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When the immune system identifies the body itself as foreign, it can attack the inner ear.

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What Is Autoimmune Inner Ear Disease (AIED)?

By Divya Chari, MD

Autoimmune disease occurs when the body's natural defense system has difficulty telling the difference between its own cells and foreign cells, causing the body to mistakenly attack normal cells. In the properly functioning immune system, cells and chemical messengers work constantly to patrol the body in search of foreign invaders, such as viruses and bacteria, and attack them once found to protect the body. If the immune system confuses the body's normal cells with foreign cells, it can launch an attack against itself. When this "self-attack" takes place in the ear, it is termed autoimmune inner ear disease (AIED).

The immune system can attack an individual organ (primary) or it can have systemic effects, affecting the entire body (secondary). When AIED is limited to the ear, it is called "primary AIED." In approximately 15-30% of cases, AIED occurs within the context of systemic autoimmune disease, it is called "secondary AIED." [1] Examples of autoimmune disorders that can cause secondary AIED include Cogan's syndrome, granulomatosis with polyangiitis (GPA), sarcoidosis, relapsing polychondritis, rheumatoid arthritis, and systemic lupus erythematosus.

The first report of AIED dates back to 1958, but modern descriptions emerged only 20 years later. [2] The first treatable AIED is credited to Brian McCabe, MD in 1979 who reported a series of 18 patients with features of AIED and described treatment with systemic corticosteroids.[3] McCabe correctly speculated that there was an autoimmune process in the inner ear that resolved after immunosuppression.[4]

WHAT SYMPTOMS OCCUR IN AIED?

AIED causes progressive sensorineural hearing loss (SNHL). Most patients suffer from bilateral disease, but the hearing loss can be asymmetric (i.e. one ear suffers worse hearing than the other). The hearing loss occurs rapidly, often within days to a few months, and sometimes patients experience fluctuating hearing thresholds. [5] Other otologic symptoms such as tinnitus and ear fullness can be present. About one-half of patients with AIED will also experience vestibular symptoms, such as imbalance, motion intolerance, ataxia (difficulty walking), and vertigo (a spinning sensation). [1,6] Some patients with systemic autoimmune disorders (e.g. granulomatosis with polyangiitis) will have a conductive hearing loss (sound cannot get through the outer and middle ear) in addition to a sensorineural hearing loss (SNHL, or damage to the inner ear) due to

disease involvement of the middle ear mucosa and/or Eustachian tube.

HOW COMMON IS AIED?

The incidence and prevalence of AIED are difficult to determine because the disease is rare – the best estimates that we have suggest an annual incidence of AIED to be fewer than five cases per 100,000 and a prevalence of 15/100,000. [7,8] AIED seems to be more common in women than men and is typically diagnosed in adulthood, from the third to sixth decades of life.

HOW IS AIED DIAGNOSED?

Diagnosis of AIED is challenging because there are no standardized diagnostic criteria or reliable tests. The diagnosis is based primarily on clinical symptoms, laboratory tests, and a favorable response to corticosteroids. Laboratory tests may be used to highlight a possible systemic autoimmune condition, but specific markers for primary AIED are not readily available. Other causes of SNHL will need to be ruled out. The early stage of AIED can be diagnostically confusing because SNHL fluctuations can be seen in other otologic disorders (e.g. Meniere's disease, idiopathic sudden hearing loss, tumors). Thus, AIED is a diagnosis of exclusion, and should be suspected in cases of rapidly progressive SNHL, when other etiologic causes have been ruled out. Many clinicians will recommend a brain imaging study to make sure there is nothing else causing the symptoms. What treatment options are available for AIED?

What treatment options are available for AIED?
The mainstay treatment for AIED is administration of corticosteroids. AIED is a potentially reversible cause of SNHL, so prompt medical treatment is essential. If hearing has improved after a 4-week course, it is typically recommended that steroids be continued for about 6 months followed by a gradual taper.

However, some patients do not respond favorably to steroids, and for these patients alternative immunosuppressive treatment has been proposed. Cytotoxic agents (methotrexate, cyclophosphamide, azathioprine), biologic agents (rituximab), and plasmapheresis have been proposed as potential treatments, but the data are mixed about the efficacy of these medications in the treatment of AIED. [6]

AIED is a rare entity and a multi-disciplinary collaboration is often necessary between otolaryngologists and rheumatologists in order to provide appropriate therapy and rehabilitation. Routine audiograms occurring monthly at first if the hearing thresholds fluctuate and then every 6 months to track hearing loss progression. Patients with permanent hearing loss may require assistive devices for auditory rehabilitation. Hearing aids are a reasonable option for patients with mild or moderate hearing loss and cochlear implantation is the most effective rehabilitative strategy for patients with profound and irreversible SNHL.

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