Ear Lesions

Congenital Lesions

Accessory Tragus

Developmental anomaly \( \rightarrow \) recapitulates normal external ear with 1) Central Cartilage (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 neurons and glia (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, not usually sampled for pathology.

Viral or bacterial infection of middle ear.
Most common in young kids. Usually self resolves.

Acute and/or Chronic inflammation with fibrosis.
Entrapped epithelial inclusions \( \rightarrow \) can be mistaken for tumor!

Otitic Polyp

Reactive response to longstanding Otitis media.

Granulation tissue 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**

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**

**Bilayered** 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