Neoplasms of the Nose and Paranasal Sinuses
Sinonasal Neoplasms

Incidence

- Rare
- 0.5% of all malignancies
- < 3% of all head and neck malignancies
- 23% H&N cancer in Japan
- Annual incidence 0.5-1/100,000 population
- Males:females ~ 2:1
- Most common in 5th and 6th decade
Sinonasal Neoplasms

- Nasal cavity (benign = malignant)
  - Benign - inverting papilloma
  - Malignant - SCCA

- Sinuses (malignant)
  - SCCA
  - Maxillary most common
Incidence by Sinus

- Maxillary sinus: 70-80%
- Ethmoid sinus: 10-20%
- Frontal sinus: <5%
- Sphenoid sinus: <5%
- Nasal cavity: 20-30%
Etiology

- Unknown
- Evidence of occupational risk
- Inhalation of metal dusts or aerosols
- Exposure:
  - Wood dust – hard wood dust exposure ie. mahogany particles <5μm
  - diameter
  - Industrial fumes, nickel-refining processes, leather tanning, chromium,
  - asbestos, formaldehyde
  - Found in Furniture, leather and textile industry
  - Reported increased rate from 20-100x
Etiology – Associations

- Nickel Workers:
  - 250x increased incidence of cancer of the sinus - (Scand J Work Environ Health 9:315-326, 1983)
  - Random biopsy from the middle turbinate showed 21% incidence of dysplasia.
- Wood/leather dust associated with adenocarcinoma.
- Tobacco smoke exposure associated with increased risk of SCC.
Histology

SCC: 60-70%  Adenocarcinoma: 10-20%  

**Epithelial**

Epidermoid/Squamous
- Carcinoma (spindle cell, verrucous transitional)

Non-Epidermoid
- Adenoid cystic carcinoma
- Adenocarcinoma
- Mucoepidermoid carcinoma
- Acinic cell carcinoma
- Metastases

Neuroectodermal
- Malignant melanoma
- Olfactory neuroblastoma
- Neurofibroma
- Neuroendocrine carcinoma
- Melanotic neuroectodermal tumor of infancy
**Mesenchymal**

**Vascular**
- Angiosarcoma
- Kaposi’s sarcoma
- Haemangiopericytoma

**Muscular**
- Leiomyosarcoma
- Rhabdomyosarcoma

**Cartilaginous**
- Chondrosarcoma (mesenchymal)

**Osseous**
- Osteogenic sarcoma

**Lymphoreticular**
- Burkitt’s lymphoma
  - Non-Hodgkin’s lymphoma
  - Extramedullary plasmacytoma
  - Midline destructive lesions (T cell lymphoma)
- Fibrosarcoma
- Liposarcoma
- Malignant fibrous histiocytoma
Presentation

- Similar to common problems
- 6 to 8 month delay in diagnosis
- Cranial neuropathies & proptosis
Presentation

- Oral - 30%
  - tooth pain, trismus, palatal fullness, erosion
- Nasal - 50%
  - obstruction, epistaxis, discharge, erosion
- Ocular - 25%
  - diplopia, proptosis, tearing, pain, fullness
- Facial
  - V2 numbness, asymmetry, pain
- Auditory - CHL
Advanced Disease

- Classic Triad
  - facial asymmetry
  - tumor bulge in oral cavity
  - nasal mass
- All three - 40-60%
- One - 90%
Diagnosis

- Physical exam
- Nasal endoscopy
- Biopsy
- Radiography
Computed Tomography

- Bone erosion
  - orbit, cribiform plate,
  - fovea, post max sinus wall,
  - PTPF, sphenoid, post wall
  - of frontal sinus
- 85% accuracy
- ? Tumor vs. inflammation vs. secretions
MRI

- Superior to CT
  - multiplanar
  - no ionizing radiation
- Inflammatory tissue & secretions - intense T2
- Tumor - intermediate T1 & T2
- 94% accuracy
- 98% accuracy with gadolinium
Schneiderian Papillomas

- Fungiform (50%) - septum
- Cylindrical (3%) - lateral nasal wall
- Inverting (47%) - lateral nasal wall
  - recurs, locally destructive, malignant potential
  - men, 6th-7th decades, unilateral
  - SCCA - 2-13%
  - Recurrence - 0-80%
### Inverting Papilloma

<table>
<thead>
<tr>
<th>Authors</th>
<th>Lateral rhinotomy–medial maxillectomy</th>
<th>Conservation resection</th>
</tr>
</thead>
<tbody>
<tr>
<td>Benninger et al. (1991)</td>
<td>0% (0/20)</td>
<td>36% (5/14)</td>
</tr>
<tr>
<td>Myers et al. (1990)</td>
<td>5% (1/22)</td>
<td>0% (0/4)</td>
</tr>
<tr>
<td>Peiausa and Fortier (1992)</td>
<td>7% (1/14)</td>
<td>77% (37/48)</td>
</tr>
<tr>
<td>Ouzen et al. (1991)</td>
<td>7% (3/44)</td>
<td>27% (3/11)</td>
</tr>
<tr>
<td>Lawson et al. (1989)</td>
<td>9% (7/77)</td>
<td>10% (1/10)</td>
</tr>
<tr>
<td>Segal et al. (1986)</td>
<td>10% (1/10)</td>
<td>70% (10/14)</td>
</tr>
<tr>
<td>Kristensen et al. (1985)</td>
<td>12% (7/57)</td>
<td>38% (8/21)</td>
</tr>
<tr>
<td>Phillips et al. (1990)</td>
<td>13% (9/72)</td>
<td>44% (4/9)</td>
</tr>
<tr>
<td>Smith and Guliane (1987)</td>
<td>27% (3/11)</td>
<td>57% (4/7)</td>
</tr>
<tr>
<td>Dolgin et al. (1992)</td>
<td>29% (4/14)</td>
<td>44% (4/9)</td>
</tr>
<tr>
<td>Weissler et al. (1986)</td>
<td>29% (37/126)</td>
<td>67% (103/153)</td>
</tr>
<tr>
<td>Bielamowicz et al. (1993)</td>
<td>30% (60/20)</td>
<td>74% (17/23)</td>
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<tr>
<td>Averages</td>
<td>16% (79/487)</td>
<td>60% (209/350)</td>
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</tbody>
</table>
Osteomas

- Benign, slow-growing
- 15 to 40 years
- frontal > ethmoid > maxillary
- local excision
Fibrous Dysplasia

- Normal bone replaced by collagen, fibroblasts, and osteoid material
- < 20 years
- Ground-glass appearance
- Treatment?
- No irradiation
Neurogenic tumors

- Schwannomas
  - surface of nerve fibers
  - no malignant degeneration
  - along trigeminal & ANS

- Neurofibromas
  - within nerve fibers
  - von Recklinghausen’s disease
  - malignant degeneration in 15%

- Complete excision
SCCA

- Most common - 80%
- Max > nasal cavity > ethmoids
- Males
- Sixth decade
- 90% have eroded walls of sinuses
Adenoid Cystic Carcinoma

- Palate > major salivary glands > sinuses
- Resistant to tx
- Multiple recurrences, distant mets
- Perineural spread
- Long-term followup necessary
Mucoepidermoid Carcinoma
- rare, widespread local invasion

Adenocarcinoma
- 2nd most common, 5-20%
- ethmoids
- occupational exposures
Hemangiopericytoma

- Uncommon
- pericytes of Zimmerman
- 80% of sinonasal tumors in ethmoids
- resembles nasal polyps
- average in 55 yo
- excision, XRT for (+) margins
Melanoma

- 1% originate in sinonasal cavity
- 5th-8th decades
- anterior septum
- maxillary antrum
- polypoid mass,
- pigmentation?
- 5 yr = 38%
- 10 yr = 17%
Olfactory Neuroblastoma

- Neural crest origin
- No urinary VMA or HVA
- Bimodal distribution at 20 and 50
- Locally aggressive
- Rosettes are hallmark
- Kadish staging
- Local recurrence 50-75%
- Metastasis 20-30%
Osteogenic Sarcoma
- most common primary bone tumor
- only 5% in H & N, mandible most involved
- sunray appearance

Fibrosarcoma
- rarely seen in sinuses
Chondrosarcoma
- 3rd-5th decades
- Histologic dx difficult
- Slow erosion of skull base, (+) margins

Rhabdomyosarcoma
- Most common in children
- 35-45% in H&N, 8% in sinuses
- Embryonal, alveolar, pleomorphic
- Triple tx
Lymphoma
- Bimodal presentation
- NHL
- Irradiation +/- chemo

Extramedullary plasmacytoma
- 40% in paranasal sinuses/nose
- "Benign"
- Must rule out myeloma
- Excision or irradiation
Metastatic tumors

- Renal cell carcinoma
- Lungs
- Breasts
- Urogenital tract
- Gastrointestinal tract
- Palliation necessary
Ohngren’s Line

- Suprastructure
- Infrastructure
### Staging

- **AJCC - Maxillary sinus carcinoma**

<table>
<thead>
<tr>
<th>Stage</th>
<th>Description</th>
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<tbody>
<tr>
<td>TX</td>
<td>Primary tumor cannot be assessed</td>
</tr>
<tr>
<td>T0</td>
<td>No evidence of primary tumor</td>
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<tr>
<td>Tis</td>
<td>Carcinoma in situ</td>
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<tr>
<td>T1</td>
<td>Tumor limited to the antral mucosa with no erosion or destruction of bone</td>
</tr>
<tr>
<td>T2</td>
<td>Tumor with erosion or destruction of the infrastructure, including the hard palate and/or the middle nasal meatus</td>
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<tr>
<td>T3</td>
<td>Tumor invades any of the following: skin of cheek, posterior wall of maxillary sinus, floor or medial wall of orbit, anterior ethmoid sinus</td>
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<tr>
<td>T4</td>
<td>Tumor invades orbital contents and/or any of the following: cribriform plate, posterior ethmoid or sphenoid sinuses, nasopharynx, soft palate, pterygomaxillary or temporal fossae, or base of skull</td>
</tr>
</tbody>
</table>
Treatment

- T3 and T4
- 60% local recurrence
  - Surgery
  - Irradiation
  - Chemotherapy
Surgical resection

- Unresectability (Sisson)
  - extension to frontal lobes
  - invasion of prevertebral fascia
  - bilateral optic nerve involvement
  - cavernous sinus extension
Surgical resection

- Endoscopic excision
- WLE
- Medial maxillectomy
- Total maxillectomy
- Radical maxillectomy +/- exenteration
- Craniofacial resection
Orbital Preservation

- *Harrison* - proptosis, limitation of EOM, bony erosion of orbit = exenteration
- *Conley* - save eye whenever possible
- *Sisson* - preoperative XRT, decreased exenterations without change in survival
- *Stern* - nonfunctional eye without inf/med support = exenteration
Orbital preservation

- UVA - McCary & Levine
  - 50 Gy preop XRT to orbit
  - periorbital bx
  - resect (+) periorbita
  - functional eye
Pterygopalatine Fossa

- 10-20% involvement
- Som - PTPF invasion = unresectable lesion
- Craniofacial resection (MCF)
- Postop XRT
Neck Dissection

- Retropharyngeal and jugulodigastric nodes
- 10% (+) necks
- Neck dissection
  - Palpable nodes
  - Radiographic evidence of disease
- 40% cervical mets at 4 yrs
Radiation therapy

- Primary tx only for palliation
- 10-15% improved 5 year survival
- XRT = 23% vs. Surgery + XRT = 44%
- preoperative vs. postoperative
- protection of CNS and globe
  - XRT 12-20% unilateral visual loss, 0-8% bilateral visual loss
  - Surgery 10-20% useless globes, 2X with XRT
Chemotherapy

- Palliation, unresectable disease
- (+) margins, perineural spread, surgical refusal, ECS
- Intraarterial chemotherapy
  - Robbins - 86% response of T4 lesions
  - Lee - 91% satisfactory response