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Review

Ramsey Hunt Syndrome: A Review Article

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Abstract

Ramsey Hunt Syndrome (RHS) is a disease caused by the varicella-zoster virus (VZV). After first encountering VZV infection, the virus persists in the sensory ganglia of the spinal and cranial nerves for life. Herpes zoster oticus is a condition characterized by severe otalgia and erythematous vesicular rash in the external ear canal and auricle, caused by viruses that reactivate in the facial nerve and geniculate ganglion, especially in people with weakened immune systems due to various diseases or drugs. The picture formed by the addition of peripheral facial paralysis to the table is called RHS. Other cranial nerves may also be involved through various anatomical connections. Antiviral agents and corticosteroids are useful in treatment. Early diagnosis and treatment are important.

Keywords: Facial paralysis, shingles, otalgia

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Ramsey Hunt Syndrome is a disease caused by the varicella zoster virus (VZV). When VZV first enters the body, it causes chickenpox, an infectious disease characterized by itchy vesicular eruptions, usually accompanied by fever. After VZV infection is encountered for the first time, the virus remains in the sensory ganglia of the spinal and cranial nerves for life.[1] Herpes zoster infection, known as shingles, occurs due to the reactivation of the dormant virus for various reasons. Elderly patients are at risk, especially due to the weakening of the immune system due to aging, but it can affect people of any age whose immune system is weakened due to various diseases or drugs.[2] Painful vesicular eruptions occur unilaterally along one or two adjacent dermatomes in this shingles disease. Severe otalgia and erythematous vesicular rash in the external auditory canal and auricle caused by viruses reactivated in the facial nerve and geniculate ganglion is called herpes zoster oticus.[2] This disease is defined Ramsey Hunt Syndrome with the addition of ipsilateral facial paralysis to the disease by the neurologist James Ramsey Hunt.[3,4]

Etiology and Epidemiology

VZV is a neurotropic human herpes virus belonging to the alpha herpesviridae family.[5] It can be transmitted from person to person by droplets in the airway or by direct contact with vesicular lesions.[6] After primary infection, the virus remains latent in the cranial nerves or dorsal root ganglia, then reactivates in situations of physiological stress or immunodeficiency, leading to herpes zoster known as "shingles" elsewhere in the body.[5,7] Persons with reduced cell-mediated immunity resulting from carcinoma, radiation therapy, chemotherapy, or human immunodeficiency virus (HIV) infection are particularly at risk.[8] However, it can also rarely be seen in immune-competent patients.[8] The thoracic segment is involved in the majority of herpes zoster cases.[2] It has been reported that the head and neck involvement of herpes zoster is 13-35%.[1,9] In the literature, the frequency of peripheral facial paralysis in herpes zoster oticus has been reported as 96%.[9]

The annual incidence of Ramsey Hunt Syndrome is approximately 5 per 100,000 people.[7,10] Herpes Zoster is a disease

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seen mostly in elderly people (>60 years).[8] It is stated that Ramsey Hunt Syndrome constitutes 7-12% of all peripheral facial paralysis.[11,12]

Clinical Findings

In herpes zoster oticus, vesicular eruptions can be found in the eardrum, external ear canal, or auricle.[1] In a study examining the localization of herpes zoster oticus eruptions, a rash was found in the isolated external ear in 40.6% of the cases, and in both the external ear canal and external ear in 25.3%.[13] The classical triad of Ramsey Hunt Syndrome is otalgia, vesicular eruptions in the external auditory canal or auricle, and ipsilateral facial paralysis.[14] In addition, vesicular eruptions can be seen on the tongue or palate mucosa on the affected side.[15] Depending on facial nerve involvement; On the affected side, decrease in forehead lines, low corner of the mouth, effacement in the nasolabial groove, decrease or absence of movement on the affected side in voluntary mimic movements, disturbance in the sense of taste, dry eyes can be seen. Hyperacusis may be seen due to the absence of the stapes reflex.[9] A decrease in tear and saliva secretion can be observed on the affected side.[9] There are complex anastomoses between the cranial nerves that allow the herpetic lesions to progress further, and therefore other cranial nerve involvements may be seen.[1] Vestibulocochlear nerve involvement can be seen in 40-50% of patients.[9] Hearing loss, tinnitus, and vertigo may be seen in vestibulocochlear nerve involvement.[16] Hoarseness or aspiration may be seen in vagus nerve involvement.[7] The related region may have sensory and pain problems due to trigeminal nerve involvement.[9] In the study of Kim et al., the rate of polyneuropathy was found to be 1.8% among all cases with Ramsey Hunt Syndrome.[12] In the same study, the order of cranial nerves involved in polyneuropathy cases from most to least was determined as VII, VIII, IX, X, V, and III/XII.[12] The severity of the disease depends on the cellular immunity of the patient.[8]

Other causes of peripheral facial paralysis can be counted as otitis media, local trauma to the facial nerve, post-surgical complications, neoplasms, and sarcoidosis.[17] In addition, Lyme disease may cause peripheral facial paralysis in endemic areas.[17] Ramsey Hunt syndrome can be separated from these other causes with its typical findings. The diagnosis of RHS is made clinically, but in some rare cases where skin lesions are not apparent, serological tests for VZV antibodies can be used or viral antigen detection can be performed by molecular genetic methods in an oral swab.[15]

Complications include corneal ulceration and post-herpetic neuralgia.[8] Apart from these, meningoencephalitis, myelitis, cerebral vasculopathy, paresis, and paralysis can be counted.[9]

Treatment and Prognosis

Herpetic facial paralysis generally has a worse prognosis than idiopathic facial paralysis.[18] Compared to Bell's palsy, RHS has more severe facial paralysis at the beginning.[15] It has been reported that Ramsey Hunt Syndrome is associated with severe sequelae.[12] The recovery rate of hearing loss due to cochlear nerve involvement in Ramsey Hunt Syndrome was found to be 11.1%.[12] In the treatment of RHS, intravenous administration of acyclovir in combination with corticosteroids has been recommended.[9] In another study, it is stated that acyclovir can be administered orally or intravenously.[1] It is stated that treatment with prednisone and antiviral agents may improve the outcome.[15] The addition of acyclovir to the treatment was found to be significantly more beneficial on facial nerve functions when compared to corticosteroid treatment alone.[19]

Corticosteroids have a positive effect on the treatment by reducing inflammatory edema and decompression of neurogenic structures in the facial nerve canal in the petrosal bone.[9] Generally, in adults with facial paralysis, corticosteroids started within 3 days of the onset of paralysis increase the likelihood of recovery, shorten the recovery period, and reduce synkinesis.[17] It has been reported that corticosteroids will reduce the severity of residual paralysis in adults who do not fully recover.[17] In the same study, it was recommended to use adult data for children with isolated facial paralysis.[17] Although the benefit of steroids is uncertain after the third day of paralysis, it is recommended to be given due to the possibility of benefit.[17,20] In the study of Kim et al. on Ramsey Hunt Syndrome cases with polyneuropathy, it was reported that the combination with 60 mg prednisolone (or 48 mg methylprednisolone) for 2 weeks and 1500 to 1800 mg acyclovir for 1 week was sufficient for treatment success.[12] It is stated that it is beneficial to start combined acyclovir-corticosteroid therapy as early as possible.[9]

Symptomatic treatments can be given for additional complaints such as pain and dizziness. Surgical treatment of peripheral facial paralysis of acute viral origins, such as Bell's palsy and Ramsay Hunt syndrome, is facial nerve decompression, although it is rarely used.[12] If rapid diagnosis and treatment are not started for Ramsey Hunt Syndrome, the chance of complete recovery will decrease.[11]

Conclusion

RHS is the condition where facıal paralıs is added to the table of herpes zoster oticus. In addition to facial paralysis, various anatomical connections may also affect other cranial nerves. Vesicular eruptions can isolate in the exter-

nal ear canal and tympanic membrane, even if they are not seen outside the ear. Treatment with corticosteroids and antiviral agents should be initiated promptly after diagnosis. Early diagnosis and treatment are important.

Disclosures

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