Skull Base Developmental Lesions

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Significance of Skull Base Developmental Lesions

• People basically hate the skull base
  – It is a region where people often turn off
  – “If I don’t see you, you’re not there”
• Developmental lesions are relatively common
• Developmental lesions are often “leave-me-alone-lesions”
• HOWEVER, they may resemble pathology
  – Doctors want to do things to them

Topics to Discuss

• Embryology and development of normal skull base
• Skull base foramina, fissures, and surface features
• Normal variations of skull base
• Maldevelopmental lesions of anterior, central, and posterior skull base

Skull Base Regions

• Anterior
  – CN I
• Central
  – CN II, III, IV, V, VI
• Temporal bone
  – CN VII, VIII, ICA
• Posterior
  – IJV, CN IX, X, XI, XII;

Disclosures

• I have no pertinent financial disclosures
Anterior Skull Base

- Bony elements
  - Frontal, ethmoid, and sphenoid bone
- CN I is dominant CNS feature
- Interface of CNS and sinonasal cavity

Maldevelopmental Lesions

- Sinonasal encephaloceles
- Nasal glial heterotopias
- Nasal dermoid and dermal sinus
- NF 1 bony dysplasia

Normal Nasal Development

4-6 weeks

- Frontal bone and nasal bone develop externally
- Nasal capsule (S) develops under them
  - Cartilaginous interior of frontonasal process
  - Becomes ethmoids
- Fonticulus frontalis (f)
- Dural diverticulum (d) projects anteriorly into prenasal space

After 8 weeks

- Dural diverticulum regresses
  - Remnant is foramen cecum
- Prenasal space obliterated
  - Nasal bones fuse with nasal capsule
- Fonticulus frontalis obliterated
  - Remnant is nasofrontal suture

Illustration modified from Imaging the Pediatric Head, Neck and Spine by Castillo and Mukherji.
Anterior Skull Base Encephaloceles

- Contain herniated frontal lobes
- Frontonasal are majority of lesions
  - Present as mass over nasal dorsum
- Nasoethmoidal are less common
  - Present in nasal cavity
- Nasolateral are rare
  - Herniate into orbits
- Naming convention for encephaloceles
  - Roof is first named item
  - Floor is second named item

Frontonasal Encephalocele

- Brain herniates into persistent dural diverticulum
- Encephalocele presents as nasal mass
- Mass in superior nasal cavity – LOOK AT SKULL BASE
Nasoethmoidal Encephalocele

Illustration modified from Imaging the Pediatric Head, Neck and Spine
Castillo and Mukherji

Nasal Glial Heterotopia
“Nasal Glioma”

• Development analogous to encephalocele
• Defect closes leaving CNS tissue outside:
  • Frontonasal encephalocele
    – Extranasal glial heterotopia
  • Nasoethmoidal encephalocele
    – Intranasal glial heterotopia

Extranasal Glial Heterotopia

Diagnosis may be unsuspected and may be misdiagnosed by pathology. Immunoreactive for GFAP and S100.

Intranasal Glial Heterotopia

Courtesy of Rick Wiggins, MD
Nasal Dermoid/Dermal Sinus

- Dural diverticulum persists longer than normal in contact with nasal dermal structures
- Can drag nasal dermal structures back with it as it later regresses
- Ectoderm lines dermal sinus
  - Occasional dermoid or epidermoid masses along track

Illustration modified from Imaging the Pediatric Head, Neck and Spine Castellano and Mukherji

Nasal Dermoid

Intracranial dermoid at foramen cecum

Nasal Dermal Sinus

- Patients may have characteristic nasal pit
- Can occasionally express fluid

Nasal Dermal Sinus

Bony Dysplasias

- Hypodevelopment or maldevelopment of sphenoid
- Accompany NF
- Rare isolated dysplasia

Courtesy of Stephen Park MD
**Central Skull Base Anatomy**

- **Central skull base**
  - Posterior to lesser wing of sphenoid / planum sphenoidale
  - Anterior to petrous ridge/dorsum sella

**Foramen and fissures**
- Optic canal
- Superior orbital fissure (SOF)
- Inferior orbital fissure (IOF)
- Carotid canal
- Foramen rotundum
- Foramen ovale
- Foramen spinosum
- Foramen lacerum
- Vidian canal

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**Lateral Sphenoid Defect**

**Central Skull Base Anatomy**

The “Holey Land”
Central Skull Base Maldevelopmental Lesions
- Variant foramina
- Encephaloceles
  - Canal of Sterng
- Vascular hypoplasia/aplasia
- NF 1 bony dysplasia
- Sphenoid benign osseous lesion
  - AKA "Non-pneumatized sphenoid"

Variant Foramina of Skull Base
- More than you can count
- Often no more than anatomic curiosity when perusing normal skull base
- Described variants include:
  - Foramen of Vesalius
  - Palatovaginal canal
  - Canaliculus innominatus
- More important variants may be confusing, appear as pathology

Persistent Craniopharyngeal Canal (PCC)
- Midline vertical conduit in basisphenoid which may contain dura and vascular structures
  - Can be quite large and may extend to sella
  - Can contain pituitary tissue
- Larger varieties may be termed transsphenoidal canal
- May be associated with craniofacial anomalies and nasopharyngeal mass

Fossa Navicularis Magna
- Notch in basiocciput containing pharyngeal tonsillar tissue
- Likely related to notochordal remnant that inhibits development of normal basiocciput

Canalis Basilaris Medianus
- Remnant of cephalic end of primitive notochord
- Intracranial surface of basiocciput in the midline
- May be complete or incomplete
Basal Encephaloceles

- Transsphenoidal
  - Sphenoid bone into sinus or nasopharynx
- Sphenoorbital
  - SOF into orbit
- Sphenoethmoid
  - Junction of sphenoid/ethmoid into posterior nasal cavity
- Transethmoid
  - Through ethmoid into anterior nasal cavity
- Sphenomaxillary
  - IOF into pterygopalatine fossa

Lateral Sphenoid Encephaloceles

- Relatively rare
- More likely to be occult, and present later
  - CSF leaks, meningitis
- Clinical and radiological diagnosis may be difficult
  - Unsuspected
- “Cyst” in sphenoid sinus is an encephalocele until proven otherwise

Sphenoid Encephalocele

Bone defect is lateral sphenoid wall in setting of significant pterygoid pneumatization medially and laterally to foramen rotundum and transverse to vidian canal
Sternberg’s Canal

- Lateral wall of sphenoid
- Associated with lateral "spontaneous" CSF leaks/encephaloceles
- Reported in up to 4% of adults
- Doesn’t correspond to typical lateral encephalocele
  - Sternberg’s Canal must be medial to V2

Transalar Sphenoid Encephalocele

Location corresponds to enlarged foramen ovale

Transalar Sphenoid Encephalocele

Posterior Skull Base Anatomy

- Posterior to dorsum sellae
- Basiocciput and clivus
- Foramen magnum, jugular foramen, and internal auditory canals
- CNs VII-XII

Vascular Dehiscence

Carotid Dehiscence

Posterior Skull Base Maldevelopmental Lesions

- Brief word of carotid anomalies
- Clivus anomalies and variant foramen
- Ecchordosis physaliphora
- Sphenoid and occipital hypoplasia
- NF 1 bony dysplasia
- Arnold-Chiari and Dandy-Walker malformations
Benign Developmental Fatty Pseudolesion of Sphenoid

- Referred to as “arrested pneumatization”
  - Benign intraosseous lipoma
- “Bubbly” lesion of sphenoid
- Differential diagnosis of chondroid lesions
- Characteristic presence of fat in lesion
  - Similar to “hemangioma” of vertebra
- Associated hypodevelopment of sphenoid sinus

Ecchordosis Physaliphora

- Benign variant of chordoma
- T2 hyperintense
- Key differentiation
  - Absence of enhancement
  - Smooth bony margins
  - Has common intradural component
- Described in 2-3% of population in autopsy series

Notochord Remnants Chordoma, EP & Thornwaldt Cyst
Gray Gelatinous Notochord Tissue
- T1 Hypointense
- T2 Hyperintense
- No Enhancement

Chordoma
- Bulky, lobulated, destructive mass

Midline Bone Destruction
Aberrant ICA

Congenital Absence of ICA

Occipital Bone - Normal Development of Clivus

Occipital Condylar Hypoplasia with Basiocciput Hypoplasia

Occiput-C1 Assimilation
Occiput-C1 Assimilation

Arnold Chiari I Malformation

Arnold Chiari II Malformation

Arnold Chiari III Malformation

Dandy-Walker Malformation with Myelocystocele

Conclusions

- Many normal and variant skull base foramina and knowledge of normal variants can keep you out of trouble
- Anterior skull base anomalies can present in nasal cavity
- Developmental lesions are common findings in the central skull base

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