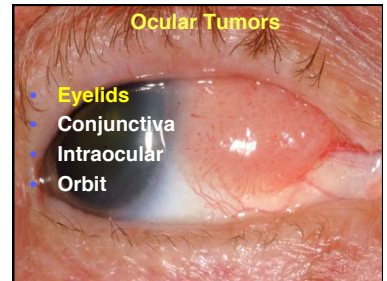
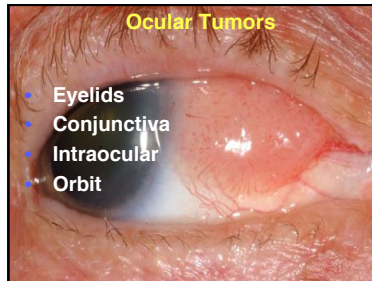


The authors have no financial interests in the materials in this presentation

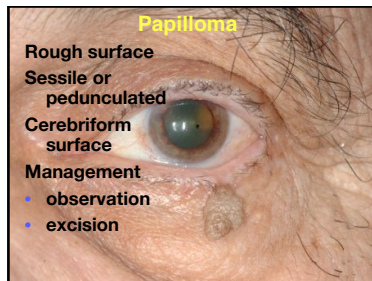


Oncology Service
Wills Eye Institute

- 5 fellows research
- 2 fellows clinical
- 5 staff
- Carlos Bianciotto MD
- Sara Lally MD
- Arman Mashayekhi MD
- Jerry Shields MD
- Carol Shields MD
- Nurses
- Technicians
- Students




- 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



Eyelid Tumors

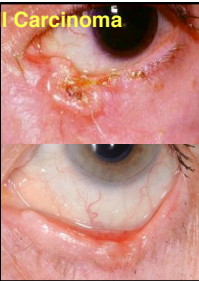
| | |
|-------------------------|-----|
| Malignant eyelid tumors | |
| Basal cell ca | 90% |
| Sebaceous ca | 4% |
| Squamous cell ca | 4% |
| Melanoma | 1% |



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

Basal Cell Carcinoma

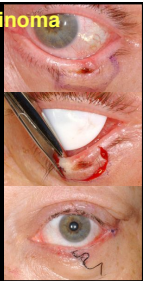
- Types
 - Noduloulcerative
 - Morpheaform
 - Pigmented
 - Cystic
- Eyelid margin
- Margins
 - Well defined
 - Poorly defined
- Central ulcer
- Loss of cilia




Basal Cell Carcinoma

Management

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



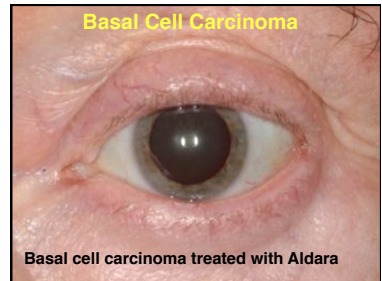
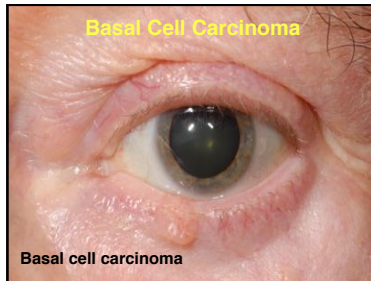
Basal Cell Carcinoma



Imiquimod 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,
Daniele Giammaria, MD,
and Emilio Balestrazzi, MD

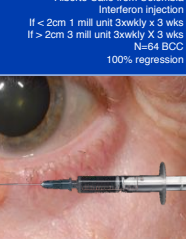


Basal Cell

International Society Ocular Oncology
Cambridge England 2009
Alberto Calle from Colombia
Interferon injection
If < 2cm 1 mill unit 3xwky x 3 wks
If > 2cm 3 mill unit 3xwky X 3 wks
N=84 BCC
100% regression

Management

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



1 J Am Acad Dermatol. 2006 Jun;54(6):1033-6.
Journal of the American Academy of Dermatology.
Long-term efficacy of interferon alfa 2b as monotherapy.

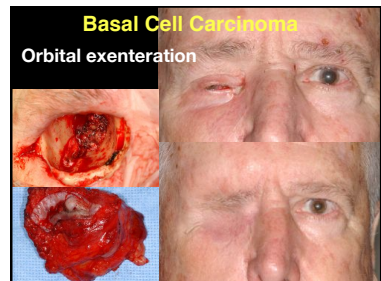
98 cases bcc

Tucker SB, Polessek JW, Perri AJ, Goldsmith EA.
Department of Dermatology, The University of Texas Medical School at Houston, Texas, USA.

BACKGROUND: Interferon alfa-2b (IFN) may be used to treat basal cell carcinoma (BCC) as an alternative to surgical or destructive methods. OBJECTIVE: The purpose of this study is to determine the long-term effectiveness of IFN treatment for BCC. METHODS: Fifty patients with 98 biopsy-proven primary superficial and nodular BCCs were treated perilesionally and intralesionally with injections of IFN between 1985 and 1992. RESULTS: Clinical cures were noted in 95 of 98 BCCs (51 nodular and 44 superficial), with a mean follow-up period of 10.5 years (9 months to 18.5 years). Of these, 35 of the 50 patients, which would include 68 of the 98 tumors, were followed up for a minimum of 10 years, with an average follow-up of 13.5 years. The 3 lesions requiring further treatment were nodular type BCC. One of these lesions showed no response to treatment, whereas the other two responded, then recurred at 4 and 154 months. Of the 68 tumors followed up for a minimum of 10 years, cure rates were 96%. Overall data analysis using Kaplan-Meier estimates showed 88% success rates at years 5 and 10, and a 96% success rate at year 15. LIMITATIONS: Since 31% of patients could not be evaluated for more than 10-year follow-up, it is possible that the long-term cure rate is lower than that found in those available for evaluation. CONCLUSIONS: Treatment of superficial and nodular BCCs with perilesional IFN is an acceptable treatment that may provide benefits over other treatment modalities depending on patient's individual needs. In addition, on the basis of this study, results of IFN treatment for BCC are comparable to most other methods of tumor destruction.

Basal Cell Carcinoma

Orbital exenteration



Sebaceous Carcinoma

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

Frequent misdiagnosis

Sebaceous Carcinoma

Growth pattern
Nodular

Sebaceous Carcinoma

Growth pattern
Diffuse

Sebaceous Carcinoma

Metastasis [n=60 pts] mean 4 yrs

| | |
|---------|-----|
| No mets | 90% |
| Mets | 10% |

Interval to mets 20 mo

Site mets

Initially lymph node 100%

Shields, Demirci, Marz, Eagle, Shields. Sebaceous Carcinoma of the Eyelids. Personal experience with 60 cases. Ophthalmology 2004.

Sebaceous Carcinoma

2 stages

- Map
- Resection

Sebaceous carcinoma

Agay-Sarraf, Eye Hospital

Sebaceous Carcinoma

Management

- Local resection
- Cryotherapy
- MMC
- Orbital exenteration

Melanocytic Tumors

- Several different types
- Different appearance
- Different implications

Melanocytic Tumors

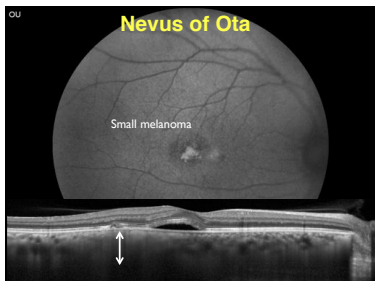
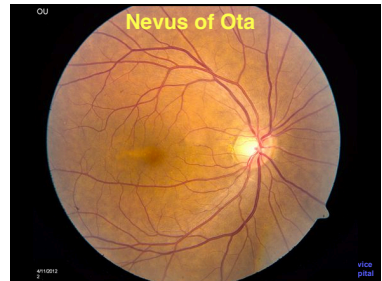
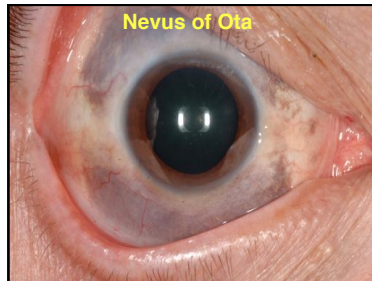
- **Nevus**
 - Lid margin
 - Stable
- **Kissing nevus**
 - Congenital
 - Skin melanoma
- **Nevus of Ota**
 - Congenital
 - Uveal melanoma
- **Lentigo maligna**
 - Acquired
 - Skin melanoma
- **Malignant melanoma**
 - Rare
 - Metastasis

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



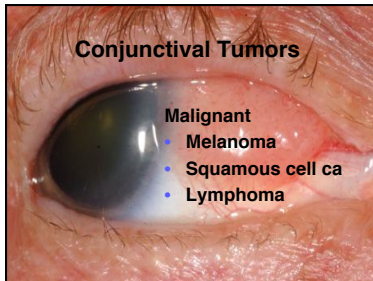
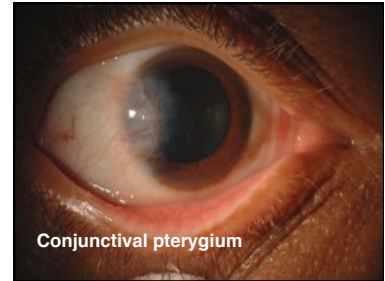
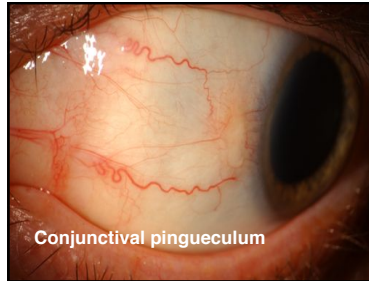
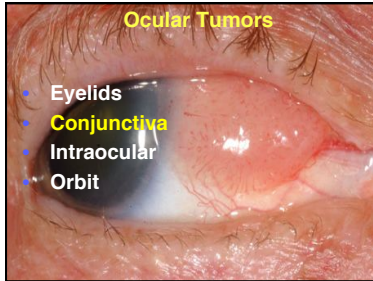
Lentigo Maligna

- 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 metastasis to regional nodes and systemically

Oncology Service
Wills Eye Hospital



Pigmented Conjunctival Tumors

... but first know the pigmented lesions of the conjunctiva

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

Oncology Service
Wilks Eye Hospital

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

Oncology Service
Wilks Eye Hospital

Primary Acquired Melanosis

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

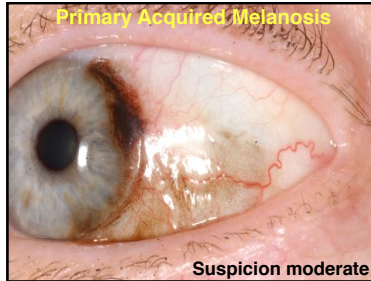
Primary Acquired Melanosis

Progression to melanoma

| | AFIP | WEH |
|--------------------|------|-----|
| Overall | 32% | 9% |
| PAM without atypia | 0% | 0% |
| PAM with atypia | 46% | 13% |

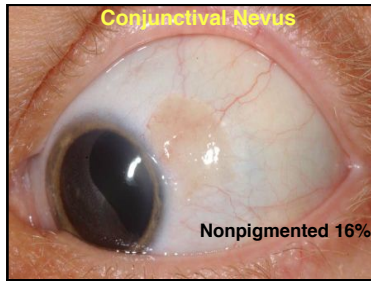
| Atypia | |
|--------|-----|
| None | 0% |
| Mild | 2% |
| Severe | 32% |

Oncology Service
Wilks Eye Hospital



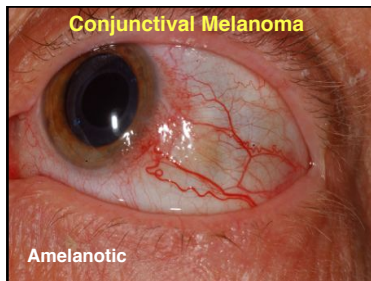
- Melanocytic Conjunctival Tumors**
- Diagnoses
- Racial melanosis
 - Primary acquired melanosis
 - Secondary acquired melanosis
 - **Nevus**
 - Melanoma
 - Metastasis
 - Simulating lesions
- Oncology Services
Wills Eye Hospital

- Conjunctival Nevus**
- Children and young adults
 - Discrete
 - Elevated
 - Cysts
 - Variably pigmented
 - Stationary
-
- Oncology Services
Wills Eye Hospital

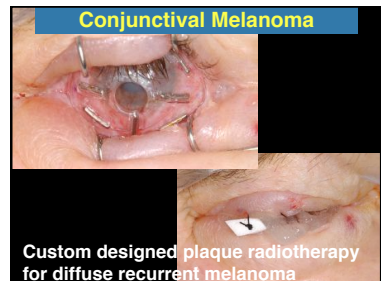
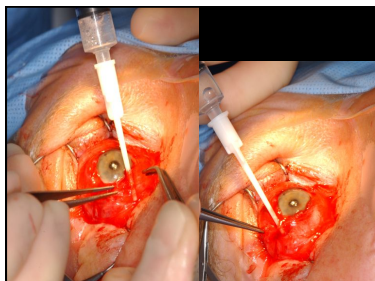
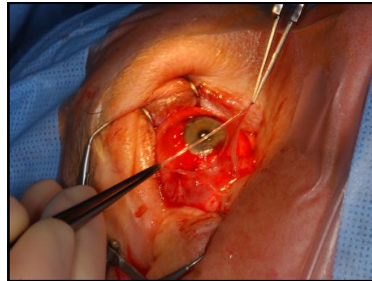
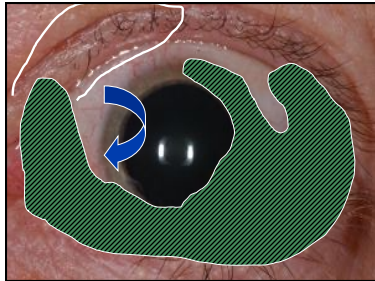
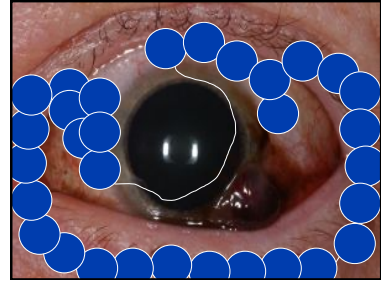
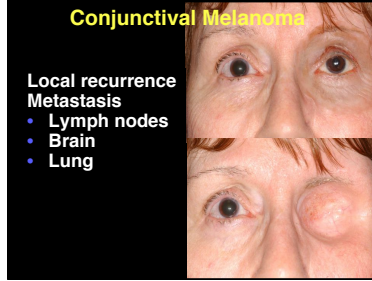
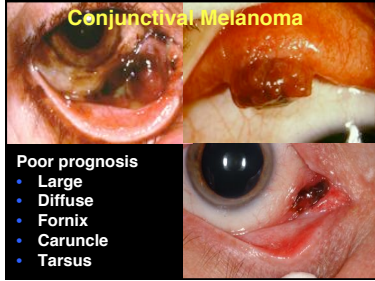


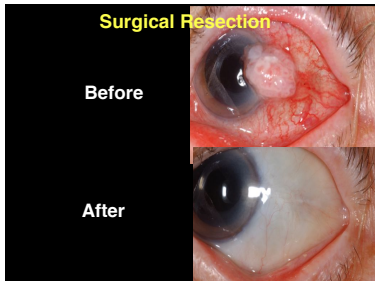
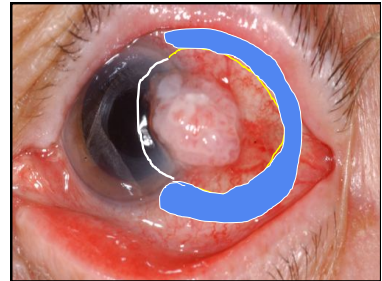
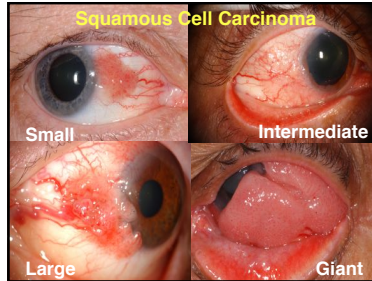
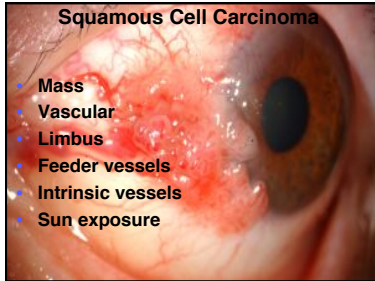
- Melanocytic Conjunctival Tumors**
- Diagnoses
- Racial melanosis
 - Primary acquired melanosis
 - Secondary acquired melanosis
 - Nevus
 - **Melanoma**
 - Metastasis
 - Simulating lesions
- Oncology Services
Wills Eye Hospital

- Conjunctival Melanoma**
- Origin
- **PAM 70%**
 - Nevus 15%
 - De novo 15%
-



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

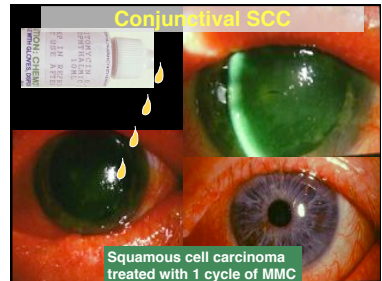




Treatment Conjunctival SCC

| | | | |
|--------------------|---|------------------------|------------------------------|
| Surgical | Alcohol epithellectomy | 2 weeks | fast |
| | Partial lamellar scleroconjunctivectomy | 0.04% qid | Toxic |
| | Cryotherapy | Punctal plugs | Dispose of bottle |
| | Closure | 3 to 12 months | slow |
| Nonsurgical | | 1 mill units/cc qid | Nontoxic |
| | Topical | | Nursing home patient |
| | • MMC | 3 weeks | fast |
| | • SFU | 3 mill units cc per wk | Multiple visits + injections |
| | • Interferon | 1 day | fast |
| | • Cidofovir | Sun exposure | expensive |
| | • Injection interferon | | |
| | • Photodynamic therapy | | |

Oncology Service
Weiz Eye Hospital



Lymphoid Tumors

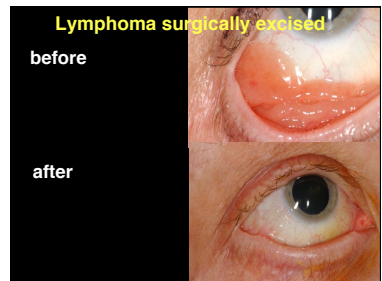
Systemic lymphoma by 30 years

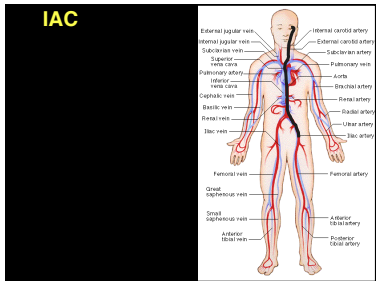
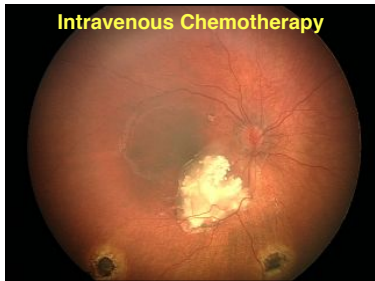
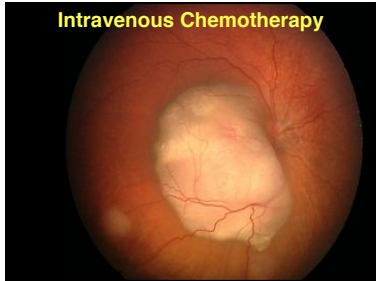
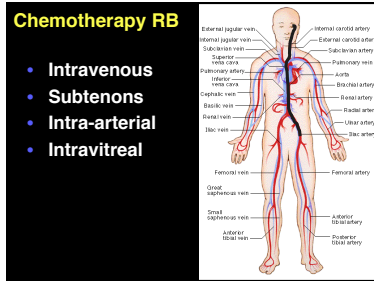
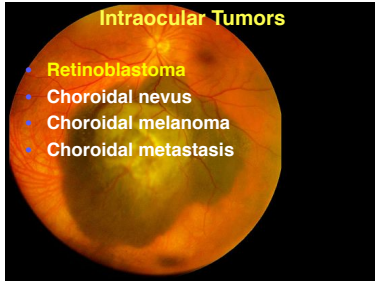
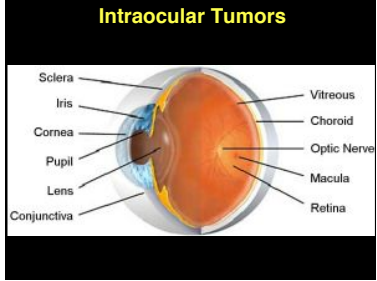
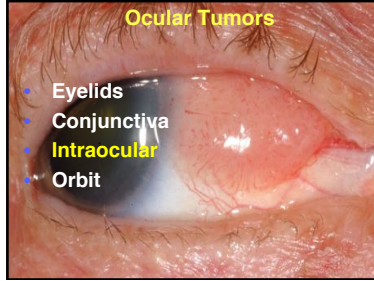
If unilateral 17%

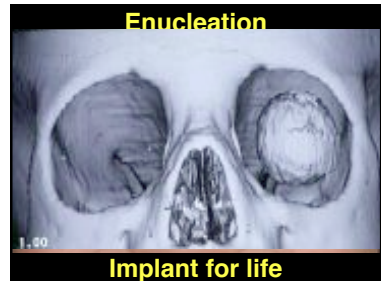
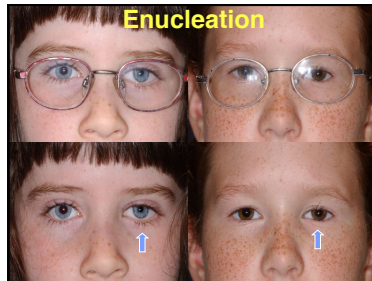
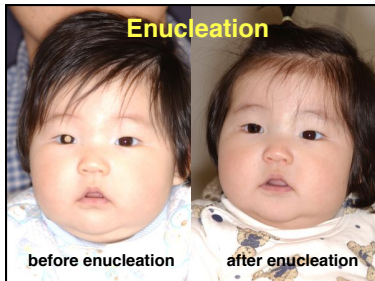
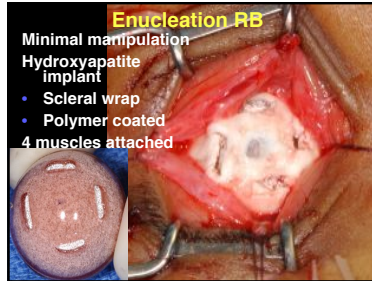
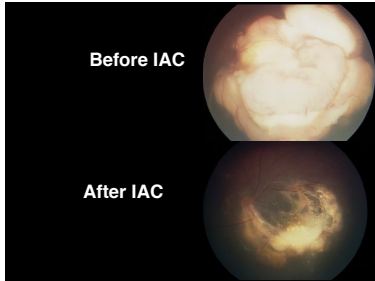
If bilateral 50%

Both
Ocular treatment
Systemic monitoring ... for life

Shields CL, Shields JA, Carvalho C, Ruddle P, Smith A. Conjunctival lymphoid tumors: Clinical analysis of 117 cases and relationship to systemic lymphoma. Ophthalmology 2001







Mysteries of intraocular tumors

- Retinoblastoma
- **Choroidal nevus**
- **Choroidal melanoma**
- Choroidal metastasis

Oncology Services
Wills Eye Hospital

Choroidal Nevus

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

Sumich P, Mitchell P, Wang JJ. Choroidal nevi in a white population: the Blue Mountains Eye Study. Arch Ophthalmol. 1998

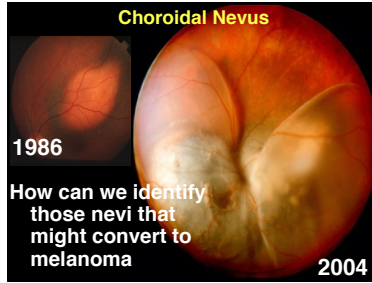
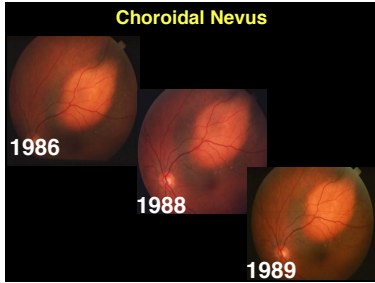
Choroidal Nevus

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

Kivela, Eskelin. Transformation of nevus to melanoma. Ophthalmology 2006.



Judge nevus vs melanoma

Lesions \leq 3 mm

To find small ocular melanoma

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

Risk Factors for Growth and Metastasis of Small Choroidal Melanocytic Lesions

Carl J. Shields MD, Jack A. Shields MD, Roger Enayati MD, David H. Miller MD, Stephen H. Park MD

Background: The recognition of small choroidal melanocytic lesions as common lesions of the eye has led to the development of the term "choroidal nevus." The clinical and histopathologic features of these lesions are well defined. However, the natural history of these lesions is still unclear. The purpose of this study was to determine the risk factors for growth and metastasis of small choroidal melanocytic lesions.

Methods: A retrospective review of 100 small choroidal melanocytic lesions (mean diameter, 2.5 mm) was conducted. The lesions were classified as choroidal nevi or melanomas based on clinical and histopathologic features. The risk factors for growth and metastasis were determined.

Results: The risk factors for growth and metastasis were thickness $>$ 2 mm, subretinal fluid, symptoms, orange pigment, and margin at optic disc.

Conclusions: The risk factors for growth and metastasis of small choroidal melanocytic lesions are thickness $>$ 2 mm, subretinal fluid, symptoms, orange pigment, and margin at optic disc.

Oncology Services
Wills Eye Hospital

Judge nevus vs melanoma

High risk for growth

- + th $>$ 2 mm
- + subretinal fluid
- + symptoms
- + orange pigment
- + margin @ disc

Judge nevus vs melanoma

Low risk for growth

- th $>$ 2 mm
- subretinal fluid
- symptoms
- orange pigment
- + margin @ disc
- drusen

Uveal Melanoma

Mortality by size

| | | |
|--------|----------|-----|
| small | $<$ 3 mm | 12% |
| medium | 3-8 mm | 25% |
| large | $>$ 8 mm | 50% |

Oncology Services
Wills Eye Hospital

Prognosis of uveal melanoma

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

Oncology Services
Wills Eye Hospital

Choroidal Melanoma

Choroidal melanoma in 6900 patients: Kaplan-Meier estimates for metastasis based on choroidal thickness

| Tumor thickness mm | @ 3 yrs | @ 5 yrs | @ 10 yrs |
|--------------------|---------|---------|----------|
| 0-1 | 2% | 5% | 5% |
| 1-2 | 3% | 8% | 10% |
| 2-3 | 2% | 5% | 12% |
| 3-4 | 3% | 8% | 17% |
| 4-5 | 8% | 15% | 26% |
| 5-6 | 9% | 18% | 28% |
| 6-7 | 9% | 15% | 28% |
| 7-8 | 12% | 21% | 41% |
| 8-9 | 18% | 33% | 48% |
| 9-10 | 19% | 31% | 45% |
| $>$ 10 | 27% | 40% | 52% |

Each mm increase adds ~ 5% increased risk for metastasis

Genotypic Profiling of 452 Choroidal Melanomas with Multiplex Ligation-Dependent Probe Amplification

Bartl Danas¹, Justyna A. Dopera², and Sarah E. Coupland²

Abstract

Purpose: Metastasis from uveal melanoma (UVM) is the leading cause of blindness in the United States. We used multiplex ligation-dependent probe amplification (MLPA) and high-resolution melting (HRM) to identify genetic alterations in UVM. We correlated these alterations with clinical and histologic risk factors.

Experimental Design: Factors were correlated between baseline risk factors and the status of the following genes: *3p*, *8q*, *10q*, *17p*, *22q*, *3p*, *8q*, *10q*, *17p*, *22q*.

Results: The primary (17p) and secondary (22q) sites had a median age of 51.4 years and a median follow-up of 3.03 years. MLPA abnormalities occurred in a wide variety of combinations. Ten-year disease-specific mortality was 0% in 133 tumors with no chromosome 3 loss, 50% in tumors with chromosome 3 loss but no chromosome 8 gain, and 71% in 140 tumors showing combined chromosome 3 loss and 8q gain. In tumors with both these abnormalities, epithelial melanoma cytogenetic profile, closed loops, and high mitotic rate correlated with poor survival as did loss of chromosome 8 gain.

Conclusions: These results support the use of MLPA for routine clinical prognostication, especially if the genetic data are considered together with clinical and histologic risk factors. We showed a wide variety of MLPA results, which suggests that chromosomal abnormalities in uveal melanoma accumulate in a variable sequence. (DOI: 10.1167/jov.2010.19.010001)

| | |
|---------------------|-------------|
| Mortality | at 10 years |
| disomy 3 | 0% |
| monosomy 3 | 55% |
| monosomy 3, 8q gain | 71% |

Clinical Cancer Research

Uveal Melanoma

Management

- transpupillary thermotherapy ttt
- plaque radiotherapy
- local resection
- enucleation
- exenteration
- photodynamic therapy
- antiVEGF
- systemic therapies

Oncology Service
Wills Eye Hospital

Uveal Melanoma

Thermotherapy

3.5 mm 1.0 mm

Uveal Melanoma

Plaque radiotherapy

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

METRIC 1

Oncology Service
Wills Eye Hospital

Uveal Melanoma

Plaque radiotherapy

Oncology Service
Wills Eye Hospital

Uveal Melanoma

Plaque radiotherapy

Oncology Service
Wills Eye Hospital

Uveal Melanoma

Eye stable
Systemic risks
Adjuvant therapy

Plaque radiotherapy

Oncology Service
Wills Eye Hospital

Uveal Melanoma

Plaque radiotherapy plus TTT

Oncology Service
Wills Eye Hospital

Uveal Melanoma

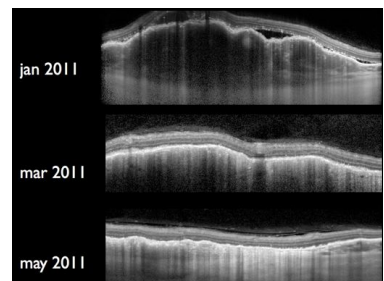
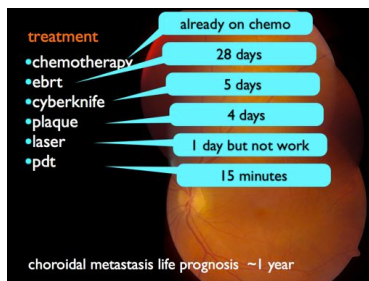
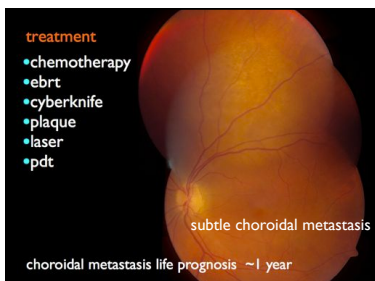
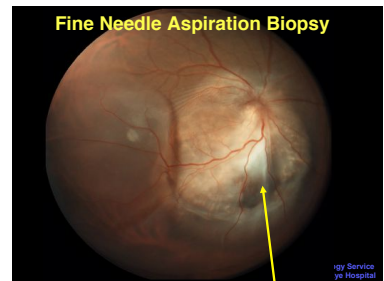
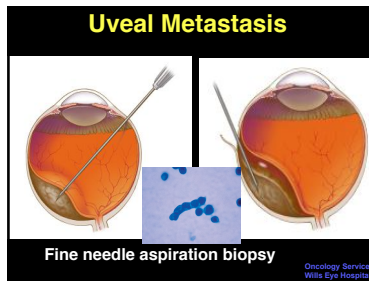
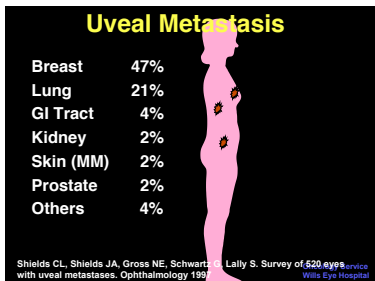
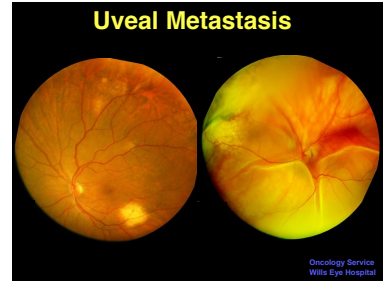
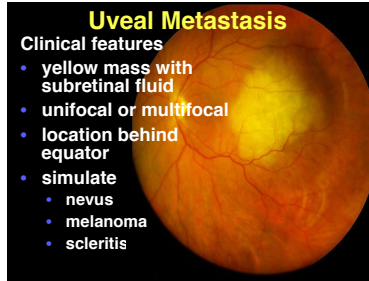
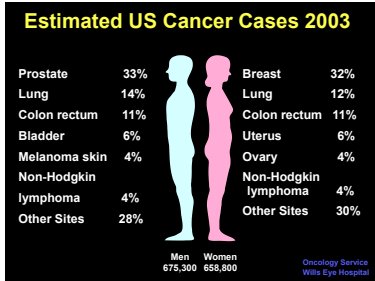
Plaque radiotherapy plus TTT

Oncology Service
Wills Eye Hospital

Mysteries of intraocular tumors

- Retinoblastoma
- Choroidal nevus
- Choroidal melanoma
- **Choroidal metastasis**

Oncology Service
Wills Eye Hospital



Orbital Tumors in Children and Adults



Carol Shields
 Oncology Service
 Wills Eye Hospital
 Thomas Jefferson
 University
 Philadelphia PA USA

Survey of 1264 Patients with Orbital Tumors and Simulating Lesions

The 2002 Montgomery Lecture, Part 1


Jerry A. Shields, MD, Carol L. Shields, MD, Richard Saravitz, MD
Objective: To determine the incidence of specific orbital tumors based on patients referred to an ocular oncology center.
Design: Retrospective, observational case series.
Participants: A total of 1264 consecutive patients referred to an ocular oncology service because of extra-ocular-involving orbital lesions.
Methods: An retrospective chart review was carried out for 1264 consecutive patients referred for a suspected orbital mass over a 30-year period. The lesions were grouped into general categories, as shown in "Results." The specific diagnosis in each case was based on clinical findings, computed tomography scan results, magnetic resonance imaging results, and histopathologic analysis results when available. The number and percentage of benign and malignant tumors per age group also was determined.
Main Outcome Measures: The incidence of orbital tumors and pseudotumors.
Results: Among 1264 consecutive patients, the number and percentage of lesions in each general category were as follows: cystic, 20 cases (1.6%); neoplastic, 273 cases (21.6%); peripheral nerve lesions, 23 (1.8%); cranial nerve and meningeal lesions, 10 (0.8%); traumatic lesions, 13 (1.0%); orbital and extra-orbital lesions, 2 (0.2%); cartilaginous lesions, 1 (0.1%); spiculate and mixed lesions, 64 (5.1%); epigeal tumors, 36 (2.8%); lacrimal gland lesions, 114 (9.0%); primary melanocytic lesions, 11 (0.9%); metastatic tumors, 91 (7.2%); lymphoma and leukemia lesions, 130 (10.3%); secondary orbital tumors, 142 (11.2%); histiocytic lesions, 17 (1.3%); thyroid-related ophthalmopathy, 67 cases (5.3%); other inflammatory lesions, 133 cases (10.5%); and miscellaneous other lesions, 13 (1.0%). The most common diagnoses were lymphoma tumor (130 cases; 11%), idiopathic orbital inflammation (136 cases; 11%), cavernous hemangioma (77 cases; 6%), lymphangioma (64 cases; 5%), meningioma (65 cases; 5%), optic nerve glioma (61 cases; 5%), metastatic breast cancer (64 cases; 5%), orbital cellulitis (61 cases; 5%), orbital melanoma (41 cases; 3%), capillary hemangioma (35 cases; 2.8%), pheochromocytoma (35 cases; 2.8%), lymphangioma (31 cases; 2.4%), hemangioid vessels (23 cases; 1.8%), dermoid cyst (20 cases; 1.6%), optic glioma (19 cases; 1.5%), and other less common lesions. Of the 1264 lesions, 810 (64%) were benign and 454 (36%) were malignant. The percentage of malignant lesions was 20% in children (age range, 0-16 years), 27% in young adults and

Table 2. Classification of 1264 Consecutive Patients with Orbital Lesions

| Category | Number of Patients (%) | Number Benign (%) | % Total Benign | Mean Age (yr, median, range) |
|-------------------------------------|------------------------|-------------------|----------------|------------------------------|
| Cystic lesions | 20 (1.6) | 20 (100) | 100 | 31 (14, 60) |
| Neoplastic lesions | 273 (21.6) | 61 (22.3) | 22.3 | 36 (19, 61) |
| Peripheral nerve lesions | 23 (1.8) | 23 (100) | 100 | 36 (16, 66) |
| Other nerve or meningeal lesions | 33 (2.6) | 33 (100) | 100 | 33 (18, 43) |
| Cartilaginous lesions | 1 (0.1) | 1 (100) | 100 | 33 (18, 47) |
| Epigeal lesions | 36 (2.8) | 1 (2.8) | 2.8 | 37 (21, 53) |
| Lacrimal gland lesions | 114 (9.0) | 86 (75.4) | 75.4 | 48 (15, 80) |
| Primary melanocytic lesions | 11 (0.9) | 11 (100) | 100 | 37 (18, 56) |
| Metastatic lesions to the orbit | 91 (7.2) | 50 (55) | 55 | 60 (40, 80) |
| Metastatic to lacrimal lesions | 112 (8.9) | 112 (100) | 100 | 60 (40, 78) |
| Miscellaneous orbital lesions total | 142 (11.2) | 133 (93.7) | 93.7 | 35 (21, 43) |
| Thyroid lesions | 2 (0.2) | 2 (100) | 100 | 46 (26, 66) |
| Conjunctival lesions | 22 (1.7) | 22 (100) | 100 | 46 (26, 66) |
| Orbital pseudotumors | 133 (10.5) | 133 (100) | 100 | 46 (26, 66) |
| Neurofibroma lesions | 6 (0.5) | 6 (100) | 100 | 47 (26, 68) |
| Other nerve lesions | 4 (0.3) | 4 (100) | 100 | 47 (26, 68) |
| Cartilaginous lesions | 1 (0.1) | 1 (100) | 100 | 47 (26, 68) |
| Spiculate lesions | 64 (5.1) | 2 (3.1) | 3.1 | 51 (27, 75) |
| Lacrimal gland lesions | 114 (9.0) | 1 (0.9) | 0.9 | 51 (27, 75) |
| Other nerve lesions | 10 (0.8) | 1 (10) | 10 | 51 (27, 75) |
| Cartilaginous lesions | 1 (0.1) | 1 (100) | 100 | 51 (27, 75) |
| Epigeal lesions | 36 (2.8) | 1 (2.8) | 2.8 | 51 (27, 75) |
| Lacrimal gland lesions | 114 (9.0) | 1 (0.9) | 0.9 | 51 (27, 75) |
| Primary melanocytic lesions | 11 (0.9) | 1 (9.1) | 9.1 | 51 (27, 75) |
| Metastatic lesions | 91 (7.2) | 49 (53.8) | 53.8 | 51 (27, 75) |
| Metastatic to lacrimal lesions | 112 (8.9) | 112 (100) | 100 | 51 (27, 75) |
| Miscellaneous lesions | 142 (11.2) | 133 (93.7) | 93.7 | 46 (26, 66) |
| Total orbital lesions | 1264 (100) | 709 (56.1) | 56.1 | 46 (26, 66) |

Orbital Tumors


- Children
- Adults



Oncology Service
 Wills Eye Hospital

Orbital Tumors

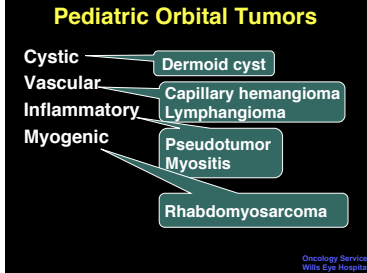
- Children
- Adults



Oncology Service
 Wills Eye Hospital

Pediatric Orbital Tumors


- Cystic
 - Dermoid cyst
- Vascular
 - Capillary hemangioma
- Inflammatory
 - Lymphangioma
- Myogenic
 - Pseudotumor
 - Myositis
 - Rhabdomyosarcoma



Oncology Service
 Wills Eye Hospital

Pediatric Orbital Tumors

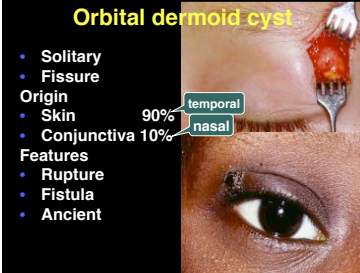
- Cystic
- Vascular
- Inflammatory
- Malignant



Oncology Service
 Wills Eye Hospital

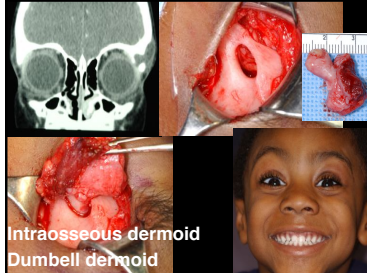
Orbital dermoid cyst

- Solitary
- Fissure
- Origin
 - Skin 90%
 - Conjunctiva 10%
- Features
 - Rupture
 - Fistula
 - Ancient



Oncology Service
 Wills Eye Hospital

Intraosseous dermoid Dumbbell dermoid




Oncology Service
 Wills Eye Hospital

Pediatric Orbital Tumors

- Cystic
- Vascular
- Inflammatory
- Malignant

Oncology Service
Wills Eye Hospital

Capillary hemangioma infancy

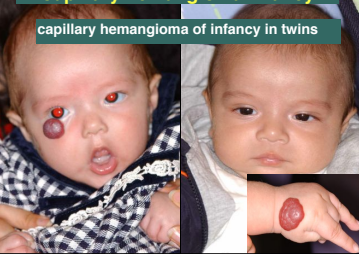


- 10% of newborns
- 20% premature children
- more common in twins
- Skin
- Kassabach Merritt syndrome
- PHACE syndrome
- Pathogenesis placental emboli

Oncology Service
Wills Eye Hospital

Capillary hemangioma infancy

capillary hemangioma of infancy in twins




Oncology Service
Wills Eye Hospital

Capillary hemangioma infancy


Management

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



Oncology Service
Wills Eye Hospital

Capillary hemangioma infancy



triplet #1

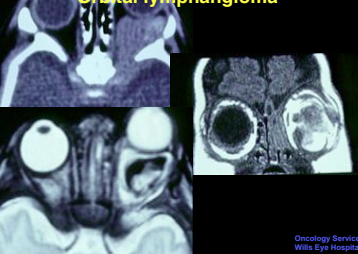
Oncology Service
Wills Eye Hospital

Orbital lymphangioma




Oncology Service
Wills Eye Hospital

Orbital lymphangioma



Oncology Service
Wills Eye Hospital

Orbital lymphangioma



Immediately after surgery


Oncology Service
Wills Eye Hospital

Pediatric Orbital Tumors

- Cystic
- Vascular
- Inflammatory
- Malignant

Oncology Service
Wills Eye Hospital

Orbital pseudotumor



Myositis
Pain on eye movement
Recurrent
Treatment

- Steroids
- NSAIDS
- Kenalog injection

Rheumatology evaluation

- SLE
- RA
- Crohns
- Churg Strauss
- Wegeners

Pediatric Orbital Tumors

Cystic
Vascular
Inflammatory
Malignant

Oncology Service
Wills Eye Hospital

Rhabdomyosarcoma

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



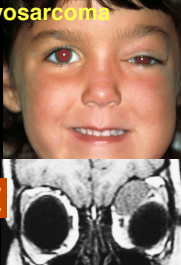
Oncology Service
Wills Eye Hospital

Rhabdomyosarcoma

Management

- Biopsy
- Chemotherapy
- Irradiation

1970 mortality 70%
2000 mortality 5%




Shields C, Shields J, Honavar, Demirci. The clinical spectrum of orbital rhabdomyosarcoma. Ophthalmology 2001

Rhabdomyosarcoma

Classification Treatment

| | |
|-------------------|-----------|
| I no residua | chemo |
| II micro residua | chemo+rad |
| III gross residua | chemo+rad |
| IV mets | |

Goal at surgery complete resection without damaging normal structures



Shields C, Shields J, Honavar, Demirci. The clinical spectrum of orbital rhabdomyosarcoma. Ophthalmology 2001

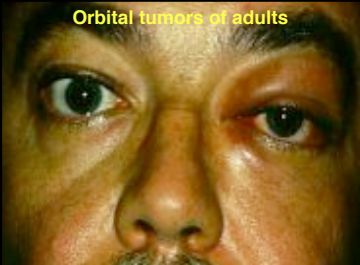
Rhabdomyosarcoma

10 years later
Recurrence
Late effects

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



Orbital tumors of adults



Orbital Tumors of Adulthood

Most common types

| | |
|--------------|----------------------|
| Inflammatory | pseudotumor |
| Benign | cavernous hemangioma |
| | meningioma |
| | optic nerve sheath |
| | sphenoid wing |
| Malignant | lymphoma |
| | metastasis |

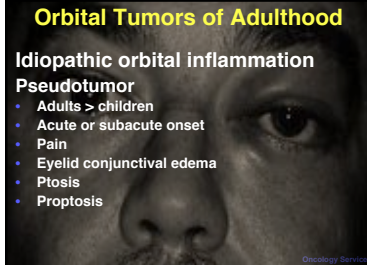
Oncology Service
Wills Eye Hospital

Orbital Tumors of Adulthood

Idiopathic orbital inflammation

Pseudotumor

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



Oncology Service
Wills Eye Hospital

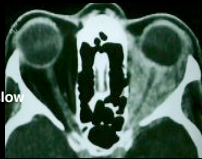
Orbital pseudotumor

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



Oncology Services
Wills Eye Hospital

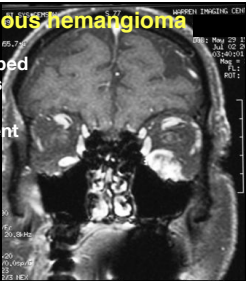
Cavernous hemangioma

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



Cavernous hemangioma

- Circumscribed round mass
- Moderate enhancement
- Intraconal



Cavernous hemangioma

Management

Observation if

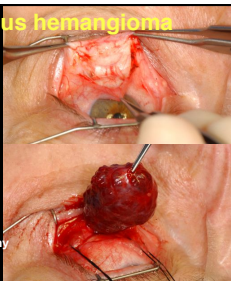
- Small
- Asymptomatic
- Posterior

Surgical excision if

- Large
- Symptomatic

Approach

- Cutaneous
- Conjunctival
- Kronlein osteotomy

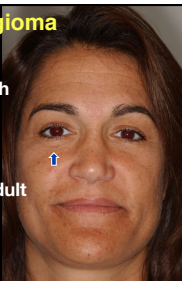


Meningioma

Two types

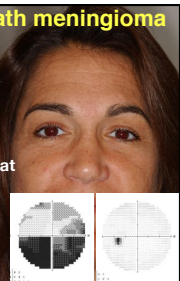
- Optic nerve sheath
- Sphenoid wing

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

CT

- Optic nerve thickening
- Calcification

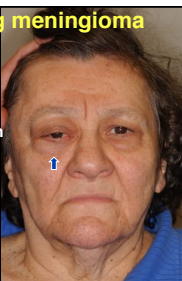
MRI

- 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



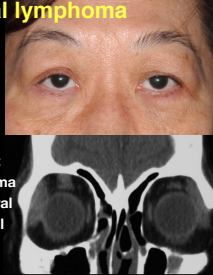
Orbital meningioma

- 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



Orbital lymphoma

Lymphoma

- ½ fresh flow cytometry
- ½ formalin permanent




Ocular Tumors

www.fighteyecancer.com

Carol Shields
Oncology Service
Wills Eye Institute
Philadelphia PA
carolshields@gmail.com



Lumps Bumps and Magic Potions Review of Ocular Tumors

Carol Shields MD
Ocular Oncology Service
Wills Eye Hospital
Philadelphia PA USA

