**OBSTRUCTIVE NASAL AND NASOPHARYNGEAL AIRWAY MASSES IN CHILDREN AND ADOLESCENTS**

Michael J. Cunningham MD  
Department of Otolaryngology and Communication Enhancement  
Children's Hospital Boston

**Nasal Airway Obstruction in Children**  
(When you hear hooves, think horses, not zebras)

- **Neonatal & Infant Airway Obstruction**  
  - Neonatal rhinitis  
  - Pyriform aperture stenosis  
  - Choanal stenosis & atresia  
  - Nasolacrimal duct cyst (dacryocystocele)  
  - Adenoid hypertrophy

- **Childhood & Adolescent Airway Obstruction**  
  - Nasal mucosal (turbinate) hypertrophy  
    - viral / allergic / non-allergic (vasomotor)  
  - Septal deviation  
  - Adenoid hypertrophy  
  - Foreign body

**CONGENITAL NASAL MASSES**  
**Relative Frequency**  
(Children's Hospital Boston 32 year retrospective review)

- Dermoid sinus tracts and cysts – 42 cases
- Gliomas – 10 cases
- Encephaloceles – 6 cases

**Nasal Dermoid Sinus Tracts and Cysts**

- Most common of the congenital nasal masses
- Midline mass typically with a sinus opening
- A protruding hair from the punctum is diagnostic
- Intermittent discharge of sebaceous material is common
- Secondary inflammation* is a significant risk
Nasal Dermoid Sinus Tracts and Cysts

- An intracranial communication must be ruled out.
- Such was confirmed in 19% of the CHB case series yet no patient had meningitis or other intracranial infection (characteristically extradural).
- Preoperative imaging crucial:
  - CT – note possible incomplete ossification of the crista galli and foramen cecum in young children
  - MRI – high intensity T1 soft tissue suggests intracranial communication

---

Nasal Dermoid Sinus Tracts / Cysts

Surgical Management

- Direct median approach via horizontal* or vertical incision
- Open rhinoplasty approach with medial crura separation
- Lateral rhinoplasty (alar base) approach
- Paracanthal approach for nasofrontal angle lesions
- Frontal craniotomy via bicoronal approach if intracranial extension
ABNORMAL EMBRYOLOGIC NASAL DEVELOPMENT

• As the neuroectodermal tract recedes, dermal attachments can follow its course, creating a dermoid sinus tract ending in a blind pouch, maintaining a fibrous connection, or extending intracranially.

• If elements of the neuroectodermal tract persist, depending on whether or not the foramen cecum closes, a glioma or encephalocele (meningoencephalocele) may result.

Nasal Gliomas

• Heterotopic neural tissue with no intracranial communication

• Clinical presentation:
  Extranasal mass – 60%
  Intranasal mass – 30%
  Combination – 10%

• CT / MRI to rule out an encephalocele

• Possible fibrous stalk and foramen cecum osseous defect

Nasal Gliomas

Surgical Management

Extranasal

• Direct median approach
• Open rhinoplasty approach
• Lateral rhinotomy (alar base) approach
• Bicoronal craniotomy if CNS involvement

Intranasal

• Transnasal endoscopic approach (triplanar image guidance)
Encephaloceles

**Extranasal Presentation (Sincipital) (60%)**
- Nasofrontal
- Nasoethmoidal
- Naso-orbital

**Intranasal Presentation (Basal) (40%)**
(Nasopharyngeal / Pterygopalatine)
- Transethmoidal
- Sphenoethmoidal
- Trans-sphenoidal
- Sphenomaxillary

- Pulsations/expansion with crying, straining or jugular vein compression
- CT/MRI for diagnosis and anatomical differentiation
Congenital Nasopharyngeal Masses

- Basal encephaloceles and meningoencephaloceles
- Teratomas “hairy polyp” variant
- Cystic developmental anomalies
  - Thornwaldt’s cyst
  - Rathke’s pouch cyst

Nasal (Sinonasal) Inflammatory Lesions

**Unilateral Polyps**
- Antrochoanal polyp
- Sphenoidochoanal polyp

**Bilateral Polyps**
- Cystic fibrosis
- Allergic fungal sinusitis
- Atopic disorders
- Idiopathic

Antrochoanal Polyp

- Accounts for 5% of polyps in adults but up to 30% of polyps in non-CF children
- Characteristic unilateral nasal airway obstruction
- Associated systemic disease predisposition unlikely
- Polypoid mass in nasal cavity, nasopharynx or oral cavity on examination
- Typically unilateral radiographic sinus opacification
- Surgical excision via a combination Caldwell-Luc and endoscopic maxillary antrotomy approach
**Nasal (Sinonasal) Neoplasms**

**Benign Epithelial**
- Papilloma

**Benign Non-epithelial**
- Hemangioma
- Angiomatous polyp
- Extranasopharyngeal angiofibroma?
- Neurogenic neoplasm (schwanomma)
Nasal (Sinonasal) Neoplasms

Malignant Epithelial
- Eshesioneuroblastoma (Olfactory Neuroblastoma)
- Metastatic Neuroblastoma

Malignant Non-epithelial
- Rhabdomyosarcoma
- Other Soft Tissue Sarcomas
- Non-Hodgkin Lymphoma

Benign Fibro-osseous
- Fibrous Dysplasia
- Ossifying Fibroma
- Juvenile (Aggressive) Ossifying Fibroma

Malignant Fibro-osseous
- Osteogenic Sarcoma
- Chondrosarcoma
NASOPHARYNGEAL NEOPLASMS

• Juvenile Nasopharyngeal Angiofibroma
• Craniopharyngioma
• Cordoma
• Non-Hodgkin Lymphoma
• Rhabdomyosarcoma
• Nasopharyngeal Carcinoma

NASOPHARYNGEAL MALIGNANCIES

• Similar clinical presentation but age and race differential
• Non-specific signs & symptoms often delay diagnosis
• Endoscopic examination often likewise non-specific
• Imaging assessment very important
• Biopsy (unless clearly JNA) and histopathologic examination crucial as lesion specific therapy

Juvenile Nasopharyngeal Angiofibroma

• Etiology
  — True neoplasm versus vascular malformation
  — Vascular hamartoma
  — Ectopic nidus of vasoformative tissue with both vascular and fibrous histopathologic components
• Epidemiology
  — Arises typically (exclusively) in adolescent males
  — Suspect an alternative diagnosis in non-adolescent males and females
• Clinical Presentation
  — Nasal obstruction and epistaxis are principal manifestations
  — Rhinorrhea / anosmia / headache / otitis media with effusion also common
  — Facial deformity / proptosis / exophthalmos / neurologic deficits such as diplopia suggest advanced disease
• Investigative work-up
  — Nasal endoscopy
  — Imaging

Juvenile Nasopharyngeal Angiofibroma

Imaging Evaluation

- Computed Tomography
- Magnetic Resonance Imaging (Magnetic Resonance Angiography)
- Angiography

- diagnosis suspicion and staging
- diagnosis confirmation and staging
- diagnosis confirmation and preoperative embolization
- # of feeding vessels
- External vs. internal carotid origin
- Ipsilateral vs. contralateral contribution
- Estimate % lesion devascularization
Juvenile Nasopharyngeal Angiofibroma

Follow-up Management

- Residual versus Recurrent Disease
  - Broad incidence range 13% - 50%
  - Wide variability depending on criteria (clinical / imaging) utilized
  - Clearly dependent on presenting stage
- Follow-up Timing
  - How soon?
  - How often?
  - Endoscopically?
  - Radiographically (MRI / MRA / CT)?
- Follow-up duration if no evidence of residual / recurrent disease?
- When to intervene if evidence of residual / recurrent disease?
  - Based on location? size? symptoms?
  - Surgical management options
  - Adjuvant therapy indications
THANK YOU

NORMAL EMBRYOLOGIC NASAL DEVELOPMENT

- The external nose forms from ectoderm, mesoderm and a deeper layer of cartilaginous capsule.
- External to the cartilaginous capsule the nasal and frontal bones form within the mesoderm via intra-membranous ossification.
- Between the frontal bones and the nasal bones is a space called the fonticulus frontalis.
- Between the nasal bones and the more posterior cartilage is the prenasal space.
- Within the prenasal space, a herniation of dura extends through the foramen cecum, contiguous with the periosteum of the nasal bones, to the skin (the neuroectodermal tract).
- During normal development, (1) skin and dura become separated by progressive ossification, (2) the dural projection obliterates, and (3) the foramen cecum and fonticulus frontalis fuse as the cribiform plates form.
Encephaloceles
Surgical Management

- Neurosurgical resection of intracranial component via frontal craniotomy
- Extranasal extracranial component resection via bicoronal incision if necessary for above or an alternative approach
- Intranasal extracranial component resection via transnasal endoscopic approach with triplanar image guidance

Juvenile Nasopharyngeal Angiofibroma

**Location of Disease and Staging**

- Primary focus suspected to be the sphenopalatine foramen
- Anteromedial extension to the nasopharynx and nasal cavities
- Posteromedial extension to the sphenoid sinus
- Lateral extension to the pterygomaxillary fossa
- Direct posterior extension to the pterygoid plates
- Subsequent extension to the ethmoid and maxillary sinuses, infratemporal fossa, orbit and/or soft tissues of the cheek
- Potential for intracranial/cavernous sinus extension typically extradural at the level of the middle cranial fossa

Juvenile Nasopharyngeal Angiofibroma

**Non-Surgical Management**

- Observational monitoring anticipating spontaneous regression
- Hormonal therapy
  - Exogenous estrogen administration
  - Testosterone receptor blockade (flutamide)
- Radiation therapy
  - Typically reserved for advanced stage primary lesions with extensive intracranial disease or residual/recurrent disease in anatomic “at risk” sites
  - Intensity-modulated radiotherapy or proton beam therapy
  - Risks of osteoradionecrosis/craniofacial growth alteration/secondary malignancy
- Chemotherapy has no established role
- Interferon therapy (angiogenesis inhibition)*

<table>
<thead>
<tr>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Stage I</td>
<td>Limited to the nasopharyngeal cavity; negligible bone destruction.</td>
<td>Confined to the nasopharynx</td>
<td>A. Limited to nose and/or nasopharynx. B. Extension into 1 sinus</td>
<td>A. Limited to nose and/or nasopharynx. B. Extension into 1 sinus</td>
</tr>
<tr>
<td>Stage II</td>
<td>Involvement of the PMF or the maxillary, ethmoid or sphenoid sinuses with bone destruction.</td>
<td>Extension to nasal cavity and/or sphenoid sinus</td>
<td>A. Minimal extension into PMF. B. Full occupation of PMF. C. Extension to ITF with or without cheek.</td>
<td>A. Minimal extension into PMF. B. Full occupation of PMF. C. Extension posterior to the pterygoid plates.</td>
</tr>
<tr>
<td>Stage III</td>
<td>IIIA. Involvement of ITF or orbit without cranial involvement. IIIB. Intracranial extrudal (parasellar) involvement.</td>
<td>Involvement of 1 or more maxillary or ethmoid sinuses, PMF, ITF, orbit and/or cheek. Intracranial involvement</td>
<td>A. Erosion of skull base - minimal intracranial. B. Erosion of skull base - extensive intracranial with or without cavernous sinus involvement.</td>
<td></td>
</tr>
<tr>
<td>Stage IV</td>
<td>Intracranial intradural tumor without (A) or with (B) infiltration of the cavernous sinus, pituitary fossa or optic chiasm.</td>
<td>Extension into cranial cavity</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
## Juvenile Nasopharyngeal Angiofibroma

### Surgical Management

- Transpalatal approach
- Transfacial approaches
  - Lateral rhinotomy
    - Associated medial maxillectomy / sphenoidectomy
    - Limited lateral exposure
  - Midface Degloving
    - Associated bilateral LeFort I osteotomies with palatal drop
    - Superior and particularly lateral exposure limitations
  - Facial Translocation
    - Requires lateral rhinotomy with subciliary incision
    - Maxilla removal and reinsertion
    - Good lateral exposure

### Infratemporal Fossa Approaches

- Lateral extended preauricular or hemicoronal incision with zygomatic osteotomy and subtemporal craniectomy
- Applicable to combined craniofacial resection or open craniotomy

### Transnasal / Transantral Endoscopic Approaches (Expanded Endonasal Approach)

### Combination Endoscopic-assisted or Microscopic-assisted Open Approaches

### Stage-Dependent Operative Approach

<table>
<thead>
<tr>
<th></th>
<th>Endoscopic</th>
<th>Transpalatal</th>
<th>Lateral Rhinotomy / Partial Maxillectomy</th>
<th>Midfacial Degloving</th>
<th>Facial Translocation</th>
<th>Infratemporal Approach</th>
</tr>
</thead>
<tbody>
<tr>
<td>I A</td>
<td>x</td>
<td>x</td>
<td>(x)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>I B</td>
<td>x</td>
<td>(x)</td>
<td>x</td>
<td>x</td>
<td></td>
<td></td>
</tr>
<tr>
<td>II A</td>
<td>x</td>
<td>(x)</td>
<td>x</td>
<td>x</td>
<td></td>
<td></td>
</tr>
<tr>
<td>II B</td>
<td>x</td>
<td>(x)</td>
<td>x</td>
<td>x</td>
<td>(x)</td>
<td></td>
</tr>
<tr>
<td>II C</td>
<td>x</td>
<td>(x)</td>
<td>x</td>
<td>x</td>
<td>x</td>
<td></td>
</tr>
<tr>
<td>III A</td>
<td>x</td>
<td>(x)</td>
<td>x</td>
<td>x</td>
<td>x</td>
<td>x</td>
</tr>
<tr>
<td>III B</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>(x*)</td>
<td>(x*)</td>
</tr>
</tbody>
</table>
**Nasal Airway Obstruction in Children**


