

RAMSAY HUNT SYNDROME

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فرع التشخيص الفمي / طب الفم



OBJECTIVES:

1-Identify the clinical signs and symptoms of Ramsay Hunt syndrome.

2- Review the evaluation of patients with suspected Ramsay Hunt syndrome.

3-Outline the management options available for Ramsay Hunt syndrome.





WHAT IS RAMSAY HUNT SYNDROME?

Ramsay Hunt syndrome

also known as herpes zoster oticus or geniculate ganglion herpes zoster, is a late complication of varicella-zoster virus (VZV) infection, resulting in inflammation of the geniculate ganglion of cranial nerve VII. The syndrome is named after James Ramsay Hunt (1872-1937), an American neurologist and Army officer in World War I who described three different syndromes, the most famous of which is the second, which is discussed here as "Ramsay Hunt syndrome.

- Early stages of VZV infection cause fever and diffuse vesicular rash, a condition that is commonly referred to as chickenpox. After the initial infection, the virus will often remain dormant in the body.
- Subsequent reactivation of the virus causes a "zoster" or "herpes zoster" phenomenon. This syndrome consists of pain and a vesicular rash along the involved nerve's distribution, typically corresponding to a single dermatome. The distribution and associated symptoms depend on the nerve involved. Less than 1% of zoster cases involve the facial nerve and result in Ramsay Hunt syndrome.

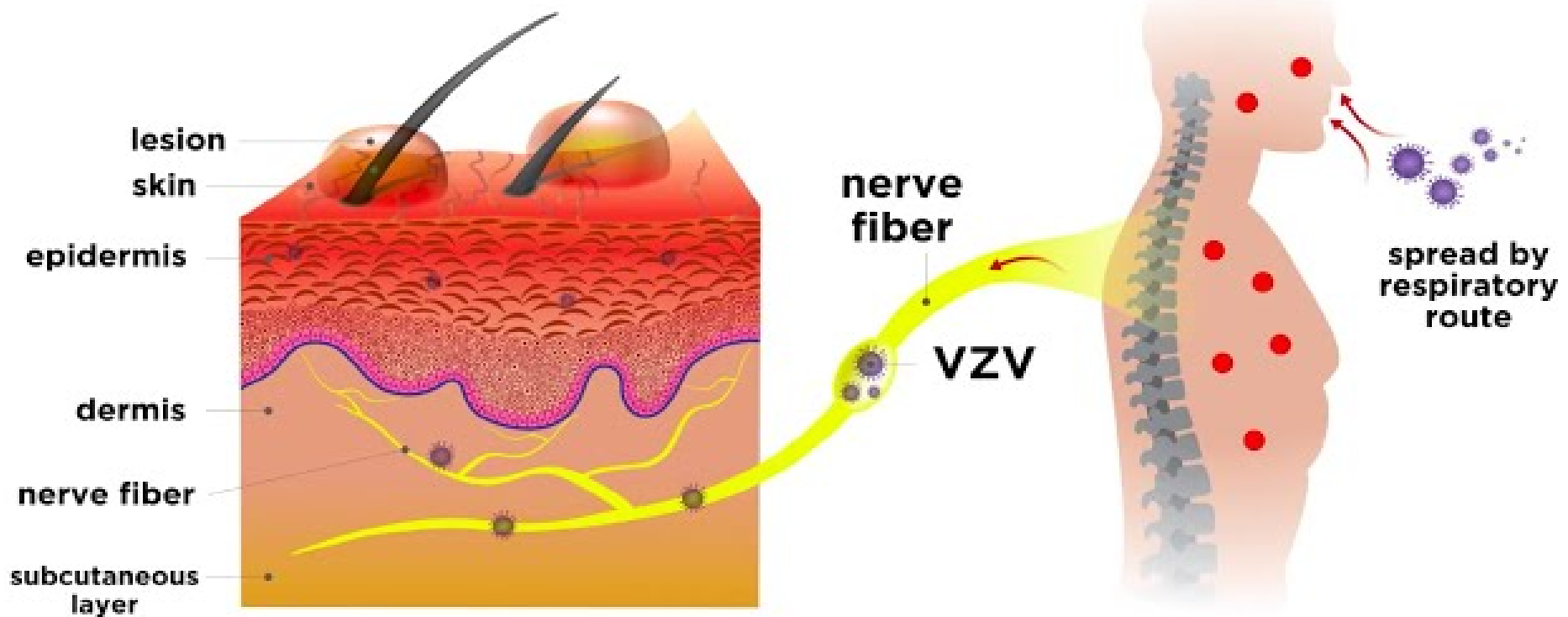
ETIOLOGY

- The causative agent in Ramsay Hunt syndrome is the varicella-zoster virus, a member of the human herpes virus family. More specifically, it is part of the alpha herpes virinae subfamily, along with herpes simplex viruses 1 and 2 (HHV-1 and HHV-2). VZV is a double-stranded DNA virus, more technically known as human alpha herpes virus 3 (HHV-3).



Varicella-Zoster Virus (VZV)

(dissimilar to HSV in method of transmission)



- Once the clinical VZV infection, chickenpox, has cleared, the virus remains latent in cranial nerves 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.

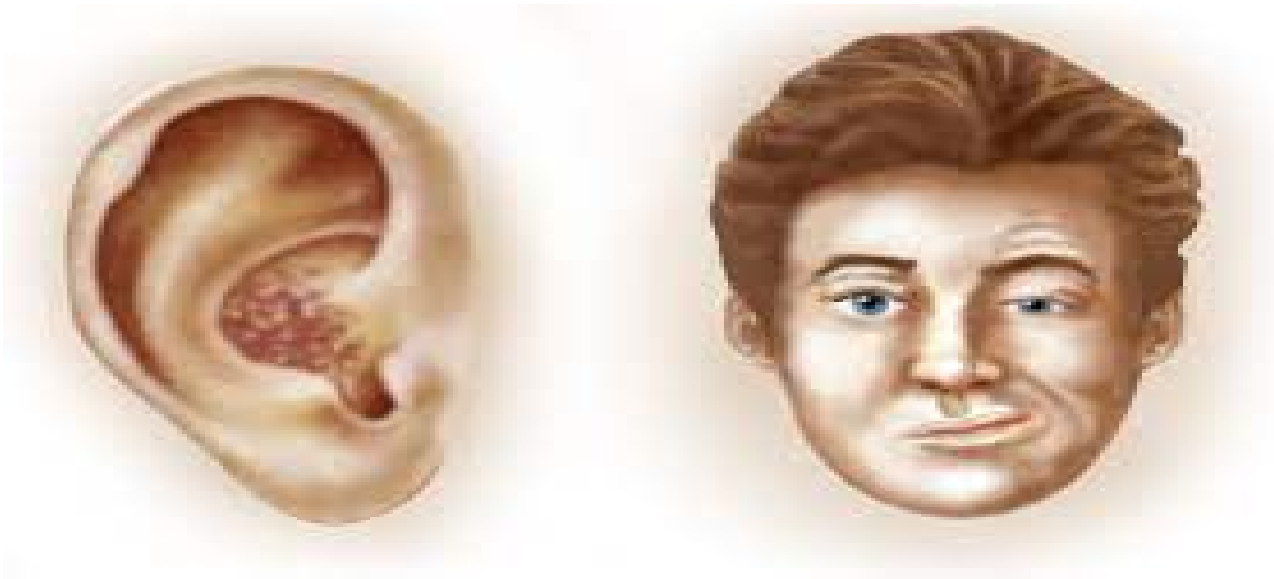


EPIDEMIOLOGY

Ramsay Hunt syndrome can occur in anyone who has had **chickenpox**. It may affect individuals of any age group, but people in the seventh and eighth decades are more susceptible to it, and it is rare in children. Ramsay Hunt syndrome is estimated to account for **16%** of all causes of unilateral facial palsies in children and **18%** in adults. It is also thought to be the cause of as many as **20%** of clinically diagnosed cases of Bell's Palsy. About **7%** of cases of Ramsay Hunt syndrome cause acute facial paralysis.

Signs and symptoms

- Ramsay Hunt patients typically present with the classic triad of
- ipsilateral facial paralysis,
- otalgia, and
- painful vesicles on the auricle



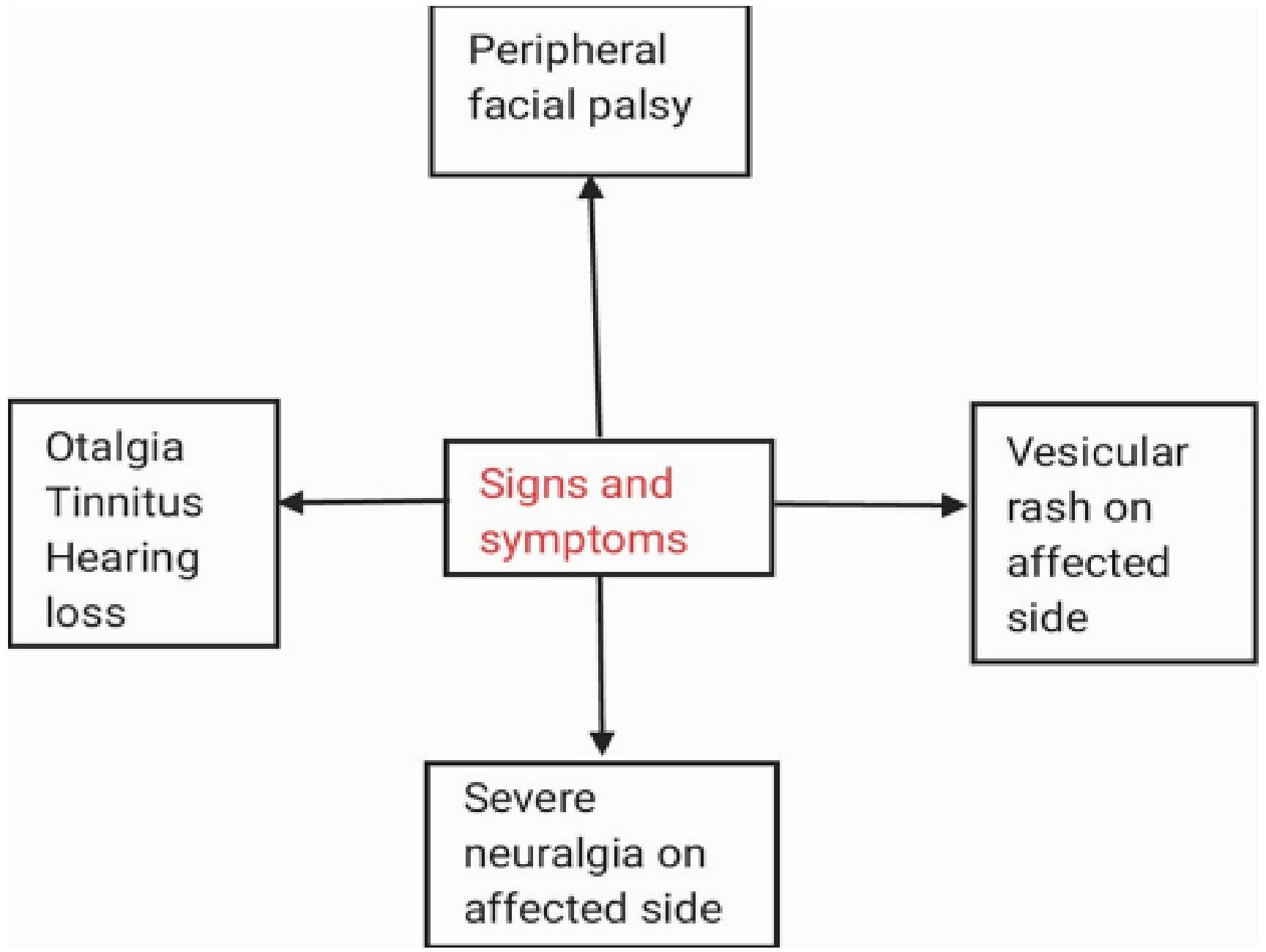
Peripheral
facial palsy

Otalgia
Tinnitus
Hearing
loss

Signs and
symptoms

Vesicular
rash on
affected
side

Severe
neuralgia on
affected side



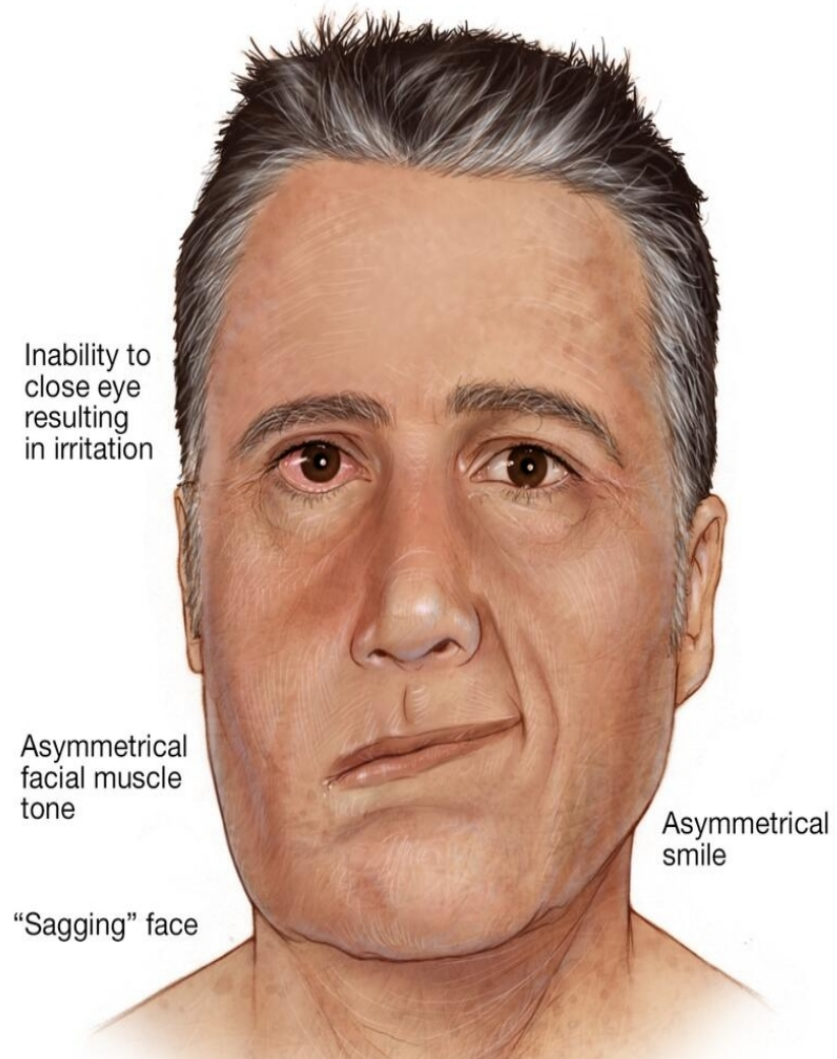
- however, patients who present early in the course of the disease may only have pain without facial paralysis or a rash. Notably, the characteristic rash correlates to the areas innervated by the facial nerve; on the auricle, this includes the conchal bowl, anti-helix, and post auricular sulcus, but the rash can involve the auditory canal, scalp, cheek, tongue, or palate.



- Hunt also contributed to findings such as the severe neuralgias experienced deep in the face scattering to the ear on the affected site,
- and it also affected the tear and saliva secretion alongside the nasal congestion.
- Changes in taste,
- dry eyes,
- hyperacusis,
- weeping,
- dysarthria, and nasal obstruction are a few uncommon symptoms.
- Most of the time, only one side of the face is affected, causing weakness in the facial muscles of that side.



- It presents as the patient's inability to smile. The severity of this only increases over time, usually a week or two after the onset. This is evident by the dribbling of saliva from the corner of the mouth and the inability to close the eyes later, causing irritation and blurring of vision



- The forehead on the affected side is smoother than the non-affected side, which has creases and folds .
- Since the orbicularis oris muscle is also paralyzed, the person cannot close the eyes, increasing the distance between the upper and lower eyelid.
- The lip axis is tilted towards the non-affected side of the face.
- Facial asymmetry is visible, difficulty in speaking develops, and the flow of secretions such as saliva and tears are reduced



- The rash is red and vesicular, filled with fluid, and associated with pain affecting the anterior portion of the pinna and the outer one-third of the external auditory canal. The rash can also be seen on some patients' mouths and soft palates. Other symptoms affecting the ear are otalgia, tinnitus, and hearing loss, which are usually temporary but can also be permanent in some rare cases



- Isolated lesions on the external ear in 40.6% of patients
- combined involvement of the outer ear and external ear and external auditory canal in 25.3%
- Both dermatological and neurological findings lead to the diagnosis of Ramsay Hunt syndrome.
- The facial nerve is predominantly a motor nerve innervating facial muscles with only a few sensory nerve fibers. Apart from facial paralysis, facial nerve palsy may also present with paralysis of the stapedius muscle of the middle ear, causing hyperacusis
- If the early presenting clinical features worsen, the probability of cranial nerve damage being permanent increases



- Ramsay Hunt syndrome is known to be a non-contagious infection. But the virus can reactivate, causing chickenpox in people who have not been vaccinated against it or who have not yet had it; thus, people are advised to avoid contact with such patients until the rash scabs off.




DIFFERENTIAL DIAGNOSES

- Bell Palsy
- Persistent Idiopathic Facial Pain
- Post herpetic Neuralgia
- Trigeminal Neuralgia
- Temporomandibular Disorders



PATHOPHYSIOLOGY

- In Ramsay Hunt syndrome, the virus lays dormant within the geniculate ganglion and primarily reactivates along the facial nerve, the initial presenting symptom is typically pain in the ipsilateral ear (55% of patients), with facial paralysis and vesicles appearing within 2 to 3 days. In 23% of patients, facial paralysis is the presenting symptom, and in only 2%, vesicles appear first. While 86% of their patients reported that the rash occurred only on the auricle, 7% only had vesicles in the oral cavity, and 8% had them in both locations.
 - Rashes have also been reported on the scalp and the cheek.
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PATHOPHYSIOLOGY

- The proximity of the facial nerve to the vestibulocochlear nerve can result in hearing loss, tinnitus, and vertigo. Sensorineural hearing loss was present in 43% of patients in Coulson's series, imbalance or vertigo in 51%, and tinnitus in 20%.
- Vagal nerve involvement is also probably more common than it appears to be. Unless the patient is symptomatic with hoarseness or aspiration, vocal cord paralysis is not usually noted because it requires mirror or fiberoptic laryngoscopy to discover. Although less frequent, other cranial nerves that can be involved include the trigeminal, glossopharyngeal, and hypoglossal nerves..



- Cranial polyneuropathy is more likely to present in immunocompromised patients, such as those with diabetes mellitus or human immunodeficiency virus infection.
- The facial paralysis resulting from Ramsay Hunt syndrome has a worse prognosis than that seen in Bell's palsy, with only 70% regaining normal or near-normal facial function compared with over 90% in Bell's palsy.

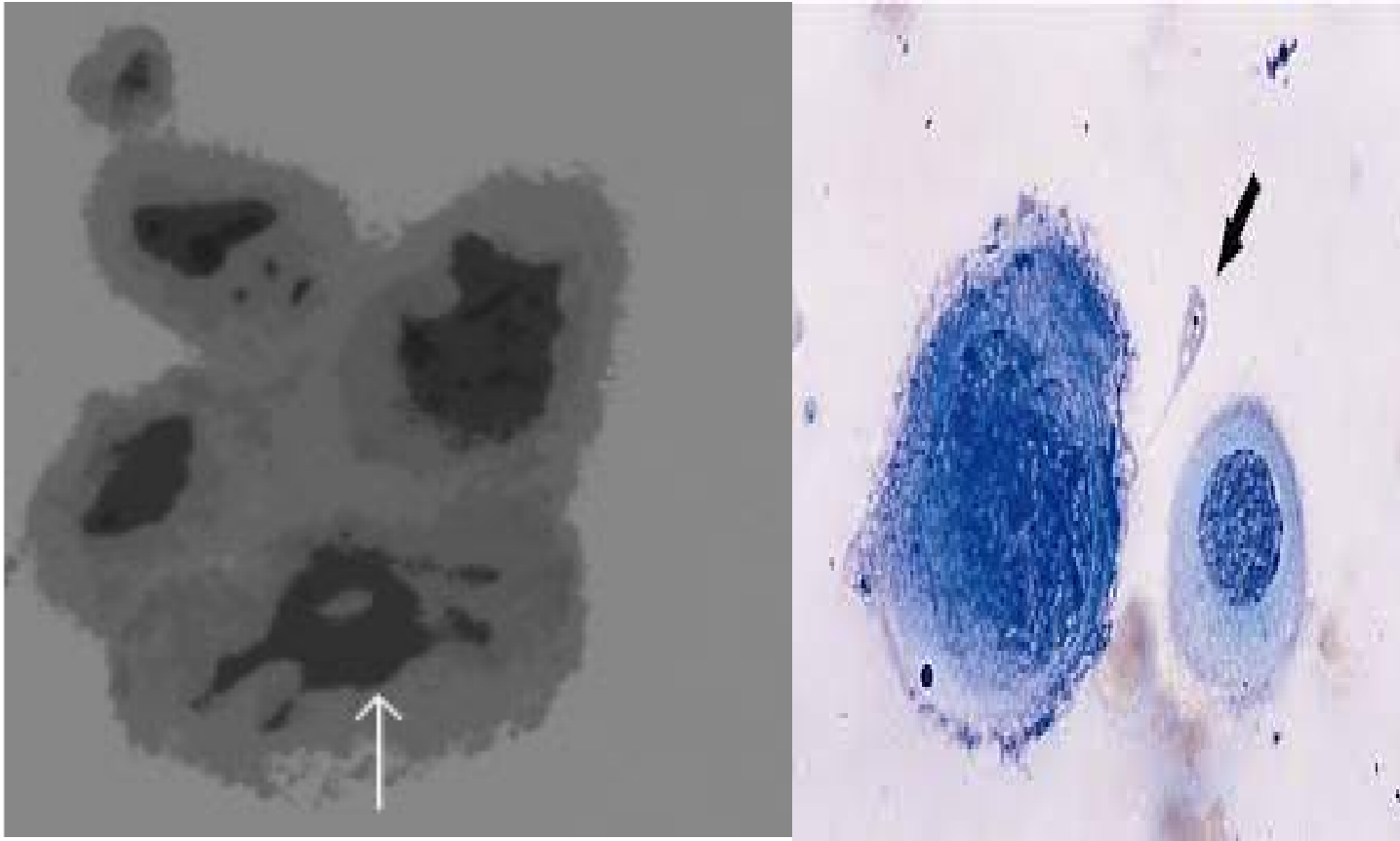


HISTOPATHOLOGY


- Microscopic evaluation with the Tzanck smear technique can be performed on the fluid obtained from the vesicles. This should reveal multinucleated giant cells using a Giemsa stain, methylene blue, or Wright's stain. The sensitivity of the test is low, but it has a high specificity when pre-test clinical suspicion is high. Distinguishing between herpes simplex, varicella-zoster, and cytomegalovirus infections can be challenging because they are all herpes viruses; the Tzanck smear is also used to identify pemphigus vulgaris, leprosy, and leishmaniasis.



MULTINUCLEATED GIANT CELLS ON TZANCK SMEAR



EVALUATION

- Routine testing is not usually recommended as the diagnosis of Ramsay Hunt syndrome is typically made based on **clinical history** and **physical examination** . Ramsay Hunt syndrome's typical features include rash, pain, and facial drooping.
 - A Tzanck smear can be performed on fluid obtained from the vesicles,
 - and PCR analysis of **tears, saliva, or fluid from the vesicles** is available in some academic settings .
 - The degree of cranial nerve involvement can be determined by investigations like audiometry, vestibular testing, and flexible fiberoptic laryngoscopy
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TREATMENT AND PREVENTION

- It heals in a self-automated way; the aims of the treatment are for us to reduce the occurrence of complications such as facial paralysis and post-herpetic neuralgia, to provide immediate relief from the pain and inflammation, and to prevent exposure to keratopathy of the cornea of the affected side as the patient is unable to close the eye



- ocular ointments have been valuable in keeping the eye lubricated, artificial tears, and wearing an eye patch.
- Clinically it has been seen over the years that preliminary treatment and good prognosis prevent post-herpetic neuralgia because the elderly population is more susceptible to being immunocompromised; hence an aggressive approach is advised for the treatment



- Capsaicin is United States Food and Drug Administration (FDA) approved for the neuropathic pain associated with post-herpetic neuralgia .
- oral antivirals and steroids effectively reduce late complications such as acyclovir, valacyclovir, and famciclovir.
- The treatment is continued for up to a week or 10 days. that delayed degeneration of the facial nerve axon took up to three weeks, antiviral therapy is preferred to be continued till the given time. The potency of antiviral treatment is disputable when it comes to the paresis of the cranial nerve .




- Corticosteroids are administered in high doses, at the same time, for three weeks. Oral prednisone 60 mg is given daily for three weeks and is stopped gradually and not suddenly to avoid acute adrenal complications. Corticosteroids are known to have various side effects. Side effects include irritability, gastric reflux, insomnia, hyperglycemia, etc.
- Corticosteroids and their value can be illustrated by relief in edema and by decompression of the neurogenic structures that are present in the facial nerve canal in the petrosal bone



- Another modality for corticosteroid treatment is via intra tympanic injection. This dehiscence allows for topical administration of a corticosteroid, sparing the patient from the adverse side effects associated with systemic steroids.



- Further treatment is aimed at providing symptomatic relief from pain and rashes;
 - **carbamazepine** is given to prevent seizures.
 - **anticholinergics and**
 - **antihistamines** are given to suppress vertigo, and
 - **diazepam** also helps relieve pain and dizziness.
 - Patients with risk factors like hypertension, diabetes mellitus, old age, and some other immunocompromised states need to be careful as they may take more time to get better and should be advised to keep their eyelid weighted on the affected side to ease the closure of the eye and prevent exposure keratopathy during treatment . Corneal hypesthesia is an indication of weight placement on the eyelid of the affected side .
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- Surgical procedures are not helpful in patients with Ramsay Hunt syndrome.
- Prevention from the VZV can be achieved by administering a vaccine against chickenpox in childhood, or if someone is 50 years or older, they are advised to get the shingles vaccine. These vaccines are remarkably effective against the VZV, which would, in turn, prevent the infection causing the Ramsay Hunt



- the management of synkinesis can be accomplished with both conservative and surgical approaches.
- Conservative approaches include massage and physical therapy as well as chemodenervation with botulinum toxin.
- Surgical management of synkinesis may involve selective neurectomy and/or myomectomy, to improve smile symmetry.



Prognosis

- Most patients recover from Ramsay Hunt syndrome, but the degree to which their recovery occurs varies from patient to patient . For patients who do not recover, their premorbid function may lead to complications like synkinesis . In general, the prognosis for Ramsay Hunt syndrome is worse than that for Bell's Palsy, as it has a lower rate of synkinesis development . The severity of facial paralysis decides the prognosis of Ramsay Hunt syndrome. However, age over 50, greater axonal damage, presence of oropharyngeal lesions, multiple cranial neuropathies, and diabetes are some other factors that may influence the prognosis of a patient .
- Besides synkinesis, a common complication of Ramsay Hunt syndrome is post-herpetic neuralgia, in which pain persists for longer than three months after the onset and is more likely to develop in patients older than 50 years

complications

- Other than the presenting symptoms of pain, rash, facial paralysis, dysgeusia, hearing loss, tinnitus, vertigo, hoarseness, dysarthria, and others mentioned above, short-term complications of Ramsay Hunt syndrome include corneal abrasion and exposure keratopathy,
- the development of synkinesis is very common.
- Other long-term complications include postherpetic neuralgia, scarring from the vesicles,
- depression and social anxiety, due to loss of facial function



Other complications

- Aseptic meningitis
- Preipheral motor neuropathy
- Myelitis
- Encephalitis
- Gullian-Barre syndrom
- Bacterial infection





Thank you