Neoplasms of the Nose and Paranasal Sinus

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May 19, 2004

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 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

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

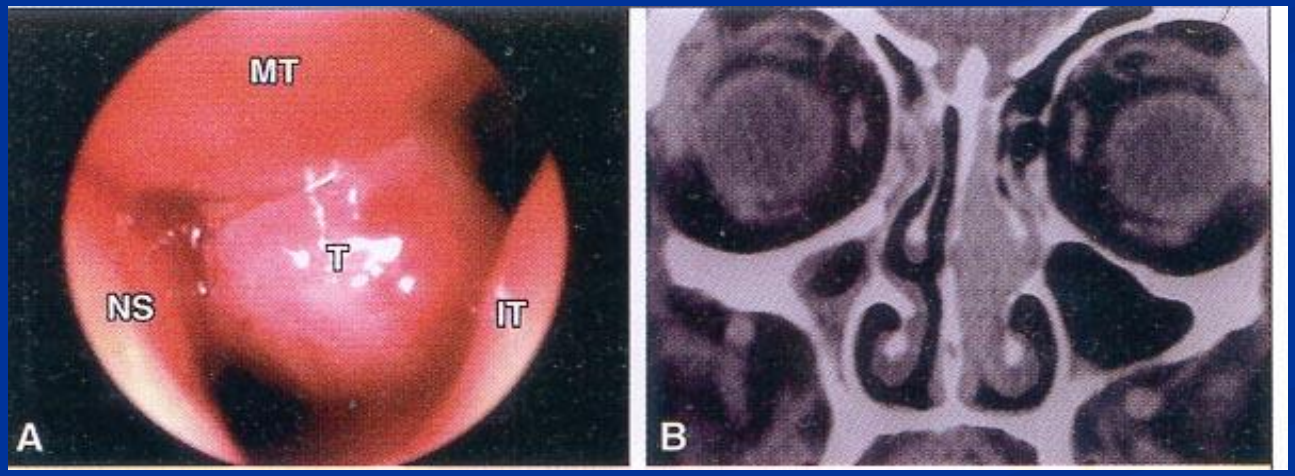
- Extremely rare
- Widespread local invasion makes resection difficult, therefore radiation is often indicated

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

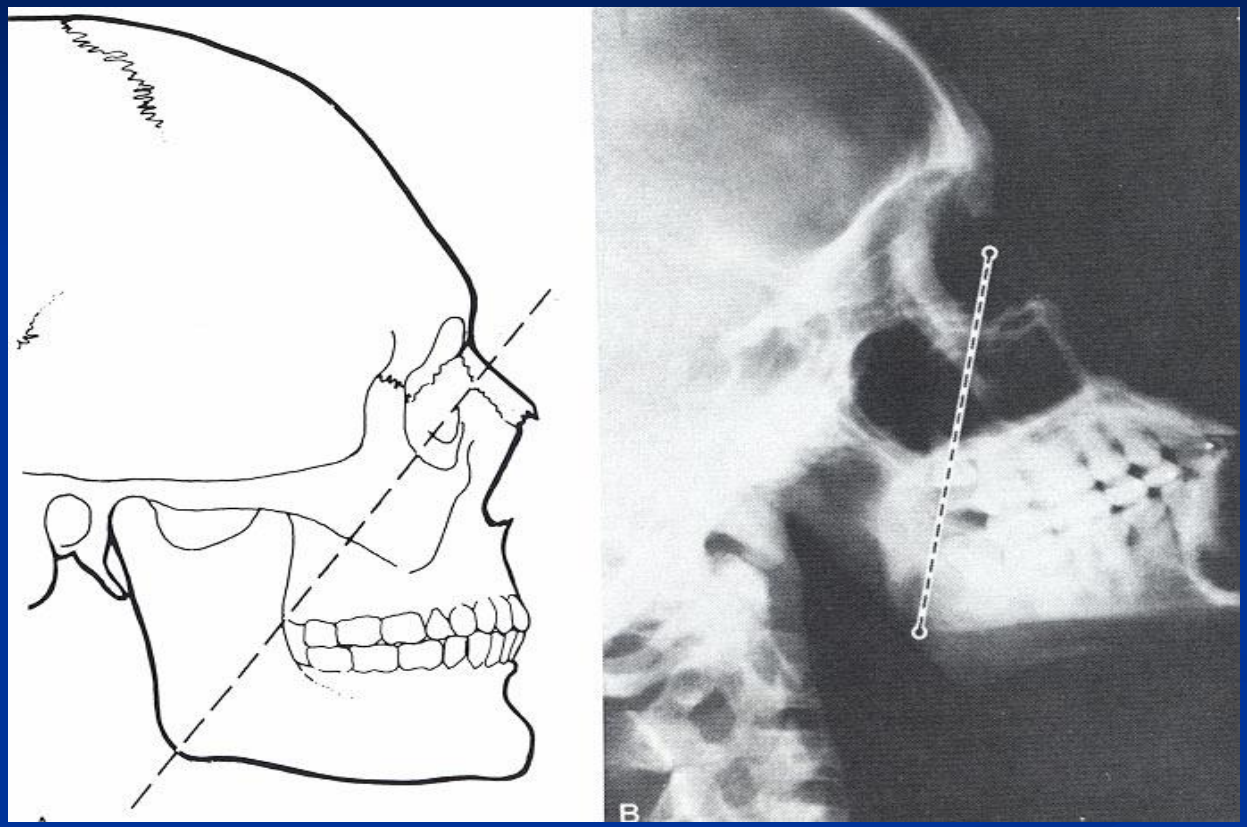
Sinonasal Undifferentiated Carcinoma

- 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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