Pediatric Ocular Tumors

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No financial interests

Retinoblastoma
CHRPE-like FAP
CHRPE

Dead 1 year
Dead 28 years
No death but blindness
No death no blindness

Pediatric Ocular Tumors
- Eyelids
- Conjunctiva
- Intraocular
- Orbit

Capillary hemangioma of infancy
- 5%-10% of all newborns
- eyelids and orbit complications
  - amblyopia
  - Strabismus
- Placental metastasis

Children with cardiomyopathy from capillary hemangioma treated with propranolol
1 day – tumor softer
1 mo – tumor smaller

Capillary hemangioma of infancy
corticosteroid treatment
- systemic
  - 4-6 mg/kg prelone or orapred
- local injection
  - triamcinolone 40 mg
  - betamethasone 6 mg
- Topical

Propranolol
- Systemic
  - 2 mg/kg
- Watch cardiac output
- Topical

Placental metastasis

Propranolol 2 mg/kg
Watch cardiac output
**Capillary Hemangioma Syndromes**

Kasabach Merritt Phenomenon
- platelet sequestration
  - hemolytic anemia
  - thrombocytopenia
  - coagulopathy
  - death

PHACE syndrome
- large facial hemangioma
- brain hemangioma
- death

**PHACE**

Consensus Statement on Diagnostic Criteria for PHACE Syndrome

**Eyelid Tumors**

Kissing nevus
- Lids fused week 9-20 gestation
- Melanocytes differentiate

Best to treat by 2-3 wks after birth with curettage

**Topical Timolol for Periorbital Hemangioma: Report of Further Study**

Children with superficial capillary hemangiomas of the eyelid may lead to amblyopia or anisometropia. Although benign, such tumors can cause irreversible visual loss if not treated promptly. Treatment options for infantile hemangiomas include both systemic and topical agents.

Nina Ni, BA
Paul Langer, MD
Rudolph Wagner, MD
Sueyin Goo, MD

Current strategy:
- If infant and flat - topical timolol bid
- If infant and thick - oral propranolol 2mg/kg
Pediatric Eyelid Tumors

Pilomatrixoma
calcifying epithelioma of Malherbe
clinical
• children or young adults
  • 40% age 10 yrs or under
• upper eyelid and brow
• movable subcutaneous nodule
• solitary
• pink to purple color

Pediatric Eyelid Tumors

Infrabrow incision
adherent to skin
complete removal

Pediatric Eyelid Tumors

Juvenile xanthogranuloma
• infancy
• rapid growth
• involution
• steroids

Pediatric Eyelid Tumors

Basal cell carcinoma
Gorlin Goltz syndrome

Pediatric Ocular Tumors

• Eyelids
• Conjunctiva
• Intraocular
• Orbit
Pediatric conjunctival tumors

- Children
- Adults

Conjunctival Tumors Children

- 262 tumors children
- Benign 97%
- Malignant 3%

Conjunctival Tumors Children

- Diagnosis
- Nevi 64%
- Dermolipoma 5%
- Lymphangioma 3%
- Capillary hemangioma 3%
- Dermoid 2%

Melanoma 2%
Lymphoma 1%
Conjunctival Nevus

- Discrete
- Elevated
- Cysts
- Pigmented
- Stationary

Cysts nevus 65%

Correlation with anterior segment OCT

Tumor color
darker 5%
lighter 8%

Growth into melanoma <1%
Beware the nevus
- Fornix
- Tarsus
- PAM
- Enlargement
- Recurrence after excision
- Family history of skin melanoma

Many children have excision and cryo by age 25 yrs
Pediatric Conjunctival Tumors

Conjunctival Tumors Children

Diagnosis
Nevus 64%
Dermolipoma 5%
Lymphangioma 3%
Capillary hemangioma 3%
Dermoid 2%

Goldenhar syndrome
- Ear
- Nose
- Soft palate
- Lip
- Mandible
- Scoliosis
- Hearing loss 10% bilateral

Conjunctival Dermolipoma

Choristoma
Fornix
- Superotemporal
- Lateral
Conjunctival fold
Goldenhar’s syndrome

Conjunctival Lymphangioma

Hemorrhage
Dilated clear lymphatics
Check palate
Contiguous orbital component
Systemic lymphangioma
- Bone fractures
- Cysts
Observation
Conjunctival Lymphangioma

- Known for 20 years
- Hemorrhage cleared over 3 months

Conjunctival Tumors Children

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Percentage</th>
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<tbody>
<tr>
<td>Nevus</td>
<td>64%</td>
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<tr>
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<td>5%</td>
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<td>Lymphangioma</td>
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<tr>
<td>Capillary hemangioma</td>
<td>3%</td>
</tr>
<tr>
<td>Dermoid</td>
<td>2%</td>
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Conjunctival Capillary Hemangioma

- Presents shortly after birth
- Red patch
- No hemorrhage
- Often skin involvement
- Rare intraocular involvement
- Observation

Conjunctival Dermoid

- Choristoma
- Inferotemporal limbus
- White spot
- Noted at birth
  - Feeder vessel
  - Hair
  - Corneal lipid ring later

Conjunctival Tumors Children

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<td>Capillary hemangioma</td>
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<tr>
<td>Dermoid</td>
<td>2%</td>
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Papilloma <1%
**Comparing nevus vs melanoma in 510 cases in children**

**Factors for conjunctival melanoma in children**
- Age older
- Thickness/Base greater
- Cyst absent
- Hemorrhage

<table>
<thead>
<tr>
<th>Factor</th>
<th>Nevus</th>
<th>Melanoma</th>
<th>p value</th>
<th>RR</th>
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</thead>
<tbody>
<tr>
<td>Age</td>
<td>12</td>
<td>15</td>
<td>0.0013</td>
<td>4.8 &gt;15 yr</td>
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<tr>
<td>Race</td>
<td>83% white</td>
<td>89% white</td>
<td>na</td>
<td>na</td>
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<tr>
<td>Sex</td>
<td>43% female</td>
<td>67% female</td>
<td>na</td>
<td>na</td>
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<tr>
<td>Location</td>
<td>49% limbus</td>
<td>56% limbus</td>
<td>na</td>
<td>na</td>
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<tr>
<td>Color</td>
<td>51% brown</td>
<td>50% brown</td>
<td>na</td>
<td>na</td>
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<tr>
<td>Base</td>
<td>4.7 mm</td>
<td>5.8 mm</td>
<td>0.01</td>
<td>4.92 per ≥10 mm</td>
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<tr>
<td>Thickness</td>
<td>1.1 mm</td>
<td>1.3 mm</td>
<td>0.004</td>
<td>1.1 per ≥1 mm</td>
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<tr>
<td>Cyst</td>
<td>67%</td>
<td>28%</td>
<td>0.0013</td>
<td>0.1</td>
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<tr>
<td>Feeder vessel</td>
<td>49%</td>
<td>72%</td>
<td>na</td>
<td>na</td>
</tr>
<tr>
<td>Hemorrhage</td>
<td>11%</td>
<td>17%</td>
<td>0.0001</td>
<td>25.3</td>
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</tbody>
</table>
Comparison of nevus vs melanoma in 510 cases in children

Factors for conjunctiva melanoma in Children
  - Age older
  - Thickness/Base greater
  - Cyst absent
  - Hemorrhage

**CATCH melanoma**

<table>
<thead>
<tr>
<th></th>
<th>BRLH</th>
<th>Lymphoma</th>
<th>p value</th>
<th>RR</th>
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<tbody>
<tr>
<td>Age</td>
<td>12</td>
<td>12</td>
<td>ns</td>
<td></td>
</tr>
<tr>
<td>Race</td>
<td>74% white</td>
<td>56% white</td>
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<tr>
<td>Sex</td>
<td>34% female</td>
<td>33% female</td>
<td>ns</td>
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<tr>
<td>Configuration</td>
<td>0% diffuse</td>
<td>22% diffuse</td>
<td>0.03</td>
<td>8.5</td>
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<tr>
<td>Location</td>
<td>82% nasal</td>
<td>22% diffuse</td>
<td>0.03</td>
<td>16.5</td>
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<tr>
<td>Base</td>
<td>8.2 mm</td>
<td>17.3 mm</td>
<td>0.0016</td>
<td>5.2 &gt;10mm</td>
</tr>
<tr>
<td>Thickness</td>
<td>2.0 mm</td>
<td>2.0 mm</td>
<td>ns</td>
<td></td>
</tr>
</tbody>
</table>

Comparison of BRLH vs lymphoma in 47 cases in children

Pediatric Ocular Tumors
  - Eyelids
  - Conjunctiva
  - Intraocular
  - Orbit

Pediatric Ocular Tumors
  - medulloepithelioma
  - retinoblastoma
  - rpe tumors
  - nevus melanoma
cyst
Pediatric Intraocular Tumors

Iris stromal cyst
Rupture
Anterior uveitis
Management
• Observation
• Needle aspiration
• Needle aspiration plus
  • alcohol
  • mitomycin
  • cryotherapy
  • Excision

If < 10 yrs more aggressive
If > 10 yrs less aggressive

Iris pigment epithelial cyst

Pediatric Intraocular Tumors

Iris nevus
Iris melanoma

Metastasis KM
@ 5yrs 5%
@ 10 yrs 9%
@ 20 yrs 11%

Pediatric Intraocular Tumors

Choroidal nevus
Choroidal melanoma

mm
Choroidal Melanoma

**Ocular melanocytosis**
- Skirf
- Sclera
- Uvea
- Orbit
- Meninges
- Palate

1/400 uveal melanoma

**Congenital melanocytosis**

- Skin
- Sclera
- Uvea
- Orbit
- Meninges
- Palate

**Pediatric Intraocular Tumors**

**Medulloepithelioma**
- Embryonal tumor of npce
- Clinical onset first decade
- Fleshy cb mass with cysts
- Leukocoria, cataract, glaucoma
- Nonteratoid and teratoid types
- Benign or malignant
- Nonhereditary
- Rarely metastasizes

**Uveal melanoma**
- Tumor diameter smaller
- Tumor thickness smaller
- Extraocular extension less
- Iris location more often
  - Kids 21% vs 7% older adults
- Metastasis less
  - Kids 9% vs 5%
  - Mid-adults 23% vs 11%
  - Older adults 28% vs 16%
- Uveal melanoma in 8033 eyes
  - Kids
  - Mid-adults
  - Older adults
  - Many statistically significant differences
- Uveal melanoma in children
  - Different features
  - Different locations
  - Different risk for metastasis and death

**Clinical Spectrum and Prognosis of Uveal Melanoma Based on Age at Presentation in 8,033 Cases**

- @10 years
  - Kids: 9%
  - Mid-adults: 23%
  - Older adults: 28%

- Death
  - Kids: 5%
  - Mid-adults: 11%
  - Older adults: 16%
Congenital hypertrophy RPE
- Solitary
- Multifocal
CHRPE-like lesions FAP
Torpedo maculopathy
Congenital simple hamartoma
Combined hamartoma
RPE adenoma adenocarcinoma
RPE hyperplasia

Pediatric Intraocular Tumors
CHRPE-like lesions with Familial Adenomatous Polyposis
- ≥ 4 lesions
- irregular margins
- white tail of depigmentation
- highly suggestive of FAP-EM (Gardner’s)

Torpedo Maculopathy
Small Parfoveal Feeder vessel
Stable

Associated with NF2
Management
• observation
• vitrectomy and membrane peeling for severe traction

Retinoblastoma
• most common intraocular malignancy of childhood
• 1:15,000 live births
• hereditary or non hereditary
• unilateral or bilateral
• second cancers

International classification RB
Group
A small ≤ 3mm
B bigger > 3mm,macula,srf
C contained seeds
D diffuse seeds
E extensive
  >50% globe
  NVI
  opaque media

International classification RB
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A small ≤ 3mm
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**Retinoblastoma**

- **Management**
  - enucleation
  - external beam irradiation
  - plaque brachytherapy
  - laser photocoagulation
  - cryotherapy
  - intravenous chemoreduction
  - intraarterial chemotherapy
  - intravitreal chemotherapy
  - chemothermotherapy
  - gene therapy
  - genetic counseling

- **Chemotherapy**
  - Intravenous
  - Subtenons
  - Intra-arterial
  - Intravitreous

- **Chemotherapy RB**
  - vincristine
  - etoposide
  - carboplatin

- **High risk**
  - Optic nerve postlam
  - chor inv >3mm
  - ant chamber inv
  - any or chor inv

- **Adjuvant chemotherapy**
  - vincristine
  - etoposide
  - carboplatin

100% survival

- **International classification RB**

<table>
<thead>
<tr>
<th>Group</th>
<th>All</th>
<th>Greater than 3mm, macula, srf</th>
<th>Contained seeds</th>
<th>Diffuse seeds</th>
<th>Extensive &gt;50% globe, NVI, opaque media</th>
</tr>
</thead>
</table>
Success of chemoreduction for RB in 250 eyes:

- A: 100%
- B: 93%
- C: 90%
- D: 47%

ICRB group:

- A: Intravenous
- B: Subtenons
- C: Intra-arterial
- D: Intravitreous

Chemotherapy RB:

- Melphalan
- Topotecan

Before Intra-arterial chemotherapy RB

After Intra-arterial chemotherapy RB
Chemotherapy RB
- Intravenous
- Subtenons
- Intra-arterial
- Intravitreous

Retinoblastoma
- Intravitreal melphalan

Pediatric Orbital Tumors
- Cystic
- Vascular
- Inflammatory
- Myogenic

80% pediatric orbital tumors
Pediatric Orbital Tumors

Cystic
Vascular
Inflammatory
Myogenic

Benign 92%
Malignant 8%

Dermoid Cyst
- Extraorbital
- Intraosseous
- Intraorbital
- Dumbell

Capillary hemangioma
- Infancy
- Observation
- Steroids
- Propranolol

Lymphangioma
- Pseudotumor
- Myositis
- Rhabdomyosarcoma

Serious adverse drug rxn - 3%
- Bradycardia
- Bronchospasm
- Hypoglycemia

Less severe drug rxn - 2%
- Nightmares
- Acycyanosis
- Hypotension
- Diarrhea

n=906 patients
Orbital lymphangioma

Contrast shows cyst and wall
Multilobulated
Fluid levels
• Best seen T2
• Can be subtle

Management
• Observation
• Aspiration
• Aspiration + tissueal
• Sildenafil
• Sclerotherapy
• Resection

Case #1

Lymphangioma Sildenafil

7 patients
2.6 injections
No effect on vision
Pediatric Orbital Tumors

Cystic
Vascular
Inflammatory
Myogenic

- pseudotumor myositis
- lupus or jra
- eosinophilic granuloma
- langerhans histiocytosis
- wegener's
- churg strauss
- microscopic polyangitis
- polyarteritis nodosa
- sarcoid
- tb
- kimura's disease
- angiolymphoid hyperplasia eos

If recurrent or persistent, think:
- rheumatologic histiocytic
- vasculitic inflammatory

Rhabdomyosarcoma

- Classification:
  I  no residua
  II micro residua
  III gross residua
  IV mets

- Treatment:
  chemo
  chemo+rad
  chemo+rad

- Goal at surgery complete resection without damaging normal structures

Lifetime Mortality from Ped CT

- If 1 year old child ...
  - Head CT  0.07%
  - Abd CT  0.18%

- Sounds little, but

- If 600,000 abd+head CTs done annually in kids <15 yrs, roughly 500 would ultimately die of radiation-induced cancer


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