# Head & Neck Tumours Part II

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### Content

- Tumours of the Ears
- Tumours of the Nose
- Tumours of the Mouth
- Tumours of the Pharynx
- Tumours of the Larynx

# Neoplasms of the Ear and Lateral Skull Base

- Lesions of the Pinna and EAC
- Lesions of the Middle Ear and Mastoid
- Lesions of the Petrous Apex and Clivus
- Lesions of the IAC, CPA, and Skull Base

# Introduction

- Generally classified by location, and occasionally by cell-type
- Causes of these neoplasms are largely unknown?

# Neoplasms of the pinna and external auditory canal

#### <u>Cutaneous carcinoma</u>

Squamous cell carcinoma Basal cell carcinoma

#### Malignant melanoma

Glandular neoplasm Ceruminous adenoma Ceruminous adenocarcinoma Pleomorphic adenoma Adenoid cystic carcinoma

#### Osteoma and exostosis

Miscellaneous neoplasm Merkel cell carcinoma Squamous papilloma Pilomatrixoma Myxoma Auricular endochondrial pseudocyst Chondrodermatitis nodularis chronica helicis (Winkler disease)

### Lesions of the Petrous Apex and Clivus

• Adenomatous neoplasm Benign middle ear adenoma Endolymphatic sac tumor

#### Chordoma

Congenital neoplasm Dermoid Teratoma Choristoma <u>Cholesterol granuloma</u> Langerhans cell histiocytosis Eosinophilic granuloma Hand-Schüller-Christian disease Letterer-Siwe disease Sarcoma Rhabdomyosarcoma Chondrosarcoma Ewing sarcoma Osteogenic sarcoma Fibrosarcoma

# Neoplasms of the internal auditory canal and cerebellopontine angle

### <u>Schwannoma</u>

Vestibular schwannoma Facial nerve schwannoma Trigeminal schwannoma Jugular foramen schwannoma Meningioma Lipoma Metastases

### Neoplasms of the Pinna and EAC

### **Cutaneous Carcinoma BCC**

- BCC (20% of ear/TB neoplasms)
- Most on pinna
- Sun exposure is initiator
- Locally infiltrative, rolled border central crusting ulcer
- May invade TB if left untreated

# Cutaneous Carcinoma SCCA

- Pinna and EAC are common
- Sun, cold, radiation are all factors
- Scaly irregular indurated maculopapular lesion, often ulcerated with sero-sang d/c
- Can be confused with OE
- Other symptoms VII, CHL, SNHL (with invasion of TB)
- Met. To LN more common than BCC

# Cutaneous Carcinoma Treatment

- Moh's micro surgery for most scc and bcc pinna lesions
- TB lesions require TB resection and RT
- Address LN in SCC

### Osteomata and Exostoses

- Benign bony growths in EAC
- Osteoma's solitary, pedunculated, smooth, round lesions arising from tympanomastoid and squamous suture
- Exostose's broad, more medial, multiple, often bilateral
- Related to cold water exposure

### Lesions of the Middle Ear and Mastoid

#### Paragangliomas

- Most common neoplasm of middle ear but still rare
- Glomus tympanicum
  - Originate on promontory of cochlea (jacobson or Arnold's nerve)
  - Fill ME space and ossicles involved
  - May extend to hypotympanum and expose jugular or petrous carotid
  - Present with HL and pulsatile tinnitus and ME mass
- Glomus jugulare
  - Arise in jugular fossa
  - Become large before symptomatic (multiple CN)

### Lesions of the Middle Ear and Mastoid

- Paragangliomas
  - Brown sign +ve pressure leads to blanching
  - Aquino sign ipsilat CA compression decreases pulsation
  - Vernet syndrome (or JF syndrome) – paresis of CN's 9, 10, 11
  - Villaret Syndrome = JF
    syndrome plus Horners





# Paragangliomas

- Rx is complete surgical excision
- If secretory must address this (alpha or beta blockade)
- Trans canal, trans mastoid-lab, trans cervical, infra temporal, intra cranial
- Pre-op embolization is a neccessity
- If you think it invades the ICA, balloon occlusion studies must be done
- RT or stereotactic radiosurgery can halt disease in up to 90%

### Lesions of the Petrous Apex and Clivus

#### Cholesterol granulomas

- Most common lesion of the petrous apex
- Negative pressure in lumen causes hemorrhage
- Expansile lesion
- Hearing loss, tinnitus, vertigo, facial twitching
- HRCT
- MRI diagnostic
  - T1 and T2 hyperintense



### Lesions of the Petrous Apex and Clivus



# **Cholesterol Granuloma**

- Causes: poor drainage of ME, hemmorhage, obstruction of ventilation, FB reaction to cholesterol crystals from HB catabolism
- Rx is surgical drainage

### Lesions of the IAC, CPA, and Skull Base

- Schwannomas (no longer acoustic)
  - Arise from sheaths of cranial nerves
  - Vestibular, facial, trigeminal, jugular
  - Varied presentation
  - HRCT
    - Inhomogeneous enhancement
    - Smooth mass effect
  - MRI definitive diagnosis
    - T1- low intensity
    - Marked enhancement with gadolinium on T1



# Neoplasms of the Nose and Paranasal Sinus

- Introduction
- Benign Lesions
- Malignant lesions

# Neoplasms of Nose and Paranasal Sinuses

- Very rare 3%
- Delay in diagnosis due to similarity to benign conditions
- Nasal cavity
  - $-\frac{1}{2}$  benign
  - ½ malignant
- Paranasal Sinuses
  - Malignant

# Neoplasms of Nose and Paranasal Sinuses

- Multimodality treatment
- Orbital Preservation
- Minimally invasive surgical techniques

# Epidemiology

- Predominately of older males
- Exposure:
  - Wood, nickel-refining processes
  - Industrial fumes, leather tanning
- Cigarette and Alcohol consumption
  - No significant association has been shown

# Location

- Maxillary sinus
  - 70%

### • Ethmoid sinus

- 20%

### • Sphenoid

- 3%

### • Frontal

### Presentation

- Oral symptoms: 25-35%
  - Pain, trismus, alveolar ridge fullness, erosion
- Nasal findings: 50%
  - Obstruction, epistaxis, rhinorrhea
- Ocular findings: 25%
  - Epiphora, diplopia, proptosis
- Facial signs
  - Paresthesias, asymmetry



- Papillomas
- Osteomas
- Fibrous Dysplasia

# Papilloma

- Vestibular papillomas
- Schneiderian papillomas derived from schneiderian mucosa (squamous)
  - Fungiform: 50%, nasal septum
  - Cylindrical: 3%, lateral wall/sinuses
  - Inverted: 47%, lateral wall

# **Inverted Papilloma**

- 4% of sinonasal tumors
- Site of Origin: lateral nasal wall
- Unilateral
- Malignant degeneration in 2-13% (avg 10%)

# Inverted Papilloma Resection

### • Initially via transnasal resection:

- 50-80% recurrence

### • Medial Maxillectomy via lateral rhinotomy:

- Gold Standard
- 10-20%

### • Endoscopic medial maxillectomy:

- Key concepts:
  - Identify the origin of the papilloma
  - Bony removal of this region

### • Recurrent lesions:

- Via medial maxillectomy vs. Endoscopic resection
- 22%

### Osteomas

- Benign slow growing tumors of mature bone
- Location:
  - Frontal, ethmoids, maxillary sinuses
- When obstructing mucosal flow can lead to mucocele formation
- Treatment is local excision

# Fibrous dysplasia

- Dysplastic transformation of normal bone with collagen, fibroblasts, and osteoid material
- Monostotic vs Polyostotic
- Surgical excision for obstructing lesions
- Malignant transformation to rhabdomyosarcoma has been seen with radiation

# Malignant lesions

- <u>Squamous cell carcinoma</u>
- Adenoid cystic carcinoma
- Mucoepidermoid carcinoma
- Adenocarcinoma
- Hemangiopericytoma
- Melanoma
- Olfactory neuroblastoma
- Osteogenic sarcoma, fibrosarcoma, chondrosarcoma, rhabdomyosarcoma
- Lymphoma
- Metastatic tumors
- Sinonasal undifferentiated carcinoma

# Squamous cell carcinoma

- Most common tumor (80%)
- Location:
  - Maxillary sinus (70%)
  - Nasal cavity (20%)
- 90% have local invasion by presentation
- Lymphatic drainage:
  - First echelon: retropharyngeal nodes
  - Second echelon: subdigastric nodes

# Staging of Maxillary Sinus Tumors



# Staging of Maxillary Sinus Tumors

- T1: limited to antral mucosa without bony erosion
- T2: erosion or destruction of the infrastructure, including the hard palate and/or middle meatus
- T3: Tumor invades: skin of cheek, posterior wall of sinus, inferior or medial wall of orbit, anterior ethmoid sinus
- T4: tumor invades orbital contents and/or: cribriform plate, post ethmoids or sphenoid, nasopharynx, soft palate, pterygopalatine or infratemporal fossa or base of skull

### Treatment

- 88% present in advanced stages (T3/T4)
- Surgical resection with postoperative radiation

- Complex 3-D anatomy makes margins difficult

Olfactory Neuroblastoma Esthesioneuroblastoma

- Originate from stem cells of neural crest origin that differentiate into olfactory sensory cells.
- Kadish Classification
  - A: confined to nasal cavity
  - B: involving the paranasal cavity
  - C: extending beyond these limits
Olfactory Neuroblastoma Esthesioneuroblastoma

- Aggressive behavior
- Local failure: 50-75%
- Metastatic disease develops in 20-30%
- Treatment:
  - En bloc surgical resection with postoperative XRT

# **Oral Cavity Cancer**

- Introduction
- Premalignant Lesions
- Malignant Lesions

# Epidemiology

95% are squamous cell carcinoma

## Risk factors

- Smoking (depends on dosage and type)
- Alcohol
- Snuff dipping / tobacco chewing
- HPV (subtype 16)
- Reverse cigar smoking (India)
- Betel-nut chewing (Asia)
- ?Poor dentition / mechanical irritation (dentures)

# Epidemiology

- 75% of cases occur on 10% of mucosal surface area
  - Area from ant FOM along gingivobuccal sulcus and lat border tongue to retromolar trigone and ant tonsil pillar
  - Flow and pooling of carcinogen-contaminated saliva here
- Incidence 4% cancers in males, 2% in females (increasing in females)

## **Evaluation and Diagnosis**

- Lesions generally easy to see
- Simple biopsy under local anesthesia
- Important goals:
  - Stage full extent of disease
  - Rule out synchronous primary
  - Evaluate for possible metastatic disease
- CT or MRI for T2 or greater
- Staging endoscopy

# AJCC TNM Staging

### • Primary Tumor (T)

- Tx: unassessable
- T1: tumor 2cm or less in greatest diameter
- T2: tumor 2-4cm
- T3: tumor >4cm
- T4: tumor invades adjacent structures
  - Cortical bone, deep tongue musculature, maxillary sinus, skin

# **Differential Diagnosis**

- Granular cell myoblastoma
- Minor salivary gland neoplasm
  - Adenoid cystic, mucoepidermoid, adeno-ca.
- Sarcomas (rhabdo, lipo, MFH, leiomyo)
- Hodgkin and NH lymphoma
- Malignant melanoma
- Hairy leukoplakia, Kaposi sarcoma

   HIV, immunocompromised

# **Premalignant Lesions**

- Leukoplakia
  - Hyperkeratosis, dysplasia
  - Malignant transformation greater in non-smokers
  - Treatment:
    - Surgical or laser excision
    - Topical bleomycin, retinoids,

## • Erythroplasia

- Greater risk of malignancy

## **Prognostic Factors**

- Poor prognostic tumor factors include
  - Tumor thickness (3mm FOM, 5mm tongue)
  - Stage
  - Perineural invasion
  - Lymphatic invasion
  - Vascular invasion
  - Neck/distant mets
  - DNA ploidy
  - Pathology

#### <u>Treatment and posttreatment follow-up:</u> <u>neoplasms of the oral cavity</u>

#### SURGERY

- Primary
  - Resection with adequate margins; frozen section as needed
  - Tracheostomy as needed
  - Feeding tube optional
  - Surgical orientation of specimen for pathologist
- Neck
  - Modified/radical dissection for unilateral metastatic disease and bilateral dissections for metastases in both necks
  - Suction drainage

#### Perioperative care

- Antibiotics
- Hospitalization for 3–10 days
- Tube feedings
- Suction drainage for necks(s)—remove when output <25–30 mL/24-h period
- Suture removal 5–10 days postoperatively

## **Tumours of Pharynx**

- Nasopharyngeal Carcinoma
- Oropharyngeal Carcinoma
- Hypopharyngeal Carcinoma

# Nasopharyngeal Carcinoma Introduction

- Rare in the US, more common in Asia
- High index of suspicion required for early diagnosis
- Nasopharyngeal malignancies
  - SCCA ("nasopharyngeal carcinoma")
  - Lymphoma
  - Salivary gland tumors
  - Sarcomas

# Classification

## • WHO classes

- Based on light microscopy findings
- All SCCA by EM
- Type I "SCCA"
  - 25 % of NPC (in North Amer population)
  - 1-2 % NPC of endemic populations
  - moderate to well differentiated cells similar to other SCCA (keratin, intercellular bridges)

## Classification

## • Type II - "non-keratinizing" carcinoma

- 12 % of NPC
- variable differentiation of cells (mature to anaplastic)
- minimal if any keratin production
- may resemble transitional cell carcinoma of the bladder
- Lumped with Type III in 1991 WHO revision

# Classification

## • Type III - "undifferentiated" carcinoma

- 60 % of NPC in North Amer population, majority of NPC in young patients, and 95% of endemic cases
- Difficult to differentiate from lymphoma by light microscopy requiring special stains & markers
- Diverse group
  - Lymphoepitheliomas, spindle cell, clear cell and anaplastic variants

# Epidemiology

- Chinese native (esp Guangdong province) > Chinese immigrant > North American caucasian
  - Both genetic and environmental factors
- Genetic
  - HLA histocompatibility loci possible markers
    - HLA-A2, B17 and Bw46



### • Environmental

- Viruses
  - EBV- well documented viral "fingerprints" in tumor cells and also anti-EBV serologies with WHO type II and III NPC
  - HPV possible factor in WHO type I lesions
- Nitrosamines salted fish
- Others polycyclic hydrocarbons, chronic nasal infection, poor hygiene, poor ventilation

## **Clinical Presentation**

### • Often subtle initial symptoms

- unilateral HL (SOM)
- painless, slowly enlarging neck mass (70%)
  - Lymphatic channels cross midline in NP, bilateral disease common

### Larger lesions

- nasal obstruction
- epistaxis
- cranial nerve involvement

# **Staging EUCC**

### • T1 – tumor confined to NP

### • T2 – tumor extends to soft tissue

- T2a: into OP or nasal cavity with no parapharyngeal extension
- T2b: with parapharyngeal extension (beyond the pharyngobasilar fascia)
- T3 Tumor invades bony structures and/or paranasal sinuses
- T4 intracranial extension, involvement of cranial nerves, infratemporal fossa, hypopharynx, orbit or masticator space

## Treatment

### External beam radiation

- Dose: 6500-7000 cGy
- Primary, upper cervical nodes
- Consider 5000 cGy prophylactic tx of clinically negative lower neck

## Treatment

## Adjuvant Chemotherapy

- Stardard of care
- Cisplatnium (hematologic sideeffects therefore not overlapping toxicity)
- 5-FU

# Oropharyngeal Cancer Introduction

- Relatively uncommon
- 6<sup>th</sup> and 7<sup>th</sup> decades mainly
- Increasing in 4<sup>th</sup> and 5<sup>th</sup> decades
- Male predominance
- SCC = 90%
- Tobacco and alcohol
- Complex, multimodal treatment
- Team approach

## Anatomy



- Connects nasopharynx to hypopharynx
- Ant
  - Circumvallate papillae
  - Anterior tonsillar pillars
  - Junction of hard and soft palates

## Anatomy

### Pharyngeal walls

 Mucosa, submucosa, pharyngobasilar fascia, constrictor muscles, buccopharyngeal fascia

### • Tonsils sit in tonsillar fossa

### • Soft Palate

- Palatine aponeurosis
- Tensor veli palatini
- Levator veli palatini
- Uvular muscle
- Palatoglossus
- palatopharyngeus

# **Etiology**

- SCC arise from the accumulation of multiple genetic alterations to genes important to the regulation of cell growth and death
- Cells have selective growth advantage
  - Genetic
  - Environmental
    - Tobacco and alcohol
    - Dose related
    - Synergistic
  - HPV and EBV
  - Dietary factors
  - Immunosuppression

# Histopathology

### Premalignant lesions

- Leukoplakia
- Erythroplakia
- Lichen planus
- SCC and variants >90%
- Spindle cell clinically and biologically similar to SCC
- Verrucous fungating and slow growing, with well differentiated keratinizing epithelium and rare cellular atypia or mitosis
- Both invade deeply with rare mets

# Histopathology

## Lymphoepitheliomas

- Grow rapidly and readily mets
- Tonsillar region
- Younger patients without risk factors
- Adenoid squamous, adenosquamous, and basaloid SCC are rare and highly aggressive (latter two have early mets)

## Treatment

### Team approach

- Surgeons and Radiation Oncologists
- SLP
- Oral Surgeon
- T1 and T2 surgery or radiation
- T3 and T4 combined modality
- Neck
  - N0 and N1 surgery or XRT
  - N2 and N3 combined modality
  - Both necks treated with central lesions
  - Retropharyngeal nodes are always treated

# Hypopharyngeal Cancer

- Incidence 5-10% of all upper aerodigestive cancers (0.5% of all malignancies)
- M>F: males have 8X increased risk
- Females with Plummer-Vinson\*\*
  - Large increase in risk of developing SCC of the postcricoid region

# Hypopharyngeal Cancers

### Risk Factors

- Smoking
- EtOH
- Chronic reflux disease

#### • Treatment Challenge:

- Patients often present with advanced disease
- May be complicated by severe malnutrition

# Hypopharynx - Anatomy

- Abuts the oropharynx at the level of the hyoid, extends to the level of the inferior border of the cricoid
- 3 sub-sites: piriform fossa(e), post-cricoid region, posterior pharyngeal wall



## Hypopharynx - Anatomy

 Piriform apex – junction between the post-cricoid area and the inferior aspect of the piriform fossae



## **Staging Endoscopy**

- Most important component of procedure (secondary to obtaining Bx samples for diagnosis) is determining the inferior limit of the tumour
- Common site: piriform fossae, post pharyngeal wall, post-cricoid region



# **Pathology**

~95% of cancers of the hypopharynx are SCC

#### Lymphomas

- Angiocentric T-cell lymphoma
- MALT (mucosa associated lymphoid tissue)
- Non-hodgkins lymphoma

#### Adenocarcinomas

– May originate in the minor salivary glands of the hypopharynx

#### • Benign lesions

– Limpoma: < 1%, usually resected due to risk of airway obstruction

# Surgical Tx Options

### Hypopharynx

- Based on Site of Involvement
  - Piriform Fossa (64%)
  - Posterior Pharyngeal Wall (30%)
  - Post-cricoid (4%)
- Treating the Neck
  - Hypopharynx
    - Neck mets in 75%
    - In N0 neck risk of occult nodes ~30-40% (all patients get neck dissections)\*\*
    - Risk of disant mets at presentation ~20%

# Hypopharynx Tx Surgical Options

Procedure	T stage	Reconstruction
Partial Pharyngectomy	T1, T2	Primary closure
Partial Laryngopharyngectomy	T1, T2, T3	Regional or free flap
Supracricoid hemilaryngectomy	T1, T2, T3	Primary closure
Endoscopic CO2 laser resection	T1, T2 (possibly T3, T4)	Secondary intention
Total Laryngectomy with partial-total pharyngectomy	Т3	Primary closure vs. regional or free flap
Total Laryngo-Pharyngo- esophagectomy	Τ4	Gastric pull-up
## Laryngeal Tumours

- Introduction
- Benign Lesions
- Malignant Lesions



- 11 600 new cases laryngeal cancer per year in USA
- ~1% of all cancers (excluding skin)
- **79% occur in** *∂*
- >90% are squamous cell carcinomas (SCC)

## **Etiology**

- EtOH supraglottic
- Tobacco glottic
- GERD chronic laryngeal irritation
- Viral infection
- Asbestos
- Nickel
- Wood
- Isopropyl alcohol
- Radiation

#### Laryngeal Papillomatosis

- Most common benign laryngeal tumor, HPV etiology
- Vocal folds and subglottis most common laryngeal sites

## Laryngeal Papillomatosis

- More prevalent in children, less common in individuals over 30 years of age
  - HPV is transmitted to child through birth canal from cervix
  - Risk of transmission 1:400
- Papillomas appear multinodular, and may be either sessile or exophytic

– May resemble carcinoma-in-site or even invasive SCC

Exophytic, warty, friable, tanwhite to red growths



## Laryngeal Papillomatosis

- Most common viral subtypes are 6 or 11, but 16 or 18 have higher potential for malignant change
- Hoarseness is common early symptom followed by airway obstruction and respiratory difficulty

## Laryngeal Papillomatosis

- Laryngeal papillomas presenting in adults seem to be less aggressive than juvenile form but remission rate unpredictable
  - In adults, growth may be rapid during periods of hormone change such as during pregnancy
- Malignant degeneration of laryngeal papillomas rare and usually associated with history of radiotherapy, tobacco abuse or both

#### Treatment

#### - Surgery

- Laser microlaryngoscopy (most commonly CO<sub>2</sub> [10.6um] or Nd:YAG [1.06um]) at power setting of 2-8W pulse or continuous
- Powered microdebrider
- Always biopsy before remainder of case proceeds

#### Cont Treatment

#### Interferon

- Bad chronic side-effects (myalgias, flu-like symptoms)
- Lesions tend to return after interferon finished
- Intralesional cidofovir (acyclic nucleoside analogue)
- Indole-3-carbinol (found in cruciferous vegetables, works via inhibition of estrogen metabolism)
- Acyclovir
- Photodynamic therapy

#### Supraglottic vs Glottic Disease

- North America glottic cancer > supraglottic
  (2:1)
- France supraglottic > glottic (2:1)

#### Anatomy - Glottis

- True vocal cords
- Anterior and posterior commissures
- Superior limit apex of ventricle
- Inferior limit 1 cm inferior to line through apex

## Staging – Early Glottic

- T<sub>is</sub> no invasion beyond basement membrane
- T<sub>1</sub> confined to glottis with normal mobility
  - $T_{1a}$  tumor limited to one vocal cord
  - T<sub>1b</sub> tumor involves both cords, no limitation in mobility

## Staging – Early Glottic

- T<sub>2</sub> extend into supra- or subglottis without complete vocal cord fixation
  - T<sub>2a</sub> involve supra- or subglottis but do not impair movement
  - T<sub>2b</sub> impair movement of vocal cords, but not complete fixation

## Staging – Advanced Glottic

- T<sub>3</sub> complete vocal cord fixation, ± paraglottic space, ± minor thyroid cartilage erosion (inner cortex)
- T<sub>4</sub> extends beyond larynx, into thyroid cartilage

#### Symptoms

- Hoarseness >4 weeks investigate
- Occasionally may present without hoarseness
- Dysphagia
- Hemoptysis

- Early: irregular area of mucosal thickening
- Advanced: exophytic, fungating, endophytic, ulcerated mass
- More commonly keratinizing, well to moderately differentiated
- In situ component
- Invasive component predominantly infiltrative

Up to 20% of T<sub>1</sub> cancers have some degree of vocal cord ligament invasion



- Most tumors originate on free surface of vocal cord
- Anterior 2/3



#### Treatment

- Early Stage: Laser or Radiation
- Advance Stage: Chemo+Radiation
   Surgery+Radiation



# **Thank You**