Neoplasms of the Nose and Paranasal Sinus

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Neoplasms of Nose and Paranasal Sinuses

- Very rare 3%
- Delay in diagnosis due to similarity to benign conditions
- Nasal cavity
  - ½ 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
  - 1%
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
Radiography

- **CT**
  - Bony erosion
  - Limitations with periorbita involvement

- **MRI**
  - 94-98% correlation with surgical findings
  - Inflammation/retained secretions: low T1, high T2
  - Hypercellular malignancy: low/intermediate on both
    - Enhancement with Gadolinium
Benign Lesions

- Papillomas
- Osteomas
- Fibrous Dysplasia
- Neurogenic tumors
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 mukocele 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
Neurogenic tumors

- 4% are found within the paranasal sinuses
- Schwannomas
- Neurofibromas
- Treatment via surgical resection
- Neurogenic Sarcomas are very aggressive and require surgical excision with post op chemo/XRT for residual disease.
- When associated with Von Recklinghausen’s syndrome: more aggressive (30% 5yr survival).
Malignant lesions

- Squamous cell carcinoma
- 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
Treatment

- 88% present in advanced stages (T3/T4)
- Surgical resection with postoperative radiation
  - Complex 3-D anatomy makes margins difficult
Adenoid Cystic Carcinoma

- 3rd most common site is the nose/paranasal sinuses
- Perineural spread
  - Anterograde and retrograde
- Despite aggressive surgical resection and radiotherapy, most grow insidiously.
- Neck metastasis is rare and usually a sign of local failure
- Postoperative XRT is very important
Mucoepidermoid Carcinoma

- Extremely rare
- Widespread local invasion makes resection difficult, therefore radiation is often indicated
Adenocarcinoma

- 2nd most common malignant tumor in the maxillary and ethmoid sinuses
- Present most often in the superior portions
  - Strong association with occupational exposures
- High grade: solid growth pattern with poorly defined margins. 30% present with metastasis
- Low grade: uniform and glandular with less incidence of perineural invasion/metastasis.
Hemangiopericytoma

- Pericytes of Zimmerman
- Present as rubbery, pale/gray, well circumscribed lesions resembling nasal polyps
- Treatment is surgical resection with postoperative XRT for positive margins
Melanoma

- 0.5- 1.5% of melanoma originates from the nasal cavity and paranasal sinus.
- Anterior Septum: most common site
- Treatment is wide local excision with/without postoperative radiation therapy
- END not recommended
- AFIP: Poor prognosis
  - 5yr: 11%
  - 20yr: 0.5%
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

- **UCLA Staging system**
  - **T1:** Tumor involving nasal cavity and/or paranasal sinus, excluding the sphenoid and superior most ethmoids
  - **T2:** Tumor involving the nasal cavity and/or paranasal sinus including sphenoid/cribriform plate
  - **T3:** Tumor extending into the orbit or anterior cranial fossa
  - **T4:** Tumor involving the brain
Olfactory Neuroblastoma
Esthesioneuroblastoma

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

- Osteogenic Sarcoma
  - Most common primary malignancy of bone.
  - Mandible > Maxilla
  - Sunray radiographic appearance

- Fibrosarcoma

- Chondrosarcoma
Rhabdomyosarcoma

- Most common paranasal sinus malignancy in children
- Non-orbital, parameningeal
- Triple therapy is often necessary
- Aggressive chemo/XRT has improved survival from 51% to 81% in patients with cranial nerve deficits/skull/intracranial involvement.
- Adults, Surgical resection with postoperative XRT for positive margins.
Lymphoma

- Non-Hodgkins type
- Treatment is by radiation, with or without chemotherapy
- Survival drops to 10% for recurrent lesions
Sinonasal Undifferentiated Carcinoma

- Aggressive locally destructive lesion
- Dependent on pathological differentiation from melanoma, lymphoma, and olfactory neuroblastoma
- Preoperative chemotherapy and radiation may offer improved survival
Metastatic Tumors

- Renal cell carcinoma is the most common
- Palliative treatment only
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
Surgery

- Unresectable tumors:
  - Superior extension: frontal lobes
  - Lateral extension: cavernous sinus
  - Posterior extension: prevertebral fascia
  - Bilateral optic nerve involvement
Surgery

- **Surgical approaches:**
  - Endoscopic
  - Lateral rhinotomy
  - Transoral/transpalatal
  - Midfacial degloving
  - Weber-Fergusson
  - Combined craniofacial approach

- **Extent of resection**
  - Medial maxillectomy
  - Inferior maxillectomy
  - Total maxillectomy
Tracheostomy

- 130 maxillectomies only 7.7% required tracheostomy
- Of those not receiving tracheostomy during surgery, only 0.9% experienced postoperative airway complications
- Tracheostomy is unnecessary except in certain circumstances (bulky packing/flaps, mandibulectomy)
Treatment of the Orbit

Before 1970’s orbital exenteration was included in the radical resection

Preoperative radiation reduced tumor load and allowed for orbital preservation with clear surgical margins

Currently, the debate is centered on what “degree” of orbital invasion is allowed.
Current indications for orbital exenteration

- Involvement of the orbital apex
- Involvement of the extraocular muscles
- Involvement of the bulbar conjunctiva or sclera
- Lid involvement beyond a reasonable hope for reconstruction
- Non-resectable full thickness invasion through the periorbita into the retrobulbar fat
Conclusions

- Neoplasms of the nose and paranasal sinus are very rare and require a high index of suspicion for diagnosis
- Most lesions present in advanced states and require multimodality therapy
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