Chapter 145: Otologic Manifestations of Systemic Disease

Congrats!

The Laryngoscope

Glossopharyngeal Schwannomas: A 100 Year Review

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Objectives: To review the literature on glossopharyngeal schwannomas with a focus on clinical presentation, radiologic/audiologic characteristics, and management options, and to propose a mechanism explaining the nature of vestibulocochlear dysfunction seen with these tumors.

Study Design: Contemporary review.

Methods: English literature search for cases of primary isolated glossopharyngeal schwannomas and chart review of two new cases.

Results: A total of 42 glossopharyngeal schwannoma cases between 1908–2008 were reviewed. Of these 84% presented with vestibulocochlear symptoms whereas only 30% presented with glossopharyngeal symptoms. Tumors can occur anywhere along the CNIX; however, the majority of symptomatic cases are intracranial/intraosseous, which present with vestibulocochlear dysfunction. Reviewed cases typically described the caliber of CNVII and VIII on CT/MRI as normal. We present a tumor's location posterior and medial to CNVIII combined with the complex CNVIII tonotopic organization may account for the preferential mid-frequency hearing loss seen in these patients.

Key Words: Glossopharyngeal, schwannoma, jugular foramen, mid frequency, sensorineural hearing loss.

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INTRODUCTION

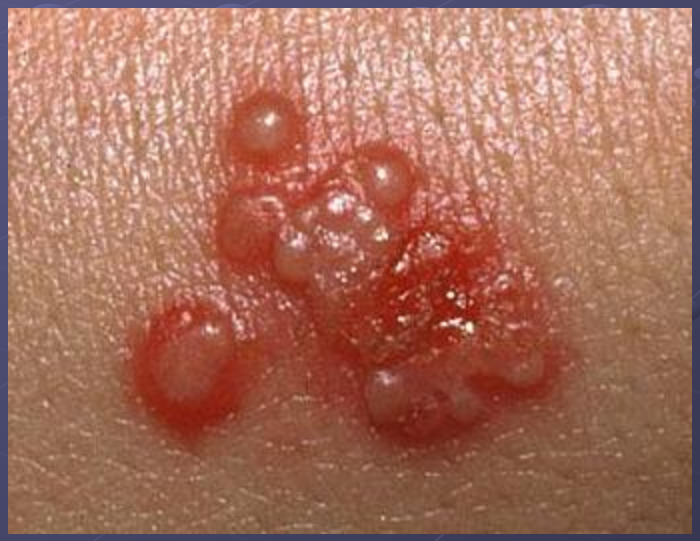
Jugular foramen schwannomas comprise only 2.9% of all intracranial schwannomas.[1] Among cranial nerves of the jugular foramen, schwannomas originate from the ninth cranial nerve the second most often.[2] We review the literature on glossopharyngeal schwannomas with a focus on correlating radiological findings with audiological characteristics and clinical presentation. Furthermore, we illustrate two more cases to contribute to the limited literature.

Otologic Manifestations of Systemic Disease

- ***Infectious***
  - Herpes simplex
  - Varicella-zoster
  - Mumps
  - Measles
  - Syphilis
  - Lyme disease
  - Leprosy
  - Tuberculosis
  - Human immunodeficiency virus
- ***Immunologic***
  - Wegener granulomatosis
  - Relapsing polychondritis
  - Discoid lupus erythematosus
  - Systemic lupus erythematosus
  - Polyarteritis nodosa
  - Rheumatoid arthritis
  - Autoimmune inner ear disease
- ***Metabolic***
  - Gout
  - Ochronosis
  - Mucopolysaccharidoses
- ***Diseases of bone***
  - Osteogenesis imperfecta
  - Osteopetrosis
  - Paget disease
  - Fibrous dysplasia
- ***Idiopathic***
  - Histiocytosis X

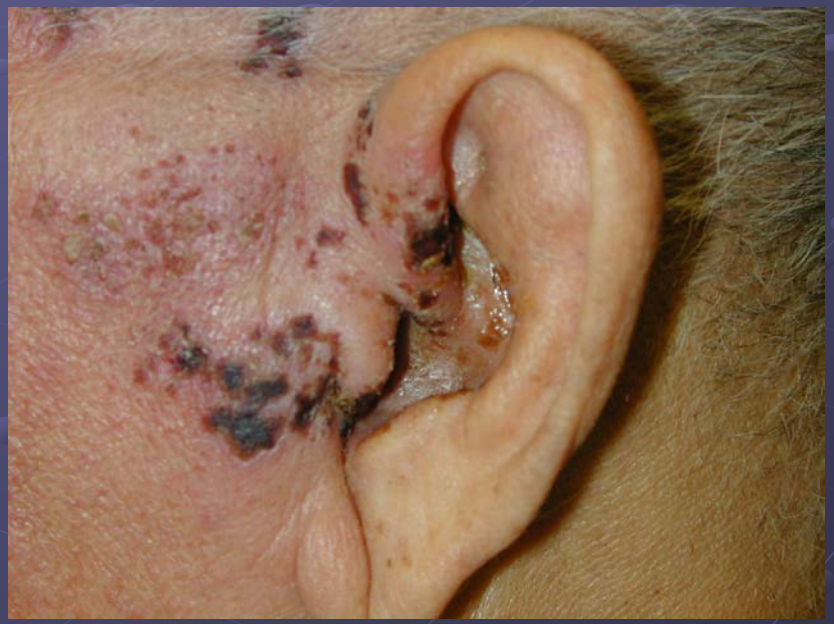
Herpes Simplex

- Caused by Herpes Simplex Virus (DNA virus)
- Primary infection occurs in patients without prior immunity (mostly infants/children)
- May present with gingivostomatitis, keratoconjunctivitis, or meningoencephalitis
- Otologic Manifestations: vesicles on skin of auricle and ear (herpetic external otitis)
- Rx: Symptomatic, though antivirals may be used and are most effective in prodromal phase of infection (Acyclovir 400mg TID for 7-10 days)



Varicella-Zoster

- Two clinical disease: varicella (chickenpox) and herpes zoster (shingles)
- Herpes Zoster Oticus results from reactivation of VZV from geniculate ganglion
- Vesicles can erupt along skin of auricle (sensory innervation of facial nerve)
- Ramsay Hunt Syndrome
  - Facial nerve paralysis (typically 1-2 days after vesicular eruption)
  - Treatment recommended even for immunocompetent patients (improved return of facial nerve function)
  - Steroids + Acyclovir (800 mg PO 5 times a day x 7-10 days)
- May also present with hearing loss and vestibular symptoms due to inflammatory reaction of 8th nerve

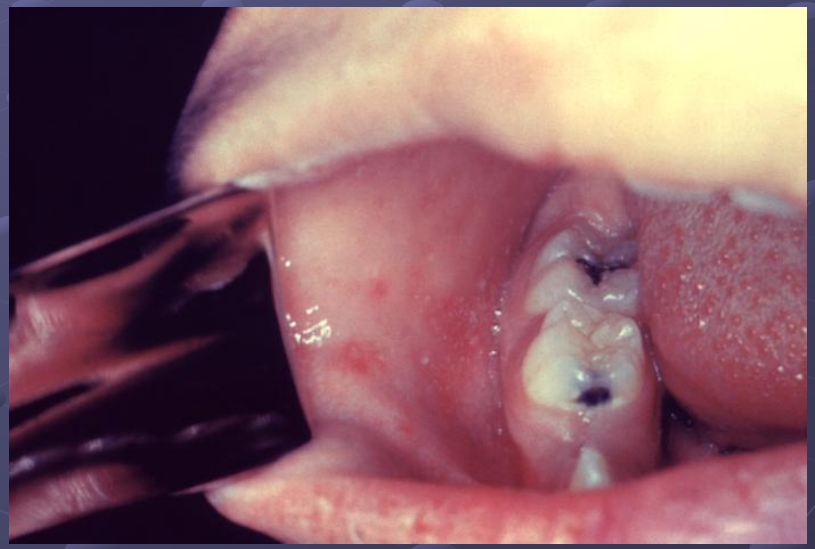


Mumps

- RNA Paramyxovirus
- Can present with salivary adenitis, epididymoorchitis, pancreatitis, thyroiditis, polyarthritis, and myocarditis
- Otologic Manifestions:
  - Hearing loss: incidence 5 out of 10,000 patients, SNHL, unilateral in 80% of cases
  - Develops towards the end of the parotitis phase
  - Tends to be more severe in the high frequency range
  - Loss is usually permanent
  - Vestibular symptoms may also be present
- Pathogenesis: direct viral invasion of the cochlea via the perilymph
- Rx: Supportive +/- corticosteroids (diminish fevers and pain from parotid and testicular swelling)

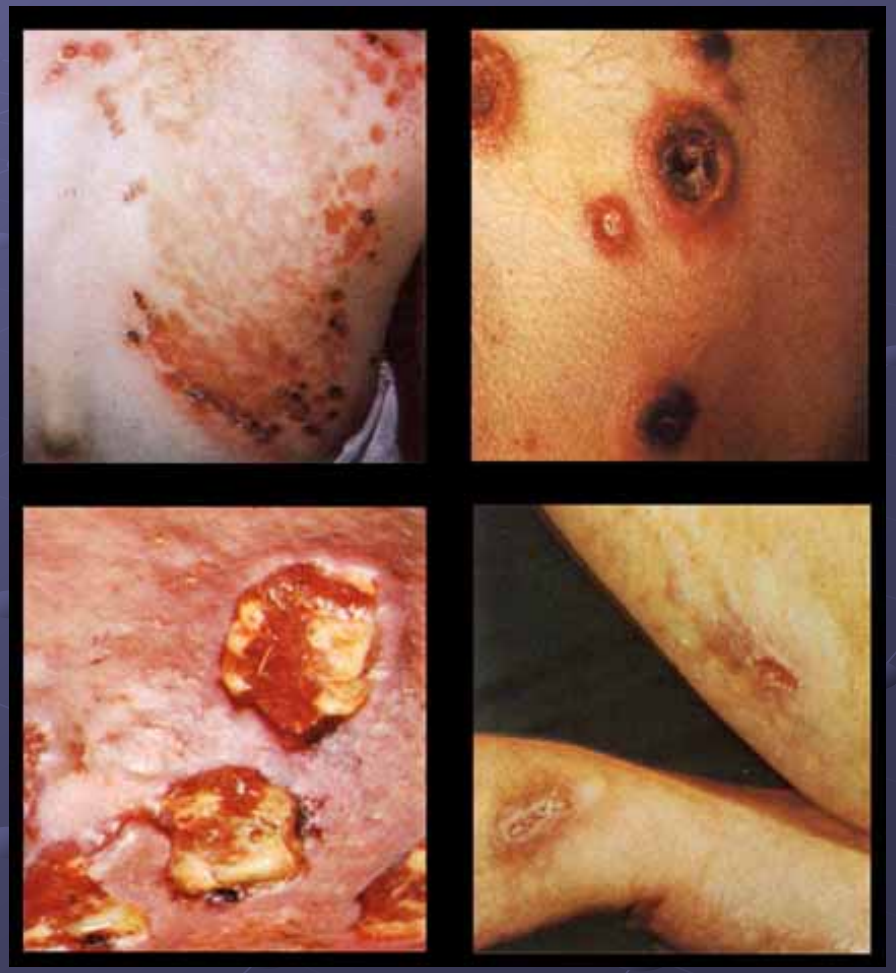
Measles

- RNA Virus
- Prior to advent of vaccination, accounted for up to 10% of acquired deafness in children (overall incidence is <0.1% of measles infections)
- Bilateral SNHL → occurs abruptly at the same time as macular rash
  - 45% of affected individuals will have profound loss
  - HL worse at higher frequencies
  - Typically permanent
- May also be associated with vestibulopathy
  - 70% of patients suffering from hearing loss having associated diminished vestibular responses



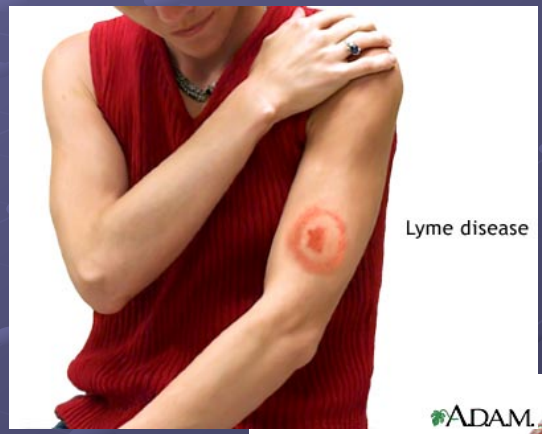
Syphilis

- Treponema palladium (spirochete)
- Primary, secondary, latent, tertiary phases
- Manifestations of Otosyphilis
  - Secondary Phase: abrupt hearing loss, bilateral, rapidly progressive
  - Tertiary Phase: related to neurosyphilis
    - Connection between CSF and perilymph via cochlear aqueduct
    - Hearing loss is typically asymmetric, fluctuating, associated with episodic vertigo (can mimick MD)
    - Positive fistula test (Hennebert sign) due to hypermobile stapes footplate
    - Gumma formation within ME, can cause TM perforation
  - Rx: PCN, prednisone can be used for hearing loss (30-60 mg/day for 3-6 months)
    - 50% of patients with HL will improve after steroids



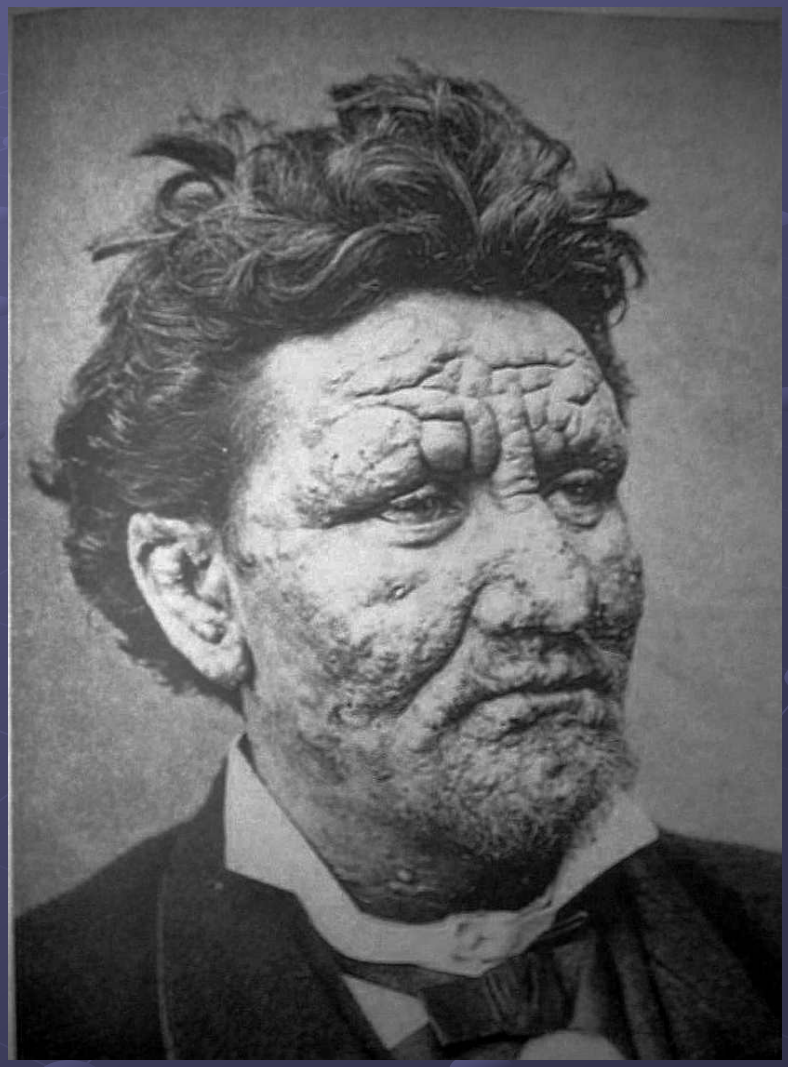
Lyme Disease

- Borrelia burgdorferi (transmitted from *Ixodes* tick bite)
- Facial paralysis (unilateral or bilateral): resolves slowly over 6-12 months
  - Also: tinnitus, SSNHL, otalgia
- Rx: single agent therapy (doxy, amoxicillin, PCN, tetracycline, cefuroxime) x 14 days



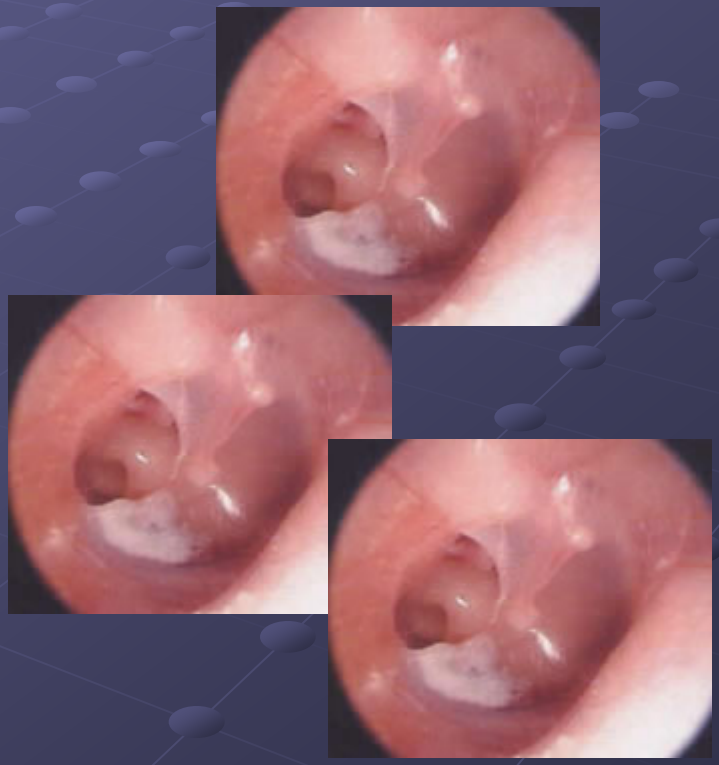
Leprosy

- Mycobacterium leprae; nasal mucosa is primary site of transmission
- Classified as tuberculoid, borderline tuberculoid, borderline, borderline lepromatous, or lepromatous
- Present with skin lesions, numbness, ulcerations, leonine facies
- Propensity to infect nerves, including *greater auricular nerve*, ulnar, radial, and peroneal
- 70% will have manifestations on the auricle (nodules, loss of cartilage, ulceration)
- Diagnosis: 1 or more of the following symptoms
  - hypopigmented or reddish patches with definite loss of sensation
  - thickened peripheral nerves and skin
  - nerve biopsies showing acid-fast bacilli in modified mononuclear or epithelioid cells called lepra cells
- Rx: oral administration of dapsone +/- rifampin (for resistant strains)



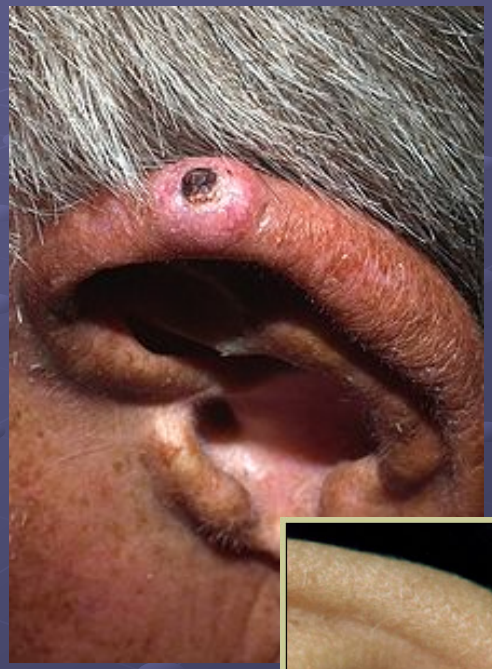
Tuberculosis

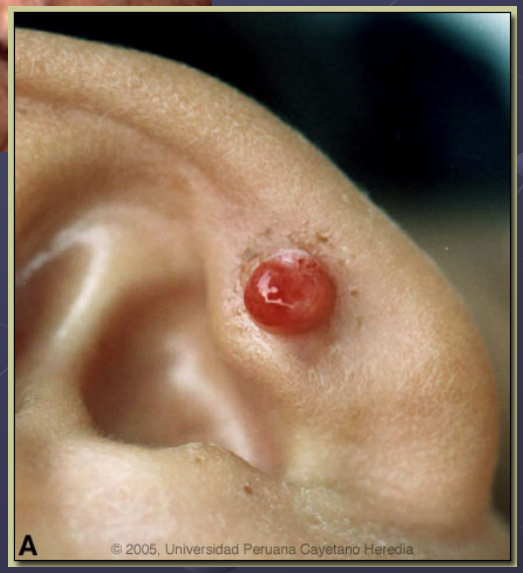
- Mycobacterium tuberculosis
- Can affect multitude of sites within H&N: larynx, pharynx, oral cavity, nasal cavity, salivary glands, cervical spine, *middle ear, and mastoid*
- Infections of ME can be caused either via hematogenous spread or ascending via the ET from the nasopharynx
- Otologic manifestations: facial paralysis, persistent aural discharge, refractory OM, perichondritis, failed otologic surgery for persistent otomastoiditis
- Middle ear infections may present as painless otorrhea and multiple TM perforations
- Can develop CHL due to ossicular erosion, and SNHL from labrynthitis at later stages of infection
- Dx: AFB stain, M.Tb cultures (definitive), newer technique is PCR analysis
- Rx: multidrug cocktail including INH, rifampin, pyrazinamide, pyridoxine +/- surgical intervention to remove pathologic tissue (if refractory to medical management)



HIV

- RNA retrovirus
- 20-30% of affected patients will report otologic symptoms, typically due to opportunistic infections
- Most commonly recurrent OM, OE, as well as SOM from adenoid hypertrophy, lymphoma, and NPC
- Similar pathogens to immunocompetent patients with addition of opportunistic organisms
- Rx: same as that in immunocompetent patients
- Also: Kaposi sarcoma vs. Bacillary Angiomatosis





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  - Mucopolysaccharidoses
- ***Diseases of bone***
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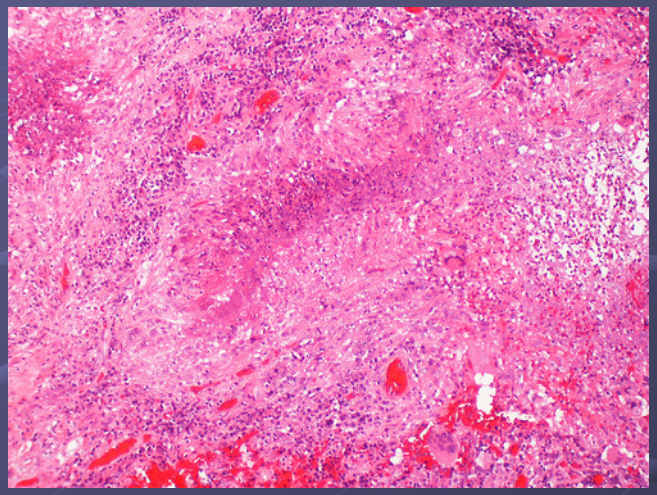
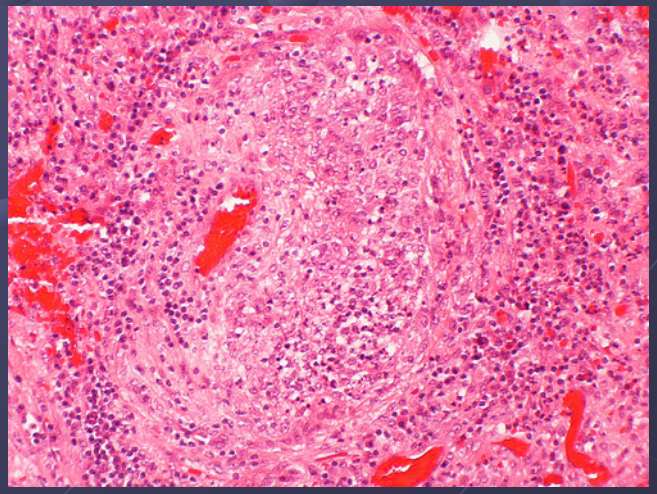
  - Osteopetrosis
  - Paget disease
  - Fibrous dysplasia
- ***Idiopathic***
  - Histiocytosis X

Wegener's Granulomatosis

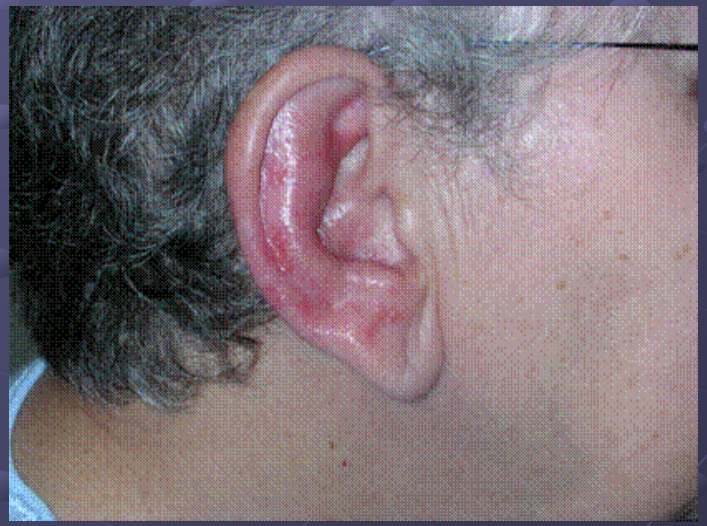
- Immunologically mediated systemic disease of unknown etiology
- Otologic involvement in ~35% of cases
  - Facial or postauricular pain
  - OM (AOM, CSOM, SOM) → TM perforations + otorrhea
  - Hearing loss, both conductive and sensorineural (from immune complex deposits in cochlea, vasculitis of the vasa vasorum of the cochlea, or pressure on the acoustic nerve by a granulomatous lesion) as well as vertigo
- Rx: corticosteroids and immunosuppressives such as cyclophosphamide, azathioprine, or methotrexate





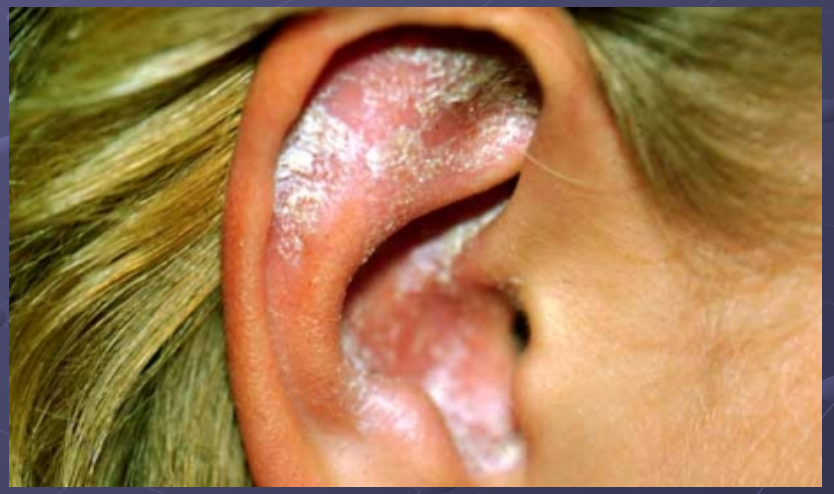
Relapsing Polychondritis

- Autoimmune origin
- Characterized by episodic inflammation of cartilaginous structures (ear, nose, trachea, larynx, ribs, and eustachian tubes) leading to atrophy, scarring, and distortion of the involved cartilage
- Otologic: red, tender, edematous pinna with sparing of the external ear canal (used to differentiate from OE)
- Dx: clinical → avoid biopsy due to risk of infection in acute inflammation; +ESR, RF
- Rx: corticosteroids



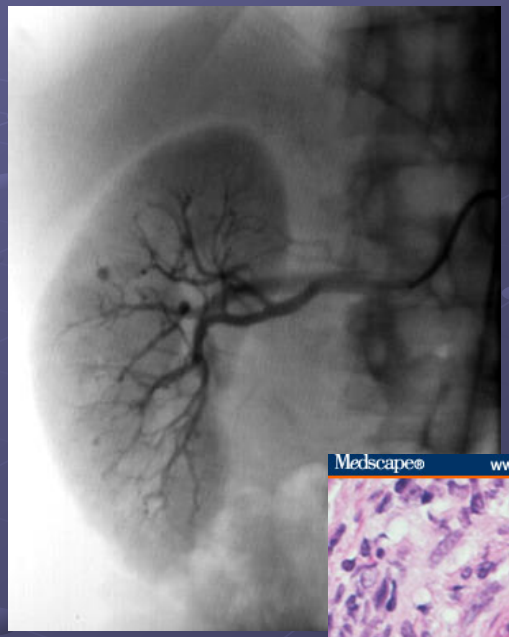
Chronic Discoid Lupus Erythematosus

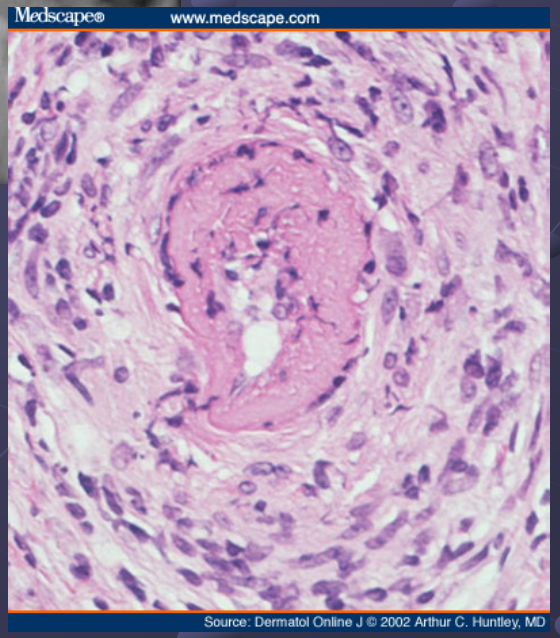
- Discoid version is limited to skin
- Immune complexes at the epidermal-dermal junction
- Can present with lesions on the external ear
- Rx: topical corticosteroids, sun exposure avoidance



Polyarteritis Nodosa

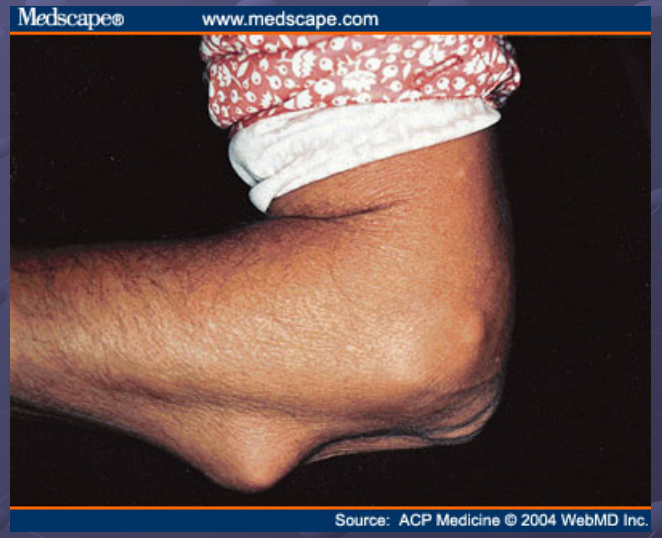
- Necrotizing vasculitis of small- and medium-diameter arteries
- Dx:
  - tissue biopsy showing PMNs invading all layers of the vessel walls with deposition of immune complexes
  - angiography of affect organisms show small aneurysms at the branch points of vessels
  - HepB surface Ag + in 30-40% of patients
- Otologic Manifestations:
  - CHL or SNHL
- Rx: corticosteroids +/- cyclophosphamide





Rheumatoid Arthritis

- Inflammatory synovitis
- Associated with HLA-DR4 expression
- Otologic Manifestations:
  - Rheumatoid nodules in external ear
  - CHL: due to involvement of the ossicular suspensory ligaments and IS joints
  - SNHL: autoimmune inner ear disease
- Rx: ASA, NSAIDs, glucocorticoids, immunosuppressives



Vogt-Koyanagi-Harada Syndrome

- Autoimmune multiorgan disease involving eyes, ears, skin, and nervous system
- Three phases
  - First phase: meningoencephalitis
  - Second phase: loss of visual acuity, *tinnitus, and hearing loss*
  - Convalescence phase
- Rx: corticosteroids + immunosuppressive drugs

Autoimmune Ear Disease

- Fluctuating, progressive SNHL, bilateral but often asymmetric, half will also have vestibular symptoms
- May have concurrent autoimmune disease such as SLE or RA
- Dx: clinical based on rapidly progressive, fluctuating hearing loss that responds to corticosteroid therapy; elevation of non-specific rheumatologic testing such as ESR, ANA
- Rx: corticosteroids versus cyclophosphamide and MTX, after hearing loss stabilized, can tape dose very slowly (some may require long term maintainence therapy)

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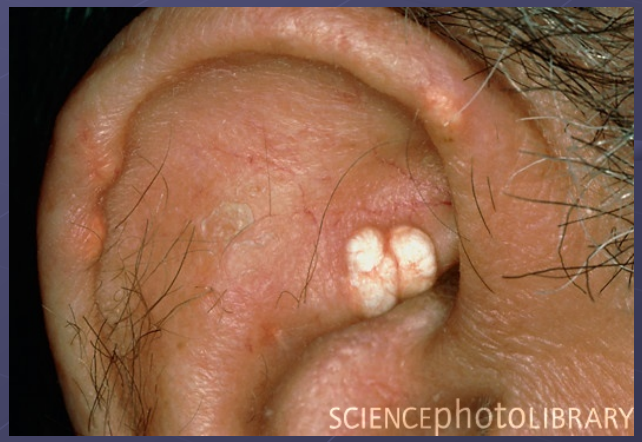
- ***Idiopathic***
  - Histiocytosis X

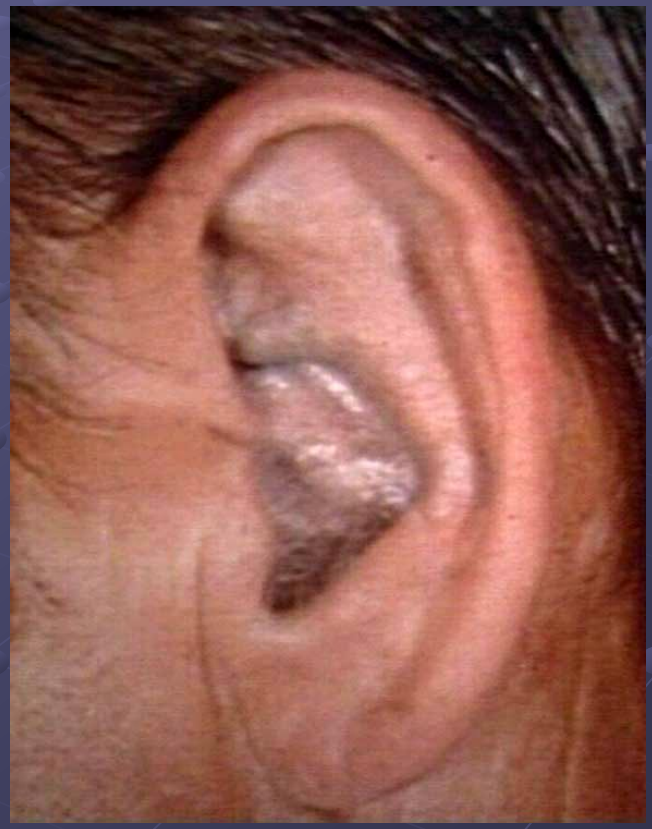
Gout

- Disorder of purine metabolism, high uric acid levels leading to deposition in soft tissues and joints
- Otologic Manifestations: auricular tophi
- Rx: allopurinol (overproducers) versus probenacid (underexcreters)



Ochronosis

- AR disorder of homogentisic acid metabolism
- Accumulation of HA, which adheres to cartilage and can discolor the ear, fingers, buccal mucosa, and nose
- Dx: clinical (urine turning black on exposure to sunlight is pathognomonic)
- Rx: low protein diet



Mucopolysaccharidoses

- MPS storage diseases (Hunter, Hurler, Morquio)

- Otologic Manifestations:
  - Mixed HL
  - Conductive component due to ETD with SOM
  - SNHL component possible due to abnormal lipid metabolism within nerve cells (not understood)

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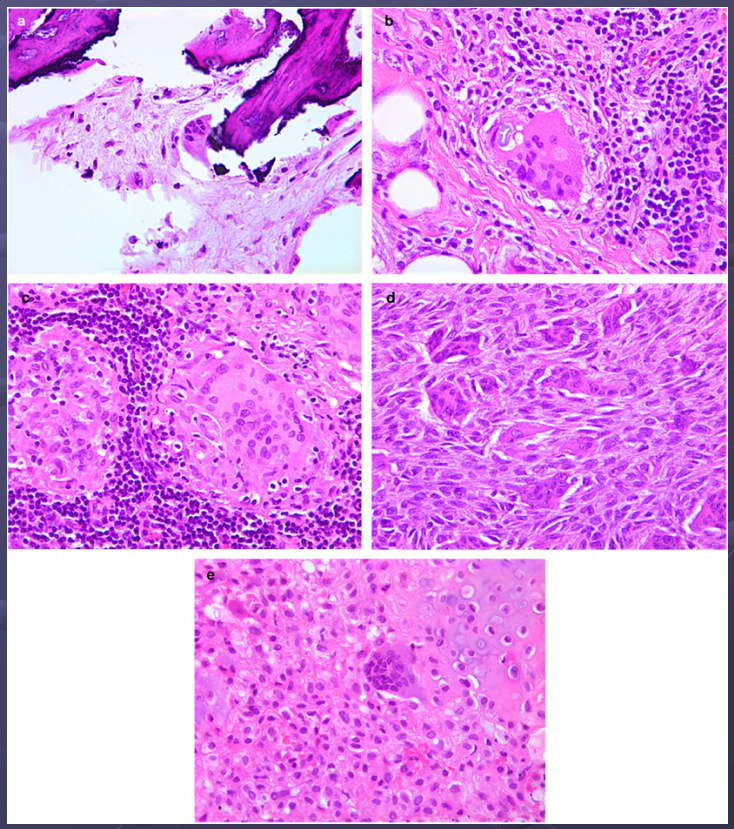
Osteogenesis Imperfecta

- AD disorder of connective tissue which causes high susceptibility to pathologic fractures
- Can vary in severity from lethal form in perinatal period to mild forms which may be subclinical
- Otologic Manifestations:
  - CHL and SNHL seen in 90% of individuals > 30 years old
  - CHL due to pathologic fractures of the ossicles, fixation of stapes footplate
  - CHL is associated with blue sclera,
  - SNHL usually is associated with gray or white sclera
- Rx: HAs versus stapedectomy

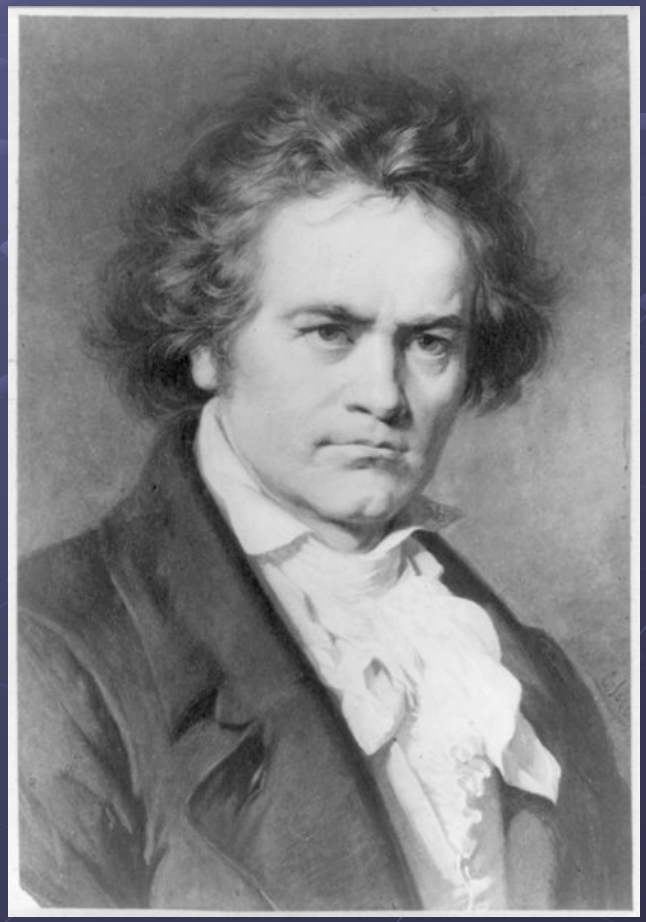
Osteopetrosis

- Inherited disroder (AD or AR) causing faulty remodeling of bone
- Otologic Manifestations:
  - Progressive cranial neuropathies due to compression at the foramina leading to SNHL, facial nerve paralysis
  - CHL can be secondary to thickening of the ossicles versus fixation of the stapes footplate



Paget's Disease

- Disorder of increased activity of both osteoblasts and osteoclasts, bony hypertrophy and remodeling
- Otologic Manifestations:
  - Mixed HL; though mechanism is incompletely understood, thought to be related to changes in bone density that dampen the normal hearing mechanism within both middle and inner ears
  - Hearing loss is not due to ossicular fixation or compression of CN8
- Rx: calcitonin and etidronate disodium to decrease osteoclast activity



Fibrous Dysplasia

- Replacement of cancellous bone by spicules of woven bone in a fibrous stroma
- Can involve the temporal bone leading to enlargement of the squamosa, mastoid, and bony EAC
- Can monostotic (70%), polyostotic, or as a part of McCune-Albright syndrome (polyostotic fibrous dysplasia, precocious puberty, and abnormal skin pigmentation)
- Otologic Manifestations:
  - CHL (80%) either due to EAC occlusion involvement of ossicular chain
  - Progressive occlusion of the external auditory canal (85%)
  - Canal cholesteatoma (40%)

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Langerhans Cell Histiocytosis

- Encompasses a group of disease including eosinophilic granuloma, Hand-Schuller-Christian disease, Letterer-Siwe diease
- Accumulation of histiocytes, lymphocytes, and eosinophils in the skin, bone marrow, lymph nodes, lung, liver, thymus, spleen, and CNS
- Cytoplasmic inclusion bodies known as Birbeck granules on EM
- Otologic:
  - Involvement of external auditory canal, temporal bone
  - Otorrhea
  - Aural granulation tissue
  - Mastoid swelling
  - Collapse of the EAC
  - SNHL, facial nerve paresis
- Rx: steroids, non-responders treated with etoposide, vincristine, or vinblastine