next speaker:
Elizabeth Loney
Bradford/UK

CONDUCTIVE HEARING LOSS WITH AN INTACT TYPANIC MEMBRANE

Dr Elizabeth Loney
Clinical Director of Radiology and Consultant Radiologist
County Durham and Darlington NHS Foundation Trust, UK

Aims
- NOT a textbook lecture on causes
- Concept-based presentation
- Memorable (hopefully in a good way!)

AKA:
A WEEK IN THE LIFE OF MY SONS’ WASHING!

What is Conductive Hearing Loss (CHL)?
- CHL- Hearing loss secondary to inadequate sound transmission through external and middle ear structures.
- with an intact tympanic membrane- problems ‘at or beyond’ the TM

How is Sound Transmitted?
What are Potential Causes?
- Any pathology that will reduce ME sound transmission -
  - Absent ossicles
  - Ankylosed or dissociated ossicles
  - Fluid or other material in the ME cavity
  - Problems at the interface with inner ear structures (round and oval windows)
- Ossicular transmission - around 60db lost if disrupted

How will we look for them?
- High resolution MDCT/ CBCT
- Multiplanar Reformats
- Pre and post contrast MRI/ DWI

Topics for this Lecture
- Malformed / absent ossicles
- Dislocated / ankylosed ossicles
- ME infection/ inflammation/ tumours
- Trauma / haemotympanum
- Aberrant vascular structures
- Congenital cholesteatoma
- Cholesterol granuloma
- Otosclerosis

‘...not so Easy’ to ‘...oh so Easy’!

My Son and his Washing!

Concept
- The middle ear is a washing machine -
Ossicular Embryology
- First branchial arch (Meckel's cartilage)
  - head of the malleus
  - body and short process of the incus
  - tensor tympani muscle and tendon.
- Second branchial arch (Reichert’s cartilage)
  - the rest of the ossicular chain (except the footplate of the stapes)
  - stapedius muscle and tendon

Normal Otoscopy
- Congenital aural dysplasia
- Congenital ossicular anomalies
- Ossicular dissociation
  - Erosion
  - Trauma
- Ossicular ankylosis
  - Congenital
  - Post-inflammation
- Previous surgery
- Non-glomus tumours
Congenital Aural Dysplasia
- Mild end of the atresia spectrum- Altmann 1st Degree Malformation
- Mild deformities of the EAC, normal or mildly hypoplastic ME cavity, deformed ossicles and aerated mastoid
- Commonest ossicular anomaly is fusion of the malleo-incudal joint
- Hypoplastic manubrium and stapes also possible

Malleo-incudal fusion

Dysmorphic Ossicles in Fanconis’ Anaemia

Congenital Ossicular Anomalies
- Bilateral- often genetic, autosomal dominant
- Unilateral- usually sporadic
- Köslng Classification of isolated ME malformations
  - Mild: Normal ME with ossicular dysplasia
  - Moderate: Hypoplastic ME with rudimentary ossicles
  - Severe: Aplastic or cleft-like ME cavity

Stapes Malformations
- Commonest types
  - Stapes fixation- 20-35%, due to fusion of the peripheral lamina stapedialis and the annular ligament
  - Incudostapedial discontinuity
  - Footplate- up to 0.25mm in depth
- DDx- otosclerosis but deafness is not progressive
- Aberrant facial nerves- dehiscence or inferior displacement of the tympanic segment
Incus Malformations
- Absence/hypoplasia of the long process with ISJ separation
- Less frequently- variable long process position or complete incus aplasia
- Congenital or acquired
- Acquired IS discontinuity - non-cholesteatomatous inflammatory erosion, previously evacuated cholesteatoma and trauma
- In 15.7% of patients with CHL and normal otoscopy
- Usually have other ossicular anomalies, especially stapes
- Main differential is otosclerosis but worse surgical outcomes

Malleus Malformations
- The malleus is less commonly involved.
- Most frequent findings-
  - deformities and hypoplasia of the head and of the manubrium of the malleus
  - fixation in the epitympanic recess
  - malleoincudal joint abnormality
  - Absent malleus

Window stenosis/ aplasia
- Oval window development requires contact between otic capsule and stapes footplate
- Theory- developing facial nerve is displaced anteriorly, coming between the two
- Oval window stenosis/ absence is often associated with anomalous VII course and stapes deformity
Facial Nerve Schwannoma and Middle Ear Adenoma
- Rare, benign, slow-growing lesions
- Middle-aged patients
- Symptoms include CHL, facial paresis and hemifacial spasm, aural fullness, tinnitus
- **CT:**
  - VII: smooth 'benign' bone expansion along the nerve
  - ME Adenoma: non-specific <3cm soft tissue lesion. No bone invasion
- **MRI:**
  - Both: well-defined Interm T1W, high T2W, enhancing lesions
- **PET-CT:**
  - ME Adenoma: focal tracer uptake in somatostatin-receptor scan due to neuroendocrine differentiation

Otitis Media with Effusion (OME)
- Presence of fluid in the middle ear without signs or symptoms of acute ear infection. Mucoid or serous
- Symptoms: hearing loss or aural fullness but typically no pain or fever
- In children, hearing loss is generally mild and is often detected only with an audiomagram
- Serous otitis media- transudate formation as a result of a rapid decrease in middle ear pressure relative to the atmospheric pressure. The fluid in this case is watery and clear.
Chronic granulation tissue which can affect any pneumatized part of the temporal bone
- M=F, young and middle-aged adults
- Tend to be clinically silent/ incidental findings
- Most middle ear and mastoid CG do not erode adjacent bone, unlike petrous apex CG
- Symptoms include headaches, cranial neuropathies, hearing, balance, speech and swallowing problems

Cholesterol Granuloma
- Causes:
  - Obstruction vacuum theory: Eustachian tube dysfunction causes repeated obstruction and episodes of bleeding
  - Exposed marrow theory: hyperplastic mucosa invades underlying bone and exposes marrow which bleeds
  - Trapped blood, chronic inflammation +/- infection
  - Yellow-brown fluid containing cholesterol crystals
  - Surrounding fragile vessels prone to rupture, reducing healing

Dehiscent Jugular Bulb
- Dehiscence of the jugular bulb may occur in up to 15% of temporal bones studied by CT and at PM
- Dehiscence occurs over time
- May mimic a middle ear mass and be inadvertently biopsied.
- Dehiscence into the round window niche can cause CHL
Temporal Bone Fractures

- Typically from high-energy, blunt trauma
  - motor vehicle accidents (45%)
  - falls (32%)
  - assault (12%)
- Around 20% with skull-base fractures also sustain a temporal bone fracture: look carefully
- Longitudinal, transverse or mixed. Otic capsule sparing or violating
- CHL - due to haemotympanum, ossicular discontinuity (ISJ and MIJ dislocation) and fractures
- VII paralysis: immediate (involved in fracture line) or delayed (swollen nerve)
- Imaging: CT to assess fracture pattern, structures involved, other intracerebral problems, MRI - cochlear haemorrhage

Battles' Sign

Ossicular Dislocation

- Meriot et al identified five types of ossicular dislocation following trauma:
  - incudostapedial joint separation
  - incudomalleolar joint separation
  - dislocation of the incus
  - dislocation of the malleoincudal complex
  - stapediovestibular dislocation

Glomus Tumours
- Paragangliomas: benign, highly vascular, from extra-adrenal neuroendocrine system. Sporadic or familial.
- Pulsatile tinnitus, CHL, vertigo and red TM
- Types
  - Jugulare: from Arnold’s nerve, permeative erosion of jugular foramen
  - Tympanicum: Jacobson’s nerve, cochlear promontory. Fisch Type A
  - Jugulare-tympanicum: involving both sites
- Surround rather than erode the ossicular chain
- MRI: low T1W, high T2W signal and intense contrast uptake with a 'salt and pepper' appearance due to haemorrhage, slow flow and arterial flow voids
- Angiography: hypervascular tumour blush and early draining veins
- Nuclear Medicine: Indium-111 labeled octreotide accumulates due to somatostatin receptors

Aberrant ICA
- Rare congenital vascular anomaly secondary to an underdeveloped ICA cervical segment
- Resultant collateral pathway: enlarged inferior tympanic artery coursing into the ME cavity via an expanded inferior tympanic canaliculus
- May lead to pulsatile tinnitus and CHL
- DDx glomus tumour: Do not biopsy!
Persistent Stapedial Artery

- Persistent stapedial artery is a rare congenital anomaly
- Normally transient in the embryo between the ICA and MMA/ECA
- Failure to regress is associated with an aberrant ICA, absent foramen spinosum and thick tympanic facial canal
- Passes between stapes crura, over the oval window
- May be associated with CHL, pulsatile tinnitus and vertigo

Day 4...

Tympanosclerosis

- Deposition of hyaline and calcific deposits in the TM and/or ME structures
- Thought to be a complication of chronic otitis media
- TM: high density foci often associated with membrane thickening (myringosclerosis).
- ME involvement:
  - high density foci on the surface of the ossicles
  - thickening of the stapes crura and footplate
  - thickening and increased density of suspensory ligaments and muscle tendons
  - areas of heaped up new bone
**Congenital Cholesteatoma**

- epidermoid cyst, differing only in name and location
- Intraosseus inclusions of ectoderm comprised of keratin and cholesterol, usually located in the petrous apex
  - T1 - low signal
  - T2 - high signal (usually slightly brighter than CSF)
  - T1 + C - no central enhancement; thin peripheral enhancement may be seen
  - DWI - restricted diffusion

**Kazahaya & Potsic** suggested subdivision into four stages.

- **Stage I**: one quadrant of TM affected; no ossicular involvement or mastoid extension
- **Stage II**: multiple quadrants affected; no ossicular involvement or mastoid extension
- **Stage III**: ossicular involvement; includes erosion of ossicles and surgical removal for eradication of disease; no mastoid involvement
- **Stage IV**: mastoid extension (regardless of findings elsewhere)

**CT in Congenital Cholesteatoma**

Usefulness of Computed Tomography Hounsfield unit Measurement for Diagnosis of Congenital Cholesteatoma. Y.-W. Kim, S. K. Baik. [http://dx.doi.org/10.1594/ecr2014/C-1070](http://dx.doi.org/10.1594/ecr2014/C-1070)

**Schwartz’s Sign**
Otosclerosis/ Otospongiosis
- Genetically mediated metabolic bone condition of unknown aetiology
- 'Clastic' spongy phase followed by 'Blastic' sclerotic phase
- CHL > SNHL. Middle age. F>M
- Fenestral and cochlear (retrofenestral types). Bilateral > unilateral
- Demineralisation of the otic capsule and new bone formation around oval window in particular. Enhances on MRI
- Differential includes osteogenesis imperfecta, Paget Disease, syphilis and bilateral lytic metastases

CT Grading of Otosclerosis
- Symons and Fanning 2005
  - Grade 1 - Fenestral
  - Grade 2 - patchy localised cochlear disease
    - 2A - basal turn
    - 2B - middle/apical turns
    - 2C - both
  - Grade 3 - confluent involvement of otic capsule ('double ring sign')

Fenestral Otosclerosis- Grade 1

Fenestral and Patchy Capsular Otosclerosis- Grade 2C

Capsular Otosclerosis- Grade 2C/3

Summary - what does the TM look like?
- Normal - congenital anomalies and tumours other than glomus
- Blue - glue ear, cholesterol granuloma and dehiscent jugular bulb
- Red - Glomus tumours, haemotympanum and aberrant ICA
- White - tympanosclerosis and congenital cholesteatoma
- Pink - otosclerosis
Thank you!