Autoimmune Inner Ear Disease

(February 2016)
The author proposes the existence of a new entity, autoimmune sensorineural hearing loss, on the basis of diagnostic study and treatment experience with a series of 18 patients. In each case the clinical pattern did not fit with known entities and thus seemed to merit distinctive categorization. In the one patient in whom tissue was available, a vasculitis was evident, a feature of autoimmune disease. All patients responded to treatment for an autoimmune disease, namely, chronic cortisone and cyclophosphamide therapy. The author suggests that all otolaryngologists should be aware of the possibility of this condition, because it is one of the few forms of sensorineural deafness for which we have a treatment.
Epidemiology

Rare (less than 1% of hearing impairments)
Female predominance & more common between 20 – 50 years
**Immunopathology**

- **Specific Antibody for**
  - 68-kDa bovine inner ear antigen
  - HSP-70
  - CTL2 (cochlin)

- **Vestibular end organs:** fibrous tissue and new bone formation
- **Sensory end organs:** degeneration and collapse of membranous labyrinth
- **Lymphocytic infiltration**

**Endolymphatic sac is important in inner ear immune response**
Clinical Findings

Hallmark: Bilateral Progressive SNHL during weeks to months

- May initially be **unilateral** (take months for bilateral involvement)
- May have **fluctuate** hearing loss (but overall trend is progressive)
- **Vestibular symptoms** in 50-80% (20% have vertigo like Meniere)
- **Tinnitus & Aural fullness** in 25–50% (can fluctuate)
Secondary AIED

The *most common* autoimmune diseases that affect the inner ear are:

- Cogan syndrome
- Polyarteritis nodosa (PAN)
- Rheumatoid arthritis
- Wegener granulomatosis
- Inflammatory Bowel Disease (*Beethoven!* )
Cogan Syndrome

- Bilateral autoimmune disorder - more in 4th decade
- 50% present with Sudden SNHL

Definition of Disease

- Meniere-like cochleovestibular symptoms (hydrops) and
- Nonsyphilitic Interstitial keratitis (pain, scleral redness, photophobia)

David G. Cogan
Ophthalmologist
1908 - 1993
Differential Diagnoses

- Sudden SNHL
- Meniere’s Disease
- Vestibular Schwannoma
- Otosyphilis
- Other intracranial pathologies [Malignancy with dura involvement (Metastatic disease, Lymphoma), Meningitis, Multiple Sclerosis]
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Clinical Workup

Appropriate history taking should include questions for:

- Recurrent or chronic ocular disease
- Nephritis
- Arthritis
- Pneumonitis or Sinusitis
- Inflammatory bowel disease

In order to differentiate between primary AIED and secondary AIED
Paraclinical Workup

Necessary assessments in any patients suspected to AIED:

- CBC-diff
- ESR, CRP
- ANA
- Anti-ds DNA
- RF
- ANCA
- Anti SS-A, SS-B
- Anti gliadin Ab
- Anti Phospho. Ab
- Raji cell assay
- Serum C3 and C4
- TFT (free T3 & T4)
- FTA-ABS
- CPA MRI with Gd
- HIV assessment (?)
**Autoimmune Inner Ear Disease**

- **Background**
- **Diagnosis**
- **Management**

**Prednisolone**

1 mg/kg PO for 4 weeks

Audiometric data at the end of 4 weeks in comparison to baseline:

- **Steroid Responders**
  - 15 dB improve at one frequency
  - 10 dB improve at ≥2 consecutive freq.
  - 10% Improvement in WRS
  - Stabilization of fluctuation

- **Non-Responders**
**Autoimmune Inner Ear Disease**

### Background

### Diagnosis

### Management

**Prednisolone**

- **Steroid Responders**
  - Continue full dose steroid until no change in monthly audiometry
  - Taper to 10–20 mg every other day in 8 weeks &
  - Continue treatment duration for at least 6 months

- **Non-Responders**
  - Taper off in 10 days
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Background

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**Prednisolone**

**What to do in Relapse?** Restart management cycle (high dose steroid for 4 weeks)

**Result:** Overall response rate is 60% (better in vestibular symptoms)

**Precaution:** significant risk of long term steroid therapy including:

- Peptic ulcer
- Weight gain
- Mood changes
Methotrexate

**Indication:**
for replacing steroid due to adverse effects

*Not beneficial in Nonresponders group*

Prednisone sparing effects of MTX may take 1-2 months

*Maintaining steroid until effects appeared*

**Administration (Oral):**
Initial: 5-7.5 mg/week
not to exceed 20 mg/week
Methotrexate

Precaution:
- renal & hepatic adjustment
- error in use (daily instead of weekly)

Contraindications:
- Sensitivity, Pregnancy, breast feeding, blood dyscrasia, liver at risk

Follow up assessments:
- CBC, RFT, LFT

Administration (Oral):
Initial: 5-7.5 mg/week
not to exceed 20 mg/week
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Cyclophosphamide

**Indication:**
Nonresponders or Refractory to wean from steroids

**Precaution:**
- renal & hepatic adjustment, not use partial pill

**Complications:**
- Hemorrhagic cystitis, Bone marrow suppression

**Follow up assessments:**
- CBC, RFT, LFT

**Administration (Oral):**
- 1 – 2 mg / kg / day
- 4 – 6 weeks
Other Managements

- Etanercept (recombinant Anti TNFα antibody)
- Azathioprine
- Mycophenolate mofetil (CellCept)
- Intratympanic injections (steroid, TNFα inhibitor)
- Plasmapheresis
- Immunotherapy
- Cochlear Implant
11-13 May 2016
Tehran - Iran

www.apon2016.com

Main Topics:
- Middle Ear & Ossicular Surgery
- Implantable Hearing Devices
- Approach to Dizzy Patient
- Facial Nerve Management
- Tinnitus Updates
- Lateral Skull base Surgery
- Endoscopic Ear Surgery
- Temporal Bone Imaging
- Audiovestibular Study

Workshops:
- Temporal Bone Dissection
- Endoscopic Ear Surgery
- Implantable Hearing Aids
- Practical Management of - Dizzy Patient

Thanks a lot for your Attention

Current presentation content is available at: http://tums.ac.ir/faculties/s-dabiri