Sinonasal Tumors

Objectives
- Incidence and demographics of sinonasal tumors
- Separating tumors from inflammatory changes
- Common and notable histologic types of sinonasal tumors
- Staging of sinonasal tumors

Incidence of Paranasal Sinus Tumors
- Rare tumors:
  - Yearly US incidence is approximately 3 / 1,000,000
  - 3% of all head and neck malignancies
  - 75% arise in the paranasal sinuses, 25% in the nasal cavity

Paranasal Sinus Tumors
- 40% of patients present with advanced disease
- Average 6 month delay between onset of pain and diagnosis
- Improved prognosis is associated with surgical resection
- Advanced disease can render surgical excision excessively morbid, or cosmetically unacceptable for the patient.
- Radiologists can provide early recognition and accurate anatomic delineation to guide appropriate therapy.

Demographics of Paranasal Sinus Tumors
- Demographics – think grumpy old men…
  - 2/3rds men
  - 50% between ages 60 and 79
  - Only 4% from patients less than 40 years old.

Greg Avey, MD
Demographics of Paranasal Sinus Tumors

- Demographics – think Statler and Waldorf...
  - 2/3rds men
  - 50% between ages 60 and 79
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Sites of Paranasal Sinus Tumors

- Disease Sites
  - Maxillary Sinuses – 57%
  - Nasal Cavity - 25%
  - Ethmoids – 8%
  - Sphenoid Sinus – 3%
  - Frontal Sinus – 1.5%

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Separating Tumors from Inflammatory Changes

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  On Sinus CTs
  - Very challenging for early T stage tumors.
  - Increased CT density can help – tumors will often be more dense than inflammatory changes.
  - Narrow CT windows and high index of suspicion are key!

  Separating Tumors from Inflammatory Changes

  On Sinus CTs
  - Contrast enhancement can help, but is rarely performed for screening exams.
  - For more advanced tumors, osseous destruction, expansion of the sinus, or infiltration of the adjacent fat leads to the diagnosis.
Due to the low water content of most nasal / paranasal neoplasms, the majority will be isointense to hypointense on T2 images.

Exceptions include minor salivary gland tumors, schwannomas, and polypoid tumors such as inverted papillomas.

Contrast can also help distinguish tumor from adjacent secretions.

Incidental MRI findings:
In general, benign collections will either be high in T2 signal intensity (increased intracellular and intercellular water), or high in T1 signal intensity (due to increased protein in inspissated collections).
A mass which is low in signal intensity on both T2 and T1 requires further investigation.

Squamous Cell Carcinoma
- Makes up 52% of the tumors of the paranasal sinuses and nasal cavity
- Associated with industrial exposures such as wood dust, nickel, and chromium
- 15% present with nodal metastasis – especially if there is spread to the premaxillary soft tissues, orbit or pterygooids
- Local recurrence is reported in 20% to 50% of cases, with most recurrences occurring (80%) within one year of therapy

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PN Squamous Cell Carcinoma

- 5 year survival is dismal (30%), and hasn’t improved much over the last 40 years.


Inverted Papilloma

- A benign process, but carries a 15% risk of developing squamous carcinoma ex inverted papilloma
- Can be locally recurrent, and has traditionally been managed through an en-bloc resection
- Pathologically consists of squamous epithelium replacing the mucosal glands and ducts
- Typically originates on the lateral nasal wall
Inverted Papilloma

- Often erosive – leading to bone fragments within the mass (40%)
- A focus of hyperostosis is often present at the site of origin – helpful both in identifying the potential for an inverted papilloma, and for guiding surgery.
- Classically has a cerebriform appearance on T2 or postcontrast T1 MRI – however this is not universal!

Middle Turbinate

Inverted Papilloma

Adenocarcinoma

- 13% of all sinonasal cancers (SCC + adeno = 65%)
- Wood dust exposure increases risk ~500x over male population, and 900x over the general population

WARNING
THIS AREA CONTAINS A CHEMICAL (WOOD DUST) KNOWN TO CAUSE CANCER
**Adenocarcinoma**
- Can be more T2 hyperintense than SCC
- Similar demographics as SCC – mean age 64 years, 75% males
- 5 year survival ~50%
- Increased predilection for nasal cavity and ethmoids
- Should be on the differential of most nasal / paranasal masses, especially with a history of woodworking and a nasal cavity or ethmoid sinus location

**Salivary Tumors**
- 100s of small salivary glands / rests are present within the mucosa of the nasal cavity and paranasal sinuses
- Follows the general rules of the salivary neoplasms: the smaller the gland the greater the chance of a malignant neoplasm
- Adenoid cystic is the most common pathology, with most presenting with T3 or T4 stage (77%), and few having local or distant metastasis (N0 98%, M0 97%)
- Five year survival rates are good (71%), but local recurrence and late recurrences are common

**Adenoid Cystic Carcinoma**

**Olfactory Neuroblastoma**
- The tumor formerly known as esthesioneuroblastoma
- Bimodal peaks in the 2nd and 6th decades
- Arises from olfactory mucosa along the superior nasal cavity, often with intracranial extension
- Peritumoral cysts at the margin between the tumor and the brain parenchyma are suggestive of an olfactory neuroblastoma

**Olfactory Neuroblastoma**
- ENB / ONB has its own staging system
  - Kadish Staging System
    - A: Nasal Cavity Involvement only
    - B: Nasal Cavity and Paranasal Sinus
    - C: Intracranial Involvement
    - D: Nodal Metastasis
- 30% present with metastasis, 30% have a late recurrence (can be 15 years or longer)
Olfactory Neuroblastoma

- ENB / ONB can be difficult to diagnose on pathology
- Exists on a continuum with sinonasal neuroendocrine carcinoma and sinonasal undifferentiated carcinoma
- Can mimic other small round blue cell tumors (SNUC, NHL, Ewings, Melanoma, rhabdomyosarcoma)
- Prognosis is generally good, with 75% five year survival

Box 1: Differential diagnosis for sinonasal pathology conditions with a similar presentation (clinical and histopathology) as ENB

- Nasal undifferentiated carcinoma (NUC)
- Sinonasal undifferentiated carcinoma
- Neuroendocrine carcinoma (NEC)
- Melanoma
- Ewings sarcoma
- Metastatic pulmonary small cell NEC
- Small cell lymphoma
- Alveolar soft part sarcoma
- Rhabdomyosarcoma
- Phaeohyomelanoma
- Melanoma

The least differentiated of the tumors on the olfactory neuroblastoma <-> sinonasal neuroendocrine carcinoma <-> sinonasal undifferentiated carcinoma spectrum

Extremely rare and very aggressive: high propensity for metastasis and aggressive intracranial extension
Sinonasal Undifferentiated Carcinoma

Sinonasal Undifferentiated Carcinoma

Sinonasal Undifferentiated Carcinoma

Sinonasal Undifferentiated Carcinoma

Sinonasal Melanoma

- Rare tumor, most commonly presenting within the nasal cavity. 40% are amelanotic.
- Melanotic melanomas are typically T1 hyperintense to grey matter on MRI.
- Both melanotic and amelanotic melanomas will be T2 hypointense.
- Very poor prognosis, 17% 5 year survival rate. staging starts at T3. Systemic metastasis are common.

Sinonasal Melanoma

s/p resection x 2, radiation and proton beam therapy.

Sinonasal Melanoma
Sinonasal Melanoma

Ewings
- Small round blue cell tumor, extracranial PNET equivalent
- Presents with a permeative mass – often with osseous destruction
- 90% of patients < 20 years old
- Often treated with surgical resection (if possible), radiation if resection is not feasible, and chemotherapy
- Survival is good for local disease (75%), much worse with metastatic disease (15%)

2 yo female with vision loss
Juvenile Nasopharyngeal Angiofibroma

- Benign, hypervascular mass arising from the sphenopalatine foramen
- Almost exclusively young males (8-25 years)
- Presents with nasal obstruction and epistaxis
- Preoperative embolization is often performed to minimize blood loss during surgery
- Commonly presents with enlarged ECA branches (imax, ascending pharyngeal, facial) and ICA branches (ant and post ethmoids, vidian artery)

A 13 yo male with recurrent epistaxis

Juvenile Nasopharyngeal Angiofibroma

A female infant was noted to have left cheek/intraoral swelling shortly after birth.

A maxillofacial CT was obtained for further evaluation.
The greater palatine artery was ligated via a palatal incision.
Following ligation, there was a notable reduction in the volume of the mass.
Open biopsy and debulking was performed with minimal blood loss.

Surgical Findings
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Maxillary Sinus Staging

**Ohngren’s line**

- Patients with disease in the posterior superior half of the maxillary sinus have a worse prognosis.
- Formed the early basis of staging.
- We still maintain an increased stage for those tumors with involvement of the posterior wall of the maxilla, pterygoid plates, or orbital floor.

Maxillary Sinus Staging

**T Stage Description**

<table>
<thead>
<tr>
<th>T Stage</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>T0</td>
<td>No evidence of primary tumor</td>
</tr>
<tr>
<td>T1</td>
<td>Tumor limited to maxillary sinus mucosa with no erosion or destruction of bone</td>
</tr>
<tr>
<td>T2</td>
<td>Tumor causing bone erosion or destruction including extension to the hard palate and/or middle nasal meatus, except extension to the posterior wall of the maxillary sinus and the pterygoid plates</td>
</tr>
<tr>
<td>T3</td>
<td>Tumor involves any of the following: bone of the posterior wall of the maxillary sinus, subcutaneous tissue, the floor or medial wall of the orbit, pterygoid fossa, ethmoid sinuses</td>
</tr>
<tr>
<td>T4a</td>
<td>Tumor involves anterior orbital contents, skin of the cheek, pterygoid plates, infratemporal fossae, orbitofrontal plates, sphenoid or frontal sinuses</td>
</tr>
<tr>
<td>T4b</td>
<td>Tumor involves any of the following: orbital apex, dura, brain, middle cranial fossae, cranial nerves other than maxillary division of the trigeminal nerve (V2), nasopharynx, or clivus</td>
</tr>
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Maxillary Sinus Staging

**Maxillary Sinus take-home points**

- T staging determined by involved sites
- Osseous involvement T1 -> T2
- Osseous involvement of the posterior wall of the maxillary sinus T2 -> T3
- The anterior orbit and orbital apex are treated differently for staging
- Named CN involvement other than V2 results in T4b staging

Ethmoid Sinus / Nasal Cavity Staging

**Based on sites and subsites**

<table>
<thead>
<tr>
<th>Sites</th>
<th>Subsites</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ethmoid Sinus and Nasal Cavity</td>
<td>Left, Right, Septum, Floor, Latera Wall, Vestibule</td>
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### Ethmoid Sinus / Nasal Cavity Staging

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<td>T0</td>
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<td>T1</td>
<td>Tumor restricted to any one subsite, with or without bony invasion</td>
</tr>
<tr>
<td>T2</td>
<td>Tumor invading two subsites in a single region within the nasoethmoidal complex, with or without bony invasion</td>
</tr>
<tr>
<td>T3</td>
<td>Tumor extends to invade the medial wall or floor of the orbit, maxillary sinus, palate, or cribiform plate</td>
</tr>
<tr>
<td>T4a</td>
<td>Tumor invades any of the following: anterior orbital contents, skin of nose or cheek, minimal extension to the anterior cranial fossa, pterygoid plates, sphenoid or frontal sinuses</td>
</tr>
<tr>
<td>T4b</td>
<td>Tumor invades any of the following: orbital apex, dura, brain, middle cranial fossa, cranial nerves other than V2, nasopharynx or clivus</td>
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- **Ethmoid / Nasal Cavity Staging take-home points**
  - Tumor which extends outside the nasal cavity -> T3 (orbit, palate, maxillary sinus, cribiform plate)
  - The anterior orbit and orbital apex are treated differently for staging
  - Named CN involvement other than V2 results in T4b staging

### Frontal Sinus and Sphenoid Staging

- There is no frontal sinus or sphenoid staging system!
- These tumors are rare. However, they do occur and should be kept on the differential.

### Frontal Sinus NUT Carcinoma

- 44 year old woman with headache and blurry vision
The tumor initially responded well to chemotherapy, but then presented with increasing pain, concern for recurrence. PET/MRI was ordered to help to differentiate recurrent tumor from granulation tissue from prior surgery.

Multiple rounds of chemotherapy were attempted, with only minimal effect on the tumor. Orbital exenteration was subsequently performed in an attempt to excise the remaining disease. The patient subsequently presented with aphasia and concern for stroke.
Frontal Sinus NUT Carcinoma

NUT Midline Carcinoma
- Genetically defined, highly lethal cancer resulting from rearrangement of the NUT gene on chromosome 15q14.
- This gene is typically only expressed in the testis.
- Arises from midline locations – head, neck, mediastinum.
- Mean survival is less than one year.

Take Home Points
- Sinonasal neoplasms are rare, but important entities
- Think grumpy old men for demographics
- Inverted papillomas may have a focal area of hyperostosis at the origin of the mass
- Olfactory neuroblastomas can mimic other entities, both on imaging and pathology

Take Home Points
- NUT midline carcinoma, sinonasal melanomas and SNUC share poor prognosis
- High quality imaging and detailed anatomic knowledge are key to guiding appropriate therapy