AUTOIMMUNE HEARING LOSS

Sensorineural Hearing Loss in Autoimmune Disease

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Autoimmune hearing loss, which is a medical emergency, can occur as a distinct condition or as an accompanying feature of other autoimmune disorders.

Discovery

In 1979, the physician McCabe first described a group of patients with sudden sensorineural hearing loss (related to the inner ear) who improved after being treated with corticosteroids. This favorable response to corticosteroids suggested that hearing loss in these patients was caused by an autoimmune mechanism.

Autoimmune Cause

Since, other researchers have elaborated on these findings to create a more concise picture of autoimmune inner ear disorders. Now it is known that the perisacular tissue surrounding the inner ear’s endolymphatic sac contains the necessary components for an autoimmune reaction. Suspected autoimmune triggers include trauma and infectious agents. Autoimmune hearing loss is considered a medical emergency. Without prompt treatment, permanent deafness can result.

Who Is Affected?

Autoimmune hearing loss typically occurs in adults between the ages of 20 and 50 years. Females are affected more often than men, and patients may have another autoimmune disorder or have a prior diagnosis of Meniere’s syndrome. Autoimmune hearing loss is responsible for less than 1 percent of the incidence of hearing loss in the United States.

Symptoms

The hallmark of autoimmune sensorineural or inner ear hearing loss is its rapid, and sometimes fluctuating, progression to hearing loss in both ears occurring over a period of weeks to months. Accompanying symptoms of Meniere syndrome such as vertigo (spinning sensation), generalized imbalance, and ataxia (uncoordination), may also occur. Ear fullness may also accompany the hearing loss. In addition, a ringing, hissing, or roaring sound in the ear may be experienced. With treatment, symptoms of hearing loss can fluctuate and stabilize at a certain level.

In nearly 80 percent of cases, both ears are affected. In patients with hearing loss occurring in one ear, the other ear is often affected at a later time, usually within one
About 50 percent of patients have vestibular symptoms, including disequilibrium, ataxia, and motion intolerance.

**Diagnosis**

There are no specific tests used to diagnose autoimmune hearing loss although the lymphocyte transformation test and Western blot, which are performed at select clinics, are helpful in suggesting autoimmune hearing loss. Blood tests, including a sed rate, CBC, metabolic panel, coagulation tests, lipid levels and thyroid function tests are used to determine if hearing loss has a specific viral or metabolic origin or is associated with another autoimmune condition.

Other causes of autoimmune sensorineural hearing loss such as rheumatoid arthritis, systemic lupus erythematosus, sarcoidosis, Cogan syndrome, and the following conditions of vasculitis: polyarteritis nodosa, Buerger’s disease, polyarteritis nodosa, temporal arteritis, and Wegener granulomatosis, should be ruled out before a diagnose of autoimmune inner ear disease is made.

**Treatment**

Corticosteroids are used to reduce inflammation and are effective in the early stages of inner ear disease. If hearing loss becomes refractory to steroids, patients may be treated with cytotoxic drugs such as cyclophosphamide or methotrexate. In some cases plasmapheresis has been used with good results. In patients unresponsive to medical treatment, surgical treatments such as cochlear implantation or endolymphatic sac decompression are used.

**Resources**


Autoimmune Inner Ear Disease, University of Pennsylvania Health System Balance Center.

Autoimmune Inner Ear disease, American Academy of Otolaryngology, Head and Neck Surgery,


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