

Ramsay Hunt Syndrome: An Overview on Unwelcomed Disease.

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ABSTRACT

Acute Herpes Zoster is a form that includes Ramsay Hunt Syndrome (RHS), includes peripheral facial palsy (FP). The Cranial Nerves in the head and neck region are severely affected. It is an extremely uncommon neurological condition.

J. Ramsay Hunt discusses the different clinical facial manifestations of face paralysis, as well as rashes, hearing loss, nausea, vomiting, and excruciating pain on one side of the face.

The Ramsay Hunt Syndrome is a virus brought on by the varicella zoster virus (VZV). Compared to Bell's Palsy, a patient with Ramsay Hunt Syndrome, a severe form of paralysis, has very little chance of fully recovering (Facial paralysis without rash).

In the treatment of Ramsay Hunt Syndrome two main drugs are considered important. Prednisone and Acyclovir are these two drugs which can improve the outcomes. If the medications are started within 72 hours of Ramsay Hunt Syndrome Diagnosis, it results in either totally recovered facial palsy or with only slight consequences in over 80% of the affected area. Approximately 15-20% of patient had their blisters in certain hidden places i.e., ear canal or mouth.

In the treatment of Ramsay Hunt Syndrome use of different combinations of drugs are preferred. They contain some of the anti-viral agents and steroids, which is recommended by experts. Additionally, early diagnosis of the Ramsay Hunt Syndrome is a vital factor which is used to improve nerve response which are damaged in Ramsay Hunt Syndrome, which initiates treatment to be recovered as soon as possible.

Keywords: Facial Paralysis, Herpes Zoster, Herpes Zoster Oticus, Ramsay Hunt Syndrome, Varicella Zoster Virus (VZV)

INTRODUCTION:

James Ramsay Hunt, an American neurosurgeon, founded it for the first time in 1907. A vesicular rash on the external ear and the

ipsilateral two-thirds of the tongue are among the self-described constellation of symptoms that are associated with VIIth cranial nerve paralysis^[1]. A condition known as Ramsay Hunt Syndrome (RHS) causes cranial nerve peripheral facial palsy. This area contains the VIIIth cranial nerve. There are also head and neck herpes blisters are present. Here, all the nerves that connect with the face nerves are involved including,

- the cervical nerves C2, C3, and C4 and
- the cranial nerves V, VIII, IX, and X^[2].

Reactivation of varicella-Zoster virus (VZV) give rise to Ramsay Hunt Syndrome (RHS). And also, the cranial neuropathy which includes many additional cranial nerves (CN's) i.e., III, XI, XII are excluded from the original definition of the Ramsay Hunt Syndrome.



Fig.1: Ramsay Hunt Syndrome

There were 178 incidences of Ramsay Hunt Syndrome among the 3015 people with facial palsy (FP). Regarding the time of the association between blisters and facial palsy, there are three primary possibilities in the clinical picture of RHS. Ramsay Hunt Syndrome can be easily recognised if the blisters appear early or concurrently with the palsy. Blisters soon disappear and are easy for inspectors to miss^[3,4,5].

Vesicles may occur after the onset of neurological symptoms; however, pain is typically

the first sign to manifest. Although it can be observed on the affected side, scalp, tongue, and palate, vesicles primarily develop on the auricle. However, other patients' vesicles are absent, and their main complaints are facial palsy and excruciating pain. This variation is known as Zoster sine herpete, and clinically, it can be challenging to distinguish it from Bell Palsy^[6].

Hunt's research on herpetic inflammation of the geniculate ganglion described the relation between the geniculate ganglion and somatic sensory function in the ear^[32]. Before Hunt's superficial description of the facial nerve sensory system, ear, rash and pain had been attributed to inflammation of the gasserian ganglion. Hunt

concluded that the Somatic sensation from portions of the ear was not received by afferent fibres of second and third dorsal root ganglion^[33].

He discovered that the distribution of vesicular discharge matched the areas of sensation spread by surgery after reviewing 60 cases of zoster oticus. He came to the conclusion that the geniculate ganglion of the facial nerve, and the plexiform nerve of the vagous nerve were responsible for the ear's sensation^[34].

On the combination of his clinical knowledge with anatomy, Hunt suspected that the geniculate zone included the concha, antihelix, antitragus and the portion of lobule^[35].

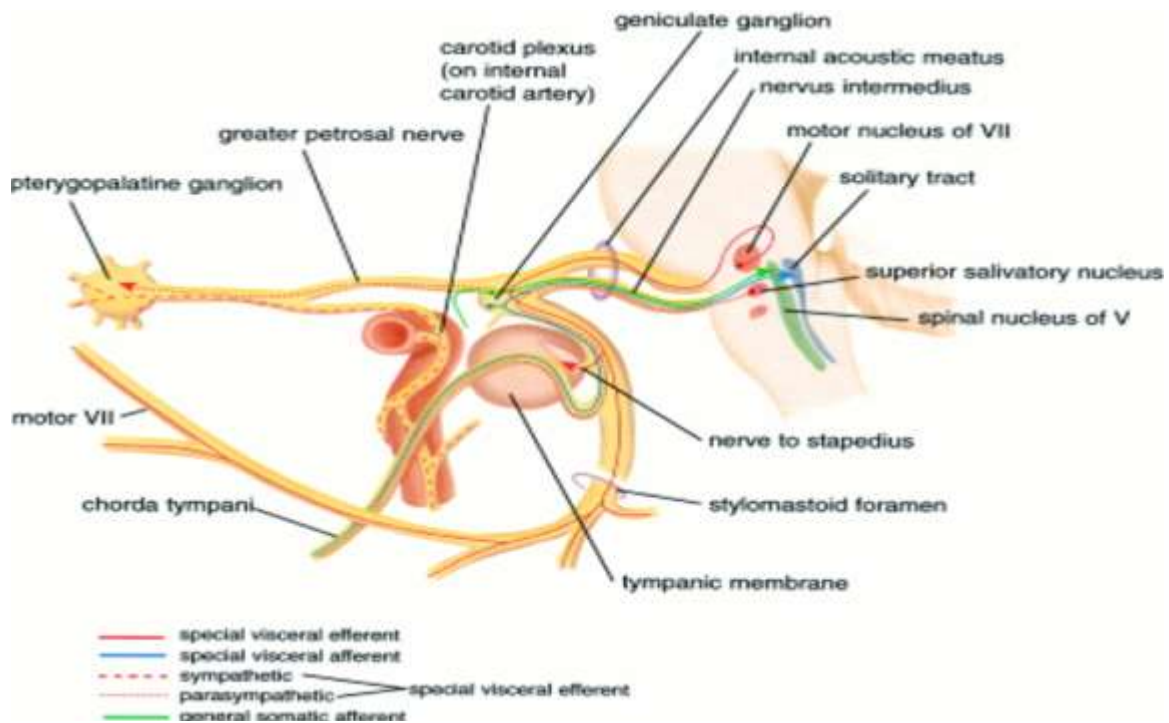


Fig.2: Anatomy of the facial and intermedius nerves

Pathophysiology

The Ramsay Hunt Syndrome (RHS) and the Varicella-Zoster virus (VZV) initially cause fever and body rashes, a condition known as chickenpox. During the acute stage of the infection, droplets found in the respiratory system disseminate this virus. The virus reactivates within the nerve distribution where it was dormant during physiological stress and low immune defence. This virus is reproducing and is being expelled from the host cell where it is doing so from the vesicle. A virus can proliferate from an organism's saliva and tears, and it can also be detected through PCR

testing. Reactivation of the virus occurs via the cranial nerve via Geniculate Ganglion in Ramsay Hunt Syndrome^[11].

Vagal nerve involvement is more common than it appears to be. Unless the patient is suffering with the symptoms like hoarseness or aspiration, vocal cord paralysis is not usually noted since it requires mirror or fiberoptic laryngoscopy to discover^[12,13].

Rarely, other cranial nerves can be involved as well, including the trigeminal, glossopharyngeal, and hypoglossal, although cranial polyneuropathy is more likely to present in

immunocompromised patients, such as those with diabetes mellitus or human immunodeficiency virus infection^[14,15,16,12]. The facial paralysis (FP) resulting from (RHS) Ramsay Hunt Syndrome has a worse forecast than that seen in Bell's palsy, with only 70% regaining normal or near-normal facial function compared with over 90% in Bell palsy^[17,18].

After Bell palsy, the rate of synkinesis development is roughly about 16% but is closer to 40% after Ramsay Hunt Syndrome^[19,20].

Etiology

The Varicella Zoster Virus (VZV), a member of the human herpes virus family, is the cause of Ramsay Hunt Syndrome (RHS). And more particularly, it, together with herpes simplex viruses 1 and 2, belongs to the alpha-herpes-virinae subfamily. Technically referred to as human alpha herpes virus 3 (VZV), varicella zoster virus is a double stranded DNA virus^[21].

Once the clinical (VZV) Varicella Zoster Virus infection, chickenpox, has cleared, the virus remains latent in cranial nerve or dorsal root ganglia and may subsequently reactivate in times of physiological stress or immunocompromise, leading to herpes zoster, known as "shingles" anywhere on the body or Ramsay Hunt Syndrome when facial paralysis is involved^[22].

Although there has been a significant decline in the incidence of both chickenpox and shingles since the live attenuated Varicella Zoster Virus (VZV) vaccine became widely available in 1995, shingles and Ramsay Hunt Syndrome have still been reported in patients who have never had chickenpox but have received the vaccine^[23,24,25].

Signs and Symptoms

When Varicella Zoster Virus (VZV) is reactivated then patients may also experience symptoms as like flu accompanied by abnormalities like deep, dull and aching sensation in the ear which often progress to sharper, neuritic type pain for the geniculate ganglion distribution^[7].

The Neuropathic pain in the studied Ramsay Hunt Syndrome which can resolve as the skin lesions heal, in the major problems of the patients. In minor patients the pain persists beyond lesions healing. This pain is referred to as postherpetic neuralgia. This shows a high effect on younger counterparts than in their elderly patient has much higher rate^[8].

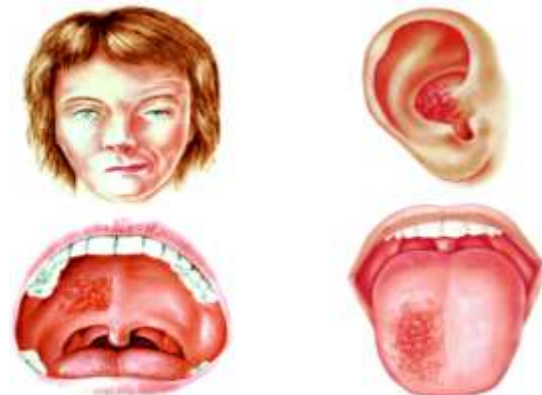


Fig.3: Facial Palsy, Ear Lesions, Tongue and Soft Palette

Additional Symptoms are vomiting, nausea and the sensations. Complications possible rare cases of Ramsay Hunt Syndrome includes,

- Loss of vision cause by eye damage
- Corneal ulcer and Infections
- Due to nerves growing abnormal reactions to facial movements
- Facial weakness
- Drowsiness
- Headache
- Change in taste
- Limb weakness^[9].

Causes and Risk factors

The Varicella-Zoster Virus (VZV) is what causes Ramsay Hunt Syndrome (RHS). The individual who has chickenpox as a child continues to have this virus in a healthy state for decades. The Ramsay Hunt Syndrome is brought on by the virus's many effects on the facial nerves and reactivation of those nerves^[10].

Risk Factors

- Person preliminary suffering from chicken pox.
- Patient older than 60 years.
- Pursuing with weak or compromised immune system.

Evaluation

Routine testing is not a factor in the diagnosis of Ramsay Hunt syndrome because it is based on the clinical history and physical examination. Rash, discomfort, and facial droop are pathognomonic symptoms of the illness. The damaged facial nerve should show signs of inflammation on magnetic resonance imaging (MRI), although computed tomography typically

won't help with the diagnosis and the MRI isn't necessarily necessary^[26].

When accessible, electrodiagnostic testing such as electroneurography (ENoG) and electromyography (EMG) may provide useful prognostic information by quantifying the extent of nerve damage more precisely than is possible with a physical examination alone, predominantly in the case of House-Brackman grade VI paralysis^[27,28].

Diagnosis

The history and neurological examination remain the same for diagnosing Ramsay Hunt syndrome. Examination of CSF and gadolinium enhanced MRIs have no diagnostic or prognostic value^[29].

The Varicella Zoster Virus (VZV) Polymerase Chain Reaction (PCR) to identify it in transude from the geniculate zone of the ear is more sensitive than the Varicella Zoster Virus (VZV) PCR to detect it in tears or blood mononuclear cells. In the same study, it was discovered that 2/3 (71%) of the patients had PCR positive ear exudates prior to the emergence of vesicles. The geniculate zone (VZV) Varicella Zoster Virus Polymerase Chain Reaction (PCR) may be used to discriminate between Bell's palsy and early Ramsay Hunt syndrome patients, while more research is required^[30].

Ramsay Hunt syndrome is frequently diagnosed by a doctor based on a patient's medical history, physical examination results, and the syndrome's characteristic signs and symptoms. A sample of fluid from one of the rash blisters in your ear may be taken by the doctor for testing in order to confirm the diagnosis^[31].

Treatment

Efficient treatment of Ramsay Hunt Syndrome can ease pain and decreases the risk of long-term complications. Medications can include:

- **Corticosteroids:** A short administration of high dose prednisone appears to increase the effect of antiviral drugs in Ramsay Hunt Syndrome.
- **Anti-anxiety medications:** Drugs such as diazepam (Valium) help in relieving vertigo.
- **Antiviral drugs:** Medications like acyclovir (Zovirax), Famciclovir (Famvir) or Valacyclovir (Valtrex) often help cure the chickenpox virus.
- **Pain relievers:** The pain co-related with Ramsay Hunt Syndrome (RHS) can be severe.

Prescription of pain medications must be needed.

➤ Lifestyle and Home remedies:

The following can help decrease the discomfort of Ramsay Hunt Syndrome:

-Affected area (by the rash) should be maintained clean.

-Application of cool or wet compresses to the rash can ease the pain.

-Take an over-the-counter (OTC) medication or anti-inflammatory drug, such as ibuprofen (Advil, Motrin IB, Others)^[36].

HOME REMEDIES:

➤ Green Nerve Repair Juice

Content-Italian cucumber (1)

English cucumber (1/2)

Head of celery (1)

Med apples (3)

Chunk of ginger (1)

Peeled lemon (1)

Juice from cucumber or celery can be used as a source of favourable copper. Similar to many other important minerals, copper enhances health by triggering cell-based enzymes. Copper-dependent enzymes promote immune system and neuron function in addition to fuelling your lifestyle by assisting in energy production.

A diet high in both copper and iron helps prevent anaemia and supports the health of red blood cells. Fresh cucumber juice has 102 micrograms of copper in a serving size of 1cup. Celery stems are a great source of vitamins B1, B2, B6 and C as well as potassium, folic acid, calcium, magnesium, iron, phosphorus, sodium, and vital amino acids. Celery leaves are high in vitamin A.

For those with bell's palsy and Ramsay Hunt Syndrome, B vitamins are crucial.

Many B vitamins are necessary for the healthy operation of the nervous system, so adding a basic B- Complex vitamin to your daily regimen may be a good idea while you're recovering. Some Bs that could be especially useful include:

B1- may prevent muscular Atrophy and improve circulation(circulation is diminished in inactive muscles).

B6- Helps produce the amino acids that are necessary for the production of new cells.

B12- when taken as part of a "B-complex" vitamin, vitamin B12 may aid to lessen inflammation and boost the immune system.Methyl cobalamin is a kind of vitamin B12 that is not

found in the essential B-complex vitamins, is crucial for development of nerve cells.

Stones removal and prevention: Celery juice is a fantastic toxin remover from the body, helping to break up and flush out gall-bladder and urinary stones. This delightful drink prevents persons who have previously had stones from developing them^[37].



Fig.4: Green Nerve Repair Juice

Vitamin B	for nervous system
Vitamin B1	Prevent muscular atrophy
Vitamin B6	Production of new cells
Vitamin B12	Boost immune system

Ramsay Hunt Syndrome treated with Acyclovir

➤ Case Reports

1. A 40-year-old woman came in with painful, two-day-old vesicular lesions on her right ear. She had lost the capacity to close her right eye, developed facial asymmetry, and started drooling liquids from her right angle of the mouth one day before being admitted. She also described a day of uncomfortable solid food swallowing in her past. Additionally, there was a history that would have pointed to tinnitus and impaired hearing in the right ear. Clinical examination vital indicators were within normal limits. Her right pinna and external auditory meatus both had vesicular lesions, according to a central nervous system (CNS) examination (Fig 5). Tympanic membrane on the right was undamaged. In the right ear, there was a corresponding sensory hearing loss. Ulcers were found on her soft palate, throat, and right facial pillar during an oral cavity examination (Fig 6). Complete right infranuclear facial palsy was also

seen (Fig 7). The Ramsay Hunt syndrome was diagnosed clinically. Investigations revealed normal biochemical and haematological markers. HIV testing came back negative.



fig.5: Vesicular lesions right pinna Fig.6: Ulcers on soft palate



Fig7: Right intranuclear facial palsy

Within 48 hours of the onset of symptoms, oral acyclovir 800 mg, taken five times daily, was started as a treatment. Along with supportive care, this was administered for seven days. Over the next 48 hours, she displayed a good response, with lesions involving the pinna, external auditory meatus, and oral cavity healing (Fig 8). She was monitored for a year and throughout that time her facial nerve function had improved. At the conclusion of her follow-up, she exhibited very little residual facial palsy, manifested as a small

asymmetry in her facial motions. She didn't have hearing loss or post-herpetic neuralgia, though^[38].

2. A famous Canadian pop-singer Justin Bieber has condition of Ramsay Hunt Syndrome, which is a rare condition at the age of 28. Affects left side of the face and ear, his eye is not blinking (fig 8).

He was started a treatment of Acyclovir by parenteral route of drug administration, within 24 hr. for a week after diagnosing and due to facial exercise, now he is completely recovered.

he will be taking time to rest and recover in order to return to his music career^[39].



Fig.8: Justin Bieber Facial palsy

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