

Autoimmune inner ear disease secondary to Hashimoto's thyroiditis: a case report

Beenish Fayyaz  and Sunita Upreti

Internal Medicine, Greater Baltimore Medical Center, MD, USA

ABSTRACT

Autoimmune inner ear disease is an important yet incompletely understood cause of hearing loss which can present to different medical disciplines. Its diagnostic significance is indicated by the fact that it is a reversible medical condition if recognized early with an excellent response to immunosuppressive agents. Therefore, it should be considered in the differential diagnosis of hearing disorders especially in the context of another autoimmune disease such as Hashimoto's thyroiditis.

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1. Introduction

Patients with Hashimoto's thyroiditis (HT) are at higher risk of developing audiological abnormalities as compared to the healthy individuals [1]. This auditory dysfunction can be multifactorial as indicated by case studies done on HT. However, autoimmune inner ear disease (AIED) should be considered as a possible cause when an individual with the history of HT presents with acute-onset vestibulocochlear symptoms due to a similar pathophysiology. Various cases have been reported in the medical literature documenting an association between these two autoimmune disorders.

Given the absence of specific tests, the diagnosis of AIED may be difficult and mainly depends on an appropriate clinical presentation, exclusion of the other causes, and a positive response to steroid therapy. Although poorly understood, this disease is one of the few reversible forms of hearing loss with a favorable outcome.

We describe a case when a female with a history of HT was diagnosed with AIED in the setting of an idiopathic refractory sensorineural hearing loss.

2. Case description

A 61-year-old female with a history of HT with positive antithyroid peroxidase (anti-TPO) antibodies, on thyroxine replacement therapy was referred to the ENT clinic by her primary care physician due to progressive right-sided hearing loss for 2 weeks. She denied having any vertigo, nausea, or vomiting but did give a history of mild flu-like symptoms associated with an aural fullness. She was noted to have right tympanic membrane dullness and was treated with antibiotics along with nasal steroids due to a suspected sinus/ear infection. A right-sided myringotomy and pressure equalization tube insertion

was performed due to a persistent right middle ear effusion. However, her symptoms continued to worsen and involvement of the left ear was noted as well. At that time, on pure tone audiometry, she had an asymmetric moderate left-sided sensorineural and right-sided mixed hearing loss pattern. An MRI of the brain and internal auditory canal with and without contrast did not show any lesion or mass although some nonspecific bilateral enhancement of the cochlea was noted.

As she did not respond to conventional management, her diagnosis was reviewed and it was suspected that she might have an autoimmune etiology of her symptoms. Her thyroid function tests were normal at that time which was followed by a complete immunological workup. She was noted to have C-reactive protein (CRP) of 58.3 mg/L, ESR of 67 mm³/h, and rheumatoid factor of 29.2 IU/ml. Anti-nuclear antibodies (ANA) was found to be negative. No additional testing was done.

Based on her blood workup results, she was started on Prednisone 60 mg once daily. The patient reported an improvement within 1 week of the corticosteroid therapy and thus was confirmed to have AIED. The symptoms completely resolved after 3 weeks of treatment while on a prednisone dose taper. She continued to maintain remission on low-dose prednisone while following in the ENT clinic.

3. Discussion

AIED is a distinct category of hearing disorders characterized by an immune-mediated vestibulocochlear dysfunction. It was first postulated as a unique clinical entity in 1979 by McCabe based on the treatment response to steroids in 18 patients [2]. The mechanism

responsible for this clinical entity is proposed to be T-cell mediated inflammation which has been indirectly confirmed by the success of immunosuppressive therapy and although better understood now is still not exclusive. Therefore, the current incidence of AIED as less than 1% of all the hearing loss causes is considered to be an underestimation [3].

As the name indicates, AIED causes inflammation of the inner ear structures. The hallmark symptom is fluctuating/progressive sensorineural hearing loss with up to 50% of the patients having tinnitus and vertigo. That is why AIED is also known as autoimmune sensorineural hearing loss as it most commonly involves the cochlea leading to similar symptoms as seen in our patient who presented with bilateral acute onset hearing loss [4]. The nature of the hearing loss was determined to be sensorineural based on audiological tests as discussed above while the MRI showed findings suggestive of cochlear inflammation.

In the absence of a uniform diagnostic criteria, AIED is defined as bilateral sensorineural hearing loss of 30 dB or more with a decline in at least 1 ear on 2 serial audiograms performed less than 3 months apart. Positive response to steroids is another requisite clinical criterion [5]. There is no definite test for AIED till now although, a number of immunological studies such as an elevated ESR, CRP, and positive ANA or RA factor may be consistent with the diagnosis. There are other autoantibodies which have been identified in the context of this disease (Table 1), but their diagnostic and prognostic role remains controversial. MRI of the brain and brainstem auditory-evoked responses can be done to help rule out other causes such as acoustic neuroma and ototoxic neuropathy.

Primary AIED is defined as a limited disease of the inner ear while secondary AIED occurs in the context of other systemic or autoimmune diseases such as HT (Table 2). Although AIED has been frequently associated with Cogan's syndrome and ANCA-vasculitis, its occurrence in individuals with HT is relatively uncommon and still unexplained.

HT is an important cause of adult hypothyroidism characterized by the immune-mediated destruction of thyroid tissue in the presence of various autoantibodies such as anti-TPO, anti-thyroglobulin, and anti-mitochondrial antibodies. In addition to thyroid dysfunction, HT can cause various otological symptoms. Although these abnormalities can occur as a direct consequence of hypothyroidism [1], it has been suggested that the

Table 1. Autoantibodies associated with AIED in various case studies.

Studies done	Autoantibodies identified
Moscicki et al. [6]	Anti-HSP70 (Heat shock protein 70)
Baek et al. [7]	Anti-cochlin
Boulassel et al. [8]	Anti-beta actin
Nair et al. [9]	Anti-CTL2 (Choline transporter-like protein 2)
Helfgott et al. [10]	Anti-collagen type 2

Table 2. Autoimmune disorders associated with AIED [5,11].

Autoimmune disorders
• Cogan's syndrome
• Systemic lupus erythematosus
• Vasculitides
• Rheumatoid arthritis
• Systemic sclerosis
• Relapsing polychondritis
• Polymyositis-dermatomyositis
• Hashimoto's thyroiditis
• Ankylosing spondylitis
• Behçet disease
• Inflammatory bowel disease

circulating antithyroid autoantibodies can lead to T-cell mediated vestibulocochlear dysfunction. A review of the literature shows various case studies documenting the association of these two autoimmune entities [12–15] which demonstrate the existence of a clear relationship between thyroid autoimmunity and the inner ear damage, regardless of thyroid function. Furthermore, it is postulated that the immune complexes precipitation in the inner ear with concomitant autoimmune inflammation may be the pathogenic mechanism shared with HT.

In such clinical scenarios, the diagnosis of secondary AIED should be made in the context of the presenting symptoms, medical history, thyroid function tests, immunological workup, and clinical response to immunosuppressive agents. In our case, the patient had history of HT, normal thyroid function tests, and elevated inflammatory markers (ESR and CRP). She failed to respond to conventional management. The diagnosis of AIED was made as a trial of oral prednisone led to clinical improvement within 1 week. Afterwards, she was placed on chronic steroids to maintain remission as recommended [16].

Even with emerging medical data, there is much that is not known about AIED – especially how to suspect and diagnose this autoimmune disorder. Presence of other autoimmune disorders such as HT should be considered as a clinical clue for AIED in an individual with 'idiopathic' sensorineural hearing loss. In addition, HT is not a well-defined organ-specific autoimmune disease, as traditionally considered, and can cause various other systemic manifestations.

Disclosure statement

No potential conflict of interest was reported by the authors.

ORCID

Beenish Fayyaz  <http://orcid.org/0000-0002-7837-6580>

References

- [1] Anand VT, Mann SB, Dash RJ, et al. Auditory investigations in hypothyroidism. *Acta Otolaryngol.* 1989;108:83.

- [2] McCabe BF. Autoimmune sensorineural hearing loss. *Ann Otol Rhinol Laryngol.* 1979;88(5):585–589.
- [3] Marsili M, Marzetti V, Lucantoni M, et al. Autoimmune sensorineural hearing loss as presenting manifestation of paediatric Behçet disease responding to adalimumab: a case report. *Ital J Paediatr.* 2016;42(1):81.
- [4] Ryan AF, Keithly EM, Harris JP. Autoimmune inner ear disorders. *Curr Opin Neurol.* 2001;14(1):35–40.
- [5] Mijovic T, Zeitouni A, Colmegna I. Autoimmune sensorineural hearing loss: the otology-rheumatology interface. *Rheumatology.* 2013;52(5):780–789.
- [6] Moscicki RA, San Martin JE, Quintero CH, et al. Serum antibody to inner ear proteins in patients with progressive hearing loss. Correlation with disease activity and response to corticosteroid treatment. *JAMA.* 1994;272:611–616.
- [7] Baek MJ, Park HM, Johnson JM, et al. Increased frequencies of cochlin-specific T cells in patients with autoimmune sensorineural hearing loss. *J Immunol.* 2006;177:4203–4210.
- [8] Boulassel MR, Tomasi JP, Deggouj N, et al. Identification of beta-actin as a candidate autoantigen in autoimmune inner ear disease. *Clin Otolaryngol Allied Sci.* 2000;25:535–541.
- [9] Nair TS, Kozma KE, Hoefling NL, et al. Identification and characterization of choline transporter-like protein 2, an inner ear glycoprotein of 68 and 72 kDa that is the target of antibody-induced hearing loss. *J Neurosci.* 2004;24:1772–1779.
- [10] Helfgott SM, Moscicki RA, San Martin J, et al. Correlation between antibodies to type II collagen and treatment outcome in bilateral progressive sensorineural hearing loss. *Lancet.* 1991;337:387–389.
- [11] Goodall AF. Current understanding of the pathogenesis of autoimmune inner ear disease: a review. *Clin Otolaryngol.* 2015;40(5):412–419.
- [12] Arduc A, Isik S, Allusoglu S, et al. Evaluation of hearing function in patients with euthyroid Hashimoto's thyroiditis. *Endocrine.* 2015;50(3):708–714.
- [13] Brenner M, Hoistad D, Hain TC. Prevalence of thyroid dysfunction in Meniere's disease. *Archives of Oto HNS.* 2004;130(2):226–228.
- [14] Modugno GC, Pirodda A, Ferri GG, et al. A relationship between autoimmune thyroiditis and benign paroxysmal positional vertigo? *Med Hypotheses.* 2000;54(4):614–615.
- [15] Chiarella G, Tognini S, Nacci A, et al. Vestibular disorders in euthyroid patients with Hashimoto's thyroiditis: role of thyroid autoimmunity. *Clin Endocrinol.* 2014;81:600–605.
- [16] Stachler RJ, Chandrasekhar SS, Archer SM, et al. Clinical practice guideline: sudden hearing loss. *Otolaryngol Head Neck Surg.* 2012;146(3 Suppl):S1–S35.