The authors have no financial interests in the materials in this presentation.

Lumps Bumps and Magic Potions
Review of Ocular Tumors

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Eyelid Tumors
- Benign tumors of the epidermis
- Malignant tumors of the epidermis
- Glandular and adnexal tumors
- Melanocytic tumors
- Vascular tumors
- Neurogenic tumors
- Lymphoid tumors
- Xanthomatous tumors
- Metastatic tumors
- Lacrimal drainage system tumors

Papilloma
- Rough surface
- Sessile or pedunculated
- Cerebriform surface
- Management
  - observation
  - excision

Seborrheic Keratosis
- Circumscribed
- Rough-surfaced
- Elevated
- Brown to gray lesion
- Pasted button on skin
- Management
  - observation
  - excision

Ocular Tumors
- Eyelids
- Conjunctiva
- Intraocular
- Orbit
Eyelid Tumors

Malignant eyelid tumors
- Basal cell ca 90%
- Sebaceous ca 4%
- Squamous cell ca 4%
- Melanoma 1%

... but in India, sebaceous carcinoma is most common malignant eyelid tumor

Basal Cell Carcinoma

- Types
  - Noduloulcerative
  - Morpheaform
  - Pigmented
  - Cystic
- Eyelid margin
- Margins
  - Well defined
  - Poorly defined
- Central ulcer
- Loss of cilia

Management
- Excision
- Frozen sections
- Mohs surgery
- Closure
  - primary closure
  - skin flap
  - skin graft
- Cryotherapy
- Imiquimod 5% (Aldara)
- Interferon injection
- Radiotherapy
- Orbital exenteration

Imiquimid
Cream
Apply daily for 8 to 16 weeks
Stimulates intrinsic interferon

Immunotherapy With Imiquimod 5% Cream for Eyelid Nodular Basal Cell Carcinoma
Maria Antonietta Blasi, MD,
Daniela Gimmaria, MD,
and Emilio Ballestrazi, MD

International Society Ocular Oncology
Cambridge England 2009

Alberto Calle from Colombia

If < 2cm
1 mill unit 3xwkly x 3 weeks
If > 2cm
3 mill unit 3xwkly x 3 weeks

N=64 BCC
100% regression

Orbital exenteration
Sebaceous Carcinoma

- 4% of malignant eyelid tumors
- Metastasis
  - Lymph nodes
  - Distant organs
- Origin
  - Meibomian glands
  - Zeis glands
  - Caruncle
  - Multicentric

Frequent misdiagnosis

Growth pattern
- Nodular
- Diffuse

Metastasis [n=60 pts] mean 4 yrs
- No mets 90%
- Mets 10%
- Interval to mets 20 mo
- Site mets Initially lymph node 100%

Management
- Local resection
- Cryotherapy
- IAT
- Orbital exenteration

Melanocytic Tumors
- Several different types
- Different appearance
- Different implications
Melanocytic Tumors
- Nevus
- Kissing nevus
- Nevus of Ota
- Lentigo maligna
- Malignant melanoma

Nevus
- Common
- Benign
- No loss of cilia
- Variably pigmented
- <1% risk to melanoma
- Observation

Kissing Nevus
- Rare
- Benign congenital
- Lids fused 9-20 week gestation
- Best to treat by 2-3 weeks after birth with curettage
- 9 x greater risk for melanoma

Nevus of Ota
- Rare
- Benign congenital
- Lids fused 9-20 week gestation
- Best to treat by 2-3 weeks after birth with curettage
- 9 x greater risk for melanoma

Lentigo Maligna
- Melanotic freckle of Hutchinson
- Acquired skin pigmentation
- Risk for skin melanoma ~ 30%
- Associated conjunctival PAM and risk for conj melanoma

Malignant Melanoma
- Variably pigmented eyelid nodule
- Progressive growth
- Can metastasize to regional nodes and systemically
Ocular Tumors

- Eyelids
- Conjunctiva
- Intraocular
- Orbit

Conjunctival pingueculum

Conjunctival pterygium

Conjunctival Tumors

- Pigmented Conjunctival Tumors
  - Racial melanosis
  - Primary acquired melanosis
  - Secondary acquired melanosis
  - Nevus
  - Melanoma
  - Metastasis
  - Simulating lesions

- Malignant
  - Melanoma
  - Squamous cell ca
  - Lymphoma

Racial Melanosis

Non caucasians
Flat perilimbal pigment
Bilateral
Symmetric
No risk for melanoma
No treatment

... However, be aware that conjunctival melanoma can occur in noncaucasians

Melanocytic Conjunctival Tumors

Diagnoses
- Racial melanosis
- Primary acquired melanosis
- Secondary acquired melanosis
- Nevus
- Melanoma
- Metastasis
- Simulating lesions

Primary Acquired Melanosis

- Middle age
- Light complexion
- Unilateral
- Flat
- No cysts
- Risk for melanoma

Progression to melanoma
AFIP WEH
Overall 32% 9%
PAM without atypia 0% 0%
PAM with atypia 46% 13%

Atypia
None 0%
Mild 2%
Severe 32%
Primary Acquired Melanosis

- 1 clock hr
- 4 clock hrs

1 clock hr = 1
4 clock hrs = 7 x greater risk

Suspicion moderate

Primary Acquired Melanosis

Conjunctival Nevus

- Children and young adults
- Discrete
- Elevated
- Cysts
- Variably pigmented
- Stationary

Conjunctival Melanoma

Origin
- PAM 70%
- Nevus 15%
- De novo 15%

Melanocytic Conjunctival Tumors

Diagnoses
- Racial melanosis
- Primary acquired melanosis
- Secondary acquired melanosis
- Nevus
- Melanoma
- Metastasis
- Simulating lesions

Good prognosis
- Localized
- Limbus
- Bulbar
- Thin
Conjunctival Melanoma

Poor prognosis
- Large
- Diffuse
- Fornix
- Caruncle
- Tarsus

Local recurrence
Metastasis
- Lymph nodes
- Brain
- Lung

At 3 months:
Custom designed plaque radiotherapy for diffuse recurrent melanoma
Squamous Cell Carcinoma
- Mass
- Vascular
- Limbus
- Feeder vessels
- Intrinsic vessels
- Sun exposure

Squamous Cell Carcinoma
- Small
- Intermediate
- Large
- Giant

Squamous Cell Carcinoma
- Surgical Resection
- Before
- After
- Treatment Conjunctival SCC
- Surgical
  - Alcohol epitheliectomy
  - Partial tarsal sclerocorneectomy
  - Cryotherapy
  - Closure
- Non-surgical
  - Topical MMC
  - 5FU
  - Interferon
  - Cidofovir
  - Injection interferon
  - Photodynamic therapy

Squamous cell carcinoma treated with 1 cycle of MMC

Lymphoid Tumors
- Systemic lymphoma by 30 years
  - If unilateral: 17%
  - If bilateral: 50%

Both
- Ocular treatment
- Systemic monitoring ... for life
Lymphoma treated with Rituxan
before
after 4 months

Ocular Tumors

Eyelids
Conjunctiva
Intraocular
Orbit

Intraocular Tumors

• Retinoblastoma
• Choroidal nevus
• Choroidal melanoma
• Choroidal metastasis

Chemotherapy RB

• Intravenous
• Subtenons
• Intra-arterial
• Intravitreal

Intravenous Chemotherapy

Intra-arterial Chemotherapy

IAC
Oncology Service
Wills Eye Hospital

Before IAC
After IAC

Enucleation RB

Minimal manipulation
Hydroxyapatite implant
- Scleral wrap
- Polymer coated
- 4 muscles attached

Enucleation

Fresh tissue harvest

Esther Ko RB

12/02 preop
1/03 postop

Enucleation

Implant for life

Mysteries of intraocular tumors
- Retinoblastoma
- Choroidal nevus
- Choroidal melanoma
- Choroidal metastasis

Choroidal Nevus
- 7% white population
- Brown gray mass
- Thickness < 2 mm
- Drusen
- RPE
  - atrophy
  - hyperplasia

Growth to melanoma
- 1/8000 of all choroidal nevi
- 0.2% lifetime risk adjusted
- 0.8% lifetime risk by 80 yrs
1% lifetime risk by 80 yrs

How can we identify those nevi that might convert to melanoma?

- Thickness > 2 mm
- Fluid (SRF)
- Symptoms
- Orange pigment
- Margin at optic disc

Lesions < 3 mm

To find small ocular melanoma
- Thickness > 2 mm
- Fluid (SRF)
- Symptoms
- Orange pigment
- Margin at optic disc

Uveal Melanoma

Mortality by size
- small < 3 mm 12%
- medium 3-8 mm 25%
- large > 8 mm 50%

Prognosis of uveal melanoma

2009 Shields
n=8033 pts
Long-term metastasis 4 decades
All locations All sizes

Choroidal Melanoma

Each mm increase adds ~ 5% increased risk for metastasis

2009 Shields
n=8033 pts
Long-term metastasis 4 decades
All locations All sizes

Choroidal Nevus

1986

1988

1989

2004

Judge nevus vs melanoma

High risk for growth

- Thickness > 2 mm
- Subretinal fluid
- Symptoms
- Orange pigment
- Margin @ disc

Judge nevus vs melanoma

Low risk for growth

- Thickness > 2 mm
- Subretinal fluid
- Symptoms
- Orange pigment
- Margin @ disc

- Drusen

Uveal Melanoma

Mortality
- Monosomy 3 0%
- Disomy 3 55%
- Monosomy 3, 8q gain 71%
Uveal Melanoma
Management
- transpupillary thermotherapy (TTT)
- plaque radiotherapy
  - local resection
  - enucleation
  - exenteration
  - photodynamic therapy
  - anti-VEGF
  - systemic therapies

Thermotherapy
- 3.5 mm
- 1.0 mm

Plaque radiotherapy
- iodine seeds
- custom design
  - round
  - notched
  - curvilinear
  - rectangular
- 8000 cGy apex

Plaque radiotherapy plus TTT

Mysteries of intraocular tumors
- Retinoblastoma
- Choroidal nevus
- Choroidal melanoma
- Choroidal metastasis
Estimated US Cancer Cases 2003

<table>
<thead>
<tr>
<th>Cancer Type</th>
<th>Estimated Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Prostate</td>
<td>33%</td>
</tr>
<tr>
<td>Lung</td>
<td>14%</td>
</tr>
<tr>
<td>Colon rectum</td>
<td>11%</td>
</tr>
<tr>
<td>Bladder</td>
<td>6%</td>
</tr>
<tr>
<td>Melanoma skin</td>
<td>4%</td>
</tr>
<tr>
<td>Non-Hodgkin lymphoma</td>
<td>4%</td>
</tr>
<tr>
<td>Other Sites</td>
<td>28%</td>
</tr>
<tr>
<td>Breast</td>
<td>32%</td>
</tr>
<tr>
<td>Lung</td>
<td>12%</td>
</tr>
<tr>
<td>Colon rectum</td>
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</tr>
<tr>
<td>Uterus</td>
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</tr>
<tr>
<td>Ovary</td>
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</tr>
<tr>
<td>Non-Hodgkin lymphoma</td>
<td>4%</td>
</tr>
<tr>
<td>Other Sites</td>
<td>30%</td>
</tr>
</tbody>
</table>

Breast                33%
Lung                   14%
Gastrointestinal tract  6%
Kidney                 4%
Skin (Melanoma)        4%
Prostate               2%
Others                  4%

Clinical features:
- Yellow mass with subretinal fluid
- Unifocal or multifocal
- Location behind equator
- Simulate nevus
- Melanoma
- Scleritis

Uveal Metastasis

Fine needle aspiration biopsy

Treatment:
- Chemotherapy
- Cyberknife
- Plaque
- Laser
- PDT

Choroidal metastasis life prognosis ~1 year
Orbital Tumors in Children and Adults

- Children
- Adults

Pediatric Orbital Tumors
- Cystic
- Vascular
- Inflammatory
- Malignant

Orbital dermoid cyst
- Solitary
- Fissure
- Origin
- Skin
- Conjunctiva
- Features
- Rupture
- Fistula
- Ancient

Intraosseous dermoid
Dumbell dermoid

Survey of 1264 Patients with Orbital Tumors and Simulating Lesions

The 2022 Montgomery Lecture, Part I

<table>
<thead>
<tr>
<th>Cystic</th>
<th>Vascular</th>
<th>Inflammatory</th>
<th>Malignant</th>
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<tbody>
<tr>
<td>Dermoid cyst</td>
<td>Capillary hemangioma</td>
<td>Lymphangioma</td>
<td>Pseudotumor</td>
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<tr>
<td></td>
<td></td>
<td></td>
<td>Rhabdomyosarcoma</td>
</tr>
</tbody>
</table>
Pediatric Orbital Tumors

Cystic
Vascular
Inflammatory
Malignant

Capillary hemangioma infancy

- 10% of newborns
- 20% premature children
- more common in twins
- Skin
- Kassabach Merritt syndrome
- PHACE syndrome
- Pathogenesis placental emboli

Management
- MRI
- Biopsy
- Treatment:
  - Steroids - oral vs local
  - Interferon
  - Radiation
  - Resection
  - Propranolol

Orbital lymphangioma

Immediately after surgery

Pediatric Orbital Tumors

Cystic
Vascular
Inflammatory
Malignant

Shania Lawrence 0112
Orbital pseudotumor

Myositis
Pain on eye movement
Recurrence
Treatment
- Steroids
- NSAIDS
- Kenalog injection

Pediatric Orbital Tumors

Cystic
Vascular
Inflammatory
Malignant

Rhabdomyosarcoma

- Most common primary orbital malignancy of childhood
- Mean age 8 years
- Rare congenital or onset in adulthood
- Metastasis 10%
- Cure rate >90%

Orbital tumors of adults

Orbital Tumors of Adulthood

Most common types

- Inflammatory
- Benign
- Malignant

Idiopathic orbital inflammation

Pseudotumor
- Adults > children
- Acute or subacute onset
- Pain
- Eyelid conjunctival edema
- Ptosis
- Proptosis

Management
- Biopsy
- Chemotherapy
- Irradiation

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

Recurrence

Late effects
- Rad cat
- Rad ret
- Rad pap
- Amblyopia
- Bone malformation

10 years later

Orbital Tumors of Adulthood

Idiopathic orbital inflammation

Pseudotumor
- Adults > children
- Acute or subacute onset
- Pain
- Eyelid conjunctival edema
- Ptosis
- Proptosis
Orbital pseudotumor

MRI CT
- diffuse mass
dirty fat

Treatment
- Prednisone 80 mg slow taper
- If vision threatened
- IV steroids
- If no response biopsy to rule out neoplasm

Cavernous hemangioma

- Slowly progressive axial proptosis
- No inflammatory signal
- Visual acuity and ocular motility usually good

Treatment
- Prednisone 80 mg slow taper
- If vision threatened
- IV steroids
- If no response biopsy to rule out neoplasm

Cavernous hemangioma

- Circumscribed round mass
- Moderate enhancement
- Intracanal

Cavernous hemangioma

- Slowly progressive axial proptosis
- No inflammatory signal
- Visual acuity and ocular motility usually good

Meningioma

- Observation if
- Small
- Asymptomatic
- Posterior
- Surgical excision if
- Large
- Symptomatic
- Approach
- Cutaneous
- Conjunctival
- Kronlein osteotomy

Management

- Two types
- Optic nerve sheath
- Sphenoid wing

Both types have a predilection for adult females

Optic nerve sheath meningioma

- Vision loss
- Field loss
- Optic atrophy
- Later proptosis
- Optociliary shunt at disc margin
- Bilateral < 5%

Optic nerve sheath meningioma

- Vision loss
- Field loss
- Optic atrophy
- Later proptosis
- Optociliary shunt at disc margin
- Bilateral < 5%

Optic nerve sheath meningioma

CT
- Optic nerve thickening
- Calcification

MRI
- Optic nerve enhancement with gadolinium
- Silhouette [railroad track]

Sphenoid wing meningioma

- Proptosis
- Temporal fossa fullness
- Visual loss as lesion encroaches optic canal
- Optociliary shunt vessels less common

Sphenoid wing meningioma

- Sphenoid wing thickening hyperostosis
- Soft tissue mass
- Orbit
- Temp fossa
- Brain
Sphenoid wing meningioma
• Sphenoid wing thickening hyperostosis
• Soft tissue mass
  • Orbit
  • Temp fossa
  • Brain

Orbital meningioma
• Primary optic nerve sheath
• Observation
• Irradiation
• Resection

Sphenoid wing
• Observation
• Irradiation
• Resection

Orbital lymphoma
• Most common malignant tumor orbit
• Anterior orbit
• Often palpable
• Conjunctival and uveal involvement
• Systemic lymphoma
  • 12% if unilateral
  • 72% if bilateral

Orbital lymphoma
• Flat pancake
• Molds to bone and globe
• Multifocal
• Bilateral
• Rare bone invasion

Lymphoma
• ½ fresh flow cytometry
• ½ formalin permanent

Ocular Tumors
www.fighteyecancer.com

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