Chapter 143: Otosclerosis (OS)

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Intro

- Disorder of fibrous osteodystrophy of the human otic capsule
  - Abnormal resorption and deposition of bone
- Primarily causes CHL
  - SNHL and MHL are also possible
- Autosomal dominant, incomplete penetrance (25-40%), ask about family history of OS
- Incidence of clinically evident disease: ~1%
  - Average age at presentation = 33 yo
- More common in Caucasians and women
  - Associated w/ osteoporosis (COL1A1 gene); pregnancy

Embryology

4 weeks	Otic capsule begins to surround otic vesicle
8 weeks	Cartilaginous framework
16 weeks	Endochondral bony replacement of the cartilage framework

- In some people, complete bony replacement does not occur and cartilage is left behind
  - Fissula ante fenestram (anterior to OW)
    - Affected in 80-90% of patients w/ OS

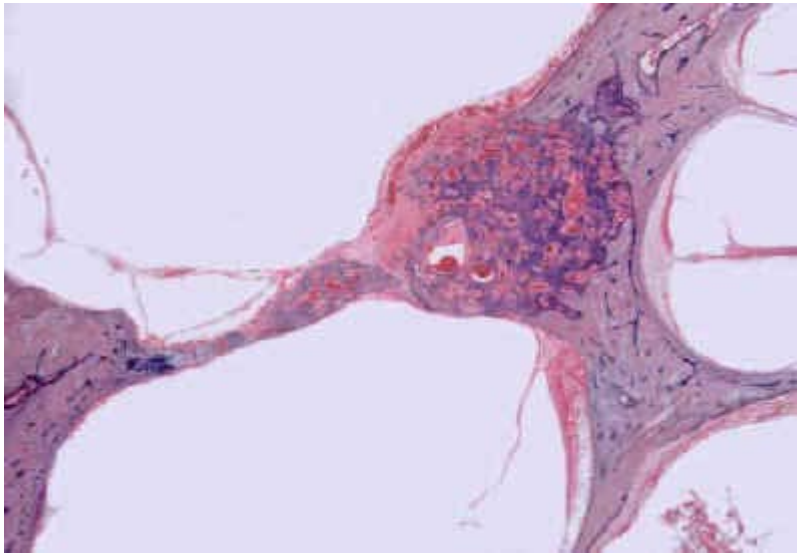
Histology

3 phases

(1) Otospongiosis (early phase):

- Osteoclasts resorb bone around pre-existing blood vessels

Widening of vascular channels → Schwartze Sign (reddish hue behind an intact TM, from dilated microcirculation on promontory)

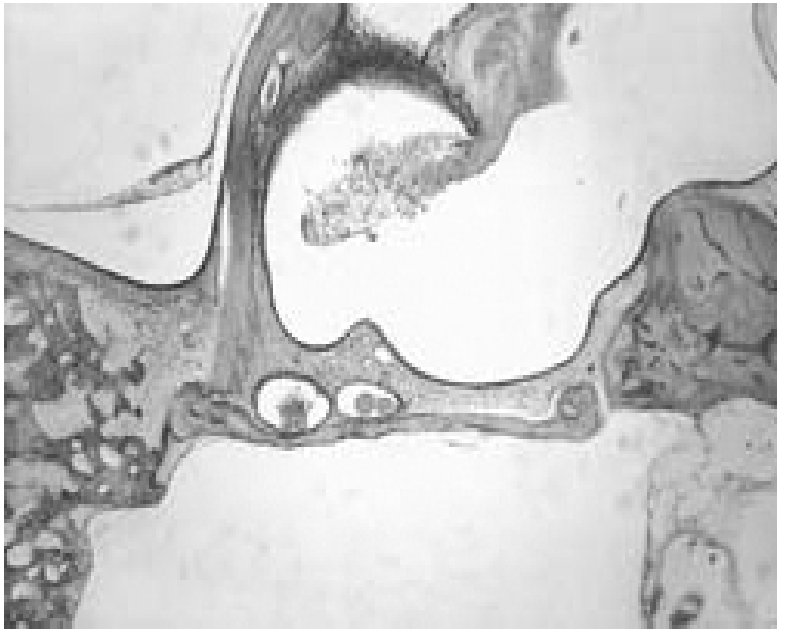


Histology

3 phases

(2) Transition Phase:

- Osteoclasts become more involved in the areas of otospongiosis
- More amorphous ground substance, less collagen → new spongy bone
- Looks blue on H&E Staining (blue mantles of Manasse)
  Up to 20% normal TB's have mantles
- Side note: H&E staining → Hematoxylin (blue; nuclei)
  Eosin (pink; cytoplasm)

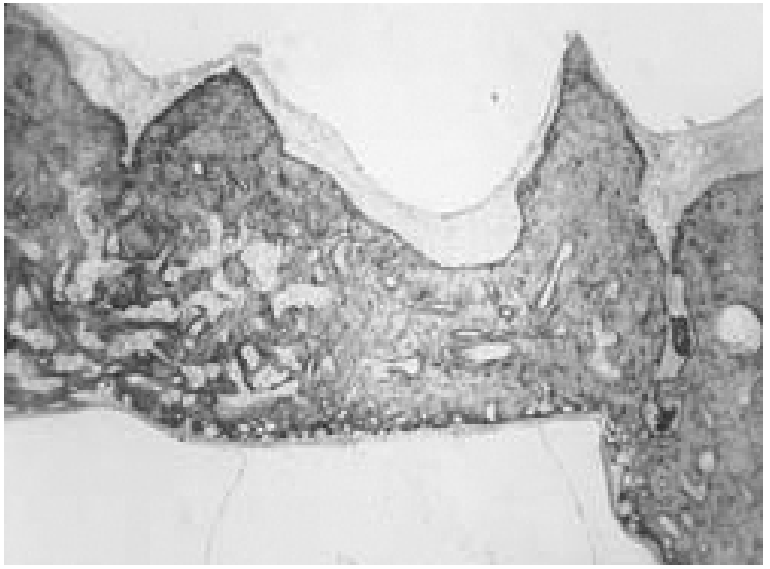


Histology

3 phases

(3) Late Phase:

- Dense, mineralized bone forms in areas of previous bony resorption
- Vascular channels which were once dilated are now narrowed



- OS begins in the endochondral bone but can progress to involve the endosteal and periosteal layers

Pathophysiology

OS focus in the fissula ante fenestram (located anterior to the OW, anterior to the stapes footplate)

Progressive involvement of the footplate and annular ligament

Can fill the oval window niche (aka obliterative OS)

If OS involves footplate but NOT the annular ligament → Biscuit footplate

Minimal fixation → footplate can be mobilized inadvertently during surgery → higher risk of SNHL while placing your prosthesis

Pathophysiology

OS and SNHL (Controversial):

- Higher severity of SNHL in patients w/ OS than in those w/o OS
- Possibly due to direct invasion of the cochlea or toxic metabolites diffusing into the inner ear
- Rare case reports of SNHL w/o CHL in patients w/ OS (“cochlear OS”)

Pathophysiology

OS and dizziness → “OS inner ear syndrome”

- Up to 30% of OS patients complain of dizziness
- dDx (in the context of dizziness): Meniere's disease, SSCD
- Meniere's Disease → episodic rotational vertigo lasting hours associated w/ nausea, vomiting; fluctuating SNHL; tinnitus; aural fullness; low frequency SNHL on audio (early)
- SSCD → Vertigo or oscillopsia (objects in visual fields appear to be moving) induced by pressure or sound; CHL or MHL with acoustic reflexes; nystagmus in the plane of the dehiscent SSC
- OS Inner ear syndrome → milder, more persistent dizziness, do not have low frequency SNHL; tinnitus; abnormal acoustic reflex (early: diphasic, late: absent)

Symptoms and Signs

Slowly progressive hearing loss over years

Paracusis (of Willis): hear speech more easily in noisy environments (CHL improves signal to noise ratio by subduing background noise)

Rinne: Bone conduction > Air conduction (negative Rinne) indicating a CHL

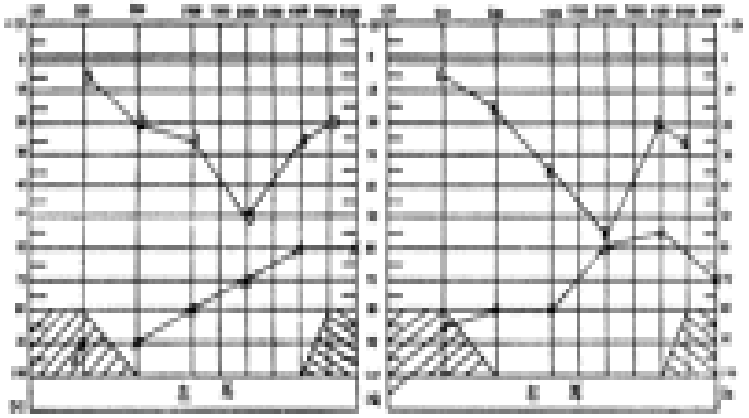
CHL seen w/ 256 Hz tuning fork first (512, 1024 later)

Weber: lateralizes to the ear w/ greater CHL (although can be affected by concurrent SNHL)

*90% of patients w/ histologic evidence of OS are asymptomatic

Audiology

- Widening of the air bone gap that begins at lower frequencies
- Variable degrees of SNHL
- Carhart's Notch
  - Depression at 2000Hz
  - Due to stapes fixation and resultant change in frequency of the bony capssule
  - Artifact of audiogram
  - Most commonly seen in OS but can also be seen in other types of CHL

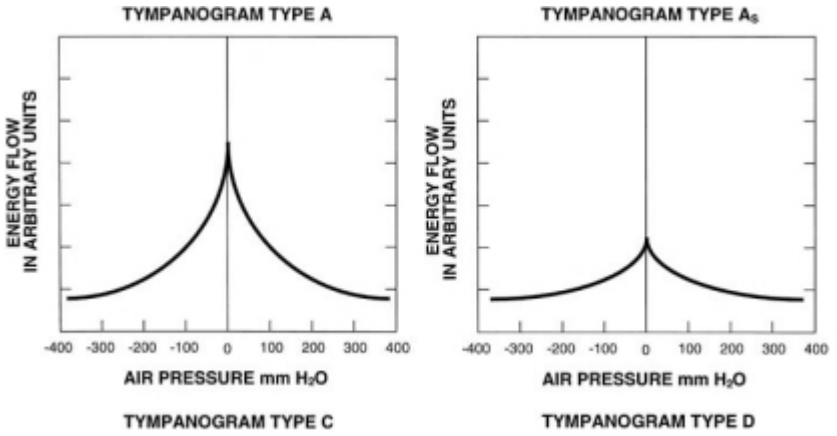


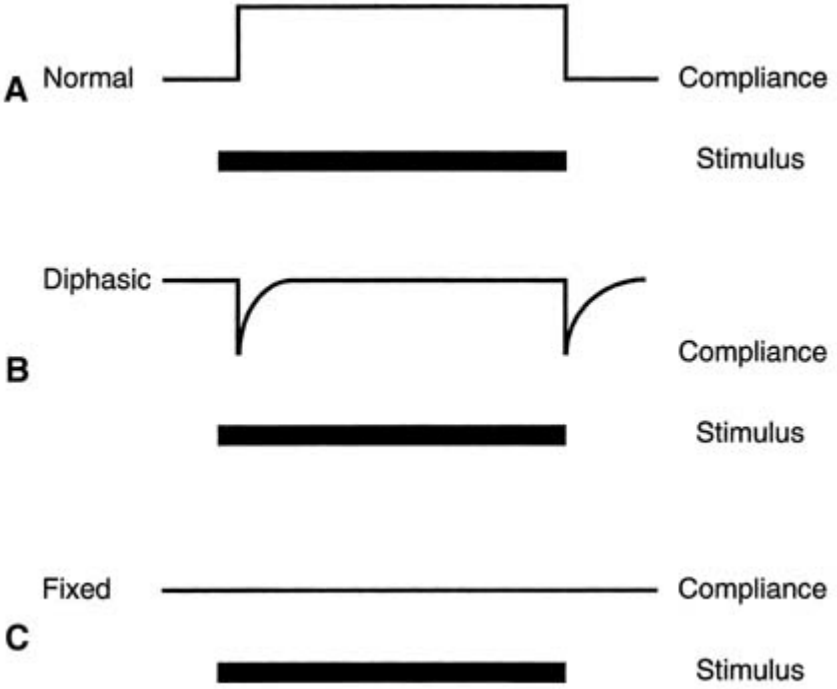
Audiology

Speech discrimination: Excellent

Tympanogram: Type A (normal) or As (pressure peak is at 0 but has a reduced amplitude from stapes fixation)

Stapedial reflex: can be normal (very early); diphasic (early OS; increase in compliance at onset and termination of the stimulus; pathognomonic as per Lalwani); absent reflex (most common finding at presentation)



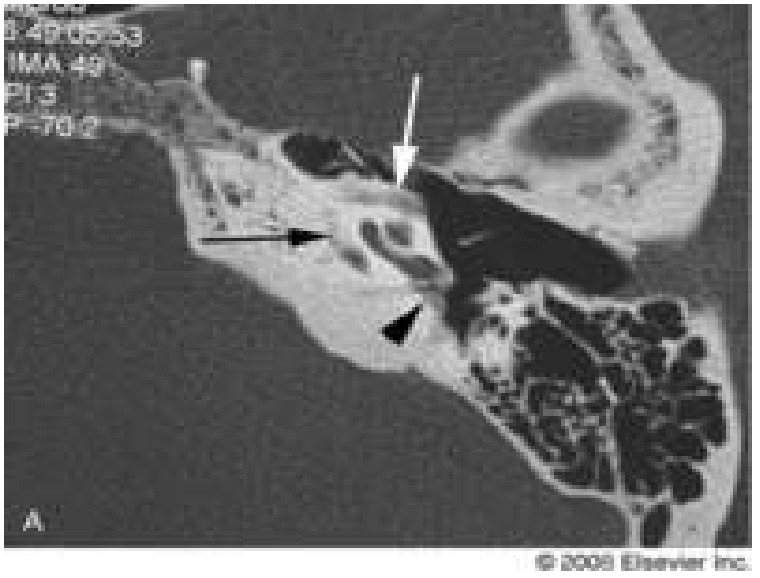


CT Imaging

"Halo Sign" → Radiolucent, demineralized area in and around the cochlea (early OS)

Diffuse sclerosis → mature OS

Negative CT scans are <u>not</u> diagnostic (OS foci can exist below CT Scan threshold)



dDx

Definitive diagnosis of OS can only be made during explor. tympanotomy

Most common conditions that mimic OS are those that:

- Result in ossicular discontinuity (e.g.: incus necrosis from recurrent chronic otitis media)
- Result in mass effect on TM or ossicles

Lateral ossicular chain fixation: malleus or incus fixation in the epitympanum (examine the whole ossicular chain)

Tympanosclerosis: can mimic OS but there should be a hx of recurrent OM or PE tubes

Paget's Disease: diffuse bony involvement resulting in enlarged and deformed bones, looks similar histologically to OS except lesions begin in the periosteal bone and involve endochondral bone last; stapes fixation is rare

Treatment

Hearing Aids (amplification)

BAHA (amplification)

Sodium fluroide

- Fluoride ions replace hydroxyl radical → more stable fluorapatite complex → which resists osteoclastic action

Severe to profound SNHL bilaterally from OS can be given Cochlear implants

Stapedectomy

Patient selection

Favorable criteria → unacceptabel CHL, negative Rinne test at 512 Hz, good speech discrimination

Pediatric patients w/ X-linked hearing loss have a higher risk of perilymph gusher

Patients w/ Meniere's Disease and OS have a higher risk of SNHL after stapedectomy (the saccule may be enlarged to the point that it adheres to the undersurface of the stapes footplate)

TM perforations should be repaired prior to stapedectomy (stage it)

ETD and cholesteatoma → not good candidates

Middle ear effusions or infections → absolute contraindications

Perform surgery on the worse hearing ear first (wait 6 months until operating on 2^{nd} ear due to small risk of sudden post-op HL)

Do not operate if it's the only good hearing ear

Stapedectomy

Procedure is nicely diagrammed in Cummings (Chapter 156)

Total Stapedectomy vs. Stapedotomy:

Generally, both involve removing the stapes superstructure and stapedius

Differ in that total stapedectomy removes the entire flootplate (older technique) while stapedotomy utilizes small fenestra (holes) in the footplate to allow placement of the prosthesis

- Stapedotomy can be done w/ microdrills or lasers (Argon, KTP, or CO2
- Distance between lateral incus and footplate is 4.5mm; between medial incus and footplate is 4.25mm (usual prosthesis size)

Problems in Surgery

1) Exposed, Overhanging Facial Nerve: covers the OW 9% of the time; sometime the nerve can be gently retraced and the procedure can continue; if the prosthesis is touching the facial nerve it's ok (as per Cummings)

2) Fixed Malleus: Rare, can be associated w/ OS; can be repaired w/ total ossicular replacement prosthesis or w/ incus replacement prosth.

3) Floating Footplate: Avoid this by creating your fenestra BEFORE fracturing the stapes superstructure

- If all or most of the footplate is depressed into the vestibule, do not attempt to retrieve it (cause more harm than benefit)

4) Perilymph gusher: profuse flow of CSF after entering the vestibule; very rare; assoc. w/ congenital footplate fixation in peds pt.; lumbar drain may be necessary to allow graft to adhere to OW

Potential Post-Op Complications

1) Facial nerve injury: < 1% chance, usually incomplete
2) SNHL: < 1% chance
3) Vertigo: Usually mild, subsides over time, conservative treatment
4) Taste Disturbance: 9% of the time; occurs more often w/ those people who had stretching of the chorda as opposed to sectioning it
5) Perilymph fistula: 3-10% risk; can occur early or late post-op period; fluctuating or progressive HL, tinnitus, vertigo. Tx initially w/ conservative measures (acetozalamide, bed restx 5 days); if symptoms persist, then explore, remove prosthesis, graft over the OW, and replace prosthesis

Conclusion

Most common cause of stapedectomy failure: prosthesis displacement w/ or w/o incus erosion

Other causes: footplate fixation, perilymph fistula, otosclerotic regrowth

Overall, can be a very successful surgery (90-95% hearing improvement w/ < 10 dB air-bone gap in the first 1-5 years; decreases to 63% after 30 years)

- Revision stapedectomy: more difficult, lower success rate