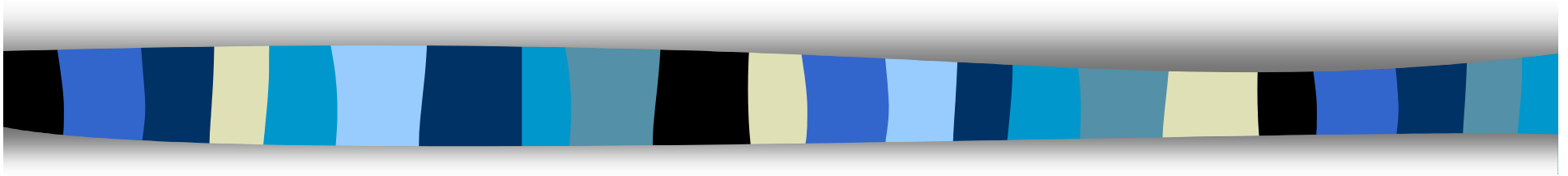


Autoimmune Inner Ear Disease



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12 January 2000



Background

- Lenhardt 1958
- McCabe 1979
- Harris 1990



Immune Function of Inner Ear

- blood-labyrinthine barrier
- maintenance of homeostasis
- little lymphatic drainage
- immunoglobulins 1/1000th of serum
- immune responsiveness

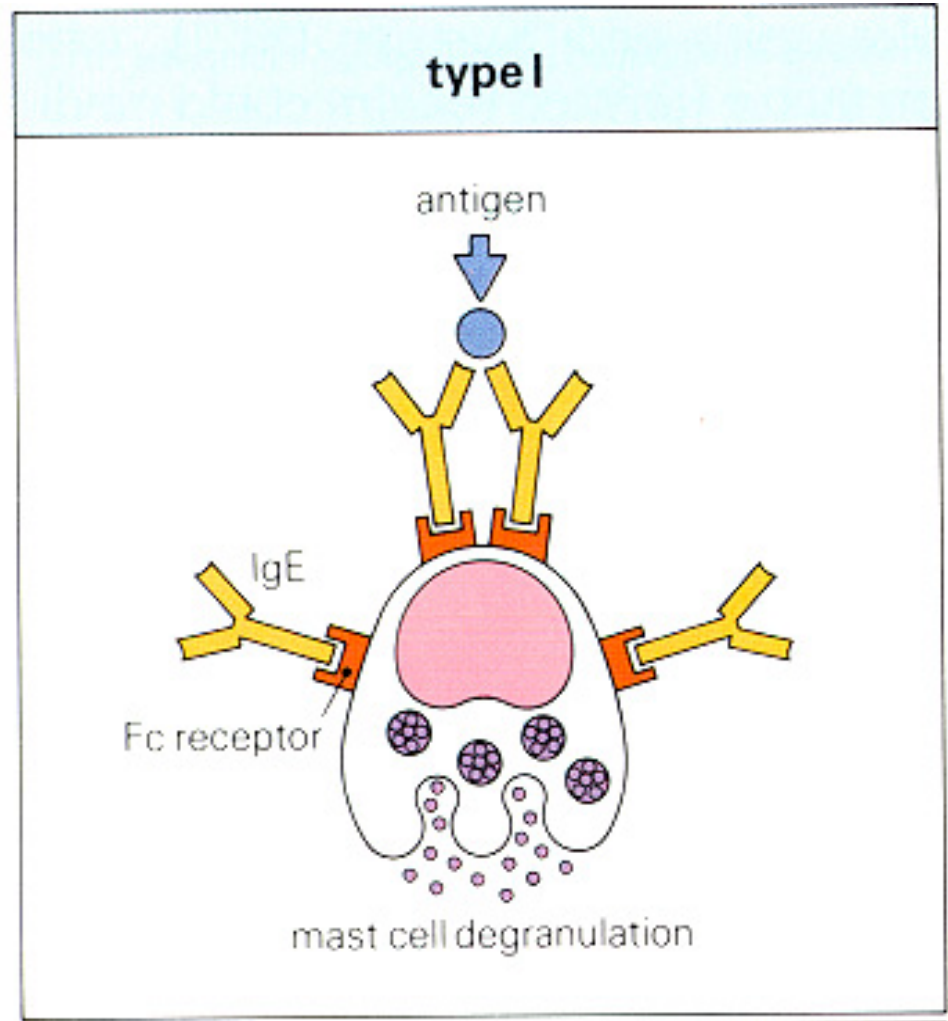


Endolymphatic Sac

- Resident lymphocytes
- immunoglobulin production
- systemic lymphocyte entry
 - spiral modiolar vein
 - intercellular adhesion molecule

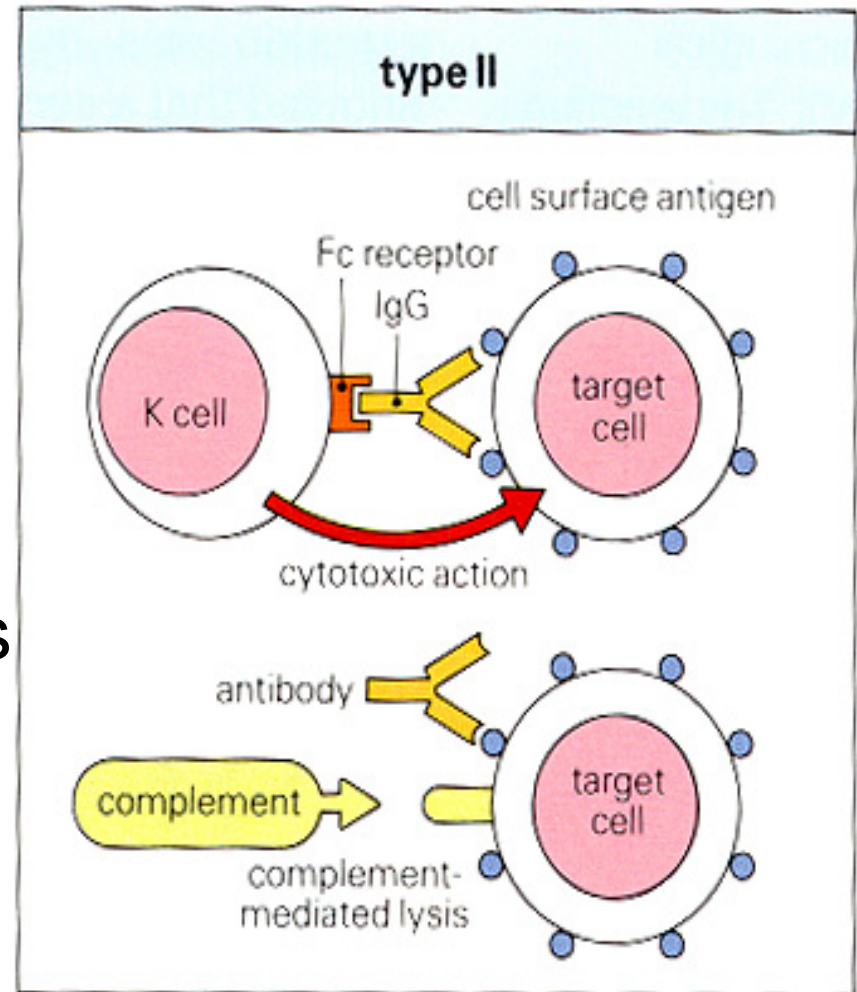
Type I Hypersensitivity

- IgE
- mast cells
- histamine
- vasodilation
- ? Hydrops → Meniere's
- inhalant allergy



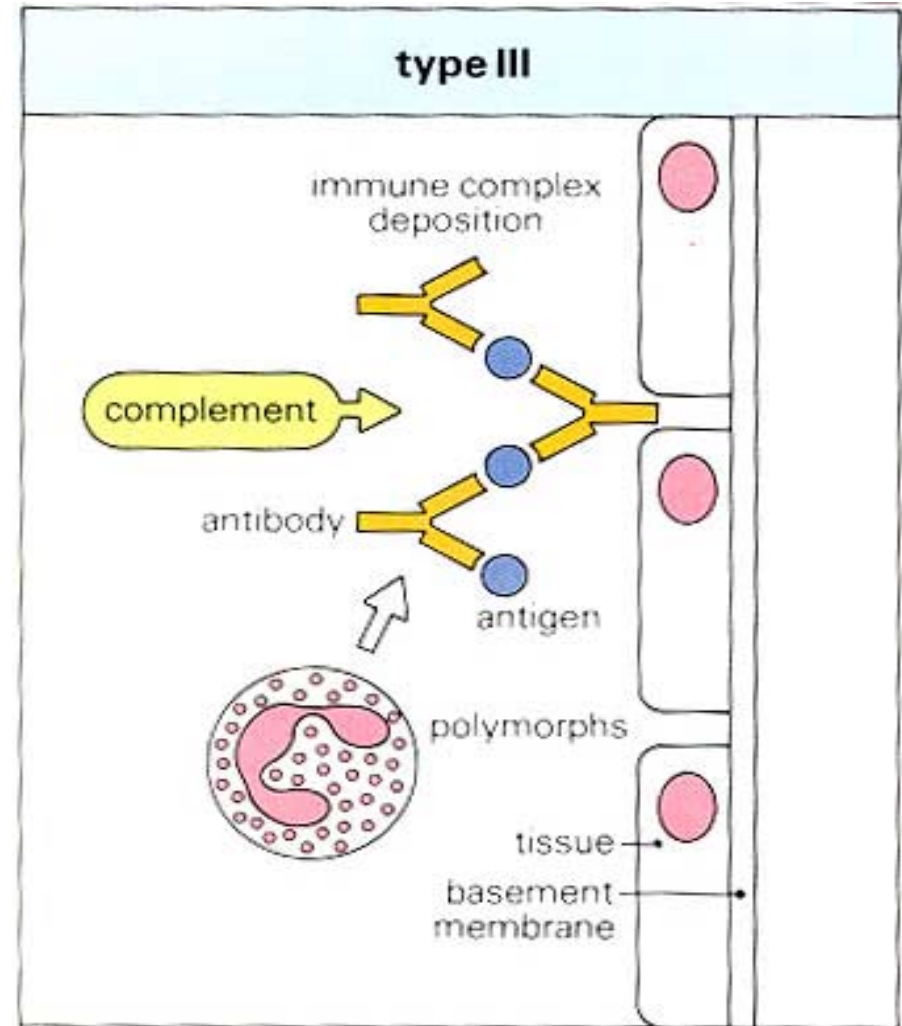
Type II Hypersensitivity

- Antibodies
- complement activation
- anti-68kDa protein antibody
- SLE, Goodpasture's



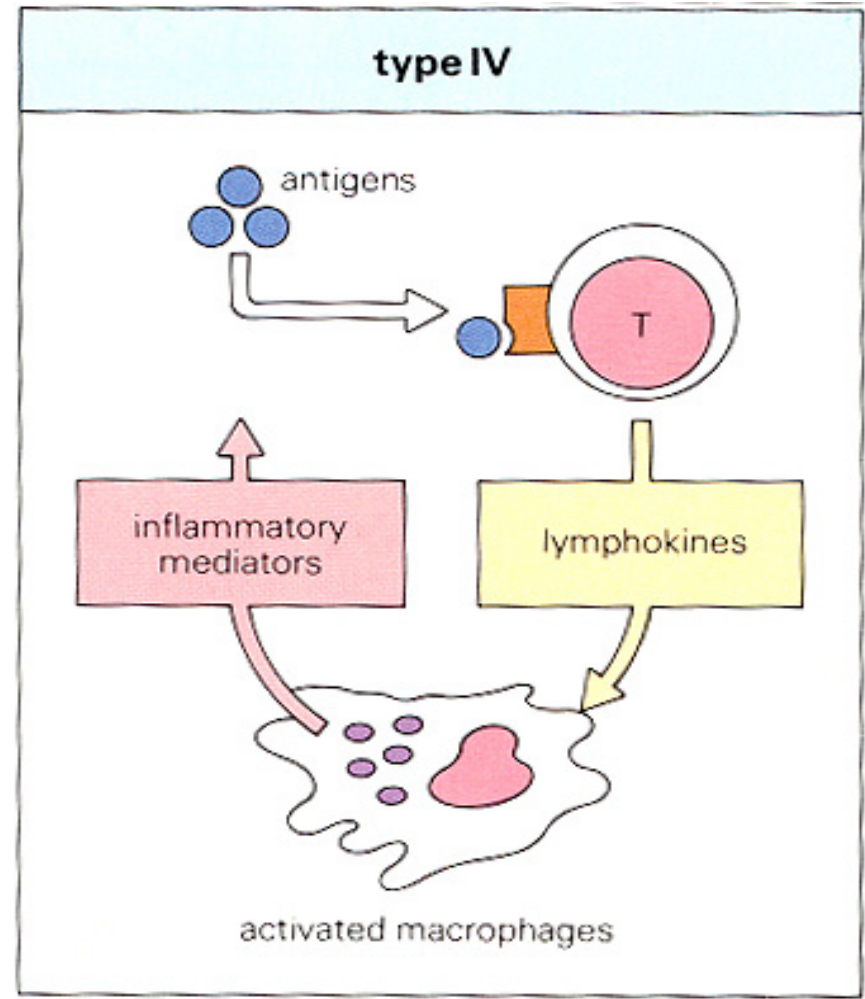
Type III Hypersensitivity

- Immune complex
- Ig deposition
- tissue injury
- Wegener's,
?Meniere's



Type IV Hypersensitivity

- T-cell mediated
- direct lysis
- lymphokine production
- lymphocyte transformation test
- Cogan's syndrome





Clinical Picture

- Middle-aged women
- progressive SNHL, weeks to months
- dizziness, aural fullness
- bilateral 79%
- $\frac{1}{3}$ no vestibular symptoms
- systemic autoimmune disease in 29%



Diagnosis

- Clinical
- LTT - 93% specific, 50-80% sensitive
- Western blot for anti-68kDa protein (hsp70)
 - 95% specific
 - insensitive
 - predictor of steroid response



Diagnosis

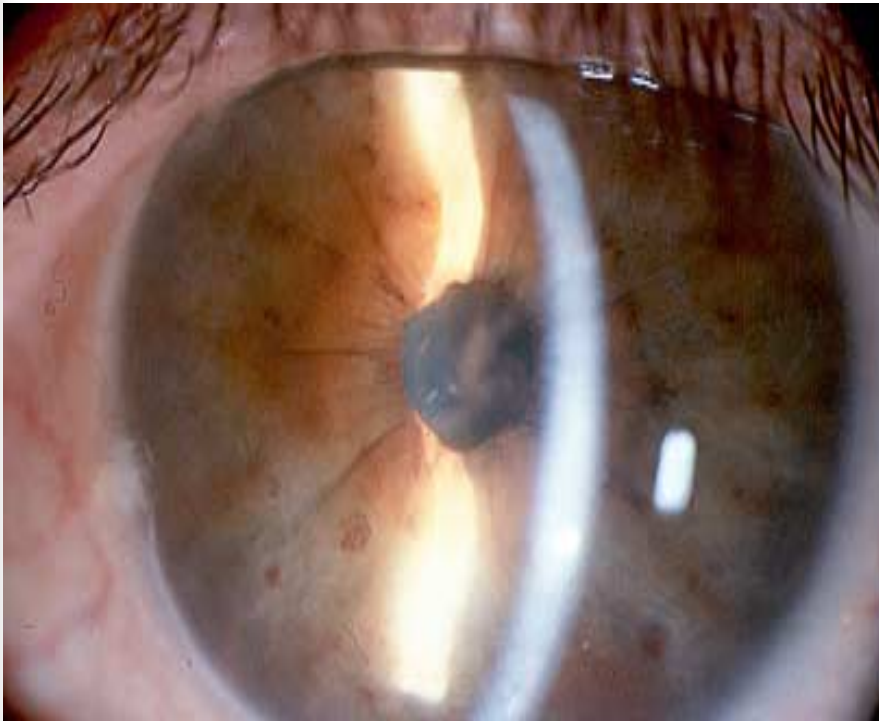
- ESR
- CRP
- C1q binding assay
- anti-cardiolipin
- ANCA
- syphilis testing
- Lyme titers
- CBC
- chemistries
- thyroid functions
- imaging



Polyarteritis Nodosa

- Vasculitis of small and medium-sized arteries
- renal and visceral
- ischemia → osteoneogenesis → fibrosis
- hearing loss rare

Cogan's Syndrome



- Interstitial keratitis
- vertigo, tinnitus, SNHL
- positive LTT to corneal antigen

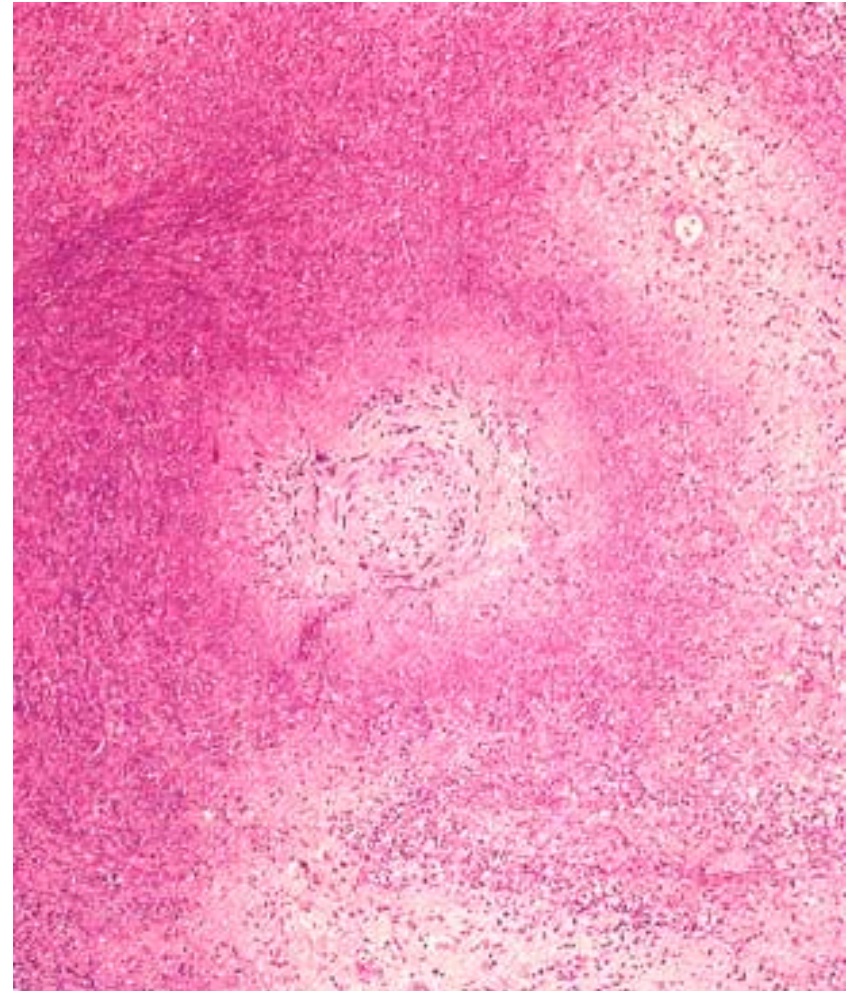


Vogt-Koyanagi-Harada (VKH) Syndrome

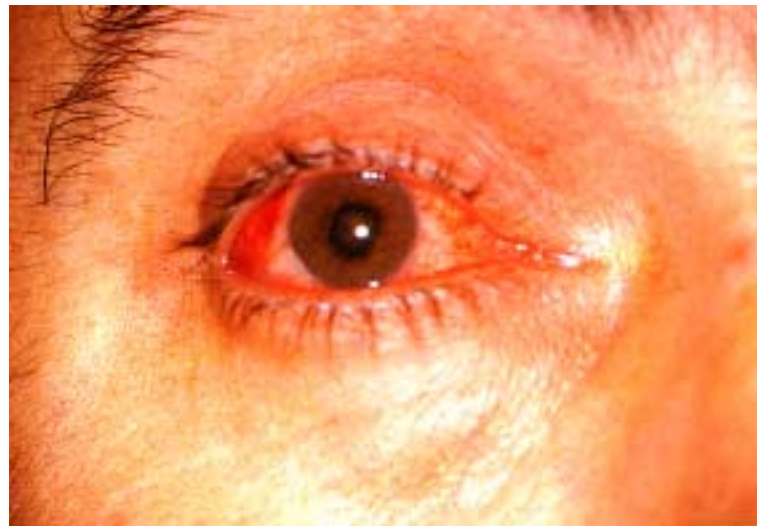
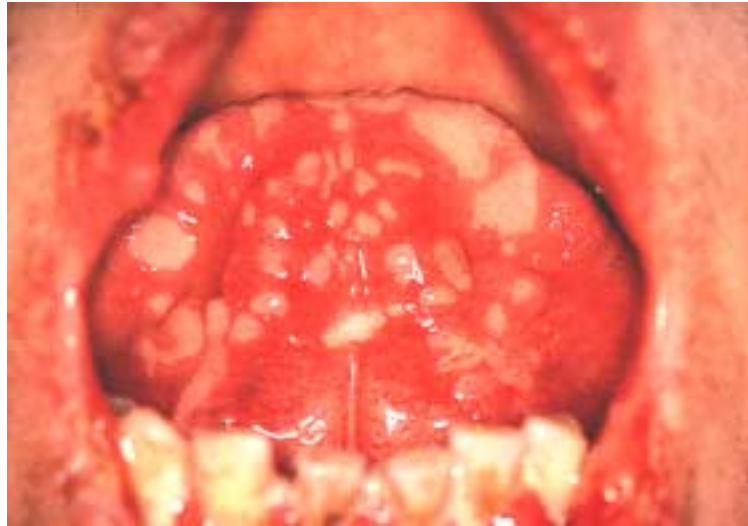
- SNHL, vestibular signs, uveitis
- periorbital hair loss, depigmentation
- aseptic meningitis
- ?autoimmunity to melanocytes

Wegener's Granulomatosis

- Necrotizing granulomata
- vasculitis
- respiratory tract and kidneys
- serous OM
- cANCA 90% specific

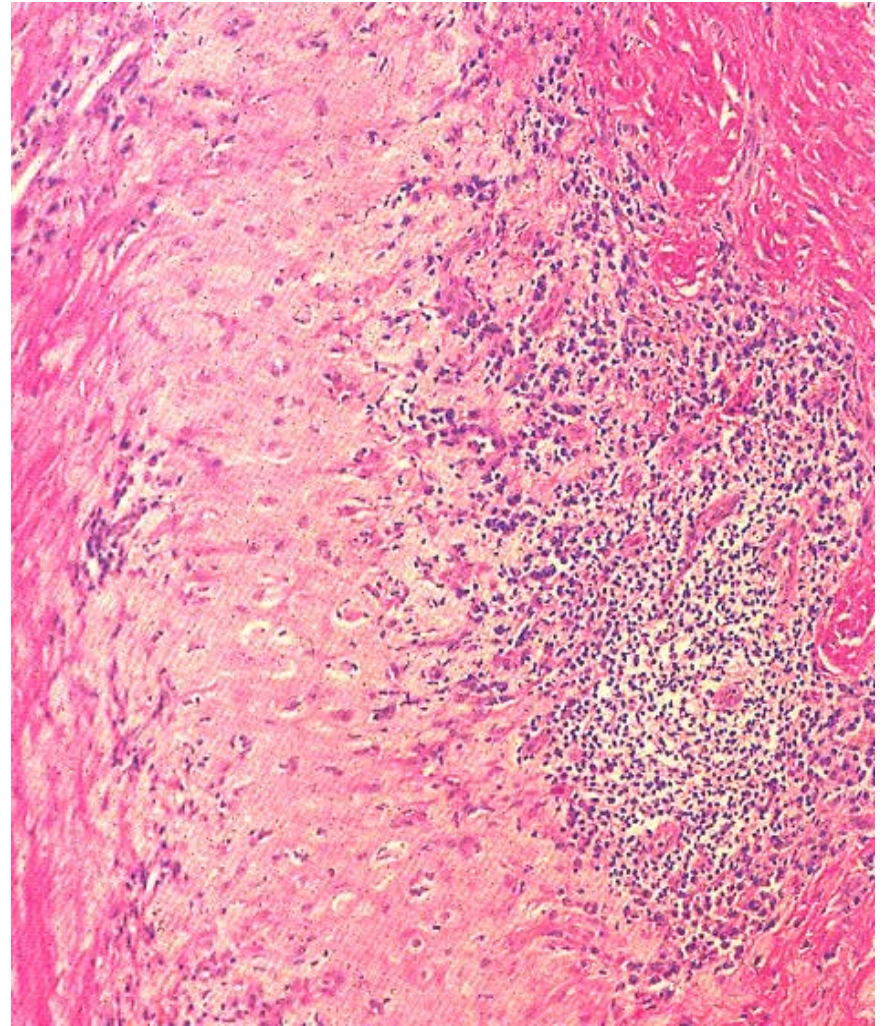


Behçet's Disease



Relapsing Polychondritis

- Recurrent inflammation of ear, nose, trachea, larynx
- autoantibodies to cartilage II & IX
- NSAIDs, steroids, dapsons



Systemic Lupus Erythematosus



- Anti-nuclear, anti-DNA antibodies
- numerous systemic manifestations
- COM with vasculitis, SNHL, dysequilibrium



Rheumatoid Arthritis

- Small joints of hands and feet
- vasculitis, muscle atrophy, subcutaneous nodules, splenomegaly
- IgM 19S and 7S, IgG 7S 75%
- 44% bilateral SNHL

Meniere's Disease

- Fluctuating SNHL, episodic vertigo, aural fullness
- ? Autoimmune etiology
 - 97% with CICs (Derebery)
 - response to immunotherapy
 - 32% with anti-68kDa antibody





Treatment

- Steroids
- Cyclophosphamide
- Plasmapheresis
- Methotrexate
 - dihydrofolate reductase inhibitor



Complications of therapy

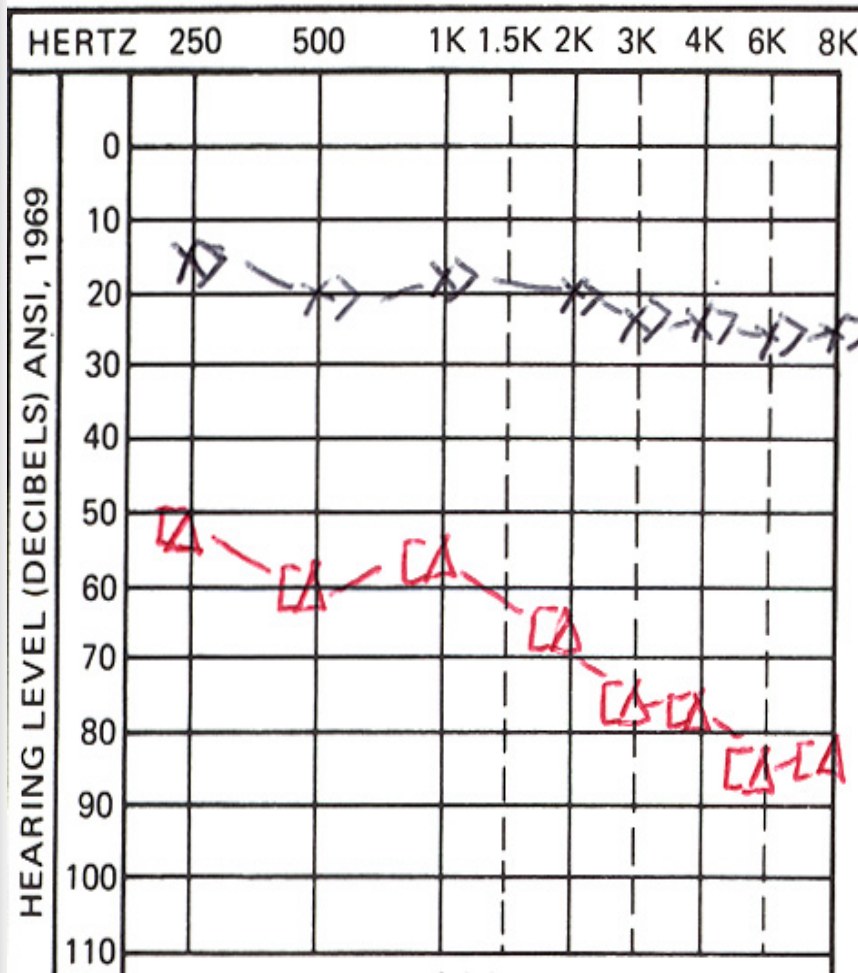
Corticosteroids	Cyclophosphamide
Sodium retention	Nausea-vomiting
Potassium loss	Alopecia, skin rash
Fluid retention	Leukopenia
Congestive heart failure	Interstitial pulmonary fibrosis
Hypertension	Hemorrhagic cystitis
Muscle weakness	Hemorrhagic myocarditis
Myopathy	Carcinogenesis (urinary bladder, myeloproliferative malignancies)
Osteoporosis	Sterility
Aseptic necrosis of femoral and humeral heads	
Peptic ulcer perforation, hemorrhage	
Glaucoma	
Increased intraocular pressure	
Cataracts	
Increased intracranial pressure	
Manifestations of latent diabetes mellitus	
Cushingoid state	
Pituitary, adrenocortical insufficiency	
Nervousness	
Insomnia	



Case Study

- 45 year old female
- right sided hearing loss and aural fullness, dysequilibrium progressive over 2 months time
- physical normal except Weber AS, Rinne positive AU

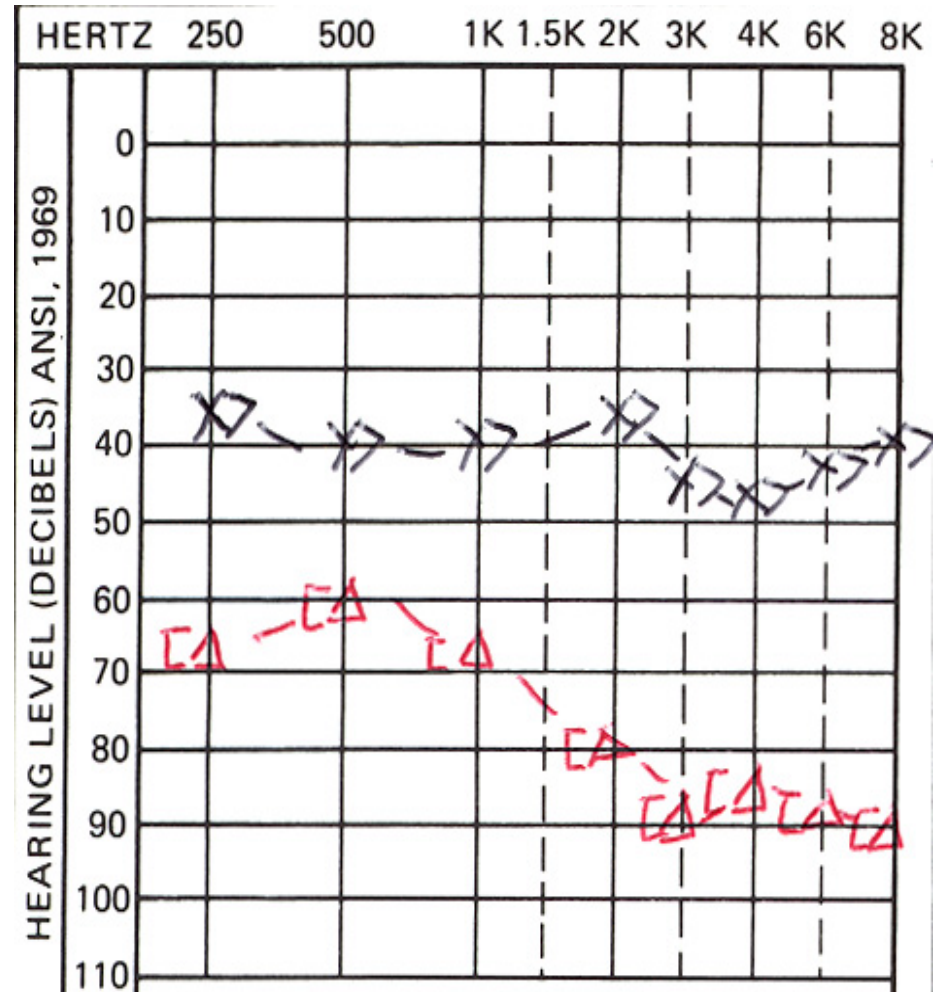
Case Study, continued



- CBC, chemistries, TFTs, RPR, ESR normal
- MRI acoustic protocol normal
- low salt diet, Dyazide

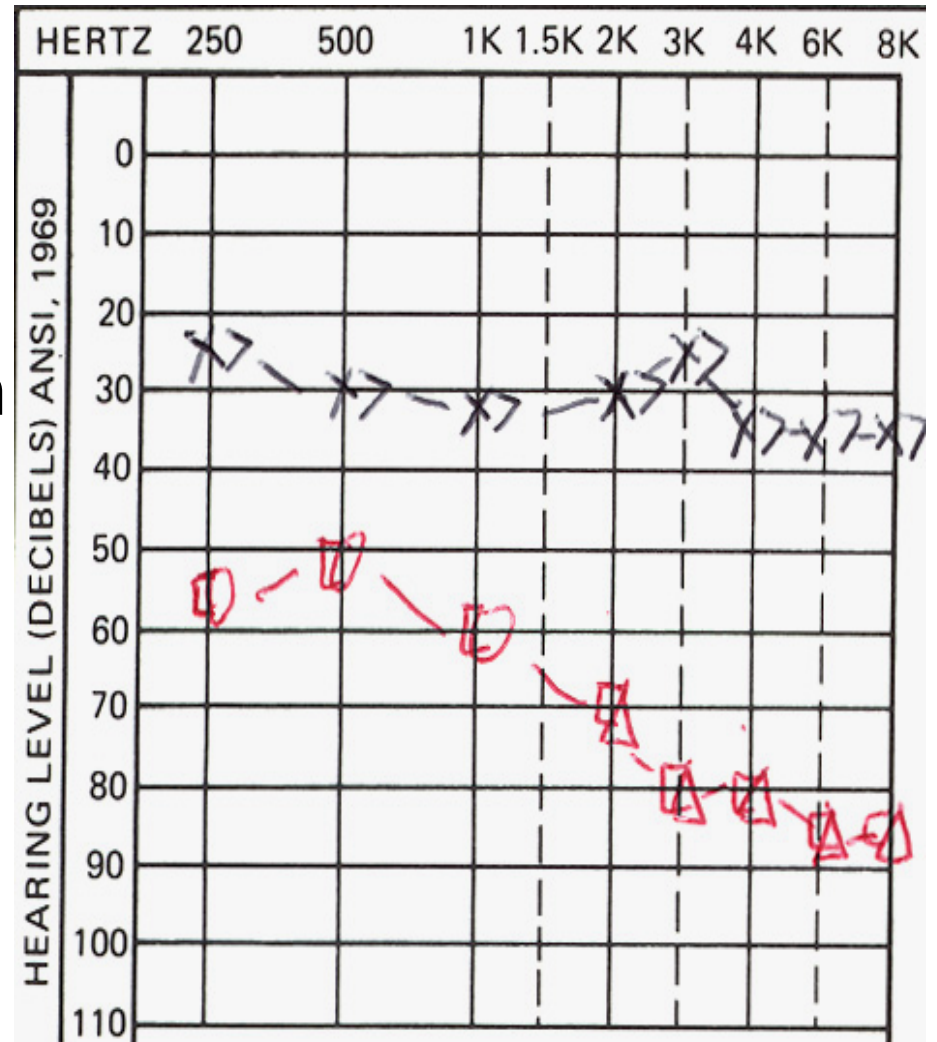
Case Study, continued

- At follow-up, AD hearing worse
- Prednisone 30 mg BID
- anti-68kDa protein positive



Case Study, continued

- Hearing improved
- steroids tapered
- one relapse, again with improvement on steroids





Conclusion

- Elusive etiology, diagnosis and treatment
- potentially treatable cause of progressive SNHL
- need less toxic therapy