Evaluation of Congenital Midline Nasal Masses

Camysha H. Wright, MD, Resident
Matthew Ryan, MD, Faculty Advisor
The University of Texas Medical Branch
Department of Otolaryngology
Grand Rounds Presentation
June 7, 2006
Outline

- Embryology of the nose
- Dermoid cysts
- Glioma
- Encephalocele/Meningocele
- Evaluation of Nasal Mass
- Imaging Studies
- Surgical Intervention
- Conclusion
Embryology

- The critical period in nasal development is in the first twelve weeks of fetal development.
- Abnormalities of development are believed to cause gliomas, dermoids, and encephaloceles.
Embryology

- Neural tube develops between the third and fourth week of gestation
- Closure of the neural tube occurs from the midline and extends cranially and caudally
- Neural tube then gives rise to neural crest cells
Embryology

- As the neural tube closes neural crest cells migrate anteriorly and laterally around the eyes to the frontonasal process
- Nose formed from the medial and lateral prominence and invagination of the nasal pit
Embryology

- In most of the body neural crest cells are involved in ectodermal components, in the face the primary role is in the formation of mesenchymal cells

- Bone, cartilage, and muscles of the face are all derivatives of neural crest cells
Embryology

- Nose develops from frontonasal processes and 2 nasal placodes
- Medial processes fuse
- Nasomaxillary groove becomes the nasolacrimal duct
Embryology

- Scanning electron micrograph
Embryology

- During formation of skull base and nose, mesenchymal structures are formed from several centers which will eventually fuse and ossify.
- Before their fusion, there are recognized spaces which are important in the development of congenital midline nasal masses
  - Fonticulus frontalis
  - Prenasal space
  - Foramen cecum
Embryology

- Fonticulus frontalis – space between the frontal and nasal bones
  - Eventually fuses with foramen cecum to create a separation between intracranial and extracranial structures

- Prenasal space is between the nasal bones and the nasal capsule (precursor of the septum and nasal cartilages)
Pediatric Anatomic Considerations

- Neonates can suffer from respiratory distress with nasal obstruction.
- Pediatric airway differs from adults in that neonates are nasal breathers.
- Epiglottis abuts nasal surface of the soft palate forming anatomic divide between the airway and digestive tracts.
- Food from oral cavity is shunted laterally into esophagus via the pyriform sinuses.
- Neonates can functionally eat and breathe concurrently.
Pediatric Airway
Nasal Masses

- Differential Diagnoses
  - Inflammatory lesions (abscess)
  - Traumatic deformity
  - Benign neoplasms (polyps, JNA)
  - Malignant neoplasms (rhabdomyosarcoma)
  - Congenital masses (teratomas, hemangiomas)
Nasal Dermoids

- Can occur as cyst or sinus
- Most common congenital midline nasal mass
- 1-3% of all dermoids
- 10% dermoids of head and neck
Nasal Dermoid

- Has ectodermal and mesodermal components
  - (ectodermal components only – epidermal cyst, ectoderm, mesoderm and endoderm - teratoma)
- May present as midline nasal pit, fistula, or infected mass anywhere from glabella to columella
- Sometimes presents as single cutaneous tract with hair at opening
- May secrete pus or sebaceous material
Nasal Dermoid

- Superonasal dermoid
Nasal Dermoid

- CNS connection variably reported (4-45%)
- Associated congenital anomalies (5-41%), however not found to be associated with syndromes

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Nasal Dermoid

Complications:

- Intermittent inflammation
- Abscess
- Osteomyelitis
- Broaden nasal root
- Meningitis
- Cerebral abscess
Nasal Dermoid abscess

- Abscess formation
Dermoid Cyst

- Development
  - During development a projection of dura projects through the foramen cecum and attaches to skin
  - Dura normally separates from nasal skin and retracts through foramen cecum losing connection
  - If skin maintains attachment to underlying fibrous tissue, nasal capsule, or dura, epithelial elements may be pulled into the prenasal space with or without dural connection
Dermoid Cyst Development
Glioma

- Glial cells in a connective tissue matrix
- Firm, noncompressible
- Red or bluish lump
- Can be found at glabella, at nasomaxillary suture, intranasally
- No connection with subarachnoid space
- Do not enlarge with crying
- Do not transilluminate
- May have telangiectasias
Glioma

- Intranasal glioma
Glioma

- Extranasal - 60%
- Intranasal - 30%
- Both - 10%
- Dural connection - 35% intranasal, 9% extranasal
- Overall 15% dural connection
- CSF rhinorrhea, meningitis possible, if dural connection exists
Glioma – Formation Hypotheses

- Similar to that of nasal dermoids
- Develop from extracranial rests of glial tissue
- Abnormal closure of fonticulus nasofrontalis
- Another theory is that they are possibly encephaloceles which have lost CSF connection
Glioma Development
Encephaloceles

- Extracranial herniation of meninges and/or brain
- Connection with subarachnoid space
- Rare at 1:35,000 births
- 30-40% associated anomalies:
  - microcephaly, hydrocephalus, microophthalmia, anophthalmia, agenesis of the corpus callosum, porencephaly, cortical atrophy, ventricular dilations
Encephaloceles

- Bluish, soft, compressible, transilluminable, pulsatile
- Enlarge with crying
- Positive Furstenberg test (enlarge with bilateral compression of internal jugular veins)
- Originate medially in the nose (cannot pass probe medially to this mass, versus with glioma generally can as they originate laterally often)
- May have associated CSF leak
Encephaloceles

- Large nasal encephalocele
Encephaloceles

- Divided into three categories:
  - Occipital 75%
  - Sincipital 15%
  - Basal 10%

- Sincipital-anterior or frontonasal (dorsum of nose, orbits, forehead)

- Basal-intranasal mass, nasopharynx, posterior orbit because they herniate through the cribiform plate or posterior to it (potential for airway obstruction in neonate)
Encephaloceles

- **Basal Encephaloceles**
  - Transethmoidal-through cribiform plate into middle meatus
  - Sphenoethmoidal-extends through cranial defect between posterior ethmoids and sphenoid to nasopharynx
  - Transsphenoidal-presents in nasopharynx
  - Sphenomaxillary-through superior and inferior orbital fissures to sphenomaxillary fossa
Encephalocele

Development

- Dural projection through fonticulus nasofrontalis
- Abnormal closure results in herniated meninges/brain
- May be closely related to glioma
Encephalocele Development
Nasal Masses
Evaluation

- Most often infants and children
- Dermoids-fistula tract, hair, pus or sebum, midline
- Gliomas-firm, noncompressible, does not transilluminate, telangiectasias
- Encephaloceles-soft, compressible, bluish or red, enlarge with crying, positive Furstenburg test
- Do not biopsy extra or intranasal mass in a child before imaging (Risk of meningitis or CSF leak if there is an intracranial connection)
Imaging

- CT and MRI most used
  - CT findings include: fluid filled cyst, soft tissue mass, intracranial mass, enlargement of foramen cecum, distortion of crista galli
  - CT findings suggestive of intracranial extension are enlarged foramen cecum and bifidity of crista galli
  - Findings valuable if absent, (when present may be false positive)
CT Imaging

- Nasal Dermoid axial ct with dermoid anterior to nasal and maxillary bones, no bony dehiscence or abnormalities noted
Nasal Dermoid with Intracranial Extension

Figure 1: CT scan axial cut showing nasal dermoid extension into the anterior cranial fossa

Figure 2: CT scan 3-D reconstruction showing defect in the anterior part of the cribriform plate
CT Imaging

- Encephalocele with defect noted in cribiform plate and herniation of brain tissue
MRI

- Better delineates soft tissue
- Ability to visualize in the sagittal plane
- Denoyelle 36 children with dermoids, 2 patients had CT suggestive of intracranial involvement not found at surgery.
- Recommended CT followed by MRI to confirm intracranial connection
MRI Imaging

- Nasal dermoid cyst
MRI-Glioma

- Saggital T1
MRI-Glioma

- Sagittal T2
MRI-Glioma

- Coronal T2
MRI-Encephalocele

- Coronal T2 weighted and sagittal T1 weighted image of sphenoid encephalocele
Workup

- **History/Physical Examination**
  - Nasal obstruction, polypoid intranasal mass, CSF leak, presence of hair or fistulous tract, compressible or firm, presence of pulsations, enlargement with crying or internal jugular compression

- **Radiologic Evaluation (CT and/or MRI)**
  - No Intracranial Extension (Dermoid/Glioma)
  - Intracranial Extension
Surgical Treatment

- Complete excision (open-transcranial vs extracranial, vs endoscopic approaches described)
- Perform early to avoid nasal distortion, bony atrophy, osteomyelitis, meningitis
- Dermoids—must excise entire tract to prevent recurrence
Dermoid

- Can be removed endoscopically or via open approach
- Transverse rhinotomy has been described
  - Small to moderately sized lesions
  - Avoids vertical scar and splaying
Nasal Dermoid with intracranial extension.

- Dermoid cyst with intracranial extension without craniotomy
  - Nasal bones removed along with anterior part of the frontal bone
  - Followed sac through cribiform plate, incised wall of sac and evacuated contents, and removed all except for its base where it was attached to dura
  - Destroyed secretory epithelial surface of remnant of sac with bipolar, replaced bone and closed wound in layers
  - Pt did well postoperative and no recurrence noted during 2 year follow up.
Glioma

- Can be removed via open or endoscopic approach
- Lateral rhinotomy or alar incisions may be used for intranasal gliomas or combined intra-extranasal gliomas
- Several authors have reported isolated cases of endoscopic excision of small gliomas with and without evidence of intracranial extension.
Encephaloceles

- In the past required combined approach with neurosurgery
- Frontal craniotomy is performed, intracranial mass excised, bone-dura defect is repaired
- Extracranial mass is then removed
- More reports describing endoscopic removal
Conclusion

- Midline nasal masses are rare but must be remembered in the differential
- Furstenberg’s test
- Don’t biopsy without imaging
- Surgical intervention necessity
Bibliography

- Hayashi T, Utsanomiya H, Hashimoto T. Transeptal encephalocele. Surg Neurol 1985; 24: 651-655
Bibliography


