Congenital Nasal Masses

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Outline

• Common Causes of Pediatric Nasal Obstruction
• Embryology
• Nasal Dermoid
• Nasal Glioma
• Encephalocele
• Nasolacrimal Duct Cysts
• Thornwaldt Cyst
• Brief Word on Pediatric Nasal Tumors
# Nasal Obstruction

<table>
<thead>
<tr>
<th>Congenital</th>
<th>Foreign body</th>
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<tbody>
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**Associated syndromes**
- Cystic fibrosis
- Kartagener
- CHARGE
- Apert
- Crouzon
- Treacher-Collins
- Fetal alcohol syndrome
- Down

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Embryology
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

- The anterior neuropore (primitive frontonasal region) develops medial to the optic recesses in the third week of fetal life. A transient fontanelle between the inferior frontal bone and nasal bone (fonticulus frontalis) exists.
Embryology

- A funnel-shaped dural projection extends inferiorly and anteriorly through a midline opening anterior to the crista galli of the ethmoid bone. This anterior skull base opening is the foramen cecum.

- The dural diverticulation extends inferior and posterior to the frontal and nasal bones and superior and anterior to the nasal cartilage (prenasal space).

- The apex of this dural diverticulation temporarily approximates the subcutaneous region of the mid-nasal bridge at the osteocartilaginous junction.
Embryology - Normal Development

(Speaker Notes for next slide)

Fig. 1 Normal anterior neuropore development.

- A: Sagittal color graphic of the fetal anterior neuropore shows the transient osteo-cartilaginous gap at the fonticulus frontalis (arrowhead) and the prenasal space (arrow).

- B: In the fourth week to the seventh week of gestation, a transient dural diverticulum extends through the plane of the foramen cecum (Arrowhead), coursing through the prenasal space and terminating at the skin of the nasal bridge (arrow).

- C: With normal regression of the dural diverticulum, a blind-ending foramen cecum is left (arrow). This lies just anterior to the crista galli
Embryology - Normal Development

- Failure of involution at these sites where surface and neural ectoderm approximate each other can lead to anomalous development including: nasal dermal sinus, encephalocele, and nasal glioma.
Normal development of the frontonasal region, MRI assessment.

- A: Sagittal T1-W image of a newborn shows isointense signal of the basal frontal lobes, nasal septal cartilage, and crista galli (arrow).

- B: At 6 months, there is normal T1 hyperintensity within the nasal bone (curved arrow) and slight T1 hyperintensity of the crista galli (arrow).

- C: Sagittal T1-W image of a normal 14-month-old boy shows a cock’s comb shape and hyperintense signal of the crista galli (arrow). Note the foramen cecum located posterior to the nasal process of the frontal bone (arrowhead)
Embryology - Normal Development
Embryology - Abnormal Development
Nasal Dermoid Cyst
Dermoid Cyst
Dermoid Cyst

- Epidemiology
  - Fewer than 10% of all dermoid cysts occur in the head and neck
  - Nearly 75% of head and neck dermoid cysts are located in the nasoglabellar region
  - Can be noted as early as at birth to within first two decades of life
Dermoid Cyst

• Embryology:
  ▫ Cysts consist of squamous cell epithelium containing epidermal appendages
  ▫ Epidermal elements are displaced during intramembranous growth phase of nasal bones
  ▫ Location can be several:
    • Fronto-temporal region
    • Orbital region
    • Nasoglabellar region
Dermoid Cyst

- During development, dura projects through the foramen cecum and attaches to skin
- Separates from nasal skin and retracts through foramen cecum
- If persistent attachment to underlying fibrous tissue, nasal capsule, or dura, epithelial elements are trapped in the prenasal space
Embryology - Abnormal Development
Dermoid Cyst
Dermoid Cyst

Nasoglabellar Cysts

- Up to 50% may have a fistula or sinus tract
- Tract traverses via the cribiform plate or foramen cecum
- Tract attaches to dura, falx cerebri, or other intracranial structures
- Diff Dx: Nasolabial cyst, premaxillary cyst, nasopalatine cyst, Jacobson organ cyst
Nasal Encephalocele
Nasal Encephalocele

- Extracranial protrusions of meninges, CSF fluid, and neural tissue

- Meningoceles present similarly without herniation of brain tissue

- May present as external or internal nasal masses

- Described by location of dehiscence in the skull base
Nasal Encephalocele

• Presentation:
  ▫ Pale
  ▫ Compressible
  ▫ Pulsatile
  ▫ Transilluminate with light
  ▫ Positive Furstenberg’s Test (expansion with compression of Jugular Vein)
  ▫ Expansion may also be triggered by crying or straining
Nasal Encephalocele
Nasal Encephalocele

Sincipital Encephalocele

Basal Encephalocele
Nasal Encephalocele
Nasal Encephalocele

- Radiological Assessment is integral
- MRI is modality
- Helps with Pre-operative planning
- Confirms need for neurosurgical involvement
Nasal Glioma
Nasal Glioma

- Encephalocele without intracranial connection

- Also known as:
  - Benign congenital nasal neuroectodermal tumor
  - Nasal cerebral heterotopia
  - Glial heterotopia
Nasal Glioma

- Hypotheses on formation
  - 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
Nasal Glioma

- Grow along with the child
- Fibrous stalk remnant noted in 15%
- Dysplastic neuroglial tissue and fibrovascular tissue
- Glabella is most common although medial canthal mass possible
Nasal Glioma

- Most identified at birth or in infancy
- Appear well-circumscribed, bluish or reddish in color, and telangiectatic surface
- Sincipital vs Basal
- 30% have intranasal presentations
  - Lateral Nasal Wall
  - Middle Turbinate
  - Nasal Septum
Nasal Glioma

- Radiological Assessment is the key
- MRI is integral to assess the lesion
- Able to better visualize if the mass has a stalk or is separate
Nasal Glioma
Treatment of Nasal Gliomas, Nasal Encephaloceles, and Nasal Dermoid Cysts
Surgical Treatment

• Steps:
  ▫ Elliptical incision around pit
  ▫ Lacrimal probe used to cannulate tract
  ▫ Small diamond burr used to drill around the tract through the nasal bones
  ▫ Evaluate and free tract from adherent dura and/or crista galli

Surgical Treatment

• For Gliomas and Encephaloceles:
  ▫ Nasal Evaluation and Pre-operative imaging
  ▫ Entities involving the dura or intracranial contents mandate neurosurgical evaluation
  ▫ Lesions restricted to the nasal cavity may be removed endoscopically or via a lateral rhinotomy approach

Surgical Evaluation

- Clipping should be performed prior to stalk resection
- Repair of the suture line and dura must be performed
- Options for closure include calvarial bone graft, fascia lata grafts, vascularized pericranial flap
Nasolacrimal duct cyst
Nasolacrimal Duct Cyst

- May occur in as high as 30% of neonates
- Many cases resolve within 1 year of life
- Proximal and distal obstruction cause formation of a dacrocytocele
- Formation of cyst causes nasal obstruction
- Distal obstruction occurs at the valve of Hasner
Nasolacrimal Duct Cyst

- Often distal obstruction cases resolve spontaneously with nasal respirations
- Bilateral dacrocystocele occur in 14% of cases
- Some children develop large enough cysts that nasal obstruction is symptomatic
Nasolacrimal Duct Cyst

• Presentation:
  ▫ Epiphora
  ▫ Facial swelling
  ▫ Blue to red discoloration inferior to the medial canthus
  ▫ Mass below inferior turbinate on endoscopy
Nasolacrimal Duct Cyst

- **Diagnosis:**
  - Can pass scope distal to cyst
  - Imaging shows cystic masses projecting into nasal cavity with superomedial displacement of the inferior turbinate
  - Imaging may show dilation of lacrimal duct and sac

- **Management:**
  - Warm compress and facial massage
  - Endoscopic probing of the duct with marsupialization of cyst
Thornwaldt Cyst
Thornwaldt Cyst

- Named for Gustav Ludwig Thornwaldt
  - 1885 – series of 26 cases
- Embryology:
  - Congenital cyst in nasopharynx
  - Results from a communication between the notochord and the nasopharyngeal endoderm
  - Incidence - 3-4% (slight male predilection)
  - Diagnosed in 2nd and 3rd decades of life
  - Triggered by infection, inflammation, complicated adenoidectomy

Thornwaldt Cyst

- Typical Clinical Presentation
  - Occipital Headaches
  - Persistent notable nasal discharge
  - Halitosis
  - Sore/Stiff cervical muscles
  - Ear fullness/Eustachian Tube Dysfunction

- Smooth, submucosal mass in nasopharynx
- Superior to the adenoid pad
- May have central dimpling and/or yellow hue

Thornwaldt Cyst

Thornwaldt Cyst

• Work-Up:
  ▫ History + Physical Exam + Imaging

• Differential Diagnosis:
  ▫ Rathke pouch cyst
  ▫ Adenoid Retention cyst
  ▫ Sphenoid sinus mucocele
  ▫ Nasopharyngeal CA

• Treatment:
  ▫ Observation if asymptomatic
  ▫ Marsupialization or resection
Quick Concluding Thoughts
Neonatal Nasal Tumors

- Congenital Tumors – 2-14 cases per 100,000 births
- Differential Diagnosis:
  - Teratoma
  - Hamartoma
  - Rhabdomyosarcoma
  - Hemangioma
  - Neurofibroma
  - Lymphatic Malformations
Nasal Obstruction and Masses

- Neonates are obligate nasal breathers
- Radiological findings help provide diagnosis
- MRI preferred over CT especially for intracranial assessment