• No financial disclosures
Dermoid Cyst

- Congenital
- Keratinized epidermis
- Dermal appendage
- Trapped during embryogenesis
  - 6% of lesions
  - 40-50% of orbital pediatric orbital lesion
  - Usually discovered in the first year of life
    - Painless/firm/subQ mass
    - Rarely presents as an acute inflammatory lesion (Rupture?)
  - **Frontozygomatic** (70%)
  - Maxillofrontal (20%) suture
Imaging - CT

- Erosion/remodeling of bone
- Adjacent bony changes: “smooth fossa” (85%)
- Dumbell dermoid: extraorbital and intraorbital components through bony defect
Imaging - MRI

- Encapsulated
- Enhancement of wall but not lumen
Treatment Options

- Observation
  - Risk of anesthesia
- Surgical Removal
  - Changes to bone
  - Rupture of cyst can lead to acute inflammation
    - Irrigation
    - Abx
    - Steroids
Dermoid
INFANTILE/Capillary Hemangioma

- Common BENIGN orbital lesion of children
- F>M
- Prematurity
- Appears in 1\textsuperscript{st} or 2\textsuperscript{nd} week of life
- Soft, bluish mass deep to the eyelid
  - Superonasal orbit
- Rapidly expands over 6-12 months
- Increases with valsalva (crying)
- Clinical findings
  - Proptosis Astigmatism
  - Strabismus Amblyopia
INFANTILE/Capillary Hemangioma

- May enlarge for 1-2 years then regress
- 70-80% resolve before age 7
- HIGH flow on doppler
- **Kasabach-Merritt Syndrome**
  - Multiple large visceral capillary hemangiomas
  - Sequestration of platelets into tumor
  - Consumptive thrombocytopenia
  - Supportive therapy and treat underlying tumor
- Complications
  - DIC
  - death
• Homogenous soft tissue mass that infiltrates throughout the orbit

• MRI: Serpiginous “Signal Voids” because of high flow rate (stimulated vs. unstimulated blood)

• Isointense to brain on pre-contrast
INFANTILE/Capillary Hemangioma Management

- Small lesions without visual compromise
  - Observation
  - 75% of lesions spontaneously resolve over 4-5 years
- Large lesions with risk of amblyopia
  - Treat
    - Intralesional steroids
      - Complications: skin depigmentation, fat atrophy, eyelid necrosis, CRAO
    - Interferon-alpha
    - Laser
    - oral steroids
    - **Propranolol**
    - Excision
Lymphatic malformation

• “Benign”
• Unencapsulated
• Hamartoma
• Composed of thinned-walled vascular channels
• Not well defined: diffuse, infiltrating
• Typically persist for life
• 10% may have involvement of other head and neck structures
  • Palatal mucosa
Lymphatic malformation

• Signs/Symptoms
  • Sudden proptosis
  • Spontaneous hemorrhage
    • “chocolate cyst”
  • Enlarge with URI
  • Enhances with contrast
  • Difficult to debulk
    • Defer surgery unless absolutely necessary
    • Drainage/Sclerotherapy/Sildenafil
Lymphatic malformation
Rhabdomyosarcoma

• Most common MALIGNANT orbital tumor in children
  • 1% of all biopsied masses
  • 4-5% of pediatric orbital masses
  • 40% of pediatric malignant orbital masses

• Average Age: 8
  • 75% in first decade
  • Up to 78 yo

• Females > Males

• Arises from pluriopotentiel mesenchymal cells

• NOT from striated EOM
Rhabdomyosarcoma

- Can originate primarily in orbit or surrounding sinus
  - Acute
  - Rapid proptosis (80-100%)
  - Unilateral
  - Superonasal location
  - Globe displacement (80%)
  - Ptosis (30-50%)
  - Eyelid swelling (60%)
    - 20% lid signs dominate
  - Pain (10%)
  - Palpable mass (25%)
Imaging

- **CT:** moderately well circumscribed but irregular, homogenous mass.
  - +/- adjacent bony destruction
  - Enhances with contrast

- **T1 MRI**
  - Isointense to EOM
  - Heterogeneous enhancement

- **T2 MRI**
  - Hyperintense to fat and muscle
Rhabdomyosarcoma – Four Types

• Embryonal
  • Most Common (80%)
  • Superonasal
  • Good survival rate

• Alveolar
  • Most Aggressive
  • Inferior Orbit

• Pleomorphic
  • Most Benign
  • Most rare
  • Typically in adults, extremeties not orbit

• Botryoid
  • Grapelike appearance
  • Extends to orbit from adjacent sinus or conjunctiva
# Rhabdomyosarcoma

## Management

- Biopsy/Surgical debulking
- Systemic evaluation to r/o metastatic dz (i.e. lung, brain, lymph nodes)
- Staging (Intergroup Rhabdomyosarcoma Study Group)
  - I  Localized disease, completely resected
  - II Microscopic disease remaining after biopsy
  - III Gross residual disease remaining after biopsy
  - IV Distant metastasis present at onset
- Chemotherapy and radiotherapy

## Prognosis

- Improved greatly since early 70’s:
  - 74% (Alveolar) 94% (Embryonal)
- Favorable location
- Tumor morphology
- Age of presentation
- Infants under 1 have worse prognosis
Metastatic Neuroblastoma

Most common met. orbital tumor in children
90% before age of 10
  Mean Age: 2
10-20% of neuroblastomas met. to orbit
  8% present with ophthalmic features
Usually from ADRENALS
  Also: retroperitoneal, mediastinum, neck
  60-72% in abdomen
Proptosis, ptosis, ecchymosis
Bilateral 60%
Paraneoplastic opsoclonus
## Metastatic Neuroblastoma

<table>
<thead>
<tr>
<th>Work up</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Palpate abdomen</td>
<td>• Radiation</td>
</tr>
<tr>
<td>• CT: orbit, neck, chest, abdomen and pelvis</td>
<td>• Chemotherapy</td>
</tr>
<tr>
<td>• Serology for catecholamines</td>
<td></td>
</tr>
<tr>
<td>• Bone Scan</td>
<td></td>
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<tr>
<td>• Bone marrow biopsy</td>
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</tbody>
</table>
Optic Nerve Glioma

• **Usually** benign
• First decade
• F>M
• 25-50% associated with neurofibromatosis 1

**Clinical Presentation**
• Gradual growth
• Painless
• Unilateral axial proptosis
• Vision loss
• APD
OPTIC NERVE GLIOMA

- Malignant ONG (glioblastoma)
  - Rare
  - Adult males
- Clinical Presentation
  - Massive swelling and hemorrhage of optic nerve head
  - Pain
- Treatment
  - Chemotherapy
  - High-dose radiation therapy
- Prognosis
  - Poor
OPTIC NERVE GLIOMA

Fusiform enlargement of ON with kinking
Optic Nerve Glioma

- Other ocular findings
  - Swollen optic disc
  - Retinochoroidal shunt vessels
  - Strabismus
  - Nystagmus
- Because of NF1 association, check:
  - Café-au-lait spots
  - Iris Lisch nodules
Optic Nerve Glioma

• Observation
  • Good vision
  • Follow closely

• Surgical excision
  • Goal is to isolate from chiasm
    • Lesions that involve chiasm and brain can be fatal
    • When hypothalamus is involved mortality increases from 5% to 50%

• Radiation
  • If tumor cannot be resected (in chiasm)

• Chemotherapy
  • May delay need for radiation
Optic nerve sheath Meningioma

- Benign neoplasms that originate from **arachnoid layer** of meninges
- **Women** (80%) in 3\textsuperscript{rd}-4\textsuperscript{th} decade
- Present
  - Gradual, unilateral painless vision loss
  - Optic atrophy, **optociliary shunts**
- Clinical association
  - Neurofibromatosis 2
  - People with NF-2 have a higher incidence but only a minority of meningiomas have NF-2
Optociliary shunt vessels

DDx:
- Optic nerve sheath meningioma
- CRVO
- Chronic Glaucoma
- Chronic papilledema
Optic nerve sheath Meningioma

“Tram tracking” on CT
Optic nerve sheath Meningioma

- Observation
- Radiotherapy
- Chemotherapy
- Surgery reserved for patients with intracranial extension or severe visual loss
Sphenoid wing meningioma

- Begins in arachnoid that lines the sphenoid
- Invades orbit from intracranial space
- Assoc with NF2
- CT:
  - Hyperostotic sphenoid bone
Sphenoid wing meningioma

• Treatment
  • Close observation
  • Surgical resection
  • Radiation
  • Hormone therapy (future)
Lacrimal Drainage System

12-15mm Lacrimal Sac

2mm vertical canaliculi

8-10 mm horizontal canaliculi with common canaliculus

12-18mm nasolacrimal duct

Valve of Hasner in inferior meatus

12-15mm Lacrimal Sac

Posterior medial Canthal ligament

Anterior medial Canthal ligament

Anterior and posterior Lacrimal crests form the Lacrimal sac fossa

Fay & Dolman
## Schirmer Testing

<table>
<thead>
<tr>
<th>Schirmer Testing</th>
<th>Basal Tear Secreation</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Schirmer I</td>
<td>• Topical anesthetic</td>
</tr>
<tr>
<td>• No topical anesthetic</td>
<td>• Schirmer strip for 5 min</td>
</tr>
<tr>
<td>• Schirmer strip for 5 min</td>
<td>• Normal = 10-15mm</td>
</tr>
<tr>
<td>• Tests basal and reflex tearing</td>
<td></td>
</tr>
<tr>
<td>• Normal &gt;10</td>
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</table>
Canalicular Probing
Dye Disappearance Test

- Fluorescein instilled into fornices
- Persistence of dye or asymmetric persistence at 5 min suggest obstruction
Jones Testing

- Dye instilled into fornix
- Jones 1 (physiologic)
  - Cotton tipped applicator at inferior meatus 2-5 min later
  - If negative test, proceed to Jones 2

- Jones 2 (non-physiologic)
  - Wipe dye from fornix
  - Irrigate with clear saline
  - No fluid on Q-tip = anatomic blockage
  - Clear fluid on Q-tip = lacrimal pump failure
  - Dye on Q-tip = functional obstruction
Lacrimal Irrigation

A. Same canaliculus clear fluid return:  
   Canalicular obstruction

B. Opposite canaliculus clear fluid return:  
   Common canalicular obstruction

C. Opposite canaliculus mucoid fluid return:  
   NLDO

D. Opposite canaliculus and nose clear fluid return:  
   Partial NLDO

E. No fluid return:  
   Patent NLD
Lacrimal Drainage Abnormalities

Congenital lacrimal-cutaneous fistula

- Intranasal to medial canthus
- Tearing from the skin
- 1/3 may have NLDO
- If simple without NLDO
  - direct surgical excision
- If NLDO
  - Add DCR

Aplasia/Hypoplasia/Stenosis

- Punctal aplasia/stenosis
  - Manage with probing and intubation
- Complete absence of canalicular system
  - CDCR (Jones tube)
Lacrimal Drainage Abnormalities

Congenital NLDO

- Membrane blocking valve of Hasner
- Canalization of NLD completed late in pregnancy or shortly after birth
- 20% of babies may have “watery eyes”
- 2-6% of babies will have clinically evident epiphora
- 95% resolve spontaneously in first year of life

Management

- Conservative management
  - Observation
  - Crigler lacrimal massage
  - Antibiotics for infections
- Surgical Management >1year
  - Probe
  - Probe and intubate
Lacrimal Drainage Abnormalities

Dacryocystocele

• Secondary to congenital NLDO with amniotic fluid collection
• Clinically swelling below the MCT
• Swelling above MCT
  • Meningoencephalocele
  • Dermoid cyst

Management

• Crigler lacrimal massage
• Probing
• If airway bilateral with obstruction
  • Urgent marsupialization
Conclusion

• Dermoid cyst is the most common orbital tumor of children
• Rhabdomyosarcoma is the most common malignant tumor of children
• Infantile hemangioms are effectively treated with propranolol
• Dacryocystocele below MCT – consider imaging if in doubt
  • Rule out meningoencephalocele

<table>
<thead>
<tr>
<th>Clinical Finding</th>
<th>Systemic Association</th>
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<tbody>
<tr>
<td>Sphenoid wing hypoplasia</td>
<td>NF1</td>
</tr>
<tr>
<td>Optic nerve glioma</td>
<td>NF1</td>
</tr>
<tr>
<td>Sphenoid wing meningioma</td>
<td>NF2</td>
</tr>
<tr>
<td>Optic nerve sheath meningioma</td>
<td>NF2</td>
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