Anatomy of the Sellar and Suprasellar Region

- Cerebral cortex
- Pineal gland
- Optic chiasm
- Pituitary
- Hypothalamus
Anatomy of the Sellar and Suprasellar Region

Coronal View

Sagittal View
Differential of Pediatric Neoplasms Involving the Optic Chiasm

- Most common tumors
  - Craniopharyngioma
  - Low Grade Glioma (WHO Grade I and II)

- Less common tumors
  - Germinoma
  - Pituitary Adenoma

Case 1

A 17 year old girl presented for a routine follow up visit in the setting of a partially resected craniopharyngioma 3 years prior.

She had a residual inferotemporal visual field defect in the left eye since the initial resection.

Examination Findings

<table>
<thead>
<tr>
<th></th>
<th>Right Eye</th>
<th>Left Eye</th>
</tr>
</thead>
<tbody>
<tr>
<td>Visual acuity</td>
<td>20/15</td>
<td>20/15</td>
</tr>
<tr>
<td>Pupils</td>
<td>Equally reactive, no rAPD</td>
<td></td>
</tr>
<tr>
<td>Color</td>
<td>Full</td>
<td>Full</td>
</tr>
<tr>
<td>Sensorimotor</td>
<td>No strabismus and full motility</td>
<td></td>
</tr>
</tbody>
</table>
Humphrey Visual Fields 24-2

OS
FL: 0/11
FP: 0%
FN: 3%

FL: 0/13
FP: 0%
FN: 13%

OD
FL: 0/11
FP: 2%
FN: 16%

FL: 1/15
FP: 5%
FN: 9%
MRI Coronal and Axial T1 post contrast
Diagnosis
Interval growth of craniopharyngioma with visual field loss

Treatment
Immediate gross total re-resection of craniopharyngioma
Continued eye exams every 6 months
Follow up MRI and Visual Fields
Pediatric Craniopharyngioma

- Most frequent suprasellar tumor in children ~ 3-15%
- Bimodal: pediatric peak <15 yrs, adult peak ~ 60-70 yrs
- Arises from epithelial remnant of embryonic tissue
- Associated with mutations in β-catenin (CTNNB1 gene) involved in the Wnt signaling pathway
- Overall survival rate is high
- Morbidity associated with visual dysfunction and endocrinopathy is significant

Clinical Presentation

- Presenting signs and symptoms
  - Headache 76% Chen et al., 2003; 31: 220-228.
  - Vision impairment 62- 84%
  - Endocrinopathy: growth delay 52- 87%
  - Nystagmus

Diagnosis

- Histologically
  - Adamantinomatous
- Neuroimaging features include
  - Heterogeneous on MRI with mixed solid and cystic features
  - May have calcifications

Treatment

- Gross or subtotal surgical excision
- With or without adjunctive radiotherapy
- Tumor recurrence rate is high

## Visual Prognosis in Children

<table>
<thead>
<tr>
<th>Study</th>
<th>N Pediatric patients</th>
<th>Mean Age (Years)</th>
<th>Post op vision &lt;20/40</th>
<th>Post op vision &lt;20/200</th>
<th>Visual Field Defect Post-op</th>
<th>Optic Atrophy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Repka et al. 1988</td>
<td>12</td>
<td>8.5</td>
<td>50%</td>
<td>-</td>
<td>50%</td>
<td>50%</td>
</tr>
<tr>
<td>Abrams and Repka 1997</td>
<td>31</td>
<td>7.7</td>
<td>3%</td>
<td>26%</td>
<td>65%</td>
<td>81%</td>
</tr>
<tr>
<td>Chen at al. 2003</td>
<td>17</td>
<td>10</td>
<td>33% (eyes)</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Guo and Heidary 2014</td>
<td>32</td>
<td>6.3</td>
<td>-</td>
<td>16%</td>
<td>87%</td>
<td>63%</td>
</tr>
</tbody>
</table>
Visual Prognosis in Children

• Risk factors for worse visual prognosis (Abrams and Repka 1997)
  - Poor pre-operative visual function
  - Age < 6 yrs at presentation

• Correlation between optic neuropathy and RNFL thinning using OCT (Bialer et al. 2013)

• Post-operative visual recovery less common* (Stark, Tychsen 1999)

• Sensory strabismus may occur: 86% (12/14 patients, Abrams and Repka, 1997)
Non Visual Morbidity

• Hypopituitarism

• Hypothalamic dysregulation
  - hypothalamic obesity

• Cognitive dysfunction

• Other neurologic sequelae
  - sensory deficits, hemiparesis
Considerations for Screening

• No consensus recommendations exist

• Consideration of continued ophthalmic monitoring because of high probability of tumor recurrence

• At BCH, we see patients every 6 months for complete ophthalmic examination
  - Visual acuity, color vision, pupillary exam
  - Visual fields (formal if possible)
  - Optic nerve assessment
  - Sensorimotor assessment
Case 2

A 11 month old girl presented with failure to thrive and a newly noted nystagmus.

Her medical history was uncomplicated until 6 months of age when she dropped from 21%ile to 8%ile in weight with normal height

Concurrently she was noted to have a delay in motor milestones. Otherwise her neurologic examination was non-focal.
## Examination Findings

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<thead>
<tr>
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<th>Right Eye</th>
<th>Left Eye</th>
</tr>
</thead>
<tbody>
<tr>
<td>Visual acuity (PLT)</td>
<td>20/94</td>
<td>20/1900</td>
</tr>
<tr>
<td>Pupils</td>
<td>Relative afferent pupillary defect left eye</td>
<td></td>
</tr>
<tr>
<td>Color</td>
<td>Unable</td>
<td>Unable</td>
</tr>
<tr>
<td>Visual Fields</td>
<td>By confrontation full with both eyes viewing</td>
<td></td>
</tr>
<tr>
<td>Sensorimotor</td>
<td>Full versions</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Sensory exotropia left eye</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Monocular nystagmus left eye</td>
<td></td>
</tr>
<tr>
<td>Posterior segment</td>
<td>Trace optic nerve pallor</td>
<td>Optic nerve pallor</td>
</tr>
<tr>
<td></td>
<td>Fovea normal</td>
<td>Fovea normal</td>
</tr>
</tbody>
</table>
MRI Sagittal and Coronal T1 post contrast
Diagnosis and Management

• Patient underwent subtotal tumor debulking with pathology confirming low grade pilocytic astrocytoma

• Adjuvant chemotherapy

• Interval growth of tumor since cessation of chemotherapy treated with second tumor debulking

• Visual exams every 2-3 months with focus on visual progress: patching right eye, works with teacher for visual impairment
Low Grade Gliomas

• Most common pediatric brain tumor ~18%
  - Optic pathway 3-5%

• Clinical signs and symptoms suggestive of chiasmal/hypothalamic involvement
  - Diencephalic syndrome (failure to thrive)
  - Nystagmus
  - Endocrinologic dysfunction
    - Short stature
    - Precocious puberty

• Role of the ophthalmologist
  - Evaluate for stigmata of NF1
  - Monitor visual function

Management of Sporadic Optic Pathway Gliomas

- Eye exams
  - Every 3 months after the initial diagnosis
  - Tumor progression is common

- Concerns for treatment
  - Progressive loss of vision (2 lines or worse)
  - Worsening of visual fields
  - Radiographic progression

- Current Treatment
  - Observation
  - Chemotherapy
  - Surgical debulking
  - Radiotherapy (*non NF1 associated OPGs)
Visual Impact of Optic Pathway Gliomas

- Approximately 1/3 to 1/2 cause vision loss in NF1

- Risk factors for poorer visual outcome
  - Sporadic etiology
  - More posterior tumor location (Balcer et al., 2001; 131: 442-445.)
  - Younger age at diagnosis
Visual Impact of Optic Pathway Gliomas

• Visual outcomes
  - Thiagalingam et al. 2004 of 54 children with NF1 OPGs, 31.5% of patients had profound visual impairment
  - Campagna et al. 2010 found that 56% of children with sporadic OPGs showed visual decline 6 yrs following dx

• Treatment outcomes with chemotherapy

<table>
<thead>
<tr>
<th></th>
<th>N patients</th>
<th>NF1 or Sporadic</th>
<th>Stabilized Vision</th>
<th>Improved Vision</th>
<th>Progressive Decline</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fisher et al. 2012</td>
<td>115</td>
<td>NF1</td>
<td>40%</td>
<td>32%</td>
<td>28%</td>
</tr>
<tr>
<td>Moreno et al. 2010</td>
<td>174</td>
<td>Both</td>
<td>47.1%</td>
<td>14.4%</td>
<td>38.5%</td>
</tr>
</tbody>
</table>

• Risk factors for worse prognosis in NF1-OPGs
  - Age <2 and > 5 years and optic atrophy Fisher et al. 2012;14:790-7.
Role of the Pediatric Ophthalmologist

• Surveillance
  - Identification of children who harbor tumors affecting the visual pathways

• Management
  - Active monitoring of visual function and contribution towards treatment plan

• Visual Prognosis