TUMOURS OF THE TEMPORAL BONE

RISK FACTORS:

- NF2 gene – Chr 22
- VHL (Von Hippel Lindau) disease – Chr 3p
- Ionizing radiation

Criteria for radiation induced temporal bone tumours by LUSTIG is as follows:

1. The second neoplasm must develop in the irradiated field.
2. A latent period of at least several years must elapse between radiation exposure and the development of a second primary.
3. The previous condition must show histological, radiographic and microscopic evidence of neoplasia.
4. The second tumour must be of a different histological type from that previously irradiated.

Chronic suppurative otitis media

- A relationship is postulated between the development of squamous carcinoma of the temporal bone and CSOM.

HPV 16, 18

CLASSIFICATION OF TUMOURS OF TEMPORAL BONE

<table>
<thead>
<tr>
<th>Cutaneous neoplasms</th>
<th>Glandular neoplasms</th>
<th>Vascular/haematological</th>
<th>Paraganglioma</th>
<th>Bone</th>
<th>Neural</th>
<th>Developmental and congenital</th>
</tr>
</thead>
<tbody>
<tr>
<td>Squamous cell papilloma and carcinoma</td>
<td>Ceruminous adenoma and adenocarcinoma</td>
<td>Haemangioma</td>
<td>Glomus</td>
<td>tympanicum</td>
<td>Osteoma</td>
<td>V, VII, VIII, IX, X,</td>
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<tr>
<td>Schwannoma</td>
<td>Chordoma</td>
<td>Pleomorphic adenoma</td>
<td>Haemangiopericytoma</td>
<td>Glomus jugulare</td>
<td>Secondary tumours</td>
<td>Meningioma</td>
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<tr>
<td>Basal cell papilloma and carcinoma</td>
<td>Adenoid cystic carcinoma</td>
<td>Leukaemia</td>
<td>Glomus vagale</td>
<td>Rhabdomyosarcoma</td>
<td>Teratoma</td>
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<tr>
<td>Malignant melanoma</td>
<td>Middle ear adenoma</td>
<td>Lymphoma</td>
<td>Plasmacytoma</td>
<td>Chondrosarcoma</td>
<td>Choristoma</td>
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<tr>
<td>Endolymphatic sac tumours</td>
<td>Langerhans' cell histiocytosis</td>
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LANGERHANS' CELL HISTIOCYTOSIS

Langerhans' cell histiocytosis (LCH), formerly called histiocytosis X, is a condition that is characterized by the proliferation of histiocytes that share the characteristic of Langerhans' cells (normally located in the dermis).
Langerhans' cell histiocytosis is now the collective name for the following conditions that were previously thought to be three distinct entities (HEL), but are now recognized as one condition:

1. **Eosinophilic granuloma**: Generally refers to osseous disease alone and may be Unifocal or multifocal

2. **Hand-Schuller-Christian disease**: A systemic disease identified by multifocal osseous lesions, with limited involvement of extra skeletal sites, such as lymph nodes and viscera.

3. **Letterer-Siwe disease**: The most serious and rapidly progressive disease, characterized by disseminated disease with multiorgan involvement.

**EPIDEMIOLOGY**

- Mean Age – 3 yrs
- M:F = 1.7:1
- The ultimate prognosis of patients with LCH depends on whether the disease is unifocal, multifocal or disseminated.

**CLINICAL FEATURES**:

- Symptoms and signs of ear and temporal bone involvement are often indistinguishable from that of CSOM and may include a conductive or sensorineural hearing loss.
- The characteristic radiological appearance of the temporal bone is that of a 'punched out' or lytic appearance.
- While extensive destruction of the temporal bone can take place, there are only limited reports of otic capsule involvement with disease often confined to the external and middle ear.

**TREATMENT**:

- Age at presentation of less than two years with organ dysfunction is an indicator of a much poorer prognosis.
- Multidisciplinary, multimodality fashion with a combination of radiotherapy, chemotherapy and Steroid therapy.

**SQUAMOUS CELL Ca OF TEMPORAL BONE**

**EPIDEMIOLOGY**

- F>M
- 7th decade
- Although rare, SCC of the temporal bone can develop in younger adults.
AETIOLOGICAL THEORIES

CHRONIC SUPPURATIVE OTITIS MEDIA

- Suggested that chronic irritation and inflammation may be a carcinogenic stimulus.
- The carcinoma may originate from the squamous epithelium lining the external auditory canal (EAC) or of a mastoid cavity. Alternatively, it could develop as a result of metaplasia of the cuboidal epithelium lining the middle ear.

RADIATION-ASSOCIATED TUMOURS

- Although fortunately rare, tumours of the temporal bone are a well-described sequela of radiotherapy for nasopharyngeal and other intracranial malignancies.

SURGICAL ANATOMY AND CLINICAL FEATURES

- Local, vascular, Neural, spread via natural foramina and fissures of temporal bone.
- LOCAL SPREAD

  - Invasion of blood vessels of EAC leads to bloody Otorrhoea.
  - The fissures of Santorini, the petroquamous fissure and foramen of Husche allow anterior extension into the temporomandibular joint, glenoid fossa, parotid and infratemporal region causing trismus, pain and preauricular swelling.
  - Continued anterior growth into the root of the zygoma, masseter muscle and mandible gives rise to pain when chewing.
  - Inferior spread into the stylomastoid foramen and Fallopian canal results in progressive facial weakness or palsy. (7th involved)
  - Inferomedial extension into the jugular foramen (9, 10, 11, 12 nerve involvement) causes dysphagia and dysphonia secondary to involvement of the glossohyaryngeal and vagal nerves.
  - Accessory nerve infiltration inflicts shoulder dysfunction
  - Hypoglossal invasion causes articulatory problems from weakness of the tongue.
  - Further involvement inferiorly through the foramen magnum and cervical vertebrae induces neck Pain, Stiffness and Fullness.
  - Medial spread into the middle ear cleft increases any conductive hearing loss.
  - Extension into the petrous apex and involvement of the internal carotid artery (ICA) is also possible but less likely since the bone of the otic capsule is highly resistant to tumour invasion.
  - Spread to the nasopharynx through the Eustachian tube is seen occasionally in patients with extremely advanced tumours.
  - Vertigo and sensorineural hearing loss may be explained by involvement of the vestibule or internal auditory canal.
  - Superior spread through the epitympanic recess and the thin bone of the tegmen tympani is a common development. Invasion of the middle fossa dura and subsequently the temporal lobe of the brain cause intractable pain and persistent headache.
  - Posterior spread into the posterior fossa through the mastoid air cells is also seen
  - Lymph node mets in <10% as they are sparse.
Distant mets rare though conversely haematogenous spread to the temporal bone from other primary tumour sites is well recognized and metastases should always be considered in the differential diagnosis of any destructive lesion of the temporal bone.

Squamous cell carcinoma is notorious for perineural invasion. The proximity of the facial nerve makes the nerve most at risk and patients often present with facial weakness.

**DIAGNOSIS AND INVESTIGATION**

- CSOM with the onset of pain and bloody or offensive Otorrhoea are both very significant symptoms.
- Multiple cranial nerve palsies

**EXAMINATION**

- Examination of the EAC and middle ear may yield little more than an ulcer, granular area or polypoid mass.
- However, an exophytic mass within the EAC is likely to be malignant and is a strong indication for an urgent biopsy. In view of the inevitable coexistence of SCC and chronic infection, multiple biopsies may be needed to confirm the disease. Superficial biopsies often show only inflammation or dysplasia and can be very misleading.
- The neck must be examined carefully, particularly in the parotid region, to determine the patient's nodal status.

**DIFFERENTIAL DIAGNOSIS**

- Pseudo-epithelial hyperplasia
- Basal cell carcinoma
- Adenocarcinoma
- Chondrosarcoma
- Melanoma
- Ewing's tumour
- Histiocytosis X
- Fibroxanthoma
- Verrucous carcinoma
- Metastatic disease

**IMAGING**

- Bone windowed CT portrays bone erosion accurately while MR images show the extent of soft tissue invasion.
- 12 specific anatomical regions as follows:
  - 1. anterior canal;
  - 2. superior canal;
  - 3. posterior canal;
  - 4. inferior canal;
  - 5. infratemporal extension;
  - 6. middle ear;
MR scans with gadolinium DTPA enhancement are better for the assessment of soft tissue disease. If CT or MR raises the possibility of (ICA) involvement, angiography with balloon occlusion should be considered, as sacrifice of the ICA may be necessary.

STAGING

Staging system: Clark's modification of Stell et al.'s proposal.

<table>
<thead>
<tr>
<th>Stage</th>
<th>Spread of Tumour</th>
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<td>T1</td>
<td>Tumour limited to site of origin</td>
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<tr>
<td>T2</td>
<td>Tumour extending beyond site of origin indicated by facial paralysis or radiological evidence of bone destruction</td>
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<tr>
<td>T3</td>
<td>Involvement of parotid gland/temporomandibular joint/skin (i.e. extracranial)</td>
</tr>
<tr>
<td>T4</td>
<td>Involvement of dura/base of skull (i.e. cranial)</td>
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</tbody>
</table>

Other classification system is Pittsberg classification.
It should be noted that the presence of metastatic lymph nodes is a poor prognostic indicator and consequently upstages the disease regardless of the T status. For example, T1 N1 = stage III, T2,3,4 N1 = stage IV.

**MANAGEMENT**

- Most favorable survival rates are achieved with enbloc extended temporal bone resection and postoperative radiotherapy.

**SURGICAL MANAGEMENT OF THE PRIMARY SITE**

- The type of resection performed is determined by the stage of the tumour.

**LATERAL TEMPORAL BONE RESECTION**

- Tumours lateral to the tympanic membrane (T1 & T2) can be managed adequately by a lateral temporal bone.
- There is considerable debate about the need to remove the entire pinna.
- There is no doubt that its removal constitutes a significant handicap both in terms of aesthetics and function for those who wear spectacles.
- A cortical mastoidectomy with a posterior tympanotomy is performed so that the facial nerve can be defined from the geniculate ganglion to the stylomastoid foramen.
- It is wise to remove the articular disc at the least and drill out the glenoid fossa as The anterior margin of the tumour is the posterior aspect of the temporomandibular joint.
- If possible, the temporal bone resection should be removed en bloc with the superficial lobe of the parotid gland and neck dissection.
- The mastoid air cell system is then exenterated, the Eustachian tube occluded and the cavity obliterated.

**EXTENDED TEMPORAL BONE RESECTION**

- Tumours that extend beyond the boundaries of the EAC i.e. T3 T4
- Total temporal bone resection is the management of choice for these tumours.
- For some, removal of the Condyle, coronoid process and the upper two-thirds of the vertical ramus of the mandible together with the soft tissue of the infratemporal fossa and parotid gland may be necessary. In a few, resection of dura and brain may be required. No attempt should be made to reserve the facial nerve as it will always be infiltrated by tumour.
- Remove pinna completely
- A posterior and middle craniotomy is performed resecting 3 cm of bone behind the sigmoid sinus and over the middle fossa. The sigmoid sinus and jugular bulb are delineated and the dissection is extended medially keeping as much of the mastoid cortex intact as possible. The labyrinth is then removed, the internal auditory canal transected and the intrapetrous portion of the carotid exposed. The middle fossa dura is retracted and the bony dissection extended forward through the root of the zygoma. At this stage the floor of the middle fossa is inspected for breach by the tumour and any extension into the dura and brain is assessed.
- The anterior margin of the resection includes the entire parotid gland.
The petrous tip medial to the carotid is resected separately after the intrapetrous carotid artery has been skeletonized.

The dural defect is repaired with fascia lata, sutured in place to achieve a 'watertight' seal.

**SURGICAL MANAGEMENT OF THE NECK**

- During the neck dissection, care is taken to preserve arterial and venous structures appropriate for reconstruction if a free tissue transfer is the method of choice.
- Parotidectomy with lymph nodes is a routine component of surgical treatment of SCC of the temporal bone.

**RECONSTRUCTION**

- The surgical defect is usually large and may require soft tissue transfer.
- They suggested that reconstructive techniques should be used in a graduated sequence as necessary:
  1. Temporal fascia:
  2. Free
  3. Pedicled rotation
  4. Fibrin glue
  5. Fat graft with or without dermis
  6. Temporalis muscle rotation
  7. Myocutaneous:
     a. Pectoralis
     b. Trapezius
     c. Latissimus dorsi
  8. Free tissue transfer.

**ADJUVANT THERAPY**

**RADIOThERAPY**

- Radiotherapy has been employed as the primary treatment modality for SCC of the ear and temporal bone.

**CHEMOTHERAPY**

- Debulk the tumour with repeated applications of 5FU

**COMPLICATIONS OF TREATMENT**

1. CRANIAL NERVE PALSIES
2. FLAP FAILURE
3. CEREBROSPINAL FLUID LEAK