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Journal of Communication Disorders, Deaf Studies & Hearing Aids

Opinion Article

Progressive Hearing Loss and Associated Complexities with Autoimmune Deafness

Anne Chan*

Department of Otolaryngology-Head and Neck Surgery, Rashtrasant Tukadoji Maharaj Nagpur University, Nagpur, Maharashtra, India

DESCRIPTION

Autoimmune deafness, also known as autoimmune inner ear disease (AIED), is a relatively rare condition characterized by progressive hearing loss resulting from an immune system dysfunction. Unlike other forms of hearing loss, which may be caused by factors such as aging, noise exposure, or genetic factors, autoimmune deafness occurs when the immune system mistakenly attacks the inner ear, leading to inflammation and damage to the auditory system. This study, will delve into the causes, symptoms, diagnosis, and treatment options for autoimmune deafness.

Causes and mechanism

The exact cause of autoimmune deafness remains unclear. However, it is believed to be an immune-mediated disorder, where the body's immune system mistakenly identifies components of the inner ear as foreign and attacks them. This immune response leads to inflammation, damage to the delicate structures of the inner ear, and subsequent hearing loss.

It is thought that autoimmune deafness may be triggered by a combination of genetic predisposition, environmental factors, and viral or bacterial infections. Autoimmune disorders such as rheumatoid arthritis, systemic lupus erythematous (SLE), or Hashimoto's thyroiditis have also been associated with an increased risk of developing autoimmune deafness.

Symptoms and diagnosis

The symptoms of autoimmune deafness can vary from person to person. Some common signs include:

Progressive, fluctuating hearing loss: Individuals may experience a gradual decline in hearing ability, which may fluctuate over time. This fluctuation can occur within hours, days, or weeks, making it difficult to pinpoint the exact extent of hearing loss.

Bilateral involvement: Autoimmune deafness typically affects both ears simultaneously, although it can sometimes begin in one ear and progress to the other.

Tinnitus: Many individuals with autoimmune deafness experience tinnitus, a perception of ringing or buzzing sounds in the ears.

Dizziness or vertigo: Some people may also experience episodes of dizziness or vertigo, which can further impact their balance and overall quality of life.

Diagnosing autoimmune deafness can be challenging since it shares symptoms with other forms of hearing loss. A comprehensive evaluation by an audiologist and an otolaryngologist (ear, nose, and throat specialist) is necessary. Diagnostic tests may include pure-tone audiometry, speech audiometry, immittance testing, otoacoustic emissions, and auditory brainstem response (ABR) testing. Additionally, blood tests, imaging studies, and sometimes a biopsy may be conducted to rule out other potential causes and confirm the autoimmune nature of the hearing loss.

Treatment

The primary goal of treating autoimmune deafness is to suppress the immune system's abnormal response and reduce inflammation in the inner ear. The treatment approach may vary depending on the severity and progression of the condition. Here are some common treatment options:

Corticosteroids: Oral or intratympanic corticosteroids, such as prednisone or dexamethasone, are often the first line of treatment for autoimmune deafness. They help to reduce inflammation and stabilize hearing loss. The dosage and duration of treatment are determined by the healthcare provider based on individual needs.

Immunosuppressants: In cases where corticosteroids alone are insufficient, immunosuppressive drugs may be prescribed. Medications like methotrexate, azathioprine, or cyclophosphamide can be used to modulate the immune response and prevent further damage to the inner ear.

Biologic agents: In some instances, biologic agents such as rituximab, which target specific immune cells, may be recommended when other treatments have been ineffective.

Correspondence to: Anne Chan, Department of Otolaryngology-Head and Neck Surgery, Rashtrasant Tukadoji Maharaj Nagpur University, Nagpur, Maharashtra, India, E-mail: annec@gmail.com

Received: 19-May-2023, Manuscript No. JCDSHA-23-24728; **Editor assigned:** 22-May-2023, PreQC No. JCDSHA-23-24728 (PQ); **Reviewed:** 06-Jun-2023, QC No. JCDSHA-23-24728; **Revised:** 13-Jun-2023, Manuscript No. JCDSHA-23-24728 (R); **Published:** 20-Jun-2023, DOI: 10.35248/2375-4427.23.11.252

Citation: Chan A (2023) Progressive Hearing Loss and Associated Complexities with Autoimmune Deafness. J Commun Disord. 11:252.