got referred to our department. On detailed examination, patient had multiple scabbed eruptions and vesicular blisters in peroral regions and left side of face, erosive blisters in upper and lower labial mucosa, diffused swelling in relation to left mandibular body with a caries exposed left mandibular second molar tooth, periauricular blisters and active pus discharge from the left ear (Figure 1 and 2). The patient also exhibited inability to frown and wrinkle the forehead and inability to blow out cheeks. The patient also experienced tinnitus. There was no other significant medical history at present, but had a past history of chicken pox at nine years of age. The patient was admitted to the hospital and IV fluids and intravenous antibiotic(cefotaxime) and antipyretic(paracetamol) was started along with antiviral therapy of Famciclovir 250mg two times a day for five days and intravenous steroid therapy with dexamethasone 8mg three times a day for 2 days and taper within 3 days. Acyclovir 2% gel was given for local applications. ENT and dermatology consultations were done and advised to continue the same line of management. The patient experienced a great relief from pain and further vesicular eruptions from third day of admissions and symptomatically relieved by sixth day. The patient was discharged by sixth day with Famciclovir 250mg two times a day for five more days, wysolone 30 mg for three days and tapered doses in later days and acyclovir ointment. On the first week review after discharge, patient had a definite improvement in his symptoms.

Patient was completely relieved from all the symptoms by third week of review without any signs of facial palsy.

Differentiation Bells Palsy From Ramsay Hunt Syndrome

The low recovery rate and potential complications of Ramsay Hunt Syndrome gave an utmost importance to get differentiated from Bell's palsy. Even though Ramsay Hunt Syndrome and Bell's palsy have clinical similarity, prognostically Bell's palsy is far better than Ramsay Hunt syndrome.

Even though Bell's palsy is idiopathic in nature, it could be viral also as per the literatures. Any fall in cell mediated immunity may cause reactivation of dormant herpes virus in such patients. Ramsay Hunt Syndrome patients have a clear history of previous varicella zoster virus infection.

In Ramsay Hunt Syndrome, patients have severe paralysis at onset and less likely to recover compared to Bells palsy. More than 90% of Bell's palsy patients recover their facial nerve function whereas those with Ramsay Hunt syndrome have only 70% recovery.[3]

Some patients of Ramsay Hunt syndrome may develop vesiscles after the onset of facial weakness making the diagnosis more difficult from Bell's palsy, so that pinna and external auditory canal must be examined in the patients with a peripheral facial nerve palsy.

Discussion

In this case presentation, there was a unusal presentation of Ramsay Hunt Syndrome with secondary infection involving a caries exposed mandibular second molar, leading to the initial presentation of cellulitis. Newer antiviral drug famciclovir and steroid dexamethasone was considered as drug of choice in this case. Antibiotic cefotaxime and antipyretic paracetamol was also administered to manage the secondary infection and associated fever as well as pain along with local application of 2% acyclovir gel.

Ramsay Hunt Syndrome(RHS) also known as Geniculate ganglion herpes or otic zoster due to the dormant VZV in the geniculate ganglions, accounts for 12% of all facial nerve palsies, which is characterized by ipsilateral lower motor neuron facial palsy, herpes blisters of auricle, external auditory canal and tympanic membrane and neurological symptoms of the inner ear forms its clinical picture[4-6]. Symptomatically patients may have fever, tinnitus, painful skin vesicles, sensorineural hearing loss, hyperacusis, otalgia, vertigo, ageusia in the ipsilateral anterior tongue & nystagmus.

The generally accepted theory of etiology is the reactivation of latent VZV which was resided in the cranial nerve ganglia of the fifth, seventh(geniculate ganglion), eighth, ninth or tenth cranial nerve or the dorsal root ganglion along the entire neural axis after the initial VZV infection of chicken pox. As it has a capacity to spread via synapse of the nervous system, related cranial neuropathies may occur in the early stages even before the occurrence of vesicular rashes and facial palsy, which may lead to misdiagnose the condition[7,8].

Another theory behind its etiology is interneuronal communication between lower cranial nerves and upper cervical nerves and interconnection of second to fourth cervical nerves of the cervical plexus with peripheral branches of the facial nerve which was illustrated by Brown et. al[9-10]. VZV and associated inflammation may spread from a primary site of reactivation, through these anastomoses, resulting in simultaneous activation of multiple ganglia accounts for its clinical picture.

Last theory regarding its etiology is that of cerebrospinal fluid (CSF)/ hematogenous spread. Gold reported pleocytosis in the CSF in 38% of cutaneous herpes zoster cases, whereas Haanpaa et al. VZV was isolated from

CSF in 21% of patients with cutaneous lesions, but without meningeal or encephalitis symptoms[11,12].

VZV infection typically occurs during childhood in temperate climates and during early adulthood in tropical areas with life long latency period. Increased age group, immunocompromised conditions like HIV infection, malignancies and chemotherapeutic conditions may provoke the reactivation of the latent virus.

The cases of severe facial pain prior to the vesicular eruptions and facial palsy may often get misdiagnosed for trigeminal neuralgia. Throat ulcers with upper respiratory tract symptoms were misdiagnosed as an upper respiratory tract infection. Ramsay Hunt Syndrome is commonly get misdiagnosed as Bell's palsy in the cases of facial palsy symptoms even before the appearance of vesicular rashes. RHS patients have more severe neuropathies as well as sequelae compared to Bell's palsy. Cranial polyneuritis may occur as a serious sequelae in RHS patients, if timely medical intervention is not administered, which increases the risk of cerebral ischemia, cerebral apoplexy and progressive cognitive decline. As per the study of Kim et. al, in 2010, cranial polyneuritis have an incidence of 1.8% of Ramsay Hunt syndrome patients[1,13].

The associated symptoms of Ramsay Hunt Syndrome may vary depending on the physical condition of the patient and route of infection, which may appear before or after the main symptoms. This may complicates the diagnosis and delays the required treatment.

Even though Ramsay Hunt Syndrome can be diagnosed clinically by a triad of symptoms including unilateral facial weakness, otalgia and herpetic eruptions in any cranial dermatome, it cannot be applied to atypical presentation with absence of skin lesions or multiple nerve involvement. Facial weakness can be identified by facial drooping, a widened palpebral fissure, absence of the frontal wrinkles and decreased smile on the affected side where as the associated pain is of dull aching characteristic with associated allodynia. Polymerase chain reaction of saliva, skin or middle ear fluid samples is considered to be the gold standard for diagnosing VZV reactivation[1].

Pharmacological management of RHS is still debatable, which is under ongoing researches. Even though there were lack of data regarding the management of RHS, antivirals and corticosteroids still remain the mainstay of the management considering the possibility of life long facial paralysis and auditory impairement. Aizawa et al studied the prognosis of RHS while administering acyclovir and prednisone therapy, in which nearly 70%-86% of complete recovery was achieved while administering acyclovir and prednisone therapy within three days of symptomatic onset compared to 20% of patients receiving no treatment. 50% of complete loss of facial nerve response in the patients who are not treated within first three days[5,14].

Acyclovir inhibits viral DNA polymerase and thereby DNA replication after getting activated to acyclovir triphosphate by viral thymidine kinase. Kinishi et. al in 2001 reported a successful improvement in facial paralysis and nerve function by acyclovir therapy, which was measured by nerve excitability testing[15]. Even though acyclovir is the drug of choice for VZV infections, newer drugs such as famciclovir, valacyclovir, penciclovir and brivudine has been recently used for the management of RHS in view of reporting an increasing resistance to acyclovir therapy. All these antivirals have been reduced the duration of acute symptoms and associated nerve damage. Although there is no statistically significant difference between oral and IV Acyclovir, IV acyclovir is generally preferred in

immunocompromised cases[16]. But in our case, famciclovir orally and acyclovir in gel form was used.

Famciclovir is the oral form of penciclovir, a highly selective antiherpesvirus agent. Famciclovir undergoes rapid biotransformation to the active antiviral compound penciclovir, which has inhibitory activity against herpes simplex virus types 1 (HSV-1) and 2 (HSV-2) and varicella zoster virus (VZV). In HSV or VZV infected cells, penciclovir get phosphorylated to monophosphate form by viral thymidine kinase and then converted to penciclovir triphosphate by cellular kinases. This penciclovir triphosphate inhibits HSV-2 DNA polymerase competitively with deoxyguanosine triphosphate, thereby inhibits the herpes viral DNA synthesis and its replication[17].

However the efficacy of steroids in the management of RHS is still debatable, it can reduce the inflammation and edema of facial nerve, thereby reducing the damage and improve its recovery. As per Randomised Contolled Trials (RCT), corticosteroids as an adjunctive option reduces the incidence and severity of pain and enables rapid healing of the rash during VZV infectons without reducing the severity of post herpetic neuralgias. Finsterer et al reported the successful use of dexamethasone over prednisolone due to its high antiedematous effect and better neuro recovery rate[18]. In our case also dexamethasone was used during the course of admission, later changed to prednisolone(wysolone) during discharge. Hill et. al reported increased risk of developing severe varicella infection and its dissemination after steroid therapy during the VZV incubation period especially in pediatric patients with immunocompromised conditions like Acute Lymphoblastic Leukemia[19].

Recent studies recommend a combination therapy of antivirals and steroids within first 72 hours of symptomatic onset for the management of RHS. 800 mg acyclovir 5 times/day for 7–10 days and Prednisone 1 mg/kg for 5 days and taper is the usual regimen used in the recent studies, which shows an improved outcome.

Eye patches, taping the eye closed, artificial tears and oral analgesics can be used as an adjuvant in the management of RHS to prevent ocular injuries and pain, thereby attaining a better symptomatic relief.

Considering the risk of life long facial paralysis, physical therapy have a vital space in the management of RHS for a better prognosis. The physical therapy treatments are combinations of transcutaneous electrical stimulation, massage, neuromuscular re-education and exercise[8]. Brach and Van Swearingen described the physical therapy approach based on stages of progression which includes initiation, facilitation, movement control and relaxation. Flores reported a decrease in recovery time after faradic electrical stimulation. Brach and Van Swearingen describe defective neuromuscular re-education by accurate facial movement patterns and isolated muscle control, as opposed to mass muscle contraction needed for full expression[20]. But in the cases of persistent neuralgias, microvascular decompression and rhizotomy are the reserved therapeutic options.

Complications of RHS[7]

1. Corneal abrasions and ulcers, if eye-lid closure is impaired
2. Secondary infection with bacteria (cellulitis)
3. Postherpetic neuralgia
4. Permanent facial paralysis
5. Long term ipsilateral hearing loss and tinnitus

Conclusion

Ramsay Hunt syndrome is a rare and serious disease in the form of multiple cranial nerve involvement and facial paralysis with vague symptoms and atypical

presentations. The rapid synaptic spreading capacity of VZV, low spontaneous recovery rate and its potential sequelae make its early diagnosis, timely medical intervention and periodic follow up more vital to significantly improve its prognosis. As per the literatures the early intervention with antivirals and steroids remains the mainstay of the treatment of Ramsay Hunt Syndrome.

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Legend Figure

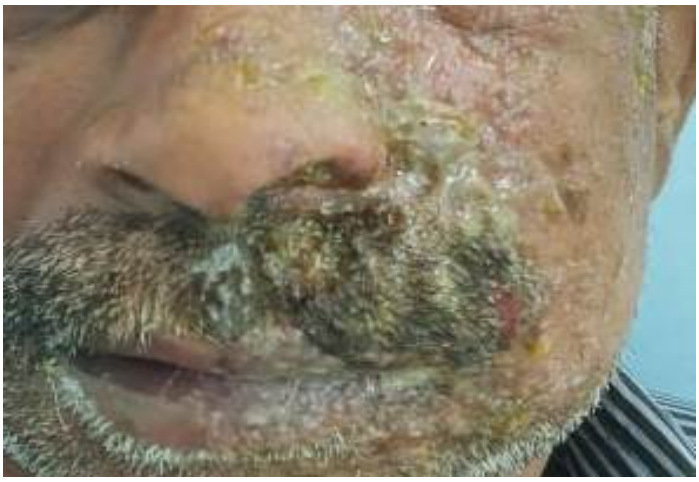

Figure 1: Perioral blisters with cellulitis on left mandibular body region

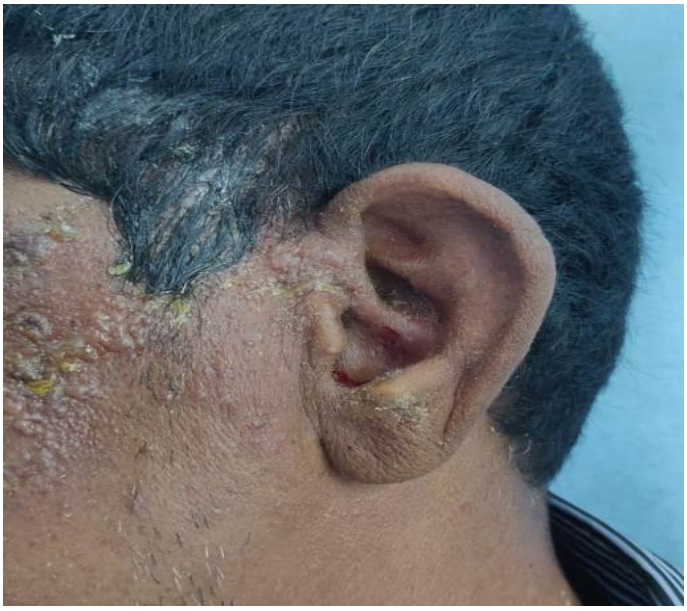

Figure 2: Periauricular blisters

Abbreviations

VZV - Varicella Zoster Virus

RHS - Ramsay Hunt Syndrome

CSF - Cerebro Spinal Fluid

HSV 1 - Herpes Simplex Virus Type 1

HSV 2 - Herpes Simplex Virus Type 2

DNA - Deoxy ribonucleic Acid