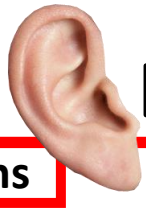
Last updated: 3/9/2022

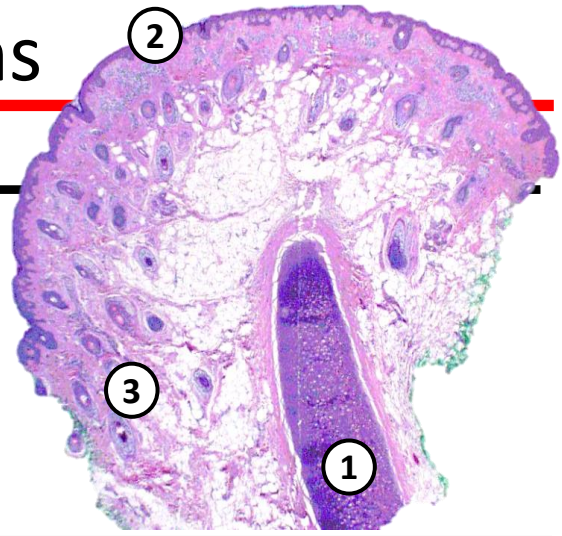
Prepared by Dr. Kurt Schaberg



Ear Lesions

Congenital Lesions

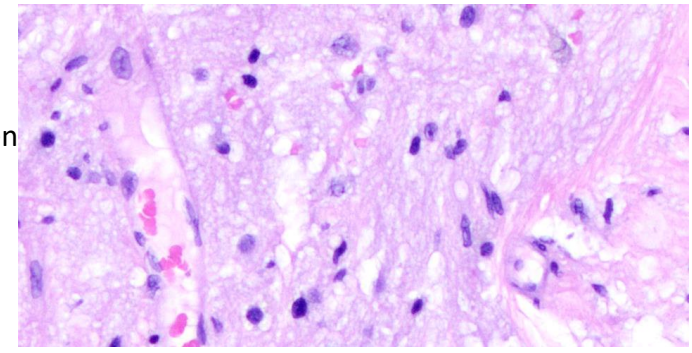
Accessory Tragus



Developmental anomaly→ recapitulates normal external ear with **1) <u>Central Cartilage</u>** (*usually*)**, 2) Surface skin, 3) Adnexal structures**

Located on skin surface **anterior to auricle**.
Usually identified in **childhood**.
Cured by excision.

Encephalocele

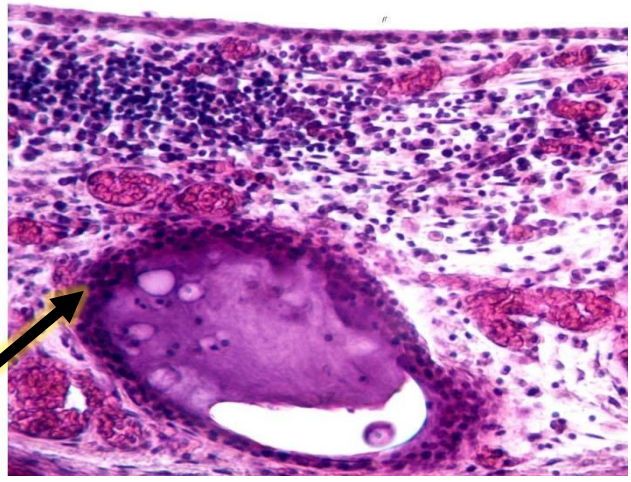


Herniation of brain tissue (continuous with brain)
Variable proportion of **<u>neurons and glia</u>** (GFAP+) often with gliosis, chronic inflammation, and fibrosis
Meninges absent usually.

Histologically indistinguishable from glial heterotopia (which discontinuous from brain)

Inflammatory/Reactive Lesions

Otitis Media

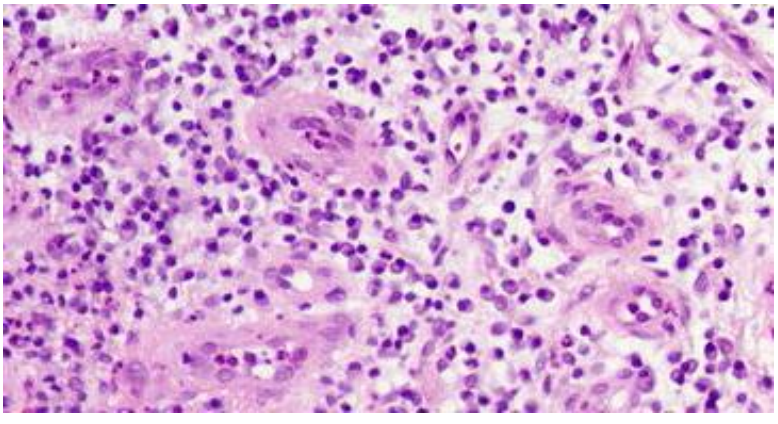


Extremely common clinically, <u>*not*</u> usually sampled for pathology.

Viral or bacterial infection of <u>*middle*</u> ear.
Most common in young kids. Usually self resolves.

Acute and/or Chronic inflammation with fibrosis.
<u>Entrapped epithelial inclusions</u> (→) can be mistaken for tumor!

Otic Polyp



Reactive response to **longstanding Otitis media.**

<u>Granulation tissue</u> with dense **chronic inflammation**.

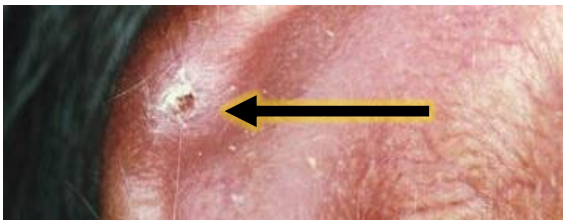
May have entrapped surface epithelium, cholesterol clefts, or calcifications.

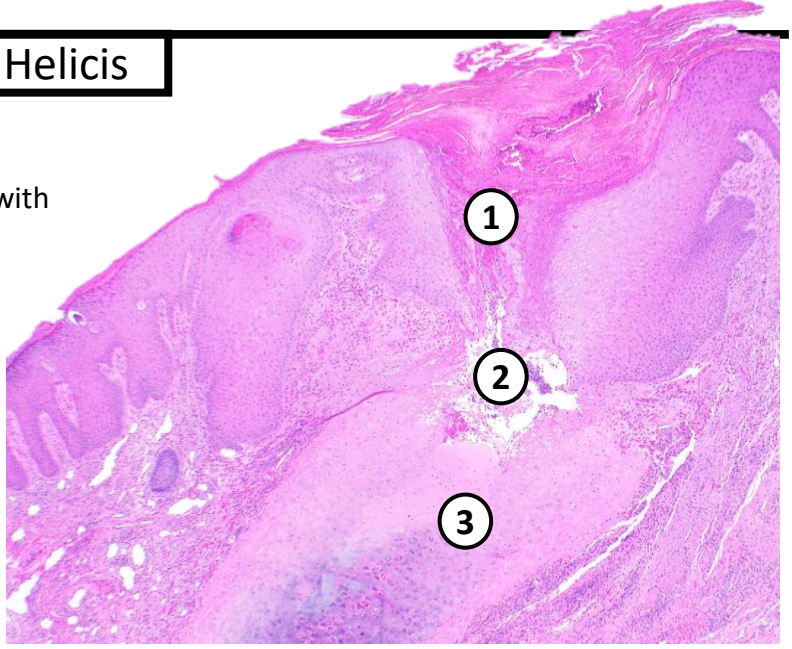
Chondrodermatitis Nodularis Helicis

aka "CNH"

1) Surface hyperplasia surrounding **ulceration** with **keratin plug,** 2)**Dermal fibrinoid necrosis**, and 3)**Necrotic Cartilage** (usually)

Helix or anti-helix of ear
Localized injury. Clinically mistaken for SCC.



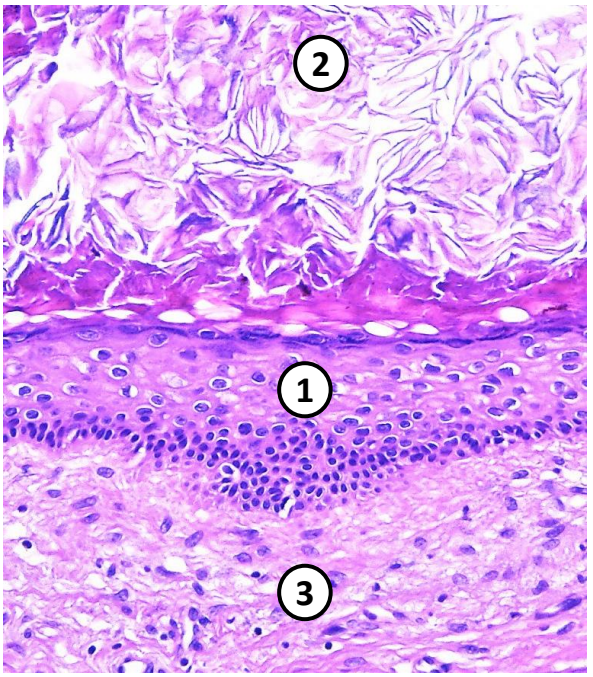


Cholesteatoma

Keratinizing cyst in middle ear→ destroys ossicular chain → conductive hearing loss and foul-smelling discharge

Three required components
1) Stratified squamous epithelium with a granular layer (derived from external auditory canal—middle ear epithelium is cuboidal/columnar)
2) Keratinaceous debris (flakes, anucleate squames…)
3) Inflamed fibrous stroma
Frequently associated cholesterol clefts and foreign body giant cell reaction (cholesterol granuloma)

Can be secondary to chronic otitis media or congenital.
Can be locally destructive and recur.

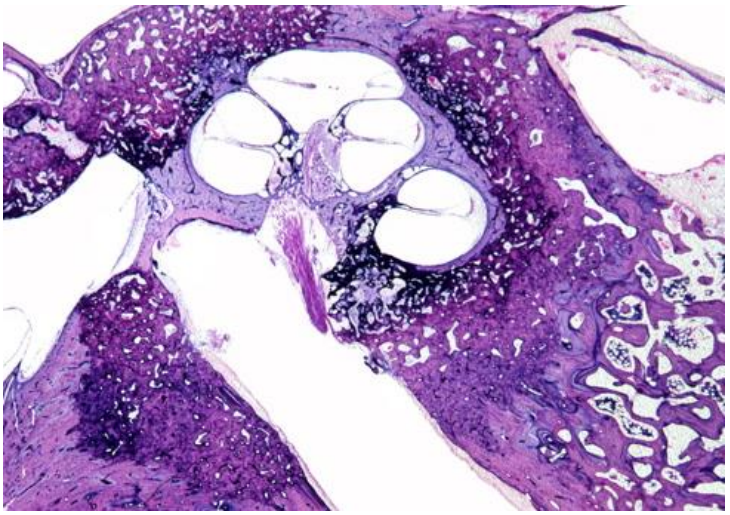


Otosclerosis

Bone overgrowth→ fixation of ossicular chain → conductive hearing loss.
Unclear etiology.

Immature **trabecular bone** and vascular stroma. (varying appearance in different phases)

Usually **bilateral** and symmetrical.



Benign Neoplasms

Middle Ear Adenoma

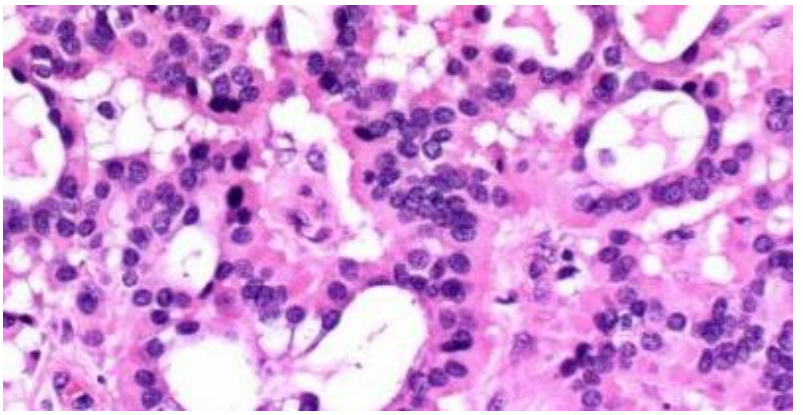
*aka "**MeMeNET**"*
Middle Ear Mixed Epithelial Neuroendocrine Tumor

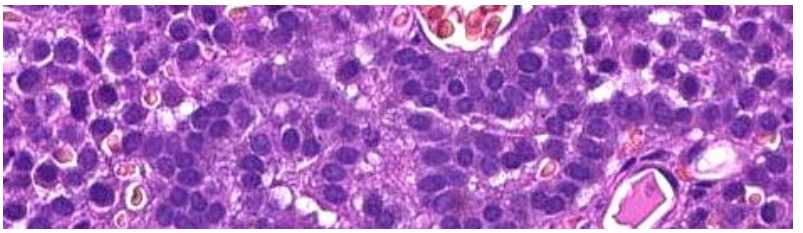
Rare neoplasm with ***dual*** **neuroendocrine and mucin-secreting differentiation**.

Infiltrative, unencapsulated.
Multiple patterns of growth (glandular, trabecular, diffuse, nested, etc…)
Ducts show dual cell population:
Inner: luminal flattened eosinophilic cells with secretion
Outer: cuboidal/columnar cells
Delicate "salt and pepper" chromatin

IHC: (+) CK, Synaptophysin, Chromogranin
Mucin stains with PAS and Alcian blue.

Found in middle ear (hence the name!)



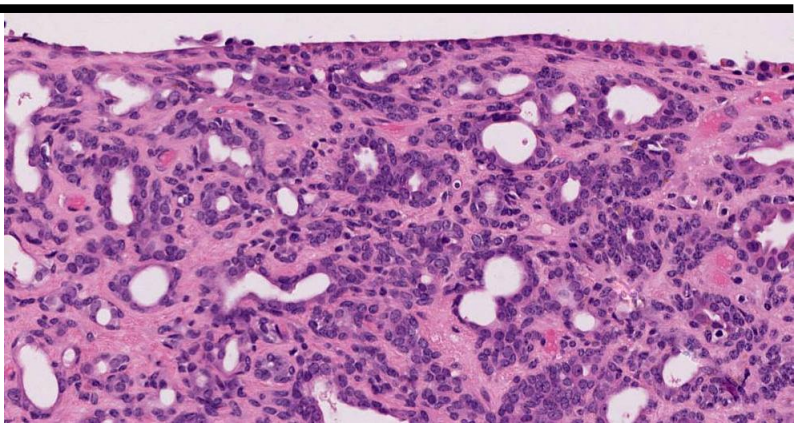


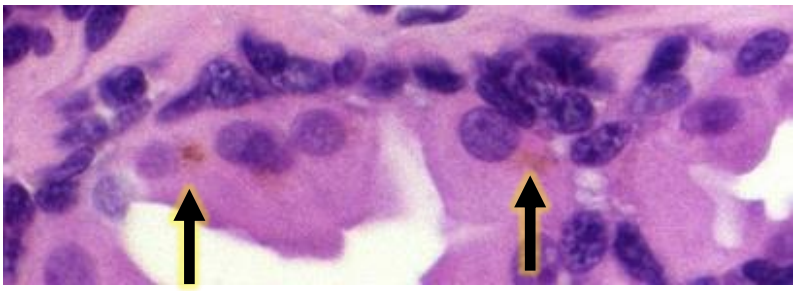
Ceruminous Adenoma

Benign tumor of the **wax-producing glands** of the **external auditory canal**

Unencapsulated, circumscribed
Bilayerd glands in fibrous stroma
Inner luminal secretory cells with abundant granular cytoplasm and yellowish ceroid secretions (→) (+CK7)
Basal myoepithelial cells (+p63, S100)

A variety of growth patterns: can overlap with pleomorphic adenoma and syringocystadenoma papilliferum.





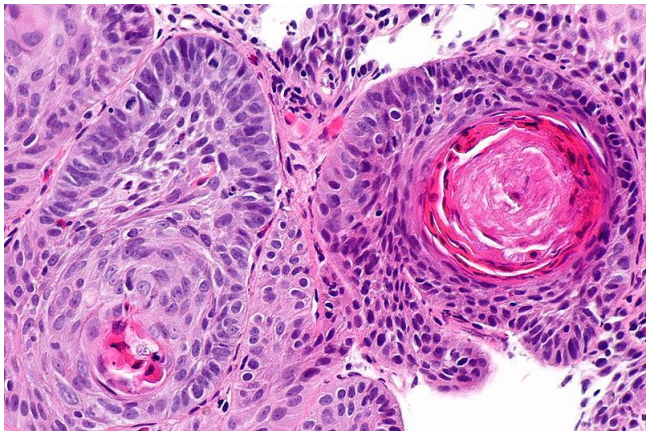
Other tumors

Other, non-unique, tumors and tumor-like lesions include:

Paraganglioma
Meningioma
Schwannoma
Exostosis
Malakoplakia
Keloid
Langerhans's cell histiocytosis
Synovial Chondromatosis
Epithelioid hemangioma

Malignant Neoplasms

Squamous Cell Carcinoma

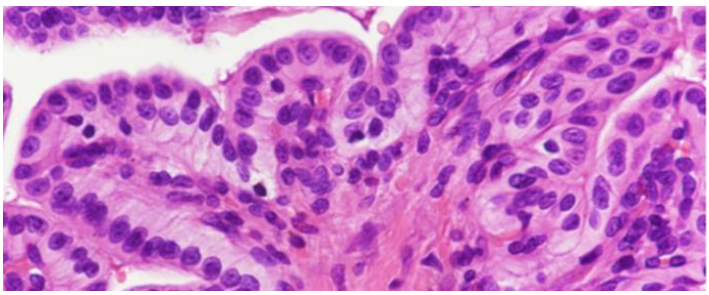


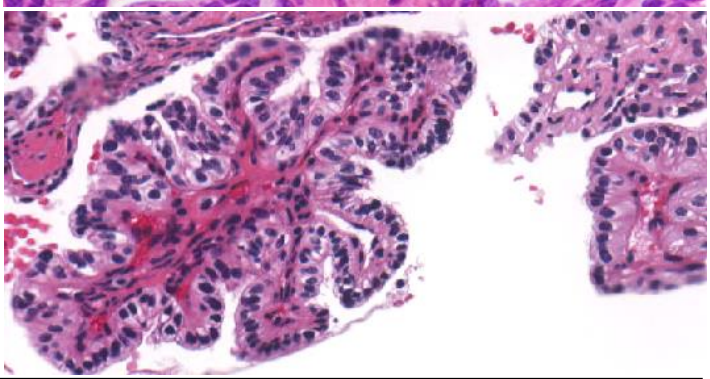
Malignant neoplasm of squamous epithelium

Similar to SCC elsewhere on the skin
Invasive carcinoma with frequent keratinization and inflamed desmoplastic stroma

Most cases are **external ear and UV-related**
Usually elderly with pre-existing actinic change

Endolymphatic sac tumor





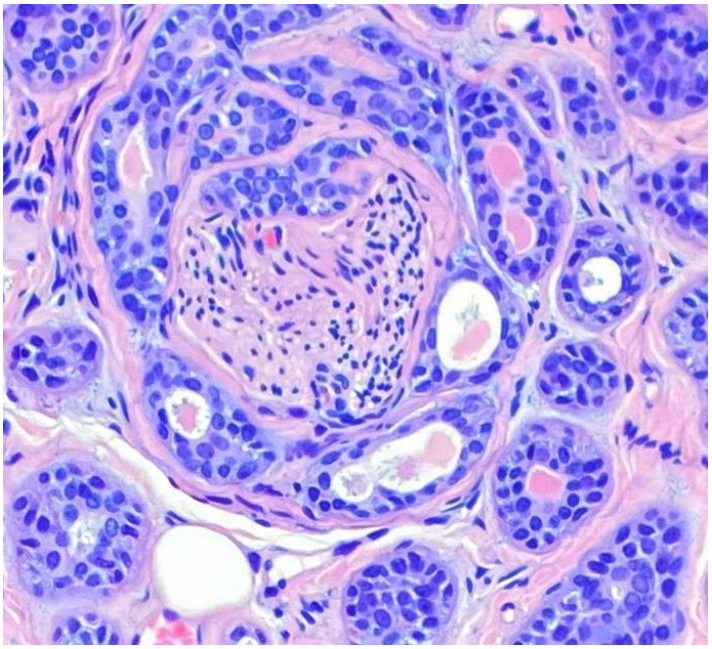
Low-grade malignant (locally destructive) tumor **arising from the endolymphatic sac** in the petrous temporal bone. Rare.

Associated with **von Hippel-Lindau** in 1/3 of cases

Unencapsulated, invasive
Papillary and cystic architecture
Clear to pale pink cells with eccentric nuclei
Usually arranged in single layer
Small, round, hyperchromatic nuclei
May have pink, PAS+ secretions

IHC: (+) CK, EMA, S100/SOX10, PAX8, CAIX;
(-) CD10, RCC, TTF1

Ceruminous adenocarcinoma



Rare. Malignant neoplasm derived from the ceruminous glands of the external auditory canal.

Infiltrative.
Variable architecture (solid, cystic, cribriform, glandular, etc…)
Frequently perineural invasion and necrosis.
Biphasic cell populations (like benign counterpart)
Cytologic pleomorphism. Mitoses.
No ceroid pigment.

Can differentiate to/be histologically identical to Adenoid cystic carcinoma or mucoepidermoid carcinoma