Pediatric Neuroradiology
Interesting Cases

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Disclosures

I do not now and have not within the last 12 months had a financial interest or other relationship with a commercial organization that may have an interest in the content of the educational activity.

Nothing to hide!
Case 1: A 16-month-old boy with delayed milestones (unable to walk or support himself in a standing position) and spasticity of the lower extremities.
Initial Reading:

- Semilobar holoprosencephaly?
Further Observations

- The callosal body is absent
• The callosal body is absent
• The middle forebrain failed to lobate
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• The Sylvian f. is deviated anteriorly and is communicating across the midline over the vertex
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• The middle forebrain failed to lobate
• The Sylvian f. is deviated anteriorly and is communicating across the midline over the vertex
• Anterior and posterior intercerebral fissure are present
• The callosal body is absent
• The middle forebrain failed to lobate
• The Sylvian f. is deviated anteriorly and is communicating across the midline over the vertex
• Anterior and posterior intercerebral fissure are present
• The anterior horns are hypoplastic with absent septum pellucidum
Diagnosis
Middle Interhemispheric Variant

- 4th variant of holoprosencephaly
- Abnormal dorsal forebrain separation (failure of the roof plate function)

### MIV

### Classic HPE

- Alobar, semilobar, lobar
- Abnormal ventral forebrain separation (failure of the floor plate function)

Pulitzer, AJNR 2004  
Simon, AJNR 2002
Middle Interhemispheric Variant

**MIV**

- Hypothalamus/LFN are normally separated
- Low incidence of endocrinopathies, hypothalamic dysfunction and choreoathetosis

**Classic HPE**

- Hypothalamus/LFN are incompletely separated
- Facial induction and optic apparatus often affected

Pulitzer, AJNR 2004  
Simon, AJNR 2002
• The callosal body is absent
• The middle forebrain failed to lobate
• The Sylvian f. is deviated anteriorly and is communicating across the midline over the vertex
• Anterior and posterior intercerebral fissure are present
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Callosal Defects

Partial agenesis  Classic HPE  MIV
Take Home!
Middle Interhemispheric Variant

1. MIV should be differentiated from classic HPE
2. MIV has a low incidence of endocrinopathies, hypothalamic dysfunction and choreoathetosis

(Thomas Geissmann)
SAM1: Which one of the following structures is more often affected in the middle interhemispheric variant (MIV) when compared to classic holoprosencephaly (HPE)?

- A. Body of the corpus callosum
- B. Hypothalamus
- C. Anterior cerebral fissure
- D. The 3rd ventricle
- E. Ventral frontal cortex
SAM1: Which one of the following structures is more often affected in middle interhemispheric variant (MIV) when compared to classic holoprosencephaly (HPE)?

- A. Body of the corpus callosum
- B. Hypothalamus
- C. Anterior cerebral fissure
- D. The 3rd ventricle
- E. Ventral frontal cortex
Case 2: A 6-day-old full term male with generalized petechiae, tachypnea and grunting. His neurological exam was completely normal.
• A large hemispheric mass with subacute/chronic hematoma
• No contrast-enhancing nodule
• Minimal mass effect and surrounding edema
Operation/Pathology

• Hemorrhagic tumor was suspected
• A solid greenish mass tinged with brownish-red surrounded by fluid was evacuated
• Pathology revealed recent and remote hemorrhage without tumor
• The mother had anti-platelet antibodies and patient’s platelets were 7,000 at presentation
Diagnosis
Neonatal Alloimmune Thrombocytopenia

• Maternal-fetal platelet Ag incompatibility, analogous to Rhesus hemolytic disease
• 10% of neonatal thrombocytopenia (1/2,000-5,000)
• Widespread petechiae or purpura and intracranial hemorrhage in 20% (*in utero/neonatal*)
• Imaging Dx important for management and guidance for subsequent pregnancy (no formalized antenatal screening)
Imaging: Neonatal Alloimmune Thrombocytopenia

- Hemispheric cerebral hemorrhage (*in utero/ neonatal*), extra-axial/intraventricular hemorrhage
- Hemispheric porencephaly
Take Home!
Neonatal Alloimmune Thrombocytopenia

(Hematoma drainage may not be needed?)

1. NAIT is an important differential diagnosis of fetal and neonatal lobar hematoma, SDH and porencephalic cyst
2. Imaging Dx is important for management and guidance because no formalized antenatal screening is yet available
SAM 2: Which of the following statements regarding neonatal alloimmune thrombocytopenia (NAIT) is incorrect?

A. NAIT is caused by paternal-fetal antigen incompatibility.
B. NAIT accounts for approximately 9% of neonatal thrombocytopenia.
C. Intracranial hemorrhage occurs in 10 – 30% of NAIT, half of which occur in utero.
D. Parenchymal hemorrhage, extraaxial hemorrhage, intraventricular hemorrhage and porencephalic cysts are all seen in NAIT.
E. There is no cost-effective screening program of primiparous women and neonates for this disease.
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E. There is no cost-effective screening program of primiparous women and neonates for this disease.
Case 3: A 19-month-old girl with pharyngitis on antibiotics developed fever, dysphagia and vomiting.
• Deep neck collection extending into the mediastinum below T2
Diagnosis
Danger Space Abscess

- Deep neck collection extending into the mediastinum below T2
Anatomy: Danger Space Abscess

1. Retropharyngeal space (skull base-T2)  
   --Alar fascia--*

2. Danger space (skull base-diaphragm)  
   --Prevertebral fascia--*

3. Prevertebral space (skull base-coccix)

* Deep layer of DCF
Retropharyngeal Abscess

- Unique to infants and young children (6m-6yr)
- URI → retropharyngeal adenitis → abscess
- Retropharyngeal nodes are abundant in young children
- Mixed aerobic and anaerobic organisms
- In adults and older children, parapharyngeal and peritonsillar abscess are more common
“Danger-ness” of Danger Space Abscess

- Descending necrotizing mediastinitis
- Jugular vein thrombophlebitis/thrombosis
- Carotid artery rupture
- Pericarditis, tamponade
- Bronchial erosion, pyopneumothorax
Take Home!
Danger Space Abscess

1. Consider DSA when deep neck abscess extends below T2
2. You should suggest more aggressive treatment
3. Potentially fatal complications: mediastinitis, carotid a./jugular v. compromise, pericarditis and pyothorax

(Thomas Geissmann)
All the statements regarding infections of the deep neck are correct except:

A. The danger space is dorsal to the retropharyngeal space and ventral to the prevertebral space.

B. The danger space is separated from the retropharyngeal space by the alar fascia and from the prevertebral space by the prevertebral fascia.

C. The danger space extends from the skull base to the coccyx.

D. Retropharyngeal space abscess most commonly originated from lymphadenitis and seen predominantly in infants.

E. Parapharyngeal and peritonsillar abscesses are more common among adults and older children.
SAM 3: All the statements regarding infections of the deep neck are correct except:

A. The danger space is dorsal to the retropharyngeal space and ventral to the prevertebral space.
B. The danger space is separated from the retropharyngeal space by the alar fascia and from the prevertebral space by the prevertebral fascia.
C. The danger space extends from the skull base to the coccyx.
D. Retropharyngeal space abscess most commonly originated from lymphadenitis and seen predominantly in infants.
E. Parapharyngeal and peritonsillar abscesses are more common among adults and older children.
Case 4: An 8-year-old boy with 6-month history of progressive headache and bilateral abduction deficit/esotropia
• Diffuse sclerosis of the skull base involving the orbits and sinuses
• Orbital conal mass
- Enhancing orbital mass (T2-dark) extending to the cavernous sinus
- Pineal mass
• Foci of T2 prolongation in the cerebellar peduncle and brain stem
• Meta-/diaphyseal sclerosis of the long bones
- Diffuse osteosclerosis of the skull base
- Orbital mass extending to the cavernous sinus (dark on T2), pineal mass, parenchymal foci of T2 prolongation
- Meta-/diaphyseal sclerosis of the long bones
Differential Diagnosis

- Langerhans cell histiocytosis
- Non-Langerhans cell histiocytosis
  - Hemophagocytic lymphohistiocytosis
  - Rosai-Dorfman disease
  - Erdheim-Chester disease
- Lymphoma
- Inflammatory pseudotumor
Neurosurgery proposed cavernous sinus biopsy
Sinus wall biopsy

Chronic sclerosing inflammatory process with a mixed B- and T-lymphocyte infiltration and abundant macrophages. No Langerhans histiocytes are present.
Diagnosis
Inflammatory Pseudotumor

• Rare benign process mimicking malignancy
• Most commonly involve the orbits and lungs but found in nearly every site in the body
• Children and young adults are often affected
• A single or multiple inflammatory mass(es) with polymorphous inflammatory cells (B-/T-lymphocytes, plasma cells, myofibroblasts) and fibrosis
• Respond to steroid: accurate Dx avoids unnecessary surgery and other invasive interventions
Take Home!
Inflammatory Pseudotumor

1. IP can affects children and young adults
2. Multisystemic T1-dark, T2-dark mass(es)
3. Radiologists can prevent unnecessary surgery/intervention

(Thomas Geissmann)
Case 5: A 14-year-old girl presented for preoperative evaluation for scoliosis. She had 2-year history of low back pain.
- Holocord, intradural extramedullary mass
  - T1-iso, T2-hyper, avidly enhancing
  - Expansile, bone erosion, cord atrophy
Round to ovoid tumor cells with inconspicuous nucleoli. Vascular proliferation with pseudorosette formation. Mucinous degeneration of the stroma is highlighted with Alcian blue stain.
Diagnosis
Myxopapillary Ependymoma

- Compared to the more common cellular ependymoma, which occurs in the cervical cord of the older patients, ME occurs in the conus and filum of the younger patients.
- T1-iso, T2-hyper, homogeneously enhancing (occasionally T1-hyper in tumors with mucin).
- Expand spinal canal and neural foramina with bone erosion.
- CSF dissemination common in pediatric age (80% in Fassett series of 5 patients, 2005).
Take Home!
Myxopapillary Ependymoma

1. Consider ME when you see a large intradural extramedullary avidly enhancing thoracolumbar mass with bone erosion
2. Look for CSF dissemination in pediatric age
Case 6: A 7-year-old boy with 3-month history of headache and referred diagnosis of “chordoma”
- Asymmetric basisphenoid
- Nonexpansile bone lesion with osteosclerotic borders, internal fat and curvilinear calcifications
- Neural foramina are preserved
• Nonexpansile basisphenoid mass
• T1-hyper, T2-hyper ,+/- enhancement
Operation

• ENT-neurosurgery combined operation
• Sphenoid sinus was widely opened and the clival “mass” was removed
Fragments of bone, cartilage and respiratory type mucosa. The scant nature of the specimen precludes diagnosis. The differential diagnosis includes psammomatoid ossifying fibroma and meningioma. Recommend correlation with radiologic findings.
Diagnosis
Arrested pneumatization of the sphenoid sinus

- Located at the site of normal pneumatization, most common at the sphenoid sinus
- Nonexpansile basisphenoid lesion with osteosclerotic borders, internal fat and curvilinear calcifications
- T1-hyper, T2-hyper, Gad-variable
Pneumatization of the Basisphenoid

- Fatty marrow conversion of the presphenoid plate antedates to pneumatization
- T1 signal of the presphenoid plate:
  - Hypo- ⇔(2 yr) ⇔Hyper- ⇔(4 yr) ⇔pneumatize

(Yonetsu, 2000; Aoki, 1989)
Take Home!
Arrested Pneumatization of the Skull Base

Radiologists should have prevented surgery!!

(Thomas Geissmann)

- Benign developmental variant, nonexpansile basisphenoid lesion
- CT: Nonexpansile, osteosclerotic borders, internal fat and curvilinear calcifications
- MR: T1-hyper, T2-hyper
- TOUCH ME NOT!: Don’t mistake as chordoma, osteomyelitis, fibrous dysplasia
Case 1

- Dx: Middle Interhemispheric Variant
Case 2

T1                 T2                 Gred              Gad
Case 2

Dx: Neonatal Alloimmune Thrombocytopenia
Case 3
Case 3

- Danger Space Abscess
Case 4
Case 4

Dx: Inflammatory Pseudotumor
Case 5

Dx: Myxopapillary Ependymoma
Case 6

T1                     T2                       Gad
Case 6

Dx: Arrested pneumatization of the sphenoid sinus