External ear is a relatively common location for skin cancers

SCC (squamous cell carcinoma) has a high metastatic rate and higher death rate than other cutaneous sites

The temporal bone is rarely the site of primary malignancy

Parotid gland, TMJ, and infratemporal fossa tumors can erode into the ear canal and middle ear and require temporal bone approach management
Incidence

EXTERNAL EAR

- USA --- 1 million persons develop cutaneous malignancies/year
  - Population-based study: ear as 1st site of SCA in 12/100000 men and 0.6/100000 women
- Caucasian men, in their 6-7th decades of life
- Cutaneous SCC is up to 17 times more common in men than women
TEMPORAL BONE

✓ Account for about 0.2% of all head and neck cancers

✓ Cancer is the underlying cause in only 1/5000-20000 PTs with an otology complaint

✓ Temporal bone is more likely to be affected secondarily from advanced cancers of...
  ✓ External ear
  ✓ Periauricular skin
  ✓ Parotid gland

✓ Older PTs --- carcinomas

✓ Younger PTs --- sarcomas
**Etiology**

**Leading Cause of External Ear Cancer is Ultraviolet Solar Radiation**

**Other entities related to external ear cancer**
- Radiotherapy
- Burn chondritis
- Chronic scarring lupus erythematosus

**Entities related to SCC of the middle ear/ear canal**
- Radiotherapy
- HPV16 has been found in a small number of temporal bone SCCs.
The outer ear represents 5-10% of all skin cancers.

For H&N, the external ear is the 2nd most common site of cutaneous SCC.

The subset of PTs with METASTATIC SPREAD OF CUTANEOUS SCC OF THE H&N
- Outer ear accounts for 20%, whereas lip for a 15% and cheek 12%.

Most common malignancies are basal cell carcinoma and SCC.
- Incidence is nearly equal (1.3 BCC: 1SCC).
- One review study (outer ear cancer)
  - SCC 55-67%
  - BCC 28-32%
  - Melanoma 1-5%
Histologic types – external ear

SCC from the auricle has the highest death rate (47%) in one study.

High-risk area for BCC and SCC as documented in the latest National Comprehensive Cancer Network (NCCN) guidelines and AJCC staging.

Table II
Definition of cutaneous squamous cell carcinoma tumor (T) staging system in 7th edition of American Joint Committee on Cancer

<table>
<thead>
<tr>
<th>Stage</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>TX</td>
<td>Primary tumor cannot be assessed</td>
</tr>
<tr>
<td>T0</td>
<td>No evidence of primary tumor</td>
</tr>
<tr>
<td>Tis</td>
<td>Carcinoma in situ</td>
</tr>
<tr>
<td>T1</td>
<td>Tumor ≤ 2 cm in greatest dimension with &lt;2 high-risk features*</td>
</tr>
<tr>
<td>T2</td>
<td>Tumor &gt;2 cm in greatest dimension with or without one additional high-risk feature, or any size with ≥ 2 high-risk features*</td>
</tr>
<tr>
<td>T3</td>
<td>Tumor with invasion of maxilla, mandible, orbit, or temporal bone</td>
</tr>
<tr>
<td>T4</td>
<td>Tumor with invasion of skeleton (axial or appendicular) or perineural invasion of skull base</td>
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</tbody>
</table>

*High-risk features include: depth (>2-mm thickness; Clark level ≥ IV), perineural invasion, location (primary site ear, primary site nonglabrous lip), and differentiation (poorly differentiated or undifferentiated).

A new American Joint Committee on Cancer staging system for cutaneous squamous cell carcinoma: Creation and rationale for inclusion of tumor (T) characteristics.

SCC and BCC account for >50% of the tumors if all primary tumor sites are considered.

If excluded, SCC accounts for 60-80% of the tumors in ear canal, middle ear or mastoid cavity.
Signs and symptoms

√ Close scrutiny of...

<table>
<thead>
<tr>
<th>External ear</th>
<th>Ear canals</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tympanic membranes</td>
<td>Parotid gland</td>
</tr>
<tr>
<td>Periauricular skin</td>
<td>Cervical lymph nodes</td>
</tr>
<tr>
<td>Cranial nerves</td>
<td></td>
</tr>
</tbody>
</table>

**External ear**

√ Typical presentation --- non-healing ulcer

√ Itching + occasional bleeding

√ If advanced --- skin erosion and cartilage exposure

√ Facial paralysis/facial numbness --- perineural spread
Signs and symptoms

Temporal bone

- Most common symptoms: OTORRHEA + OTALGIA + HEARING LOSS
  - Triad 10% PTs
- Suspect should arise when benign conditions do not respond to standard therapy
  - Pathology evaluation!
- Ddx should include: skull base osteomyelitis, pseudoepitheliomatous hyperplasia and carcinoma
- Advanced stage disease symptoms: trismus, facial weakness, dysphagia or hoarseness
- Temporal bone/ear canal are rare location for MTX (if... lung, breast, prostate or kidney primaries)
Signs and symptoms

✓ SCC
  ✓ Exophytic/ulcerated appearance
  ✓ Erythematous skin and granulation tissue

✓ BCC
  ✓ Ulcerated appearance with rolled edges

✓ Some tumors... subcutaneous spread
Diagnostic Imaging

When?

- Small, early-stage external ear cancers usually do not require imaging studies
- Late-stage skin cancers, spread to parotid gland or lymph nodes require imaging studies

Which?

- CT scan and MRI: complementary details
- CT: soft tissue + bony anatomy
- MRI: dural involvement or perineural spread suspected
Review the following spaces/locations systematically:

<table>
<thead>
<tr>
<th>4 Ear Canal Quadrants</th>
<th>Mastoid</th>
</tr>
</thead>
<tbody>
<tr>
<td>Infratemporal Fossa</td>
<td>Jugular Foramen</td>
</tr>
<tr>
<td>Middle Ear</td>
<td>Carotid Canal</td>
</tr>
<tr>
<td>Otic Capsule</td>
<td>Tegmen/Middle Fossa</td>
</tr>
<tr>
<td>Posterior Fossa</td>
<td></td>
</tr>
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</table>
Diagnostic Imaging
FALSE-NEGATIVE specimen are an important consideration in temporal bone carcinoma

- ↑ % infected secondarily
- Superficial biopsy specimens may reveal chronic inflammatory changes

What to do if initial biopsy negative?

- BIOPSY AGAIN!
  - Deep tissue biopsies in an operating room to ensure good samples are taken
STAGING – External Ear

✓ The external ear does not have a unique staging system — considered a high risk factor in the 2010 AJCC TNM for cutaneous malignancy
✓ 2 cm continues to be an important demarcation for staging

Table I

<table>
<thead>
<tr>
<th>Nonmelanoma skin cancer (including cutaneous squamous cell carcinoma) staging system in 6th edition of American Joint Committee on Cancer</th>
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<td><strong>Primary tumor (T)</strong></td>
</tr>
<tr>
<td>TX</td>
</tr>
<tr>
<td>T0</td>
</tr>
<tr>
<td>Tis</td>
</tr>
<tr>
<td>T1</td>
</tr>
<tr>
<td>T2</td>
</tr>
<tr>
<td>T3</td>
</tr>
<tr>
<td>T4</td>
</tr>
<tr>
<td><strong>Regional lymph nodes (N)</strong></td>
</tr>
<tr>
<td>NX</td>
</tr>
<tr>
<td>N0</td>
</tr>
<tr>
<td>N1</td>
</tr>
<tr>
<td><strong>Distant metastasis (M)</strong></td>
</tr>
<tr>
<td>Mx</td>
</tr>
<tr>
<td>M0</td>
</tr>
<tr>
<td>M1</td>
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</tbody>
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# STAGING - External Ear

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</tr>
<tr>
<td>T1 Tumor ≤ 2 cm in greatest dimension</td>
</tr>
<tr>
<td>T2 Tumor &gt; 2 cm, but not &gt; 5 cm, in greatest dimension</td>
</tr>
<tr>
<td>T3 Tumor &gt; 5 cm in greatest dimension</td>
</tr>
<tr>
<td>T4 Tumor invades deep extradermal structures</td>
</tr>
<tr>
<td><strong>Regional lymph nodes (N)</strong></td>
</tr>
<tr>
<td>NX Regional lymph nodes cannot be assessed</td>
</tr>
<tr>
<td>N0 No regional lymph node metastasis</td>
</tr>
<tr>
<td>N1 Regional lymph node metastasis</td>
</tr>
<tr>
<td><strong>Distant metastasis (M)</strong></td>
</tr>
<tr>
<td>Mx Distant metastasis cannot be assessed</td>
</tr>
<tr>
<td>M0 No distant metastasis</td>
</tr>
<tr>
<td>M1 Distant metastasis</td>
</tr>
</tbody>
</table>

## Table II

Definition of cutaneous squamous cell carcinoma tumor (T) staging system in the 7th edition of the American Joint Committee on Cancer

| TX Primary tumor cannot be assessed                           |
| T0 No evidence of primary tumor                                |
| Tis Carcinoma in situ                                         |
| T1 Tumor ≤ 2 cm in greatest dimension with <2 high-risk features*|
| T2 Tumor > 2 cm in greatest dimension with or without one additional high-risk feature, * or any size with ≥ 2 high-risk features *|
| T3 Tumor with invasion of maxilla, mandible, orbit, or temporal bone |
| T4 Tumor with invasion of skeleton (axial or appendicular) or perineural invasion of skull base |

*High-risk features include depth (>2-mm thickness; Clark level ≥ IV); perineural invasion; location (primary site ear; primary site nonglabrous lip); and differentiation (poorly differentiated or undifferentiated).
## Final 7th edition American Joint Committee on Cancer stage grouping for cutaneous squamous cell carcinoma

<table>
<thead>
<tr>
<th>Stage</th>
<th>T</th>
<th>N</th>
<th>M</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>In situ</td>
<td>N0</td>
<td>M0</td>
</tr>
<tr>
<td>I</td>
<td>T1</td>
<td>N0</td>
<td>M0</td>
</tr>
<tr>
<td>II</td>
<td>T2</td>
<td>N0</td>
<td>M0</td>
</tr>
<tr>
<td>III</td>
<td>T3</td>
<td>N0 or N1</td>
<td>M0</td>
</tr>
<tr>
<td></td>
<td>T1 or T2</td>
<td>N1</td>
<td>M0</td>
</tr>
<tr>
<td>IV</td>
<td>T1, 2, or 3</td>
<td>N2</td>
<td>M0</td>
</tr>
<tr>
<td></td>
<td>Any T</td>
<td>N3</td>
<td>M0</td>
</tr>
<tr>
<td></td>
<td>T4</td>
<td>Any N</td>
<td>M0</td>
</tr>
<tr>
<td></td>
<td>Any T</td>
<td>Any N</td>
<td>M1</td>
</tr>
</tbody>
</table>

TNM staging is incorporated within. Further modifications for 8th edition American Joint Committee on Cancer cutaneous squamous cell carcinoma staging system will be determined by studies that rely on evidence-based medicine.
The Pittsburgh T stage (Arriaga et al. 1990, modified by Moody et al.) has been shown to predict overall survival.

**Box 176-2. Modified University of Pittsburgh Tumor-Node-Metastasis Staging System Proposed for Neoplasms of the External Auditory Canal**

**T Status**

- **T1**: Tumor limited to the external auditory canal without bony erosion or evidence of soft tissue extension.
- **T2**: Tumor with limited external auditory canal bony erosion (not full thickness) or radiographic findings consistent with limited (<0.5 cm) soft tissue involvement.
- **T3**: Tumor erodes the osseous external auditory canal (full thickness) with limited (<0.5 cm) soft tissue involvement or tumor involves middle ear or mastoid.
- **T4**: Tumor erodes the cochlea, petrous apex, medial wall of middle ear, carotid canal, jugular foramen, or dura or shows extensive (>0.5 cm) soft tissue involvement or evidence of facial paralysis.

**Nodal Status**

Involvement of lymph nodes is a poor prognostic finding and automatically places the patient in an advanced stage (i.e., stage III [T1, N1] or stage IV [T2, T3, and T4, N1] disease).

**Metastatic Status**

Distant metastasis indicates a poor prognosis and immediately places the patient in the stage IV category.

<table>
<thead>
<tr>
<th>OVERALL STAGE</th>
<th>T Status</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>T1NO</td>
</tr>
<tr>
<td>II</td>
<td>T2NO</td>
</tr>
<tr>
<td>III</td>
<td>T3NO</td>
</tr>
<tr>
<td>IV</td>
<td>T4NO and T1-4N+</td>
</tr>
</tbody>
</table>
TEMPORAL BONE TUMORS

TREATMENT
Selection of surgical procedure TEMPORAL BONE

**T_1:** Tumor limited to the external auditory canal without bony erosion or evidence of soft tissue extension

**T_2:** Tumor with limited external auditory canal bony erosion (not full thickness) or radiographic finding consistent with limited (<0.5 cm) soft tissue involvement

→ **PARTIAL TEMPORAL BONE RESECTION +/- SUPERFICIAL PAROTIDECTOMY**

**T_3:** Tumor erodes the osseous external auditory canal (full thickness) with limited (<0.5 cm) soft tissue involvement or tumor involves middle ear or mastoid

→ **PARTIAL TEMPORAL BONE RESECTION VS. SUBTEMPORAL BONE RESECTION**

**T_4:** Tumor erodes the cochlea, petrous apex, medial wall of middle ear, carotid canal, jugular foramen, or dura or shows extensive (>0.5 cm) soft tissue involvement or evidence of facial paralysis

→ **SUBTEMPORAL BONE RESECTION SURGERY OF CHOICE (+ RT) VS. TOTAL TEMPORAL BONE RESECTION?**
Treatment - surgery

PARTIAL (LATERAL) TEMPORAL BONE RESECTION

- TNM: T1 and T2
- Medial limit of resection: facial nerve, stapes and promontory
- Includes removal of entire external auditory meatus (en bloc)

STAGES
1. Complete mastoidectomy with tegmen mastoideum/sigmoid sinus identification
2. Facial nerve dissection from LSCC to SM foramen
3. Posterior tympanotomy is extended inferiorly to expose the hypotympanum
4. Antero-inferior extension of the tympanotomy is carried out
3. Objective: separate the inferior portion of the tympanic bone - medial wall of the middle ear in the area of the hypotympanum
PARTIAL (LATERAL) TEMPORAL BONE RESECTION

4. **INFERIOR** - Drilling should be extended anteriorly until the temporomandibular joint is reached

5. Mastoid tip is dissected away (take care to facial nerve)

6. Incudostapedial joint is disarticulated to avoid SN hearing loss

7. **SUPERIOR** - Drilling in the attic area + atticotomy is extended anteriorly until the TMJ is opened
8. The tensor tympani tendon attachment to the malleus is sharply cut + ligamentous attachments of the ossicles

9. THE ANTERIOR PORTION of the external ear canal is the only attachment

8. Fracture the bone with gentle pressure/osteotome
PARTIAL (LATERAL) TEMPORAL BONE RESECTION

- If PTB resection as part of STB/TTB resection + facial nerve involved --- proximal/distal margins of facial nerve should be examined
  - Interposicional graft --- greater auricular nerve, sural nerve...

- If parotid gland involved (primarily/secondarily), it should be removed in continuity

- Primary temporal bone rarely MTX to cervical lymph nodes
  - Prudent cervical lymph node dissection if
    - 1) secondary involvement of parotid gland
    - 2) primary cancers of salivary gland
SUBTOTAL TEMPORAL BONE RESECTION

✓ **WHEN?** If middle ear extension/facial nerve (T3-T4)
✓ STBR extends into the labyrinth, cochlear, or both
  ✓ Medial margin: internal auditory canal
✓ Skin incision: large C-shaped postauricular incision
✓ Bony dissection
  ✓ First --- Lateral/partial temporal bone resection
  ✓ After that? Piecemeal tumor dissection --- intraoperative samples
    ✓ Labyrinthectomy, jugular foramen dissection, cochlectomy...
    ✓ Carotid artery often must be dissected/decompressed
✓ Posterior/middle fossa dura is often involved
TOTAL TEMPORAL BONE RESECTION

✓ Rarely performed because...
  ✓ HIGH LEVEL OF MORBIDITY
  ✓ LACK OF WELL-DOCUMENTED SURVIVAL BENEFIT (EXCEPT VERRUCOUS SQUAMOUS CARCINOMA)

✓ Main difference STB vs TTB resection
  ✓ TTB resection involves petrous apex resection
Surgery contraindications

- The obvious... 1) unresectable disease, 2) distant metastasis or 3) poor mental health status

- **LOCAL EXTENSION REASONS**
  - Carotid or vertebral artery
  - *Carotid artery by-pass?*
    - Long-term results yield to 20% - 2 year-survival
    - Attendant risk of postoperative stroke/death
  - Cervical spine erosion
  - Significant brain invasion
Treatment - radiotherapy

✓ PRIMARY RADIOTHERAPY
  ✓ Used to treat temporal bone cancers up to 1970s
  ✓ Low overall cure rate
  ✓ OPTION FOR PATIENTS NOT CANDIDATES FOR SURGERY

✓ ADYUVANT RADIOTHERAPY
  ✓ HAS IMPROVED OVERALL SURVIVAL
  ✓ RECOMMENDED FOR T2 OR >T2
  ✓ OTHER INDICATIONS
    ✓ Recurrent tumors, positive margins, perineural spread, + lymph nodes o extracapsular spread
Treatment - chemotherapy

Only a few isolated studies have examined the role of CT for temporal bone cancers.
SURVIVAL AND RECURRENCES RATES

✓ PITTSBURGH TUMOR STAGING IS AN IMPORTANT, INDEPENDENT FACTOR FOR PROGNOSIS FOR SCC

✓ T1 and T2 can be completely excised with LTBR
  ✓ T1 surgery alone
  ✓ T2 surgery + PORT = improved outcomes

✓ T3 and T4 can no longer be excised with LTBR
  ✓ SURGERY + RADIOTherapy + CHEMOTHERAPY

  ✓ Higgins et al. (2010): 5-year overall survival dropped to 19.1% in PTs with facial palsy (vs. 59.4%),
  (regardless tumor stage)

80-100% 5-year survival rates

< 50% 5-year survival rates
The End