

Ear and Temporal Bone Cancer

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Introduction

- ✓ 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

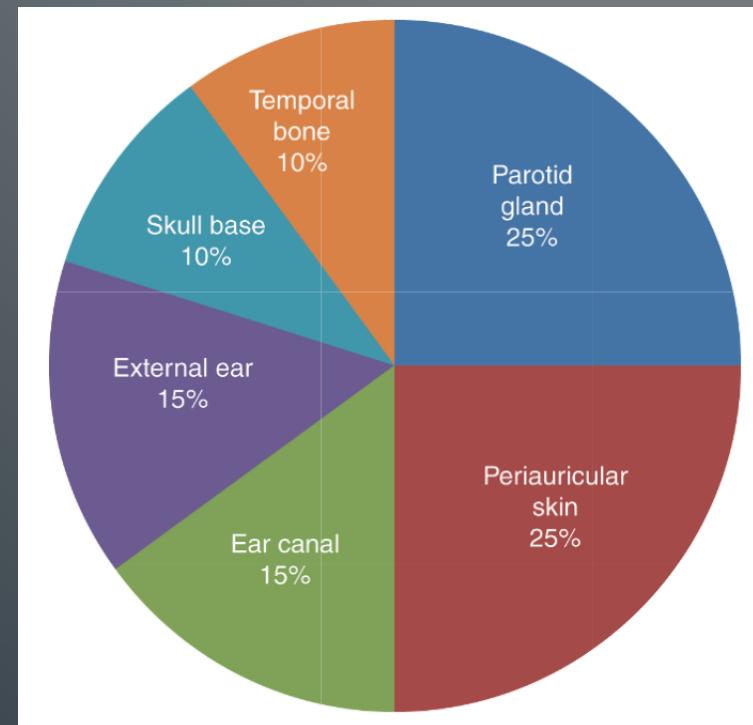
EXTERNAL EAR

- ✓ 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.

Histologic types - external ear

- ✓ 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

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

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

* 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).

Histologic types - temporal bone

Box 176-1. TEMPORAL BONE NEOPLASMS

Primary Benign Tumors

Paraganglioma
Neurofibroma/schwannoma
Meningioma
Adenoma
 Ceruminous adenoma
 Eccrine cylindroma
 Pleomorphic adenoma

Mesenchymal Neoplasms

Chondroma
Chondroblastoma
Chondromyxoid fibroma
Hemangioma
Lipoma
Myxoma
Fibroosseous tumors
 Ossifying fibroma
 Fibrous dysplasia
 Giant cell granuloma
Aneurysmal bone cysts
Osteoblastoma
Osteoma/exostosis
Unicameral bone cyst
Teratoma

Dysontogenic Tissue

Choristoma
Inverted papilloma
Glioma

Metastatic Tumors

Prostate
Breast
Gastrointestinal
Renal cell
Lung
Multiple myeloma
Lymphoma
Leukemia (chloroma)

Primary Malignant Tumors

Epidermal Carcinomas
Squamous cell carcinoma
Verrucous carcinoma
Basal cell carcinoma
Melanoma

Adenocarcinomas

Ceruminous adenocarcinoma (low and high grade)

Adenoid cystic adenocarcinoma

Mucoepidermoid adenocarcinoma

Sebaceous cell adenocarcinoma

Papillary cystadenocarcinoma

Mesenchymal Neoplasms

Rhabdomyosarcoma

Fibrosarcoma

Chondrosarcoma

Osteosarcoma

Liposarcoma

Dermatofibrosarcoma protuberans

Fibrohistiocytoma

Angiosarcoma

Osteoclastoma

Chordoma

Plasmacytoma

Contiguous tumor invasion

Neuronal structures

 Meningioma

 Glioma

 Neurofibroma/schwannoma

Choroid plexus papilloma

Parotid gland neoplasm

Periauricular Skin Carcinoma

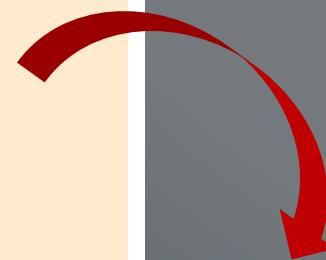
Pituitary tumors

Craniopharyngioma

Chordoma

Scalp tumors

Nasopharyngeal carcinoma



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...

External ear	Ear canals
Tympanic membranes	Parotid gland
Periauricular skin	Cervical lymph nodes
Cranial nerves	

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**

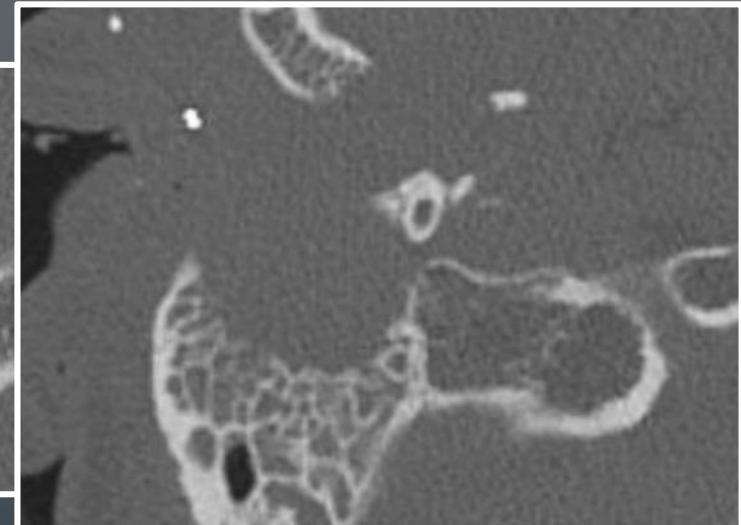
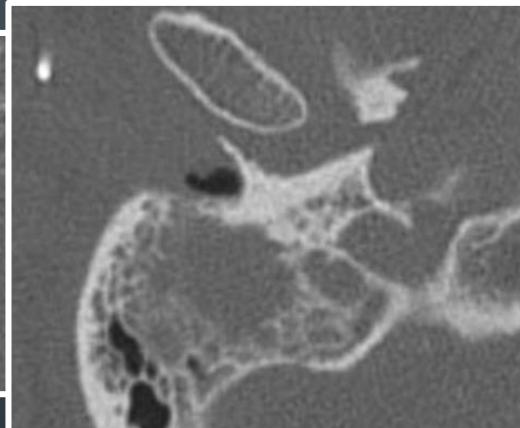
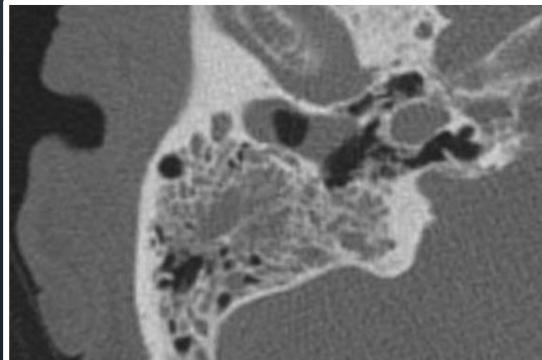
Which?

- ✓ CT scan and MRI: complementary details
- ✓ CT: soft tissue + bony anatomy
- ✓ MRI: dural involvement or perineural spread suspected

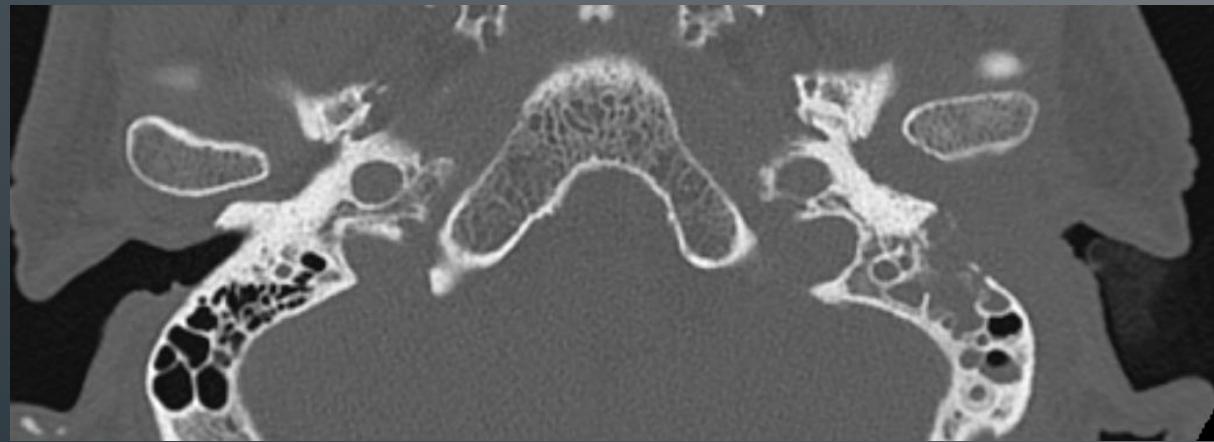
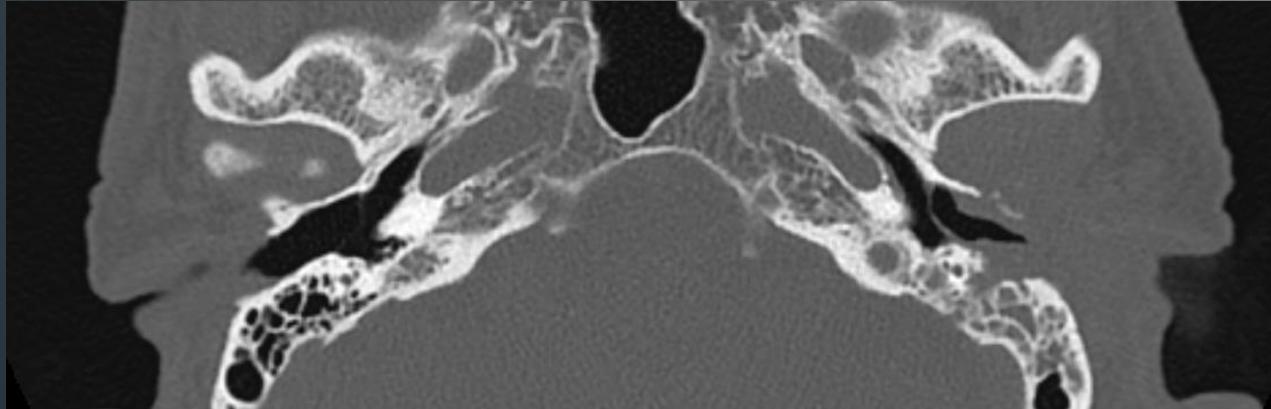
Diagnostic Imaging

✓ REVIEW THE FOLLOWING SPACES/LOCATIONS SYSTEMATICALLY

4 EAR CANAL QUADRANTS	MASTOID
INFRA TEMPORAL FOSSA	JUGULAR FORAMEN
MIDDLE EAR	CAROTID CANAL
OTIC CAPSULE	TEGMEN/MIDDLE FOSSA
POSTERIOR FOSSA	



Diagnostic Imaging



Biopsy

- ✓ 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

Nonmelanoma skin cancer (including cutaneous squamous cell carcinoma) staging system in **6th edition of American Joint Committee on Cancer**

Primary tumor (T)

- TX Primary tumor cannot be assessed
- T0 No evidence of primary tumor
- Tis Carcinoma in situ
- T1 Tumor ≤ 2 cm in greatest dimension
- T2 Tumor >2 cm, but not >5 cm, in greatest dimension
- T3 Tumor >5 cm in greatest dimension
- T4 Tumor invades deep extradermal structures

Regional lymph nodes (N)

- NX Regional lymph nodes cannot be assessed
- N0 No regional lymph node metastasis
- N1 Regional lymph node metastasis

Distant metastasis (M)

- Mx Distant metastasis cannot be assessed
- M0 No distant metastasis
- M1 Distant metastasis

STAGING – External Ear

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- Mx Distant metastasis cannot be assessed
- M0 No distant metastasis
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Table II

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

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* High-risk features include depth (>2 -mm thickness; Clark level $\geq IV$); 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

Stage	T	N	M
0	In situ	N0	M0
I	T1	N0	M0
II	T2	N0	M0
III	T3	N0 or N1	M0
	T1 or T2	N1	M0
IV	T1, 2, or 3	N2	M0
	Any T	N3	M0
	T4	Any N	M0
	Any T	Any N	M1

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.

STAGING - Temporal Bone

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 finding 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.

The **Pittsburgh T stage** (Arriaga et al. 1990, modified by Moody et al.) has been shown to **predict overall survival**

OVERALL STAGE	
I	T1N0
II	T2N0
III	T3N0
IV	T4N0 and T1-4N+

TEMPORAL BONE TUMORS

TREATMENT

Selection of surgical procedure TEMPORAL BONE

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 finding consistent with limited (<0.5 cm) soft tissue involvement



PARTIAL TEMPORAL BONE RESECTION
+/- SUPERFICIAL PAROTIDECTOMY

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



PARTIAL TEMPORAL BONE RESECTION
VS.
SUBTEMPORAL BONE RESECTION

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



SUBTEMPORAL BONE RESECTION
SURGERY OF CHOICE (+ RT) VS.
TOTAL TEMPORAL BONE RESECTION?

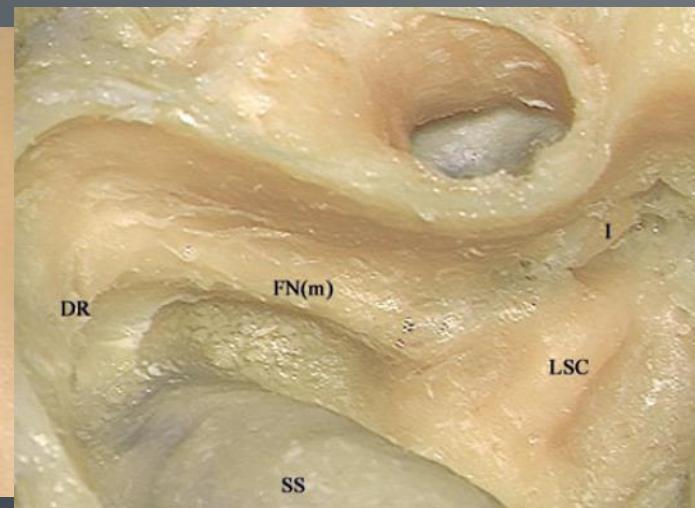
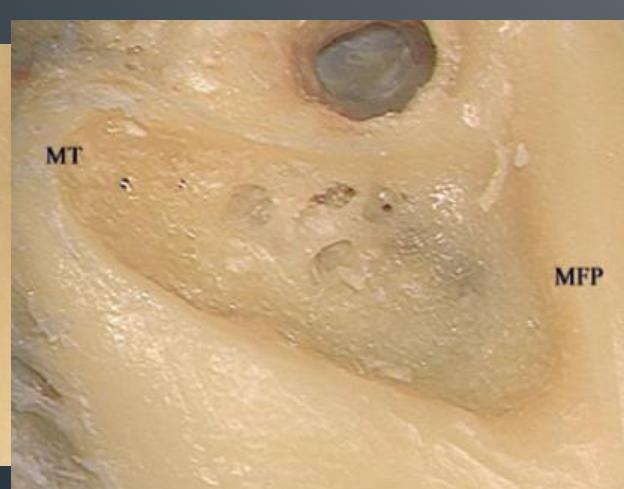
Treatment - surgery

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)

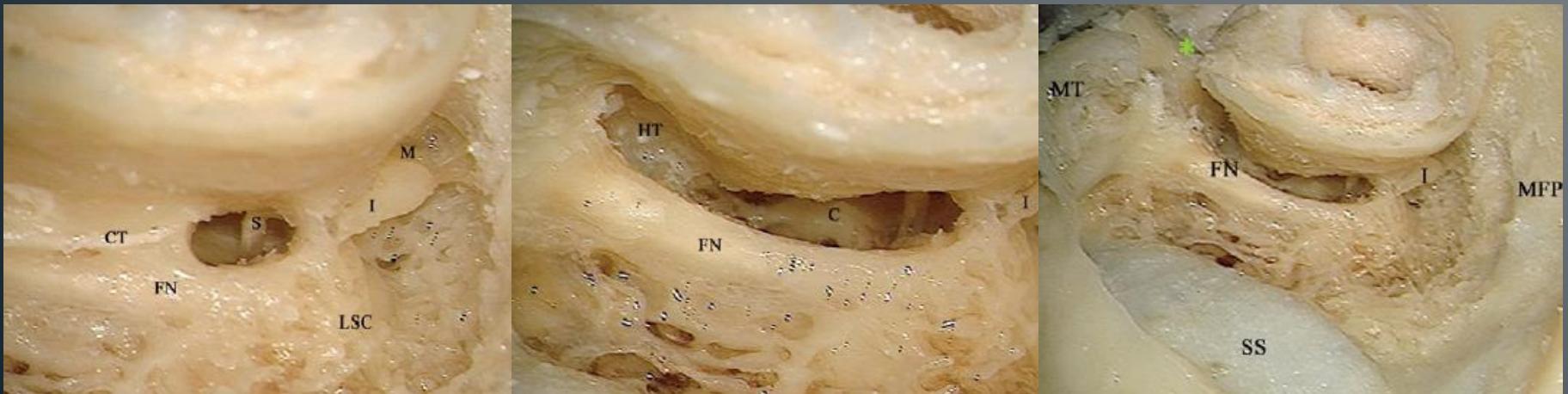
STAGES

1. Complete mastoidectomy with tegmen mastoideum/sigmoid sinus identification
2. Facial nerve dissection from LSCC to SM foramen



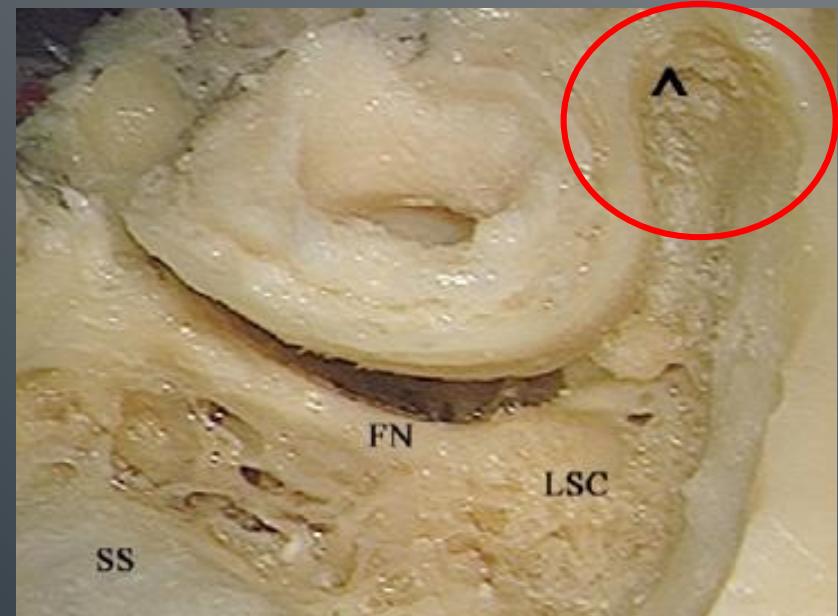
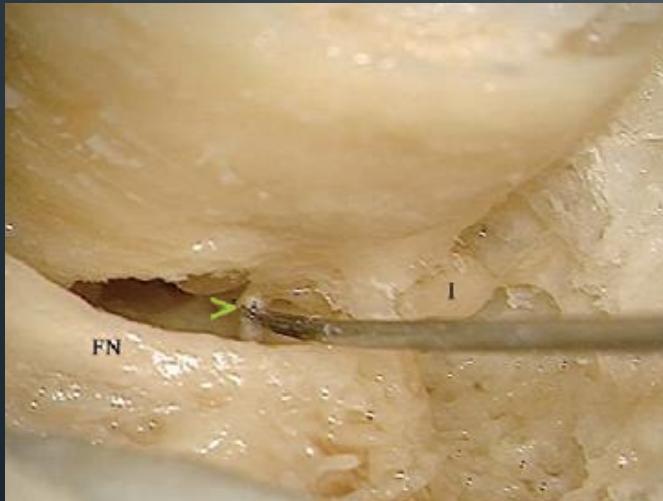
PARTIAL (LATERAL) TEMPORAL BONE RESECTION

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



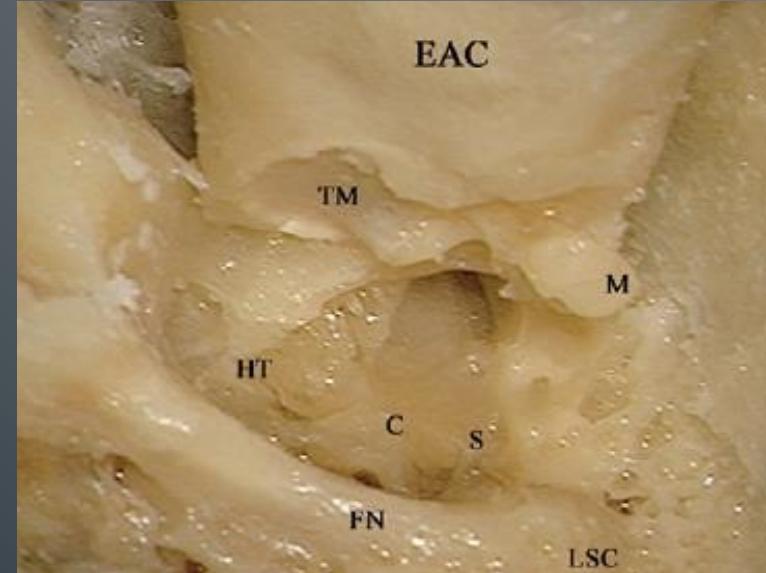
PARTIAL (LATERAL) TEMPORAL BONE RESECTION

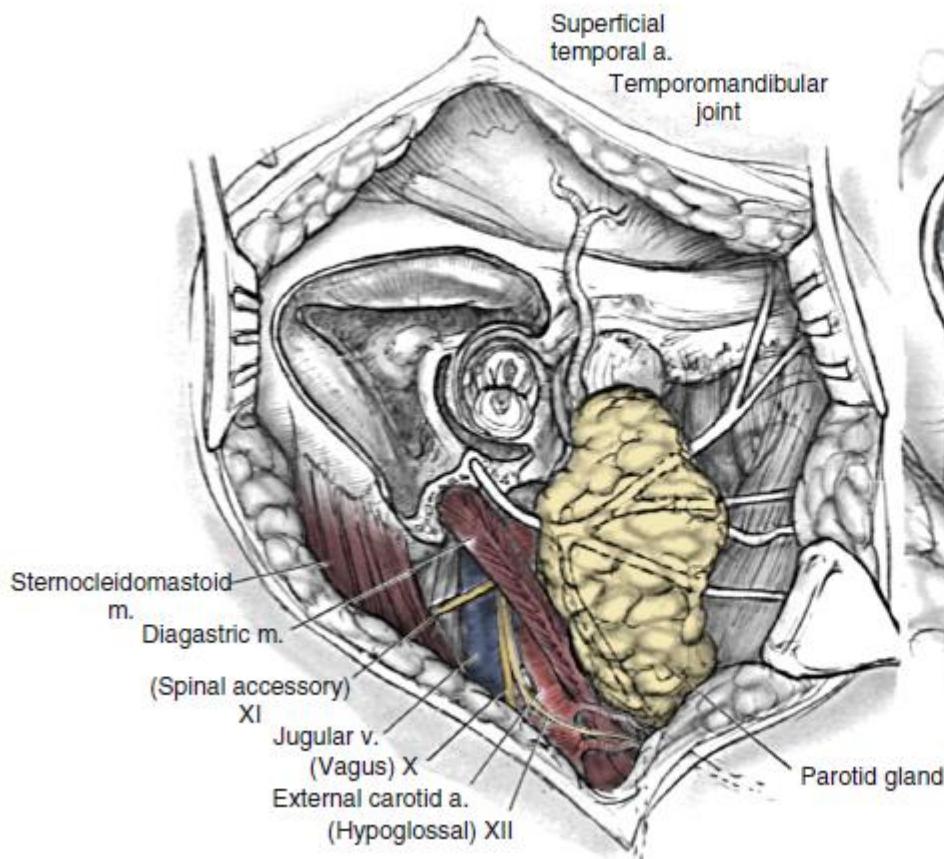
4. **INFERIOR** - Drilling should be extended anteriorly until the temporomandibular joint is reached
5. Mastoid tip is dissected away (take care to facial nerve)
6. Incudostapedial joint is disarticulated to avoid SN hearing loss
7. **SUPERIOR** - Drilling in the attic area + atticotomy is extended anteriorly until the TMJ is opened



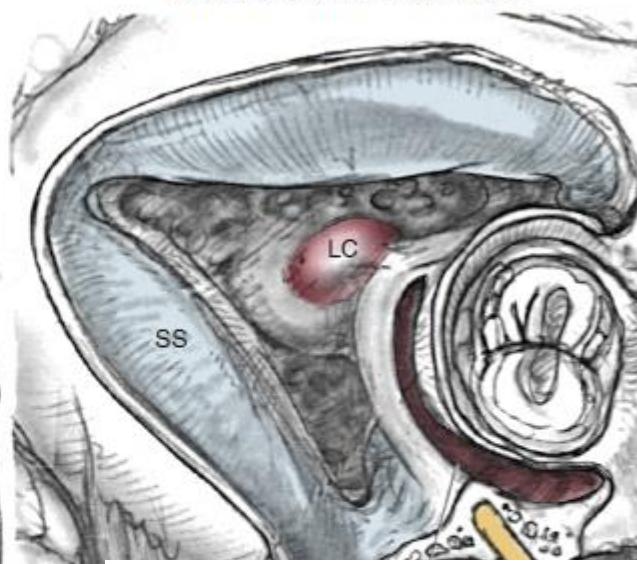
PARTIAL (LATERAL) TEMPORAL BONE RESECTION

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

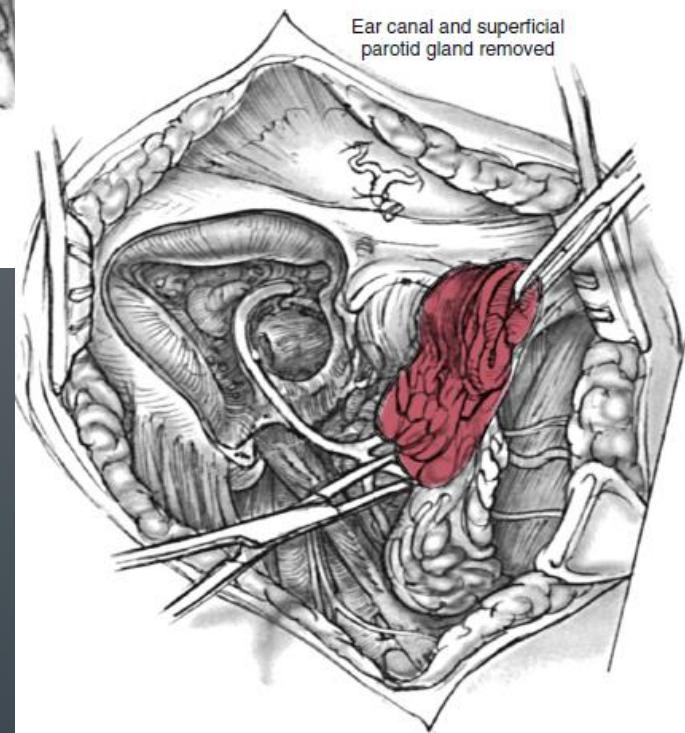


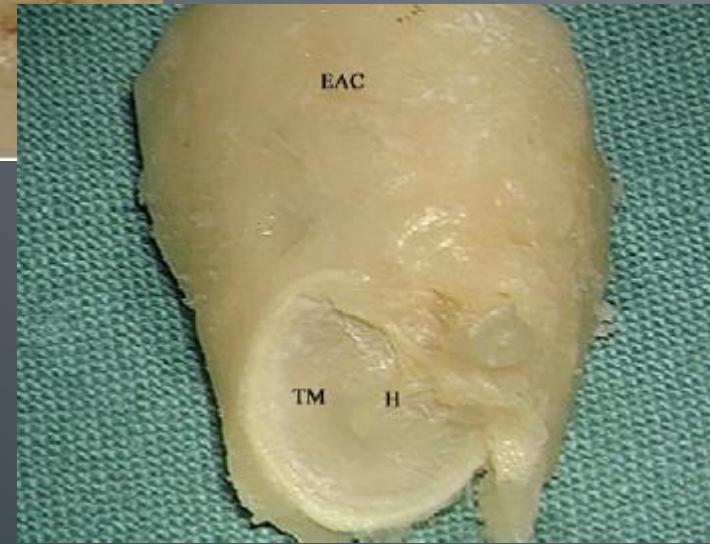
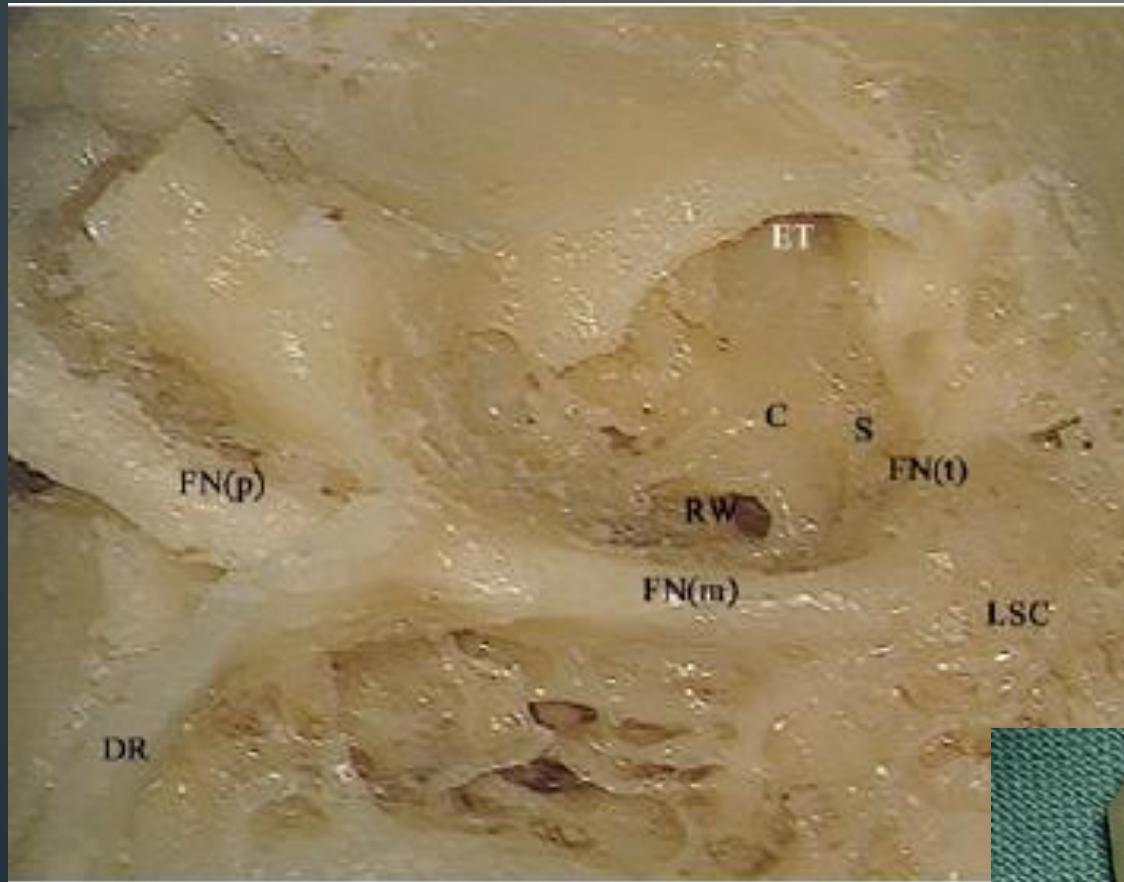


MASTOIDECTOMY ENLARGED



Ear canal and superficial parotid gland removed





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
 - ✓ Interpositional 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

Treatment - surgery

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

Treatment - surgery

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

✓ ADJUVANT 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

ORIGINAL ARTICLE

Concomitant Chemoradiotherapy as a Standard Treatment for Squamous Cell Carcinoma of the Temporal Bone

Kiyoto Shiga, M.D.,¹ Takenori Ogawa, M.D.,¹ Atsuko Maki, M.D.,¹ Masanori Amano, M.D.,¹ and Toshimitsu Kobayashi, M.D.¹

concomitant with low dose docetaxel. Nine patients with stage IV tumors were treated by CCRT using the TPF regimen (docetaxel, cisplatin, and 5-fluorouracil). As an initial treatment, all patients but one were treated by radiation therapy with or without chemotherapy. Grade 4 adverse events of patients who received CCRT using the TPF regimen

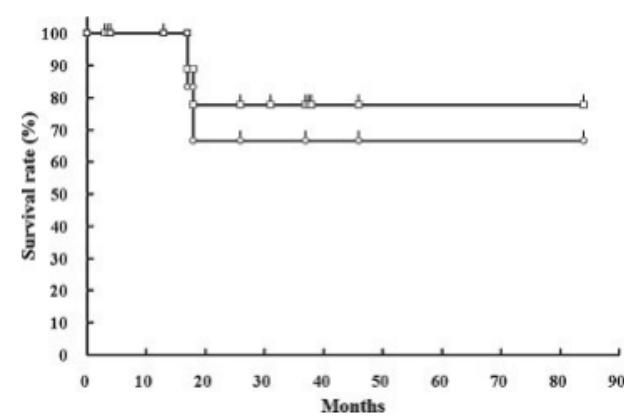


Figure 3 Survival curves of the patients. Disease-specific survival rates were calculated by the Kaplan-Meier method. The 5-year survival rate of all patients was 78% (open squares), and of T4 and stage IV patients was 67% (open circles).

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)
-
- The diagram illustrates the relationship between tumor stage and survival rates. Two brackets group the information: one bracket groups T1 and T2 under the heading '80-100% 5-year survival rates', and another bracket groups T3 and T4 under the heading '< 50% 5-year survival rates'.

The End