The Pathogenesis of Vestibule Cochlear Nerve Disease in Herpes Zoster Oticus

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Introduction

In 1907, James Ramsay Hunt first postulated that the etiology of herpes zoster oticus was the reactivation of the varicella zoster virus (VZV) in the geniculate ganglion [1]. The strict definition of the Ramsay Hunt syndrome is peripheral facial nerve palsy accompanied by an erythematous vesicular rash on the ear (zoster oticus) or in the mouth. 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. He explained these eighth nerve features by the close proximity of the geniculate ganglion to the vestibule cochlear nerve within the bony facial canal [2].

In patients with herpes zoster oticus, disorders of the eighth cranial nerve are frequently observed. Hearing loss is usually mild to moderate, and the audio logical data suggest cochlear and/or retro cochlear involvement [3]. The vestibular symptoms are sometimes severe, and a study showed that both the superior and inferior divisions of the vestibular nerve could be widely affected [4]. Kuhweide et al. reported that the infection may spread to the cochlea vestibular system through the vestibulofacial communicating branch or via perineural tissues within the internal auditory canal [5]. Evidence of inflammation within the auditory and vestibular nerves and the labyrinth has been noted on post-mortem examinations of patients with herpes zoster oticus [6]. Murakami et al., also reported that the presence of VZV in the middle ear mucosa of patients with herpes zoster oticus suggested the arrival of VZV in the labyrinth through dehiscence of the facial nerve canal, creating a passage through the oval and/or round window [7].

The diagnosis is usually made based on history and physical examination. Enhancement of the seventh and eighth cranial nerves on gadolinium-enhanced, T1-weighted MRI can be observed. Hemorrhage of the cochlear nerve and destruction of the apex of the organ of Corti have been reported. This study focused on the audio logical findings by pure tone audiometry and impedance audiometry of patients with herpes zoster oticus. The findings were compared with the clinical severity of facial paralysis and patient age to clarify the pathogenesis of vestibule cochlear nerve disease in herpes zoster oticus.

Materials and Methods

To establish a relationship between adverse affect of Herpes Zoster Oticus and Hearing system, we collected data of pure tone audiometry (PTA) and Impedance Audiometry of 60
patients with herpes zoster oticus, who had visited ASCOMS for a hearing checkup over past 2 years. The patients who had prior histories of hearing impairment or vertigo due to other causes were excluded from this study. The patients were classified according to severity of facial paralysis, based on the system proposed by House and Brackmann. Patients with symptoms of cochlear dysfunction (hearing impairment, tinnitus, and ear fullness) underwent audio logical tests. Pure tone audiometry was conducted at frequencies of 0.25, 0.5, 1, 2, 4, and 8 kHz. Acoustic thresholds of the affected ears were compared with those of the contra lateral (unaffected) ears at each frequency level.

**Results**

Sixty patients were included in the study (33 men and 27 women; age ranges 30 to 60 years). Thirty of those patients had complaint of cochlear symptoms: hearing loss, ear fullness, tinnitus, or hyperacusis. None of the patients had spontaneous or positional nystagmus on physical examination throughout hospitalization. The mean acoustic thresholds and standard errors at 0.25, 0.5, 1, 2, 4, and 8 kHz in the affected ear were calculated to be compared with those of the unaffected ear (Figure 1). The mean acoustic threshold at all frequencies showed a statistical difference between the affected ear and the normal ear. The difference of mean acoustic threshold between the affected ear and the unaffected ear increased as examination frequency was made higher.

![Figure 1: The mean acoustic threshold and standard errors of affected ears and unaffected ears of patients with herpes zoster oticus.](image)

The mean acoustic threshold at all frequencies examined exhibited a statistically significant difference between the affected ear and the unaffected ear. The difference in the mean acoustic threshold between the two increased in parallel with frequency. More severe hearing loss was evident at high frequencies (4 kHz and 8 kHz) in the affected ear. Among 30 patients with cochlear symptoms, 22 patients were male, and 8 were female. There were no significant differences noted with regard to gender and severity of facial paralysis. All the patients were undergone through impedance audiometry. There was no significant results obtained but there was elevation in both ipsi and contra lateral acoustic reflexes in the affected ear.

**Discussion**

Auditory and vestibular disturbances are common findings in patients with herpes zoster oticus. Many reports concerning vestibule cochlear nerve disease associated with herpes zoster oticus state that the classic syndrome described by Ramsay Hunt is not common and is no doubt a more complicated disease than the original description as “geniculate ganglionitis”. In our study, the incidence of patients complaining of hearing loss was 50%, and the incidence of patients who showed audio logical abnormalities was 60%. Patients with audio logical abnormalities in speech frequency range could detect hearing disturbance, but in cases of isolated high frequency loss, the patients did not have auditory symptoms in general. However, the incidence of hearing loss differs in the cases reported in the literature, ranging from 7%, to 19.7% [8], to as high as 85% [9]. The presence of cochlear and retro cochlear patterns of hearing loss has been reported by various authors.

Wayman et al., [9] in a respective study of 186 patients with herpes zoster oticus-only seven of whom were studied in audio logical detail-recognized one patient with retro cochlear involvement, five with cochlear involvement, and one with both cochlear and retro cochlear involvement. Abramovich and Prasher [10] in another detailed audio logical examination of 13 patients with herpes zoster oticus, used electrocochleography and brainstem evoked potentials in addition to classic audio logical tests. Seven of the 13 patients with aural herpes zoster who demonstrated retro cochlear involvement could be classified as sensory, with some patients showing both sensory and neural signs. However, Kaberos et al., [3] studied 15 patients with herpes zoster oticus through the use of otoacoustic emission in addition to auditory brain-stem response and demonstrated retro cochlear involvement in eight patients. Only three patients had purely retro cochlear hearing loss; the other five patients had concomitant cochlear involvement. Tinnitus was present in 14 patients (67%). Hearing Handicap was found in 86% of patients (of the 21 questionnaire responders & Tinnitus handicap in 57% (of the 14 with tinnitus) [11]. Tinnitus was present in 14 patients (67%).

**Conclusion**

The results of this study support the thesis that viral spread from the geniculate ganglion to the eighth nerve through the per neural sheath, and the connection between nerve fibber bundles in the internal auditory canal, are important factors in vestibule cochlear nerve disease in herpes zoster oticus. Clinical impact of herpes zoster could be magnified by age-related suppression in cell-mediated immunity.

**References**

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