Infections of the Labyrinth

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Infections of the Labyrinth

Labyrinthitis: inflammation of the inner ear

Multiple etiologies: infectious, autoimmune, systemic disease, trauma

Infectious agents include bacteria, viruses, fungus and protozoa.
Labyrinthitis

- Vestibular manifestations (vertigo)
- Cochlear manifestations (hearing loss)
- Both
Labrynthitis

- Infection usually occurs by one of three routes:
  - From the meninges
  - From the middle ear space
  - Hematogenous spread
Labyrinthitis

- Meningogenic: through the IAC, cochlear aqueduct, both (bilateral)
- Tympanogenic: extension of infection from the middle ear, mastoid cells or petrous apex - most common through the round or oval window (unilateral)
- Hematogenous: least common
Bacterial Infections

Two types of labyrinthitis associated with bacterial infections:

Toxic Labyrinthitis

Suppurative Labyrinthitis
Bacterial Infections

- Toxic Labyrinthitis: results from a sterile inflammation of the inner ear following an acute or chronic otitis media or early bacteria meningitis.
- Toxins penetrate the round window, IAC, or cochlear aqueduct and cause an inflammatory reaction in the perilymph space.
Bacterial Infection

Toxic Labyrinthitis produces mild high frequency hearing loss or mild vestibular dysfunction.

Treatment: Antibiotics for precipitating otitis, possible myringotomy.
Bacterial Infection

- Suppurative Labyrinthitis: direct invasion of the inner ear by bacteria.
- From otitis or meningitis
Bacterial Infection

- Suppurative Labyrinthitis: 4 stages
  - Serous or irritative: production of Ig rich exudates in the perilymph
  - Acute or purulent: bacterial and leukocyte invasion of the perilymphatic scala-end organ necrosis
  - Fibrous or latent: proliferation of fibroblasts and granulation tissue in the perilymph
  - Osseous or sclerotic: new bone deposition throughout the involved labyrinth
Bacterial Infection

- Purulent Labyrinthitis: medical emergency
- Meningitis or Otitis symptoms
- Hospitalization, hydration, vestibular suppressants and iv antibiotics
Bacterial Meningitis

- H. influenza B, N. meningitidis, S. pneumoniae

- Hib vaccine: 55% decrease in cases

- Pneumococcus now predominant org.
Bacterial Meningitis

- Postmeningitis hearing loss 10-20%
- Bilateral, severe to profound, permanent
Syphilis

- Treponema pallidum
- Diagnosis by FTA-ABS and confirmed by Western Blot.
- Congenital or Acquired
Syphilis

- Acquired: SNHL during secondary or tertiary
- Congenital:
  - Early: high fetal and infant mortality
  - Late SNHL+-/ vestibular symptoms
Viral Infections

- Congenital Infection
- Systemic viral illness
- Isolated involvement of inner ear
Cytomegalovirus

- Most common congenital infection in US
- Most common infectious cause of congenital deafness
- Low birth weight, jaundice, hepatosplenomegaly, petechiae, microcephaly and psychomotor retardation.
- 65% w SNHL-bilateral, severe to profound
CMV

- Diagnosis by isolating virus from urine during first few weeks of life.
- Virus isolation form cord blood
- No treatment: acyclovir may decrease amount of shedding, gancyclovir & foscarnet not approved during pregnancy.
Rubella

- 1969 58/100000
- 1983 0.5/100000
- Decline due to vaccine
- Congenital rubella: cataracts, heart malformations and SNHL, others
- Dx by viral culture
- No treatment; prevention only
Mumps

- Paramyxovirus
- Parotitis, orchitis, meningoencephalitis, and in 0.05% of cases-hearing loss.
- Hearing loss at end of first week of parotitis, unilateral and range from mild, high frequency SNHL to profound SNHL.
- Vestibular involvement is uncommon
Measles

- Rubeola virus
- Systemic illness w rash, conjunctivitis, and mucosal Koplik spots.
- Measles induced hearing loss is 1/1000 cases
- Measles less common 2nd to vaccine
Measles

- Encephalitis in 0.1% of cases with an overall mortality rate of 15%, with 25% of survivors with SNHL.
- SNHL seen in conjunction with rash.
- Sudden onset
- Varies from mild to profound HF SNHL
- Unilateral or bilateral
- PERMANENT
- 70% have vestibular losses also
Varicella-zoster

- Primary vzw=chicken pox
- HL w chicken pox = CHL 2nd to MEE
- Reactivation=zoster
- Herpes zoster oticus= Ramsay Hunt syndrome, reactivation from the geniculate ganglion of CN VII. Painful vesicles.
- 1/3 have auditory or vestibular symptoms- HFHL, hyperacusis, tinnitus, vertigo
Herpes simplex

- Labyrinthine infection by:
  - Reactivation in the spiral ganglion = SSNHL
  - Extension of the meningoencephalitis along CN VIII to the labyrinth = acquired SNHL
HSV

- HSV-1 & 2 can infect labyrinth. Animal models of ISSNHL. **Humans.**
- Neuroepithelial cells of the cochlea, utricle, saccule, and semicircular canals infected with HSV
- Circumstantial evidence only
Auditory and vestibular complaints rare in AIDS patients

Some with hearing loss, tinnitus and vertigo

Thought to be result of opportunistic infections (CMV, HSV), ototoxic drugs, neoplasm of inner ear.
Fungal Infections

- Fungal labyrinthitis is exceedingly rare outside the context of host immunocompromise.
- High risk: diabetics, chemo therapy, organ transplant recipients, AIDS patients
- Agents include Mucor, Cryptococcus, Candida, Aspergillus, and Blastomyces
- Hearing loss is severe and permanent
Protozoa

- Toxoplasma gondii most common
- Acquired infection usually asymptomatic
- Congenital infection may lead to severe malformations of fetus
- Triad of chorioretinitis, hydrocephalus, intracranial calcifications
- May also have microcephaly, cataracts, microphthalmia, jaundice, and hsm.
- 3000 cases annually
Toxoplasma

- 75% asymptomatic at birth
- 15% ocular problems
- 10% severe malformations
- 85% of symptomatic infants at birth will later develop decreasing visual acuity, decreased intellectual function, hearing loss or precocious puberty.
Toxoplasma

- Screening test to determine fetal infection
  - PCR analysis of amniotic fluid
  - IgM assays
  - Quantitative maternal/fetal IgG analysis of cord blood
Toxoplasma

- Treatment
  - Prenatal tx reduces both transmission and severity of illness in the fetus
  - Combination of pyrimethamine and sulfonamide
  - Neonates with documented infection should be given tx for 1\(^{st}\) year of life + folic acid supplements
Clinical Presentation

- Pts present with only auditory dysfunction - acute cochlear labyrinthitis
- Pts present with only vestibular dysfunction - acute vestibular labyrinthitis
- Both - acute cochleovestibular labyrinthitis
Clinical presentation

- Acute cochlear labyrinthitis, aka idiopathic sudden sensory neural hearing loss (ISSNHL)
- Defined as minimum of 30dB deficit in three contiguous frequencies over a period of less than 3 days in a previously healthy person.
- 3 pathologic theories: viral infection, vascular phenomenon, intralabyrinthine membrane rupture.
- Much circumstantial evidence of viral etiology
Acute cochlear labyrinthitis

- Treatment is steroids. Studies have shown no benefit of steroids and antivirals.
- 30-70% have complete recovery of hearing.
- Prognosis related to age, time from onset to presentation, type of audiogram, presence of vestibular symptoms
  - <40 years
  - Seen within 10 days
  - Started on steroids within 10 days
Clinical presentation

- Acute vestibular labyrinthitis, aka vestibular neuritis
- Defined as sudden unilateral vestibular weakness in the absence of concomitant auditory or CNS dysfunction in a previously healthy person
Acute vestibular labyrinthitis

- Diagnostic criteria:
  - An acute, unilateral, peripheral vestibular disorder w/o associated hearing loss
  - Occurrence predominantly in middle age
  - A single episode of severe, prolonged vertigo
  - Decreased caloric response in the involved ear.
  - Complete subsidence of the symptoms within 6 months
Acute vestibular labyrinthitis

- Treatment is supportive and includes hydration, antiemetics, and vestibular suppressants.
References

- Arbusow V.  HSV-1 not only in human vestibular ganglia but also in the vestibular labyrinth.  Audiology and Neuro-Otology.  6(%):259-62, Sept. 2001.
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