Tumors of the Sinonasal Cavity

C. Douglas Phillips, MD FACR
Director of Head and Neck Imaging
Weill Cornell Medical College
New York-Presbyterian Hospital
Objectives

• Discuss radiographic signs that may be useful in determining benignancy versus malignancy in sinonasal masses
• Cover organized and orderly approach to evaluating sinus disease that may be cancer
• Discuss differential points among malignant lesions on imaging examinations
Sinonasal Cancer
Signs and Symptoms

- Nonspecific signs/symptoms are rule with sinonasal disease, benign and malignant
- Imaging appearance can be misleading
- Every sinus CT may harbor cancer
- Think of red flags
  - Bone erosion or destruction
  - Clinical history of epistaxis, cranial nerve palsy, nodes
Sinonasal Cancer
Signs and Symptoms

• Nonspecific symptoms
  – Mimic RS
• Early detection uncommon
  – Average delay to diagnosis = 6 mos.
• Often advanced stage at presentation
• Surgical morbidity is high
• Poor prognosis
  – PNTS, nodes or distant mets
  – > 50% mortality
Sinonasal Cancer Epidemiology

- More common in Asia & Africa
- Incidence < 1/100K per year
  - 2000 Americans per year
- 3-4% of H&N neoplasms
- M:F 2:1
- Majority diagnosed in 6th & 7th decades
- Location
  - Maxillary (60-70%)
  - Nasal cavity (20-30%)
  - Ethmoid (10-15%)
Sinonasal Cancer Risk Factors

• **Occupational**
  – AdenoCa & SCCa
    • Leather, textile, furniture, & aluminum industries
    • Nickel dust, mustard gas, thorotrast, chromium, arsenic, etc.
    • Wood dust
      – ↑ SCCa risk 21 times
      – ↑ AdenoCa risk 874 times

• **Non-occupational**
  – HPV (subset of SCCa)
  – EBV (Asian SNUC and NK/T-cell lymphoma)
  – Inverted papilloma
    • EGFR, TGF-alpha, HPV, EBV
  – Genetics
    • Chromosomal abnormalities (SCCa, AdenoCa, ENB)
Sinonasal Cancer Imaging

• CT and MR offer unique information
  – CT for bone changes which may be subtle
  – MR is superior in evaluating soft tissue structures

• CT
  – Contrast only if neck is to be staged

• MR
  – With and without contrast

• PET-CT to assess distant disease
Sinonasal Cancer
PET and PET-CT

- Staging
  - Nodal status
  - Distant metastases
- Direct biopsy
- Therapy response
- Recurrence vs. treatment change
- Re-staging
Benign Neoplasia

- Common lesions
  - Polyps and papillomas (may be associated with chronic sinus disease)
  - Neurogenic tumors, i.e., schwannomas
  - Fibro-osseous or cartilaginous tumors
- Juvenile angiofibroma of young males
  - Other vascular lesions (hemangiomas)
- Tumors from adjacent regions
Sinonasal Polyposis

- **Associations**
  - Allergic or allergic fungal sinusitis
  - Aspirin sensitivity
  - Asthma

- Involve nasal cavity and sinuses

- May remodel or erode bone

- Variable density or signal
Sinonasal Polyposis

- Allergy-related
  - Response of mucosa to allergen
- Wide variety of inflammatory and benign polyps
- Olfactory recess often spared
  - Mucosa is of different variety
Solitary Polyps

• Most are inflammatory – Hypertrophic change in respiratory epithelium
• More common in children and teenagers
• May present with unilateral nasal obstruction
• Most common type is antrochoanal polyp – Arises in maxillary antrum – Extends through native ostium into nasal cavity
Antrochoanal Polyp
Papillomas

• Inverted papilloma most common
• Common along lateral nasal wall
• May harbor SCCa
• Imaging
  – Bony spicule at site of origin
  – Cerebriform enhancement pattern
  – Ca++ in recurrent lesions

Wenig BM, Atlas of Head and Neck Pathology, 1993
Papillomas

- Inverted papilloma most common
- Common along lateral nasal wall
- May harbor SCCa
- Imaging
  - Bony spicule at site of origin
  - Cerebriform enhancement pattern
  - Ca++ in recurrent lesions

Wenig BM, Atlas of Head and Neck Pathology, 1993
Inverted Papilloma

“Cerebriform” pattern of imaging can be distinctive

Images courtesy of Michelle Michel, MD
Inverted Papilloma

Images courtesy of Michelle Michel, MD
Inverted Papilloma

Images courtesy of Michelle Michel, MD
Inverted Papilloma

Bony spicule at base – Should suggest IPAP
Other Benign Sinonasal Neoplasia

- Tumors of mucosal origin
  - Pleomorphic adenoma
- Neurogenic lesions
- Fibrocartilaginous lesions
- Juvenile angiofibroma
Neurogenic Tumors

- 4% of PNSTs of H&N
- Nasal cavity > sinuses
- Nerves of origin:
  - V1/V2
  - Sympathetic fibers (carotid plexus) or parasympathetic fibers (sphenopalatine ganglion)
- Slow-growing, often large
- Present in middle age
- Neurofibromas in NF

Image courtesy of Michelle Michel, MD
Intranasal Schwannoma
Nasal Schwannomas

Images courtesy of Michelle Michel, MD

MISME
Pleomorphic Adenoma
Juvenile Angiofibroma

- Exclusive disease of adolescent males
- Arises in nasal cavity near SPF
- Richly vascular
- Involves nasopharynx, maxillary & ethmoid sinuses
- Extension into PPF
- Locally aggressive
Juvenile Angiofibroma

HYPERVASCULAR CHARACTERISTIC LOCATION
Juvenile Angiofibroma
Sinonasal Fibroosseous and Cartilaginous Lesions

• Share imaging features – calcified matrix
  – Cartilaginous calcifications ("arcs and whorls", "rings")
  – Fibrous calcification ("ground glass")

• Benign lesions with occasional exceptions
  – Low grade chondrosarcoma indistinguishable from benign chondromas in most cases
Fibrous Dysplasia
Fibrous Dysplasia
Ossifying Fibroma

Case courtesy of Dr. Michelle Michel
Osteoma
Imaging of Malignant Sinonasal Cancer

WHO Histologic Classification (44 types)

- Epithelial (19)
  - SCCa
  - AdenoCa
  - Adenoid Cystic Ca
- Soft tissue tumors
  - Hemangiopericytoma
  - Rhabdomyosarcoma
- Lymphomas
  - Non-Hodgkin
  - Plasmacytoma
- Tumors of Bone and Cartilage
  - Chondrosarcoma
  - Osteosarcoma
  - Ewing sarcoma
- Miscellaneous Tumors
  - Esthesioneuroblastoma
  - Melanoma
  - SNUC
Staging Questions

- Evaluate invasion of adjacent structures
  - Other sinuses, extra-sinus extension – orbit, cranial vault (dura, brain, cavernous sinus)

- Evaluate for lymphadenopathy
  - Include upper IJ nodes
  - Primary drainage is to retropharyngeal nodes
  - May drain to level II

- Distant metastases (PET-CT)
TNM Staging of Sinonasal Neoplasia

• Staging is not well established
  – Maxillary sinus and ethmoid sinus have staging system by American Joint Committee on Cancer (AJCC)
• Superior nasal vault lesions may be staged by Kadish system
• Lymphomas have several separate staging systems
Kadish Grading System

- Kadish A
- Kadish B
- Kadish C

- Disease confined to nasal cavity
- Involves one or more sinuses
- Disease extends beyond nasal cavity or paranasal sinuses
Caveat

- All may look similar – destructive lesion of paranasal sinuses
- Some features may be distinctive and allow differential to narrow
Squamous Cell Carcinoma

- Most common sinonasal malignancy
- Well, moderately, or poorly differentiated
- Develop in antrum (maxillary > ethmoid > frontal > sphenoid)
- Bone destruction is invariable
Squamous Cell Carcinoma

- Extend into orbit, contiguous sinuses, pterygopalatine fossa
- Mean 5-year survival 35%
- Poor prognostic indicators
  - Orbital involvement
  - Pterygoid fossa involvement
  - Cervical lymph node metastases
Squamous Cell Carcinoma – Maxillary Sinus
Squamous Cell Carcinoma – Maxillary Sinus
Squamous Cell Carcinoma – Maxillary Sinus
Adenocarcinoma

- About 10% of sinus malignancies
  - Minor salivary gland origin
  - “Intestinal-type” adenocarcinoma
- Cell types
  - Adenoid cystic carcinoma, adenocarcinoma, pleomorphic adenoma, mucoepidermoid carcinoma, acinic cell carcinoma, oncocytoma
- Non-specific imaging appearance
- More common in ethmoid sinus
Adenocarcinoma
Frontal Sinus
Adenocarcinoma
Ethmoid Sinus
Adenoid Cystic Carcinoma (ACCa)

- < 10% of sinonasal malignancies
- Perineural growth pattern (60%)
- Small lesions but poorly marginated
  - Surgical margins difficult to achieve
- Late recurrences and metastases
Perineural tumor spread is key feature of adenoid cystic carcinoma
Sinonasal Melanoma

- Septum or nasal cavity more than sinuses
- 50 to 70 years
- Cervical adenopathy in up to 40%
  - 2 – 3 year median survival
- May remodel bone
- Often homogeneous appearance
- T1 shortening can be indicative
Sinonasal Melanoma
Sinonasal Melanoma
Sinonasal Melanoma
Sinonasal Melanoma
Sinonasal Lymphoma

- Extranodal mucosal lymphoma
- Majority are T-cell
- Bulky soft tissue masses with variable enhancement
  - May remodel bone
  - Often homogeneous appearance
- Higher incidence in Asian population
- Associated with Epstein-Barr virus
Sinonasal Lymphoma

A: Large B-cell Lymphoma, uniform, round-to-oval nuclei with vesicular chromatin and one or multiple conspicuous nucleoli. B: tumor cells are positive for CD20

Sinonasal T-cell Lymphoma

Restricted Diffusion can be helpful
Sinonasal T-cell Lymphoma

Case courtesy of Dr. Ric Harnsberger
B-cell Lymphoma

Bone remodeling

Case courtesy of Dr. Wendy Smoker
Rhabdomyosarcoma (RMS)

- 40% occur in H&N
- Children <5 years; second incidence in 30’s-40’s
- Top of DDx list of aggressive parameningeal lesion
- Perineural tumor spread
- Heterogeneous enhancement
- Locations
  - Orbit – 36%
  - Nasopharynx – 15%
  - Middle ear/mastoids – 14%
  - Sinonasal cavity – 8%
Rhabdomyosarcoma
Rhabdomyosarcoma
Embryonal Rhabdomyosarcoma
Tumors of Superior Nasal Vault

• Primary sinonasal malignancies
  – Esthesioneuroblastoma
  – SNUC (undifferentiated neuroendocrine tumors)

• Potential for CNS disease to extend into superior nasal vault
  – Meningiomas of olfactory groove
Esthesioneuroblastoma (Olfactory Neuroblastoma)

- Arise from olfactory epithelium
- Often have cyst at brain-tumor interface with intracranial extension
- Environmental exposure important causative factor
- High success for cure with combined therapy
Esthesioneuroblastoma (Olfactory Neuroblastoma)

- Two age peaks
  - 11 – 20 years of age
  - 50 – 60 years of age
- Epistaxis
- Nasal obstruction
Esthesioneuroblastoma (Olfactory Neuroblastoma)
Esthesioneuroblastoma
Esthesioneuroblastoma
Esthesioneuroblastoma

Case courtesy of Dr. Wendy Smoker
Sinonasal Undifferentiated Carcinoma (SNUC)

- High grade sinonasal malignancy
- High propensity for distant metastases, advanced stage at presentation, rapid progression
- Often arises in anterior and superior nasal cavity
Sinonasal Undifferentiated Carcinoma (SNUC)
Sinonasal Undifferentiated Carcinoma (SNUC)
Chondroid Tumors: Chondrosarcoma of Septum
Septal Chondrosarcoma
Imaging of Sinonasal Cancer Prognosis – Overall 5-year Survival

- **ENB**
  - 72-80% 5-year
  - Despite skull base & orbit involvement

- **ACCa**
  - 70-90% 5-year
  - 40% at 15 years

- **Lymphoma**
  - 67% 5-year survival

- **AdenoCa**
  - Low grade 80% 5-year
  - High grade < 35% 3-year

- **SCCa**
  - Wide range
    - Stage 1 & 2 ≈ 78%
    - Stage 3 & 4 ≈ 35%

- **Melanoma**
  - 20% 5-year

- **SNUC**
  - 47% 2-year survival
For You Odds Players...
Density / Signal Characteristics

- High cellularity/High nuclear:cytoplasm ratio
  - Lymphoma, melanoma
  - Malignant tumors
- Calcifications
  - Inverting papilloma, Esthesioneuroblastoma
  - Chondroid or fibroosseous lesions
- T1 shortening
  - Melanoma
- Internal architecture/matrix
  - Inverting papilloma
  - Chondroid or fibroosseous lesions
- Mucoid material/protein content
  - Chondroid tumors
Tumor Margins / Bony Changes

- Well-defined/ bony remodeling
  - Neurogenic tumors
  - Pleomorphic adenoma
  - Juvenile angiofibroma
  - Melanoma
  - Lymphoma
  - Esthesioneuroblastoma

- Poorly-defined/destructive
  - Squamous carcinoma
  - SNUC
  - Esthesioneuroblastoma
  - Adenocarcinoma
Malignant Sinonasal Lesion Which May Exhibit Bone Remodeling

- Melanoma
- Lymphoma
- Esthesioneuroblastoma
Locations

• Maxillary antrum
  - Malignant tumor – Squamous cell carcinoma
  - Benign lesion – Antrochoanal polyp

• Superior nasal vault
  - Malignant tumor – Esthesioneuroblastoma, SNUC

• Nasal septum
  - Malignant tumor – Melanoma, lymphoma
  - Benign tumor – Pleomorphic adenoma

• Ethmoid sinus
  - Malignant tumor – Adenocarcinoma
Conclusions

• Maintain high level of suspicion for sinus malignancy in all sinus CT exams
  – Similar clinical presentations
  – Wide range of appearances

• An understanding of appearances of tumors aid in DDx

• CT/MR to properly stage disease