Cholesteatoma and Non-cholesteatomatous Inflammatory Disease

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Disclosures
• None

Overview
• Cholesteatoma
  – What is it?
• By location: cholesteatoma and ddx
  – EAC
  – Middle ear
  – Mastoid
  – Petrous Apex
• Non-cholesteatomatous inflammatory processes
  – Necrotizing external otitis
  – Facial nerve
  – Inner ear
  – Ossicular complications

Cholesteatoma
• Accumulation of desquamated keratin epithelium
• Acellular keratin debris surrounded by two layers
  – Inner layer (matrix): keratinizing squamous epithelium – produces keratin
  – Outer layer (perimatrix): subepithelial connective tissue – produces proteolytic enzymes that can resorb bone
• Middle ear > other pneumatized areas (e.g. EAC, mastoid, petrous air cells)

Perimatrix (connective tissue)
Matrix (epithelium)

Cholesteatoma
• CT: expansile opacified air cell
• T1: hypointense
• T2: hyperintense
• DWI: reduced diffusivity
• Need to use coronal, non-echo planar (non EPI) DWI

EAC
• EAC cholesteatoma
• Mimickers
  – Malignancies
  – Granulomatous diseases
  – Keratitis obliterans
  – Osteoradionecrosis
EAC cholesteatoma

EAC scc, looks like a cholesteatoma

EAC basal cell carcinoma

- Much less frequent than SCC
- Uniformly associated with actinic damage to epidermis
- Almost always seen in men
- Rarely fatal
- **Middle ear cholesteatoma**
- **Mimickers**
  - AOM with effusion
  - COM and its sequelae: effusion, granulation tissue, cholesterol granuloma
  - ETD with effusion
Middle ear cholesteatoma

Middle ear congenital cholesteatoma

Middle ear carcinoid – read out as cholesteatoma

Mastoid
- Mastoid cholesteatoma
- Mimickers
  - Malignancies
  - Coalescent mastoiditis

Mastoid cholesteatoma

R coalescent mastoiditis, Bezold, sigmoid sinus thrombosis (vs compressed sinus vs epidural abscess)
Coalescent mastoiditis from actinomycosis, Bezold

Petrous Apex
- Petrous apex cholesteatoma
- Mimickers
  - Mucocele
  - Cholesterol granuloma
  - Meningocele
  - Not really: effusion, asymmetric pneumatization
  - Petrous apicitis
  - Malignancies: metastasis, chondrosarcoma, etc

Cholesteatoma

Petrus apex mucocele

Cholesterol granuloma
Cholesterol granuloma

• An air space with **negative pressure** → rupture of blood vessels
• Breakdown of RBCs
• Release of cholesterol
• Foreign body giant cell reaction
• Granuloma is formed, with **cholesterol elements and blood products**
• Locations
  – Middle ear (with Eustachian tube dysfunction)
  – Petrous apex air cell (with obstructed drainage)
  – Mastoid, mastoid bowl

Ddx: expansile opacified PA air cell

• Mucocele
• Cholesterol granuloma
• Cholesteatoma
• Meningocele, arachnoid cyst

Petrous apicitis

• Infection in the mastoid spreading medially to the petrous apex
• Osteitis, disruption of cortex/septations, meningitis
• Occurs in the setting of **pneumatized petrous apex**
• *Gradenigo’s syndrome* – triad of symptoms, bacterial otitis media leading to petrous apicitis and spread of infection to meninges, Gasserian ganglion (V), Dorello’s canal (VI)
  – Otomastoiditis
  – Deep retroorbital pain in distribution of V
  – Diplopia due to VI palsy

Petrous apicitis

• CT: fluid in air cells, bone erosion
• MR: abnormal enhancement of adjacent meninges
• **Look for complications**: arterial complications, cavernous sinus/sigmoid/IJ thrombosis, epidural abscess, subdural empyema, meningitis, cerebritis

Petrous apicitis?

**Petrous apex effusion**

Opaclified petrous apex air cell, no bone erosion, no expansile margins — effusion

**Petrous apicitis?**

**Metastatic neuroblastoma**

Beware of mimics: if bone erosion is very extensive and IAC is involved, get MR. Look for abnormal soft tissue mass.
Petrous apicitis

Petrous apex effusion

Opacified petrous apex air cell, no bone erosion, no expansile margins – effusion

Petrous apex osteomyelitis

- Occurs in setting of non-pneumatized petrous apex
- Petrous marrow hyperintense on T2, enhances

Others

NEO

Facial nerve enhancement - Bell
Facial nerve enhancement – Ramsay Hunt

Facial nerve enhancement – Lyme

Labyrinthitis

- Etiology: infectious (viral, bacterial, luetic) autoimmune, toxins, post-traumatic
- Particularly associated with acute bacterial meningitis due to H. influenza or S. pneumoniae
- Children present with SNHL, vertigo; can progress rapidly to profound deafness

MR is more sensitive than CT for detection of early acute labyrinthitis

Acute labyrinthitis

Fibrous/late stage of labyrinthitis on MR
Evolution of labyrinthitis on CT

- Transverse / OCV fracture
  - Labyrinthine fractures heal slowly (paucity of vascularity) and by fibrous union
  - Complications
    - Labyrinthitis ossificans
    - Perilymphatic fistula (e.g., round window fluid, pneumolabyrinth)

Pneumolabyrinth = perilymphatic fistula

Thank you
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