Infections of the Labyrinth

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Grand Rounds Presentation
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Labyrinthitis

- Inflammatory process involving the inner ear
  - infectious vs. non-infectious
  - generalized vs. circumscribed
  - acquired vs. congenital
  - isolated vs. systemic
Pathogenesis

• Meningogenic
  – spread through IAC or cochlear aqueduct

• Tympanogenic
  – spread through round or oval windows

• Hematogenic
  – spread through vascular channels
Bacterial Infections

• Toxic Labyrinthitis
  – sterile inflammation
  – bacterial toxins penetrate perilymphatic spaces
  – mild hearing loss or mild vestibular loss
  – usually resolves without sequelae

• Suppurative Labyrinthitis
  – bacterial invasion of the inner ear
  – intense inflammatory reaction
  – progresses along four pathologic stages
  – medical emergency
Suppurative Labyrinthitis

• Presentation--acutely ill patient with severe vertigo, N/V, profound hearing loss
• Look for signs of associated meningitis or otitis media
• Hospitalization, hydration, vestibular suppressants, IV antibiotics
Bacterial Labyrinthitis

- Meningogenic
  - S. pneumoniae
  - N. meningitidis
  - H. influenzae

- Tympanogenic
  - S. pneumoniae
  - H. influenzae
  - M. catarrhalis
  - Pseudomonas
  - Proteus
  - Anaerobes
Bacterial Meningitis

- Incidence of post-meningitic hearing loss is 10-20%
- Hearing loss occurs early in the course of meningitis
- Most often bilateral, severe to profound, and permanent
- Management = Antibiotics +/- Steroids
FIG. 145-1. Follow-up protocol for the postmeningoitc child.
Syphilis

• Congenital Syphilis
  • primary maternal infection = 70-100% transmission
  • early congenital syphilis
    » symmetrical, flat, profound SNHL
  • late congenital syphilis
    » asymmetric, fluctuating, variable severity SNHL
    » lower discrim scores than expected with PTA

• Acquired Syphilis
  • secondary or tertiary disease
    » hearing loss similar to late congenital infection
Syphilis

• Diagnosis
  – non-specific screening tests
  – specific confirmatory tests

• Treatment
  – Penicillin
  – +/- Steroids
Syphilis

• Temporal Bone Findings
  – Early congenital/Acute acquired
    • round cell invasion of CN VIII, nerve fiber loss
    • degeneration of organ of Corti and spiral ganglion
    • fibrinous exudate and hemorrhage
  – Late congenital/Late acquired
    • obliterative endarteritis
    • round cell osteitis
    • gumma formation
Viral Infections

- May present as congenital syndrome, systemic illness, or isolated inner ear infection
- Definitive infection has been identified only with CMV and mumps virus
- Suspects include: rubella, rubeola, influenza, varicella-zoster, EBV, poliovirus, RSV, adenovirus, parainfluenza, herpes simplex
Cytomegalovirus

- Most common congenital infection in U.S.
  - 1% of all live births
- Infection via transplacental transmission, passage through infected birth canal, ingestion of infected breast milk
- 40% transmission rate with primary maternal infection; .15%-1.0% transmission rate from seropositive mothers
Cytomegalovirus

- 90% asymptomatic at birth
  - 10-15% develop SNHL
    - variable in severity
    - risk factors include periventricular calcifications, high maternal antibody titers

- 10% symptomatic at birth
  - 90% with cytomegalic inclusion disease
  - 65% with SNHL
    - bilateral, severe to profound, permanent
Cytomegalovirus

• **Diagnosis**
  - viral culture
  - specific antibody testing

• **Treatment**
  - acyclovir, gancyclovir, foscarnet
  - vaccine
Cytomegalovirus

• Temporal Bone Findings
  – Hematogenic Spread
    • stria vascularis
    • endolymphatic spaces
  – Meningogenic Spread
    • CN VIII, cochlear aqueduct
    • perilymphatic spaces
Rubella

- Decrease in incidence since introduction of rubella vaccine
- Transmission to fetus associated with primary maternal infection
  - First trimester = 90% symptomatic
  - Second/Third trimester = 25-50% symptomatic
Rubella

• Congenital Rubella Syndrome
  • triad of cataracts, heart deformities, SNHL

• Hearing Loss
  • 50% of symptomatic infants
  • 10-15% of asymptomatic infants
  • variable in severity, “cookie-bite” pattern on audiogram, permanent
Rubella

• **Diagnosis**
  • viral culture
  • specific antibodies

• **Treatment**
  • vaccination
  • antepartum screening
  • auditory rehabilitation
Rubella

• Temporal Bone Findings
  – Scheibe malformation
  – collapse of Reissners membrane
  – tectorial membrane abnormalities
  – atrophy of stria vascularis
Mumps

• Triad of parotitis, orchitis, meningo-encephalitis
• Primarily affects children and young adults
• Hearing loss in .05% of cases
  • presents as parotitis is resolving
  • 80% unilateral, maximal in HF, severe to profound, permanent
  • associated tinnitus and aural fullness
Mumps

- **Diagnosis**
  - viral culture
  - specific antibodies

- **Treatment**
  - vaccination
  - auditory rehabilitation
Mumps

• Temporal Bone Findings
  – Hematogenic spread
    – infection of stria vascularis and endolymph
    – degeneration of organ of Corti, tectorial membrane and cochlear neurons
  – Meningogenic spread
    – spread through CN VIII or cochlear aqueduct into perilymph
    – degeneration of modiolar neural elements
    – fibrosis/ossification of perilymph spaces
Measles

- Triad of rash, conjunctivitis, Koplik spots
- Hearing loss seen in less than 1 per 1,000 cases
  - variable in severity
  - unilateral or bilateral
  - worse in high frequencies
  - permanent
Measles

• Diagnosis
  • viral isolation
  • specific antibodies

• Treatment
  • vaccination
  • auditory rehabilitation
Measles

• Temporal Bone Findings
  – cochlear degeneration
  – atrophy of stria vascularis
  – abnormalities of tectorial membrane
  – macular degeneration
Varicella-Zoster

• Primary infection = chicken pox

• Reactivation = zoster
  – Ramsay Hunt syndrome
    • vesicles on pinnae or EAC
    • facial weakness/paralysis
    • SNHL
Varicella-Zoster

- **Diagnosis**
  - clinical presentation
  - culture of vesicular fluid

- **Treatment**
  - antiviral therapy
  - steroids
  - analgesics
Varicella-Zoster

• Temporal Bone Findings
  – facial nerve inflammation
  – vestibulocochlear nerve inflammation
  – destruction of organ of Corti
  – eventual fibrosis and ossification
Herpes Simplex

- HSV-1
  - reactivation within spiral ganglion causing SSNHL
- HSV-2
  - encephalitis with spread along CN VIII to perilymph
Human Immunodeficiency Virus

• Presentation may include sudden hearing loss, tinnitus or vertigo
• Mechanisms include direct infection of labyrinth with HIV, opportunistic infection, neoplasm, or drug ototoxicity
• Most common finding is mild SHNL
Human Immunodeficiency Virus

- Temporal Bone Findings
  - isolation of CMV, adenovirus, HSV-1
  - invasion with pneumocystis, cryptococcus
  - hair cell inclusions, viral-like particles
Protozoal Infections

• Toxoplasmosis
  – Congenital infection
    • triad of chorioretinitis, hydrocephalus, intracranial calcifications
    • first trimester infection associated with severe manifestations
    • third trimester infection associated with highest transmission rate
    • 75% are asymptomatic at birth
    • up to 85% later present with hearing loss
Toxoplasmosis

• **Diagnosis**
  - maternal infection
    » IgG seroconversion or rise in titers
  - fetal infection
    » mouse inoculation or PCR of amniotic fluid
    » umbilical cord blood IgM or quantitative IgG

• **Treatment**
  - combination therapy with pyrimethamine and sulfonamide
    » 70% reduction in transmission
    » reduction in sequelae
Fungal Infections

- Occur in immunocompromised hosts
- Reported pathogens include Mucor, Cryptococcus, Candida, Aspergillus, and Blastomyces
- Meningogenic, Tympanogenic, Hematogenic spread to the labyrinth
- Treat with appropriate antifungals
Acute Cochlear Labyrinthitis
ISSNHL

• Definition
  – 30 dB deficit
  – 3 contiguous frequencies
  – over a 3 day period

• Mechanism
  – viral infection
    – 30-50% report recent URI
  – vascular compromise
  – membrane rupture
ISSNHL

- 90% unilateral
- variable in severity
- sudden in onset
- painless
- associated with tinnitus or aural fullness
- associated vestibular dysfunction
ISSNHL

- **Differential Diagnosis**
  - autoimmune, trauma, neoplasm, ototoxic meds, vascular accidents
- **Labs**
  - CBC, ESR, glucose, FTA-ABS
- **Imaging**
  - CT, MRI
- **Auditory/Vestibular Testing**
  - audiogram, ENG
ISSNHL

• Treatment
  – Steroid therapy (Wilson, 1980)
    • double blind, controlled study
    • compared oral steroid to placebo
    • recovery rate of 61% in treatment group
    • recovery rate of 32% in control group
    • moderate hearing loss showed most improvement with steroid therapy
  – Antiviral therapy
    • interferon
    • acyclovir
ISSNHL

• Prognosis
  – 30-70% have partial/complete recovery
  – Good prognostic factors:
    • < 40 y/o
    • present within 10 days of onset
    • mild hearing loss
    • steroid therapy for moderate hearing loss
    • no vestibular symptoms
Acute Vestibular Labyrinthitis

• Diagnostic Criteria (Coatis)
  – 1. Acute, unilateral peripheral vestibular d/o without associated hearing loss
  – 2. Occurs most frequently in middle age
  – 3. A single episode of severe prolonged vertigo
  – 4. Decreased caloric response in the involved ear
  – 5. Resolution of symptoms in 6 months
Acute Vestibular Labyrinthitis

• Differential Diagnosis
  • Meniere’s disease, vestibular schwannoma, labyrinthine fistula, cerebellar infarction, multiple sclerosis, dysequilibrium of aging

• Auditory/Vestibular Testing
  • audiogram (by definition should be normal)
  • ENG

• Imaging
  • CT, MRI
Acute Vestibular Labyrinthitis

- **Treatment**
  - Supportive
    - hydration
    - vestibular suppressants
    - antiemetics

- **Prognosis**
  - Recovery/Compensation within 6 months
### Conclusion

<table>
<thead>
<tr>
<th>Type</th>
<th>History</th>
<th>Examination</th>
<th>Laboratory</th>
<th>Treatment</th>
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<tbody>
<tr>
<td>Bacterial</td>
<td>Abrupt onset of vertigo, associated with hearing loss, prior ear infections</td>
<td>May be signs of chronic otitis or acute meningitis</td>
<td>ENG: unilateral absent caloric response Audio: profound unilateral hearing loss CSF: pleocytosis CT: possible cholesteatoma</td>
<td>Local management of infection, parenteral antibiotics, vestibular suppressants, vestibular exercises ASAP</td>
</tr>
<tr>
<td>Viral</td>
<td>Vertigo develops over hours, resolves over days, prior flu-like episode, may or may not have associated hearing loss</td>
<td>Spontaneous nystagmus and imbalance for first few days; ears appear normal</td>
<td>ENG: Unilateral caloric hypoexcitability, usually returns to normal Audio: usually normal</td>
<td>High-dose steroids during acute phase, vestibular suppressants, vestibular exercises ASAP</td>
</tr>
<tr>
<td>Serous</td>
<td>Mild dizziness and disequilibrium, mild hearing loss</td>
<td>Signs of acute or chronic otitis</td>
<td>ENG: usually normal Audio: high-frequency loss CT: acute or chronic infection of middle ear</td>
<td>Parenteral antibiotics, vestibular suppressants if needed</td>
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<tr>
<td>Syphilitic</td>
<td>Recurrent episodes of vertigo lasting hours, associated tinnitus and hearing loss, prior congenital or acquired syphilis</td>
<td>May be stigmata of congenital syphilis, rarely associated signs of neurosyphilis</td>
<td>ENG: unilateral or bilateral decreased caloric response Audio: low-frequency hearing loss Serology: positive FTA-ABS CSF: usually normal</td>
<td>Penicillin IV or IM, steroids, vestibular suppressants for acute attacks</td>
</tr>
</tbody>
</table>

*ENG, Electronystagmography; Audio, audiogram; CSF, cerebrospinal fluid; CT, computed tomography; ASAP, as soon as possible; FTA-ABS, fluorescent treponemal antibody absorption test.*
23 y/o man presents to ENT clinic reporting complete loss of hearing in the right ear that he noticed upon awakening yesterday morning.
Case Presentation

• Additional History
  – no otalgia, no otorrhea
  – no associated vertigo
  – + right tinnitus
Case Presentation

• Past Medical History
  – none

• Past Surgical History
  – inguinal hernia repair age 17

• Medications
  – none

• Allergies
  – NKDA
Case Presentation

• Physical Examination
  – normal EAC and TM bilaterally
  – neurologic exam normal
  – Weber to left
  – Rinne: left AC>BC, right no response
Case Presentation

- Work-Up
Case Presentation

- Work-Up
  - Audiogram
Case Presentation

• Work-Up
  – Audio
  – Labs
Case Presentation

• Work-Up
  – Audio
  – Labs
  – Imaging
Case Presentation

• Differential Diagnosis
Case Presentation

- Differential Diagnosis
  - infectious labyrinthitis
    - viral
    - late congenital/acquired syphilis
  - autoimmune labyrinthitis
  - vascular accident
  - trauma
  - neoplasm
Case Presentation

- Treatment
Case Presentation

• Treatment
  – oral steroid taper
  – anti-viral meds
Case Presentation

• Follow-up?

• Prognosis?
Case Presentation

• Follow-up
  – repeat audio 2 weeks after presentation, steroid taper should be finished
  – repeat audio 6 weeks after presentation
Case Presentation

- **Good Prognostic Factors**
  - less than 40 y/o
  - presentation within 10 days of onset
  - no vestibular symptoms

- **Poor Prognostic Factor**
  - severe to profound loss on initial audiogram
  - associated with less than 20% chance of recovery regardless of intervention