Introduction

Sensorineural hearing loss (SNHL) is a common finding with an exhaustive list of etiologies. Rapidly progressive sensorineural hearing loss is quite distressing to patients as they are abruptly required to function with a reduced level of sensation of their environment. Autoimmune inner ear disease (AIED) is a rare diagnosis and frequently omitted from the differential diagnosis when evaluating patients with rapidly progressive sensorineural hearing loss. This is unfortunate as AIED represents a potentially treatable cause of hearing loss and vestibular symptoms. The goal of this discussion is to acquaint the reader with the clinical presentation, diagnosis and treatment of AIED.

Background

Lenhardt\(^1\) in 1958 hypothesized that anticochlear antibodies were a cause of bilateral SNHL but had no evidence to support this supposition. In 1979, McCabe\(^2\) reported a series of patients with bilateral, asymmetric SNHL that progressed over weeks to months and evidence of vestibular deficiencies. A small percentage of these patients also had facial weakness associated with the hearing loss. All patients had a response to immunosuppressive medications. This led him to describe a new syndrome of autoimmune sensorineural hearing loss. Since this original description, much investigation has further solidified the existence of such a disease and brought forth diagnostic tests that aid in making the diagnosis. Harris and Sharp\(^3\) in 1990 reported the detection of a specific autoantibody with a molecular weight of 68 kilodaltons (kDa) using Western blot analysis in 35% of their patients. The exact mechanism of labyrinthine injury has yet to be elucidated, but may involve multiple processes. Research has better defined the immune mechanisms within the inner ear but the exact cause of AIED remains to be discovered.

Immune Function of the Inner Ear

Traditional thought was that the inner ear existed as an immunoprivileged site much as the brain has been supposed to be protected from immune function. This is partly due to the existence of blood-brain and blood-labyrinthine barrier. In the brain, this barrier functions to exclude serum proteins from the extracellular fluids of the brain and CSF. Immunoglobulins within the CSF are 1/1000\(^{th}\) of the level in serum and leukocytes do not cross this blood-brain barrier under normal circumstances. More recent investigation has shown that activated T cells are capable of crossing brain capillaries. Endothelial cells within these capillaries can be induced by cytokines to express adhesion molecules that recognize receptors on lymphocytes and mediate their extravasation. It appears that the brain is subject to limited immunosurveillance, and when stimulated is capable of recruiting lymphocytes from the circulation\(^4\).
Immune function within the inner ear is similar to that in the brain. The labyrinth is separated from the circulation by the blood-labyrinthine barrier. This serves to maintain the ionic characteristics of the cochlear fluids. It is also known that the inner lacks a significant lymphatic drainage. Similar to CSF, perilymph contains immunoglobulins at a concentration of 1/1000th of that in serum. Immunoglobulin G (IgG) is the predominant antibody, while IgM and IgA are present in lesser concentrations. The inner ear shows a greater tendency to be immunoresponsive than the brain however. Harris et al. found that antigen presented to the inner ear evoked an immune response equal to immunization via a peritoneal route while exceeding the response elicited by immunization through the middle ear. It is therefore evident that antigen within the cochlea readily gains access to the systemic circulation. By contrast, animals that were systemically immunized generated immune responses within the inner ear when the antigen was placed in the labyrinth. This response was characterized by cellular infiltration, inflammation, and cochlear damage with increased antibody titers within the perilymph and local antibody production.

The endolymphatic sac (ELS) plays a significant role in the inner ear immune response. The sac contains a resident population of lymphocytes, and it has been shown that the immune response within the ear may be significantly reduced and cochlear damage decreased with the destruction of the ELS or duct. The role of the ELS is not completely clear however, as the normal cochlea does not contain lymphocytes, and lymphocytes present in the cochlea in the immune response do not originate within the ELS. The lymphocytes responding to antigenic stimulation in the inner ear enter from the systemic circulation, apparently via the spiral modiolar vein (SMV). The endothelial cells of the SMV undergo activation and express intracellular adhesion molecule during the secondary immune response, thereby attracting lymphocytes and aiding in their migration from the circulation. Antigens presented within the inner ear do appear in macrophages in the ELS and activate immunoglobulin production from the lymphocytes.

The inner ear immune response serves to protect the cochlear structures from insults such as viral labyrinthitis and other infections. The delicate nature of the inner ear tissues makes them quite vulnerable to this inflammatory response however. Darmstadt et al. showed that cochlear damage from viral labyrinthitis is decreased by immunosuppressive therapy. Injury to the cochlea may be a secondary result of an inflammatory response, but evidence is mounting that autoimmune reactions in the inner ear are the cause of cochlear damage in some cases of immune-mediated ear disease.

All four types of immune reactions described by Gell and Coombs have been hypothesized to contribute to inner ear disease. Type I immune reactions are mediated by IgE and characterized by the activation of sensitized mast cells with the release of histamine and other vasoactive substances. Inhalant allergy and anaphylaxis fall into this category of immune response. The Type I immune response has been hypothesized as a cause of disruption of ionic transport within the inner ear due to histamine induced vasodilation, resulting in endolymphatic hydrops. Other hypotheses of the role of Type I sensitivity in inner ear disease have been put forth, but none have been proved with any degree of certainty.

Type II reactions occur when antibodies directed against a specific antigen within tissues elicit the activation of complement. The antigen can be either from an exogenous source (virus or drug) or may be directed at an endogenous source as in the case of anti-DNA antibodies of systemic lupus erythematosus (SLE) or the anti-basement membrane antibodies that cause renal involvement in Goodpasture’s syndrome. Evidence is growing that Type II immunity is a cause of inner ear disease. It has been shown that 34% of sera isolated from Meniere’s patients react with preparations from the inner ear of guinea pigs implying a specific antibody directed toward an inner ear antigen. Using Western-blot analysis, a bovine inner ear protein of molecular weight 68 kDa was recognized in sera from patients with presumed autoimmune SNHL. In addition, patients with Meniere’s disease have been shown to have greater reactivity with Type II collagen than controls. While the role of Type II immune reactions remain unclear, evidence is strongly suggestive and further investigation warranted.

Type III immune reactions are the result of the deposition of intermediate-sized immune complexes in the microcirculation. The deposition of these biologically active immune complexes induces vascular injury with subsequent injury to the labyrinthine structures. Consistent with this theory has been the demonstration of deposition of immunoglobulin around an apparently occluded blood vessel as shown by
imunofluorescence of the ELS in patients with Meniere’s disease. In addition, Derebery et al. analyzed sera from 30 patients with Meniere’s disease. Ninety five percent of these patients demonstrated elevated levels of circulating immune complexes as compared with only 20% of controls. The deposition of IgG in the subepithelial area of the endolymphatic sac was demonstrated in 40% of patients undergoing ELS destruction for Meniere’s. Hearing loss and vestibular dysfunction has been demonstrated in diseases known to be mediated by systemic immune complexes including systemic lupus erythematosus (SLE) and Wegener’s granulomatosis (WG). These observations provide substantial evidence that Type III immune reactions play a role in some instances of inner ear pathology.

Type IV immune reactions are characterized by T-cell mediated delayed hypersensitivity. This is exemplified by the rejection of transplanted organs. Sensitized T-lymphocytes are activated by antigen on the target cell to either directly lyse the cell or produce lymphokines that amplify the response by attracting other inflammatory cells. Evidence supporting the role of Type IV reactions in inner ear disease is largely from laboratory testing. The lymphocyte transformation test (LTT) measures the difference in proliferation of a patient’s lymphocytes when exposed to preparations of inner ear antigens as compared to the lack of this exposure. While lacking in sensitivity and specificity, it does indicate that some patients have evidence of an inner ear antigen-specific cellular immune response. Cellular immune responses to inner ear antigens have been demonstrated in Meniere’s disease and Cogan’s syndrome. While the exact mechanism of injury in immune mediated ear disease has yet to be described, and all types of immune responses have been hypothesized to cause labyrinthine damage, a large volume of information supports immune dysfunction as a cause in at least some cases of SNHL.

Clinical Features and Diagnosis

Autoimmune inner ear disease most commonly affects middle-aged women, much like other autoimmune diseases. Of affected individuals, 65% are female and 35% male. The usual presentation is that of a progressive SNHL over weeks to months. Not uncommonly, a serous middle ear effusion is noted and the patient is diagnosed with otitis media. Treatment with antibiotics is often instituted without result. The hearing typically fluctuates and progressively worsens. Dizziness and aural fullness are common, and one quarter to one half are initially diagnosed with Meniere’s disease. Classically, the hearing in one ear becomes poor while the other remains serviceable. The diagnosis is not considered until there is a drop in hearing in the better hearing ear, prompting the clinician to consider an immune etiology and initiate steroid therapy. Hearing loss is bilateral in 79% of patients. The initial presentation usually includes vertigo similar to those in Meniere’s disease, but one third of patients do not have vestibular symptoms.

Vestibular function may be lost gradually so that acute symptoms do not occur, but patients may develop ataxia and unsteadiness more noticeable in darkness with long-term, bilateral vestibular hypofunction. Of the 18 patients in McCabe’s original description of the disease, 5 had facial paralysis. More experience with the disease has revealed this to be a rare finding and it is not considered a routine occurrence in the syndrome. Systemic autoimmune diseases occur in 29% of patients.

The diagnosis of AIED has been difficult to standardize and still largely relies on the clinical presentation. Laboratory tests that would allow the definitive diagnosis of AIED have been the area of much interest. Clinicians often treat suspected cases of AIED empirically with steroids and either continue or withhold therapy based upon response. This puts patients at risk of the complications of steroid therapy without a good estimate of the chance of benefit. In McCabe’s original description of the disease, he acknowledged the lack of testing that would allow the diagnosis to be made with any certainty. However, he did feel that the lymphocyte inhibition test offered a strong argument for autoimmune disease. This test has fallen out of favor as it has failed to demonstrate significant sensitivity and specificity. More recently, the lymphocyte transformation test has been used to demonstrate cellular mediated autoimmune reaction to inner ear antigens. The test is performed by isolating lymphocytes from patients suspected of having AIED and exposing them to inner ear antigens. If the lymphocytes are sensitized to the antigens, the lymphocytes become active and release lymphokines. This is considered a positive test suggestive of a T-cell mediated autoimmune response directed toward inner ear tissues. The test is available only at the Cleveland Clinic and has had its utility questioned. The specificity is estimated at 93% and sensitivity at 50%-80% during the active phase of the disease. Other authors have reported the LTT against type II collagen as being more useful.
Perhaps the most promising definitive test for AIED is the Western blot immunoassay to 68 kDa antibody. This test is performed by electrophoresing inner ear antigens onto a gel and incubating them for several hours with a dilute solution of the patient’s serum. Harris and colleagues found a specific anticochlear antibody reacting to an antigen in the inner ear in the molecular weight range of 62,000-68,000. This is the same range as was found in experimental autoimmune SNHL induced in animals. In addition, they were able to identify antibodies from a patient with steroid-responsive autoimmune SNHL that not only had the same molecular weight, but also the same isoelectric point as the animals with hearing loss. Mosicki identified antibodies to 68 kDa antigen in 89% of patients with active progressive bilateral SNHL suggestive of AIED. Seventy five percent of the patients who tested positive responded to steroid therapy as compared to only 18% of those who tested negative. Harris’ work now includes 279 patients with presumed AIED of whom 32% were positive for antibody to an antigen of the 68 kDa size. He concludes that the test is 95% specific for AIED, but rather insensitive when used in the general population, and states that “in a group of patients who have unexplained progressive deafness, there is about a 1/3 chance of there being an immune cause of the hearing loss.” Further investigation suggests that the antigen is heat shock protein 70 (hsp 70).

Many laboratory tests have been used in the evaluation of SNHL. In an effort to better delineate the diagnostic value of a panel of laboratory tests, Hirose et al. examined the results of their testing in suspected cases of AIED. Their test battery included erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), C1q binding assay, anticardiolipin antibody (aCL), antineutrophil cytoplasmic antibody (ANCA), microhemagglutinin assay for Treponema pallidum (microhemagglutination assay), Lyme disease titers and the Western blot for hsp 70. They found that the best test for predicting steroid responsiveness was the Western blot for hsp 70. The sensitivity was low at 42%, but specificity was 90% and positive predictive value was 91%. The ESR and CRP were considered equal at detecting acute phase reactants and the presence of a systemic inflammatory disease. The other tests were of low yield. This lead to their recommendation that the laboratory evaluation of patients suspected of having AIED consist of either the ESR or CRP and the Western blot for hsp 70. In the event of an abnormally elevated ESR or CRP, further testing with the remainder of the battery could further characterize the disease. Testing for syphilis should be reserved for those with risk factors for sexually transmitted diseases and Lyme titers used in endemic areas or when suggested by the history.

The differential diagnosis of the patient with rapidly progressive SNHL is rather extensive and includes AIED, Meniere’s disease, recessive progressive hereditary deafness, vascular insufficiency, hypercoagulability syndrome as a result of antiphospholipid antibody, numerous systemic inflammatory illnesses, large vestibular aqueduct syndrome, acoustic schwannoma and otosyphilis. As a general rule, the evaluation should be tailored to the clinical picture and the appropriate workup performed. This may include imaging if structural abnormalities or retrocochlear pathology is suggested by history, physical examination and audiogram. As research continues and a better of understanding of AIED is attained, perhaps a standardized test panel will prove reliable and efficacious, but for the time being, ESR and Western blot assay for hsp 70 appear to be the most useful tests in the diagnosis of AIED.

**Syndromes**

Polyarteritis nodosa (PAN) is a multisystem, necrotizing vasculitis of small and medium-sized muscular arteries, most commonly involving the renal and visceral vessels. Hearing loss is a rare finding as a result of the disease but has been reported by many authors. Several temporal bone studies have demonstrated vasculitis with evidence of ischemia, osteoneogenesis and fibrosis. Similar findings have been induced in the temporal bone after immunologic challenge or with interruption of cochlear blood. It thus appears that vasculitis is a potential cause of hearing loss that should be included in the differential diagnosis of the patient with unexplained profound hearing loss.

Cogan’s syndrome is characterized by vestibuloauditory symptoms in association with non-syphilitic interstitial keratitis (IK) that occurs most commonly in young adults. The IK develops suddenly with photophobia, lacrimation and pain. The symptoms gradually resolve but recur. The most common otologic symptoms are episodic vertigo, tinnitus and hearing loss. The hearing loss is progressive over a several
month period. The vestibuloauditory symptoms may proceed or follow the IK by 1 to 6 months. Atypical presentations occur with longer periods between the eye and ear symptoms, or the association with eye disease other than IK. The temporal bone pathologic findings include hydrops, infiltration of the spiral ligament with lymphocytic infiltrate, osteoneogenesis of the round window, degeneration of the spiral ganglion cells and cystic degeneration of the stria vascularis. The cause of the syndrome is unknown, but may be due to autoimmunity to the eye and ear. Cogan and colleagues demonstrated a positive LTT when a patient’s serum was exposed to a corneal antigen. Hughes et al. demonstrated similar findings when reacting sera with inner ear antigens. These findings would seem to demonstrate a cellular autoimmune phenomenon directed toward the eye and inner ear.

Vogt-Koyanagi-Harada (VKH) syndrome is similar to Cogan’s syndrome in that patients suffer with SNHL, vestibular symptoms and uveitis. They also exhibit depigmentation of periorbital hair and skin, loss of eyelashes and aseptic meningitis, whereas CSF abnormalities are rare in Cogan’s disease. The cause is unknown, but is believed to be due to autoimmunity to melanocytes with damage to tissues containing these cells.

Wegener’s granulomatosis (WG) is a systemic disorder characterized by necrotizing granulomas with vasculitis in one or more organ systems (commonly in the respiratory tract) and, in most cases, focal necrotizing glomerulonephritis. It is presumed to be due to an autoimmune process. Otologic symptoms occur in 20%-25% of patients with serous otitis media being the most common otologic manifestation. Sensorineural hearing loss responsive to steroids has been described in WG patients. The cause of the inner ear disease is unknown but may be caused by vasculitis induced damage to labyrinthine structures. The diagnosis should be considered in patients with hearing loss of unexplained etiology. The antineutrophil cytoplasmic antibody (ANCA) is useful in making the diagnosis with specificity greater than 90%.

The manifestations of Behçet’s disease include recurrent aphthous ulceration of the upper aerodigestive tract and genitalia, ocular inflammation and cutaneous vasculitis. The disease is poorly understood, but SNHL associated with the disorder has been reported. There is no known treatment.

Relapsing polychondritis is an autoimmune disorder with recurrent inflammation affecting cartilage of the ear, nose, trachea, larynx and peripheral joints. Autoantibodies to types II and IX cartilage and lymphocyte activation with exposure to cartilage form the basis for its presumed autoimmune etiology. Inner ear disease with hearing loss and vestibular symptoms is presumed to be a result of an immune response to the type II cartilage found in the tectorial membrane and otic capsule. Patients usually have elevated ESR and increased immunoglobulins. They may have false-positive results of VDRL testing. The disease is treated with non-steroidal anti-inflammatory drugs (NSAIDS), steroids and dapsone.

Systemic lupus erythematosus (SLE) is a multisystem disease caused by the production of anti-nuclear and anti-DNA antibodies. The onset may be insidious or fulminant. The classic “butterfly” malar rash is uncommon. The disease spectrum includes polyarthritis, arthritis, pleuritis, pericarditis, pneumonitis, myocarditis, endocarditis, nephritis, cranial nerve palsies, meningitis, cerebrovascular accidents, neuritis, scleritis, retinal degeneration as a result of vasculitis and inflammatory bowel disease. Chronic otitis media with necrotizing vasculitis, progressive SNHL and dysequilibrium are the common otologic manifestations.

Rheumatoid arthritis (RA) is a systemic inflammatory disorder that most commonly affects the small joints of the hands and feet. Patients may have extra-articular disease including vasculitis, muscle atrophy, subcutaneous nodules, lymphadenopathy, splenomegaly and leukopenia. Several autoantibodies have been identified in patients with RA. These include IgM 19S and 7S and IgG 7S, collectively known as the rheumatoid factors (RF) and found in 75% of patients with the disease. They react to abnormal IgG produced by lymphocytes found in the synovia. Otologic manifestations have been the subject of debate. It has been proposed that hearing loss is due to involvement of the ossicular joints with conductive hearing loss (CHL) or that SNHL can occur as a result of damage to inner ear structures by inflammation or anti-rheumatic drugs. Kastanioudakis et al. found that 44% of patients had SNHL. This was most
commonly bilateral and symmetric and showed no relationship to NSAID, D-penicillamine, plaquenil or methotrexate use.

It can be recognized that AIED may be associated with a number of autoimmune syndromes, some of which have clear-cut mechanisms for inner ear involvement, while others have more circumstantial and theoretical mechanisms for injury. The point to be remembered is that AIED should be considered in patients with other autoimmune phenomena, and patients with unexplained, rapidly progressive SNHL should undergo thorough investigation for systemic disease.

**Meniere’s Disease**

Meniere’s disease is characterized by fluctuating hearing loss, episodic vertigo and aural fullness. It has been presumed since its description to be a result of endolymphatic hydrops of unknown cause. More recent evidence suggests that the syndrome may have an autoimmune etiology. Derebery has performed a series of investigations in the matter. She examined 30 patients who met the diagnostic criteria for Meniere’s disease as set forth by the American Academy of Otolaryngology-Head and Neck Surgery (AAOHNSS) for the presence of circulating immune complexes (CIC). Ninety seven percent of Meniere’s patients demonstrated significantly elevated levels of CICs (>2 SD higher than the mean level for the control group)\(^\text{15}\). She also examined 93 patients with Meniere’s disease for the presence of inhalant and food allergy using intradermal dilutional testing (IDT) and radioallergosorbent testing (RAST) and treated those who were positive with immunotherapy \(^\text{34}\). After treatment, 62% reported a decrease in both the frequency and severity of vertigo attacks. Complete or substantial control of vertigo occurred in 86% of patients. No patients reported worsening of the disease. Gottschlich \(\text{et al.}\)\(^\text{35}\) found a 32% incidence of antibodies to 68 kDa antigen in Meniere’s patients. Antibodies to the 68 kDa antigen were demonstrated in 6 of 7 patients with the late onset of hearing loss in an only hearing ear in patients diagnosed with Meniere’s disease\(^\text{36}\). These data suggest that some patients with Meniere’s disease may have an autoimmune cause for their symptoms, or perhaps Meniere’s and AIED are two separate syndromes that are difficult to distinguish. It should be noted that while there is some evidence for immune dysfunction in Meniere’s disease, the evidence is not substantial enough at this time to warrant abandoning standard treatment regimens for therapy with steroids or other immunosuppressive drugs.

**Treatment**

Patients with progressive SNHL that is believed to be due to AIED should be considered for immunosuppressive therapy. Steroids remain the foundation of therapy. It is generally agreed that relatively high doses are needed to see a therapeutic result, but the exact dose and duration of therapy are still the matter of some debate. Patients should be started on Prednisone 1mg/kg/day (60 mg daily dose for adults). An initial therapeutic trial of 3-4 weeks may determine the response to therapy. Those who respond can be slowly tapered, with the high doses re instituted for relapses. A maintenance dose of 20 mg every other day may be required for 3-6 months. Before starting such a regimen, the potential risks and side effects associated with high dose steroid therapy should be discussed in detail\(^\text{21}\).

McCabe\(^\text{37}\) insists that cyclophosphamide is the cornerstone of therapy and advocates the institution of treatment in any patient in whom the disease is suspected. He begins with the administration of cyclophosphamide 60 mg every 12 hours with prednisolone 30 mg every other day for 3 weeks. If the patient shows some response to this test treatment, the therapy at the same doses is continued for 3 months. The cyclophosphamide is then discontinued and the prednisolone continued for 2 weeks. If there is no decrease in thresholds, the prednisolone is tapered over 2 weeks. If the hearing levels fall again, the combination is again used for an additional 3 months. The potential complications of cyclophosphamide therapy include hemorrhagic cystitis, leukopenia, sterility and malignancies of the urinary tract. In addition, it has been shown to be leukemogenic. For this reason, patients should be followed closely and have surveillance of the white blood cell counts and urinalysis periodically while on the treatment regimen. Its use in patients of child-bearing age is also to be avoided. It is prudent to enlist the expertise of a rheumatologist or hematologist/oncologist when using such a treatment regimen.
Plasmapheresis has been used in the treatment of AIED as well. Luetje and Berliner\(^{38}\) showed that 8 of 16 patients with idiopathic, rapidly progressive SNHL had improved or stabilized hearing in one or both ears. Only 25% of patients required continued immunosuppressive therapy. The conclusions from this study are limited as the sample size is small and the design lacked controls, but plasmapheresis is yet another avenue for investigation in the etiology and treatment of idiopathic SNHL. The shortcomings of plasmapheresis therapy include its expense and time consumption. Patients are required to spend several hours for multiple days. In addition, plasmapheresis alone does not reduce the underlying production of the offending antibody or cytokines, and patients frequently continue with immunosuppressive therapy while undergoing plasmapheresis\(^{21}\).

Recently methotrexate has received attention as a possible therapeutic agent in AIED for its successes in the treatment of RA. Methotrexate is an antimetabolite that inhibits dihydrofolate reductase, thereby interfering with DNA synthesis, repair and replication. Sismanis \textit{et al.}\(^{39}\) demonstrated that 69.9% of 25 patients with immune mediated cochleovestibular disease had hearing improvement and 80% of patients with vestibular symptoms had substantial improvement or resolution. Therapy was begun at 7.5 mg per week and in most patients was increased to 12.5 mg per week. Eight patients had rather mild and reversible complications including nausea, oral ulcerations, transient rash and minimal alopecia. Two patients had diffuse rashes that necessitated the discontinuation of therapy. While the sample size was small, the success of therapy is promising as the use of methotrexate in RA has proven to have mild side effects and complications. Further investigation is needed, specifically a prospective, randomized controlled trial, but methotrexate may prove to be an alternative to long-term, high-dose steroids.

\textbf{Case Study}

A 45-year-old white female with no prior medical problems presented to the Otolaryngology clinic with hearing loss, aural fullness and dysequilibrium. Her symptoms were first noticed two months prior when she awoke in the morning with the sensation of fullness in the right ear and mild right-sided hearing loss. She saw her primary care physician who prescribed antibiotics. Two weeks later she was seen by an outside otolaryngologist who noted a normal examination but found a right-sided moderate sensorineural hearing loss. Evaluation at that time included a CBC, chemistries, thyroid functions, RPR and ESR. All were within normal limits. An MRI of the internal auditory canals and cerebellopontine angle revealed no evidence of an 8\textsuperscript{th} nerve tumor. She was initially treated with low salt diet and Dyazide for presumed Meniere’s disease. On a follow-up visit, her hearing in the right ear had further deteriorated with some mild sensorineural hearing loss in the left ear that had not been previously noted. She was referred for further evaluation and treatment.

Upon initial evaluation, her examination was unremarkable. Audiogram revealed a severe SNHL on the right with a mild SNHL on the left. Blood was sent for anti-68kDa antigen and returned as positive. Therapy with steroids at 30 mg twice a day was started and continued for 4 weeks. She showed some improvement in hearing. The steroid dose was tapered over 4 months with no shift in thresholds. Hearing remained stable for 5 months when she again had a threshold shift. Steroid therapy was initiated again with good response. She was tapered from the steroids over 4 months and has had stable hearing since.

\textbf{Conclusion}

Autoimmune inner ear disease is a relatively newly described disease process of which little is known of the etiology, diagnosis and treatment. Many studies in the past 20 years have done much to further the understanding of the immune function of the ear and its involvement in systemic disease. Additional work is needed however. Clinicians should be aware of the presentation and diagnosis of the disease as it may represent a treatable cause of progressive deafness. While steroids are useful in many patients, some are unable to tolerate the doses and length of therapy required for benefit, while alternative therapies have the potential for serious complications. New medications and treatments are needed and are certain to be discovered as experience with the disease increases.
References


