




A Tour through Eye Tumors

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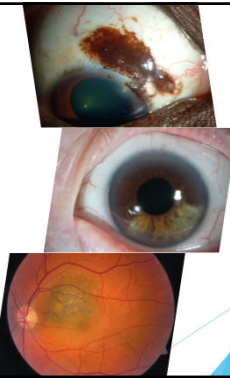
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Objectives

- ▶ Recognize key ocular tumors
- ▶ Understand how ocular tumors can impact visual acuity
- ▶ Understand how ocular tumors can impact survival

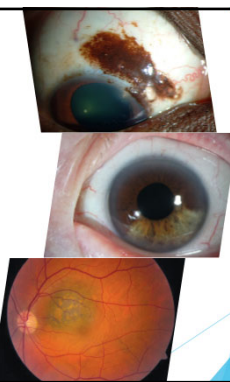
Topics: Tumor Tour

- ▶ Ocular surface
- ▶ Iris
- ▶ Posterior segment



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Ocular Surface Tumors

The Pigment Spectrum

- Conjunctival nevus
- Complexion-associated melanosis (CAM)
- Primary acquired melanosis (PAM)
- Malignant melanoma

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Conjunctival Nevus

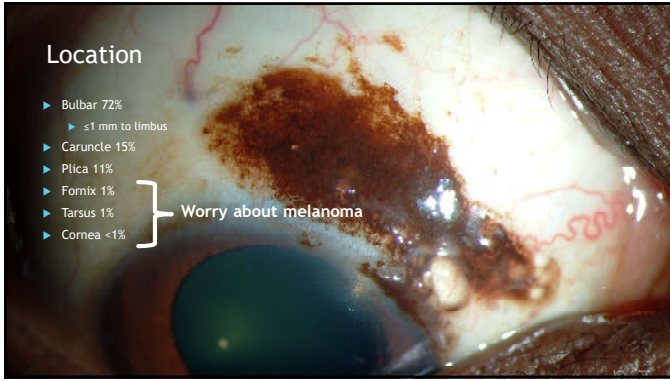
- ▶ Well-circumscribed
- ▶ Pigmented 85%
- ▶ Amelanotic 15%
- ▶ Pseudocysts 65%
- ▶ Not pathognomonic (lymphangiectasia, mucocypidermoid SCC)

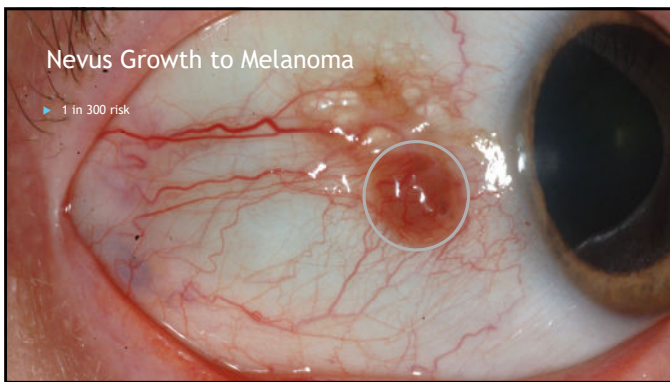


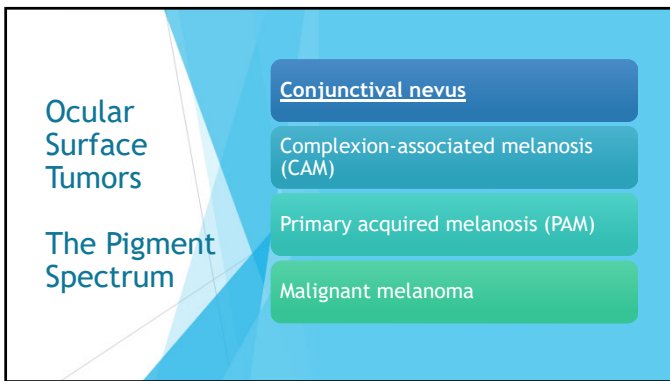
Amelanotic Conjunctival Nevus in a Child

- ▶ Up to 75% with inflammation
- ▶ Rare rapid growth









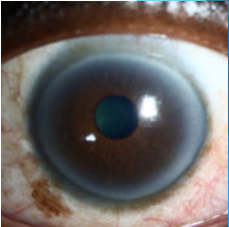
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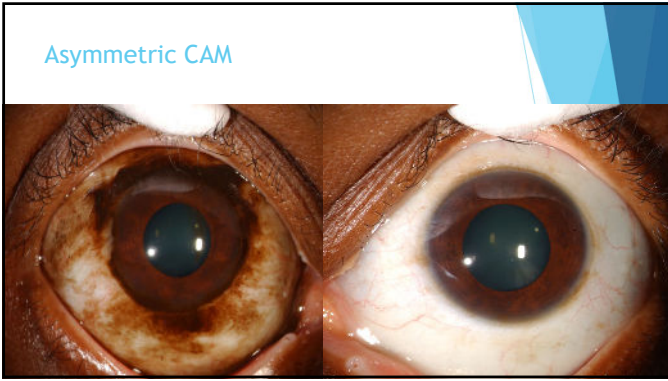
Complexion-Associated Melanosis (CAM)

- ▶ Bilateral
- ▶ Symmetric
- ▶ Flat
- ▶ Often at the limbus
- ▶ Microfolds create cobblestone appearance
- ▶ More common with darker complexion
- ▶ Can extend to fornix, rare to tarsus



Symmetric CAM





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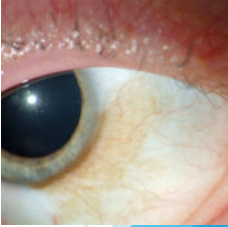
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Primary Acquired Melanosis (PAM)

- ▶ Middle age
- ▶ Light complexion
- ▶ Unilateral, asymmetric
- ▶ Flat, peppery
- ▶ No cysts, cobblestone, or microfolds
- ▶ Risk for melanoma



When to Worry

- ▶ More clock hours = 1.7-fold increased risk of growth to melanoma
- ▶ No atypia = minimal risk of melanoma
- ▶ Severe atypia = 21% risk for melanoma



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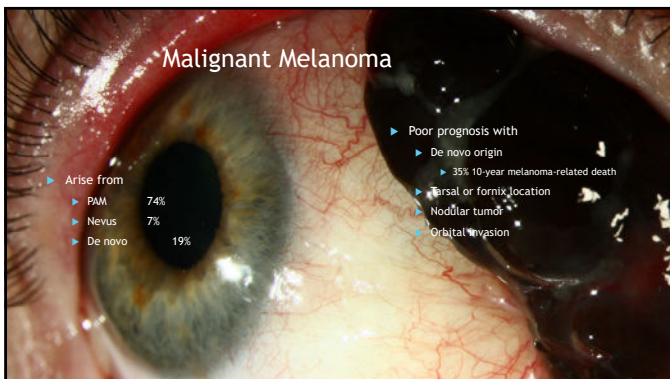
Malignant Melanoma

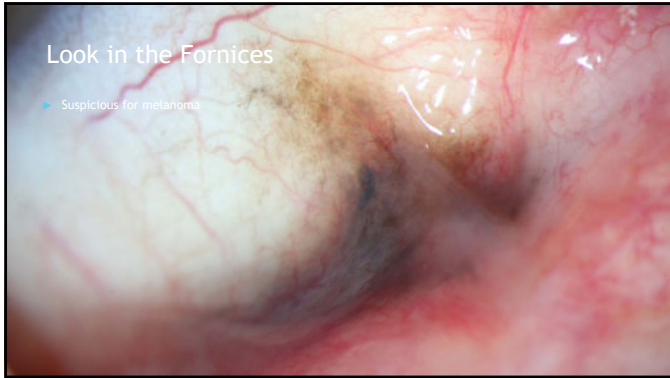
- ▶ Pigmented (but not always!)
- ▶ Elevated
- ▶ Feeder and intrinsic vessels
- ▶ 25% risk for metastasis at 10 years
- ▶ Medical oncology referral for systemic workup



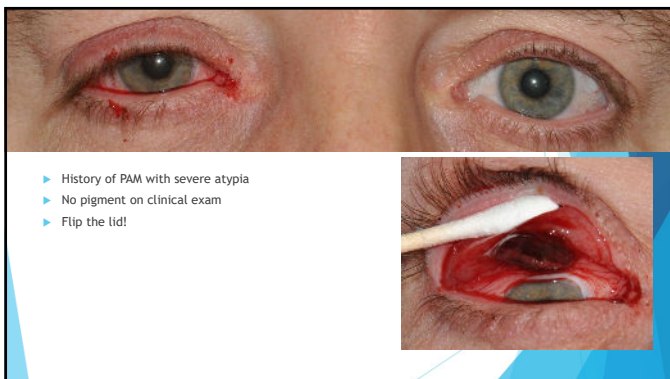
Malignant Melanoma

- ▶ Arise from
 - ▶ PAM 74%
 - ▶ Nevus 7%
 - ▶ De novo 19%
- ▶ Poor prognosis with
 - ▶ De novo origin
 - ▶ 35% 10-year melanoma-related death
 - ▶ Tarsal or fornix location
 - ▶ Nodular tumor
 - ▶ Orbital invasion









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Multimodal Imaging

- ▶ UBM to rule out scleral invasion
- ▶ AS-OCT for scleral invasion and diagnostic features in atypical cases

Normal
OSSN
Nevus
Melanoma

All contributors: Candice Meadows MD, Anne Quirk OD, MA, PhD, Carol L. Kim MD, Joseph H. Johnson MD, Jonathan Weikstein MD, and Vincent P. Fazio MD

Amelanotic Melanoma

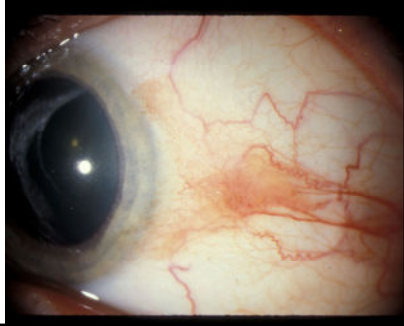
- ▶ Associated PAM
- ▶ Pigment in the fornix

Pigmented Squamous Cell Carcinoma

- ▶ Pigmented
- ▶ Cystoid spaces
- ▶ Nevus or melanoma?

What is this?

- ▶ Might have no pigment
- ▶ Can be subtle
- ▶ Always approach as if it could be melanoma



Treatment of Conjunctival Melanocytic Lesions

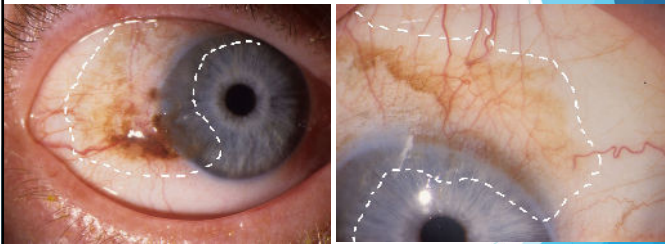
Treatment Options

- ▶ Surgery
- ▶ Topical Therapy
- ▶ Radiotherapy

Surgery

- ▶ No touch technique
- ▶ Alcohol keratectomy
- ▶ Partial lamellar scleroconjunctivectomy
- ▶ 2-4 mm margins
- ▶ Double freeze thaw cryotherapy
- ▶ First surgery is most important
- ▶ No BSS - keep it off the field
- ▶ Clean instruments with alcohol if moving to a new location
- ▶ Use different, clean instruments for closure
- ▶ SLN if > 2mm thick

Wide Margins



Topical Therapy

- ▶ Mitomycin C (0.02-0.04%)
- ▶ Alkylating agent
- ▶ For PAM
- ▶ Adjuvant for melanoma
- ▶ Wait 1-2 weeks after surgery
- ▶ Use for 7-28 days
- ▶ Side effects: keratoconjunctivitis, limbal stem cell deficiency
- ▶ Interferon alpha-2b (1 million IU/mL)
- ▶ Glycoprotein, acts at cell surface
- ▶ Adjuvant for PAM/melanoma
- ▶ Side effects: keratoconjunctivitis, less discomfort than MMC
- ▶ Less evidence than MMC

Radiotherapy

- ▶ Plaque
- ▶ Adjuvant after excision
 - ▶ Treats deep sclera
- ▶ Wait for healing after surgery
- ▶ 100 Gy, depth of 1-3 mm
- ▶ Side effects: dry eye, corneal ulcer
- ▶ Well tolerated
- ▶ EBRT/Proton beam
 - ▶ Less commonly used
 - ▶ For extensive disease
 - ▶ More recurrence
 - ▶ More metastasis
 - ▶ More radiation side effects
 - ▶ Dry eye, cataract, limbal stem cell deficiency
 - ▶ Lash loss with proton


PAM Managed with Surgery and MMC

- ▶ Biopsy - PAM with atypia
- ▶ Residual pigment
- ▶ MMC 0.04% QID for 7 days
- ▶ At 4-month follow-up

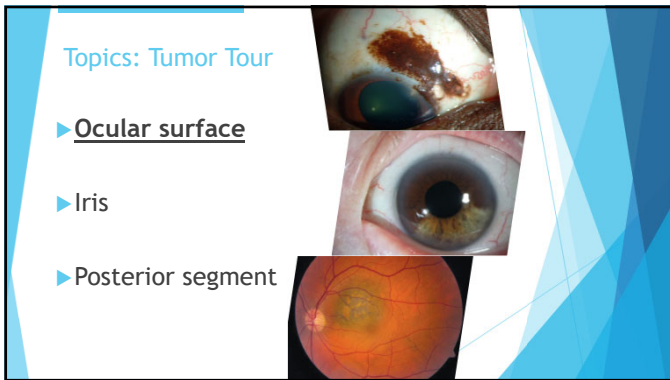


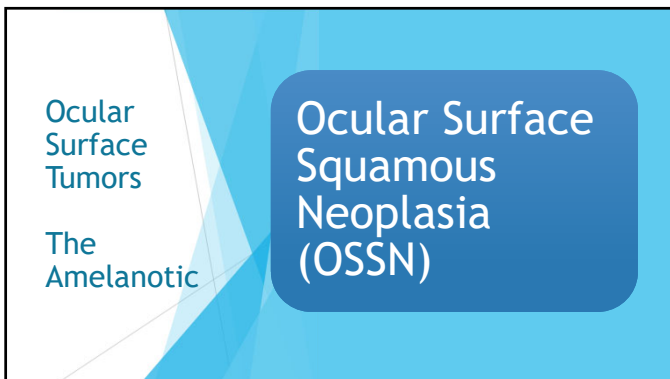
Melanoma + PAM Managed with Surgery/Cryo

- ▶ 10 clock hours = 17x risk
- ▶ Temporal conjunctiva excised
 - ▶ Small focus of melanoma
- ▶ Map biopsies and cryotherapy elsewhere
 - ▶ PAM
- ▶ Follow with MMC 0.02% 1 week on, 1 week off x3







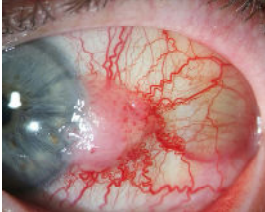


Ocular Surface Tumors

The Amelanotic


Ocular Surface Squamous Neoplasia (OSSN)

Overview



- ▶ Most common amelanotic corneal and conjunctival malignancy
- ▶ Includes
 - ▶ Dysplasia
 - ▶ Corneal and conjunctival intraepithelial neoplasia (CIN)
 - ▶ Squamous cell carcinoma
 - ▶ Invades the basement membrane
- ▶ Local invasion uncommon but can occur
- ▶ Metastasis rare: <1%

Demographics

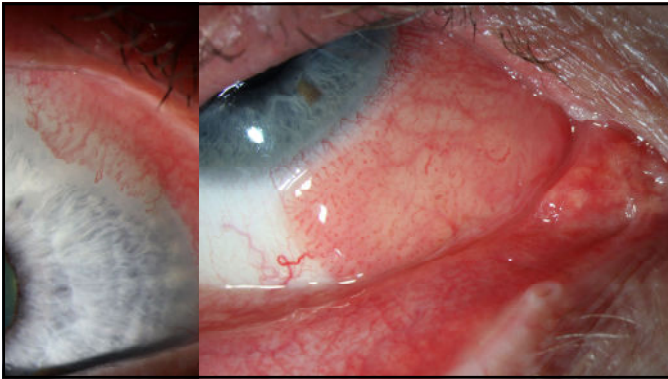


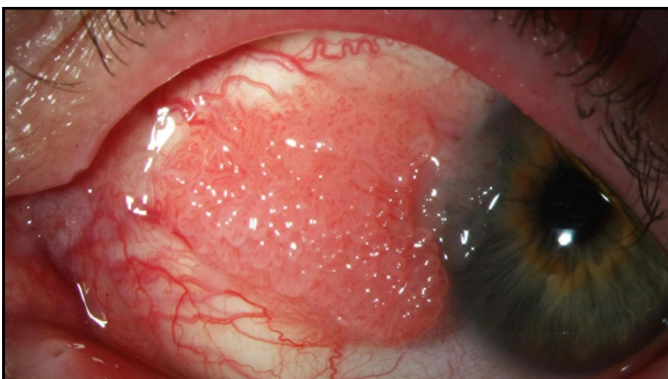
- ▶ Fair-skinned
- ▶ Middle-aged or older
 - ▶ Immunosuppression/HIV if young
- ▶ Men>Women
- ▶ Sunlight exposure
- ▶ HPV
 - ▶ 16, 18, 33
- ▶ Smokers
 - ▶ More bilateral

Clinical Features

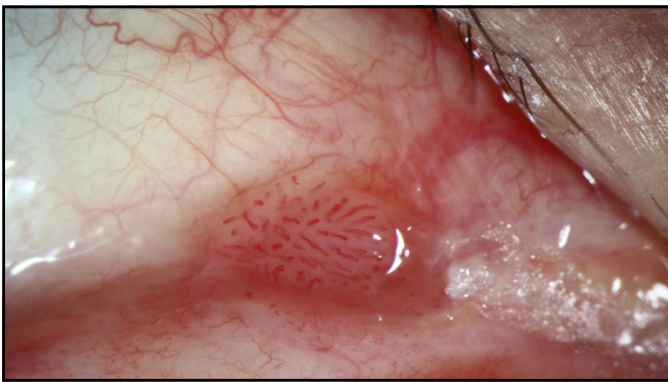
- ▶ Unilateral
- ▶ Limbus
- ▶ Interpalpebral fissure
- ▶ Fornix, palpebral conjunctival less common
 - ▶ More in smokers
- ▶ Sessile or dome-shaped, gelatinous mass
- ▶ Feeder and intrinsic vessels
- ▶ Leukoplakia
- ▶ Can involve corneal epithelium
- ▶ Amelanotic
 - ▶ Pigment common with pigmented complexion

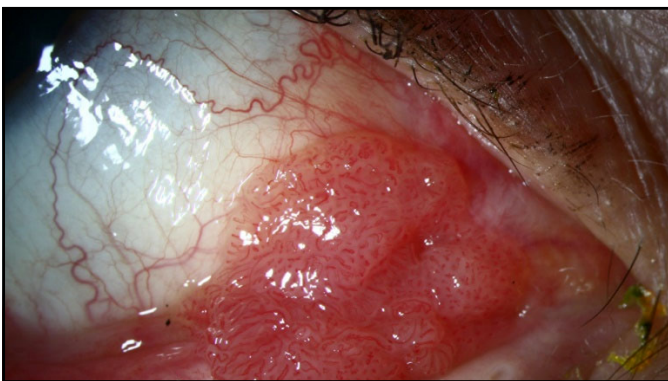


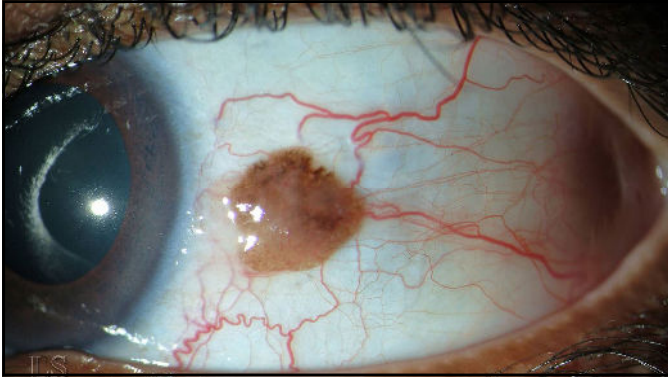


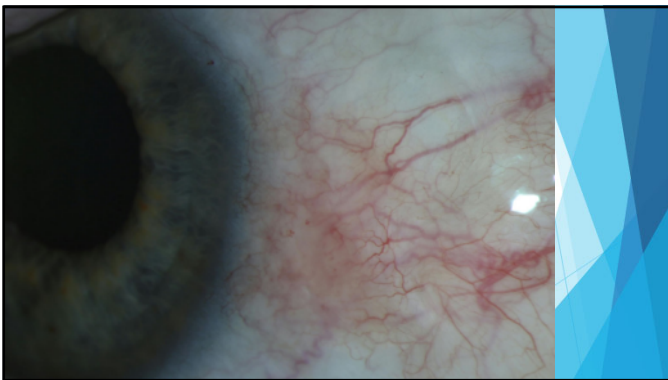






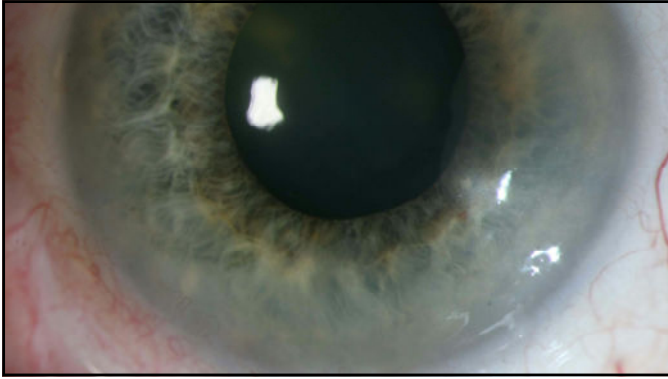


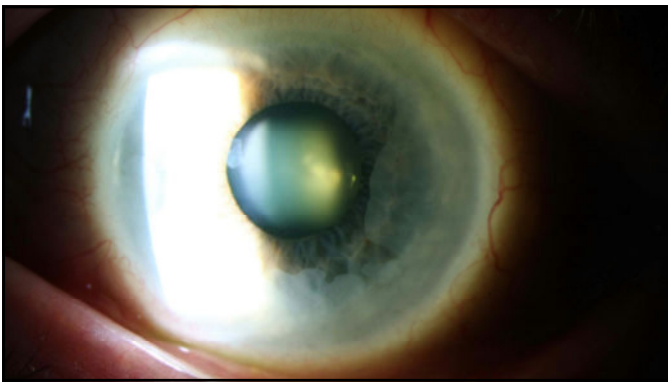


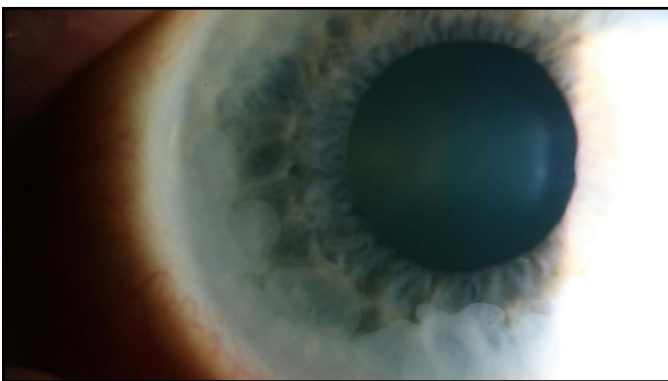


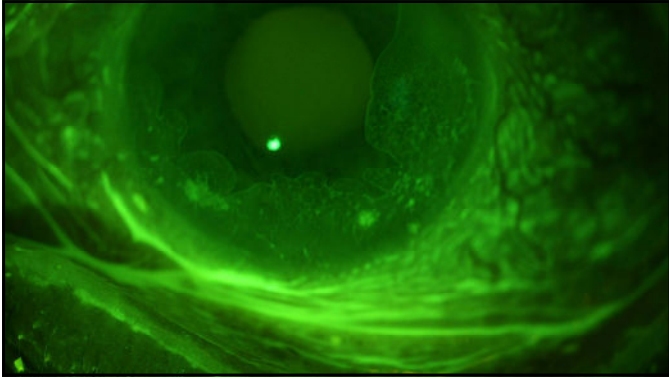
Ancillary Testing: Fluorescein with Yellow Barrier Filter

- ▶ Over cobalt blue light highlights OSSN
- ▶ Regressed after MMC 0.02%






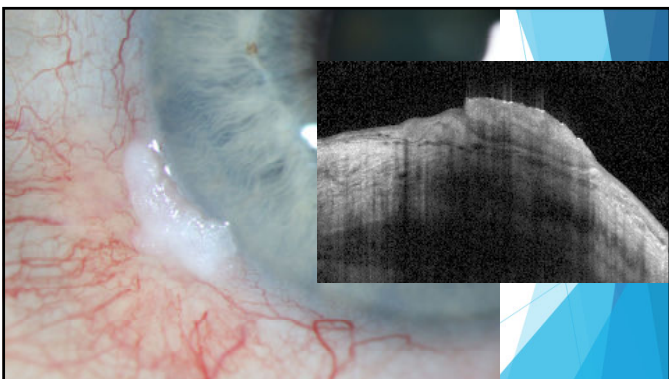




Ancillary Testing: AS-OCT




- ▶ Epithelial thickening
- ▶ Epithelial hyperreflectivity
- ▶ Abrupt transitions between normal and abnormal tissue



Treatment of OSSN

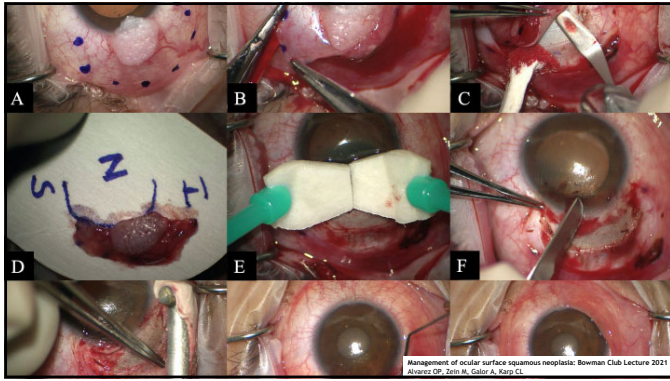
Surgical Excision

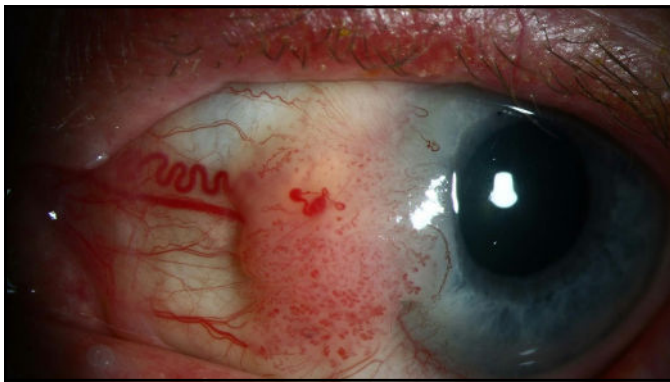
- ▶ Traditional gold standard
- ▶ Complete excision important
 - ▶ >50% recurrence if positive margins
 - ▶ 0-21% recurrence with current techniques
- ▶ Side effects
 - ▶ Conjunctival scarring
 - ▶ Symblepharon
 - ▶ Conjunctival hyperemia
 - ▶ Limbal stem cell deficiency
- ▶ Requires less time, less follow-up

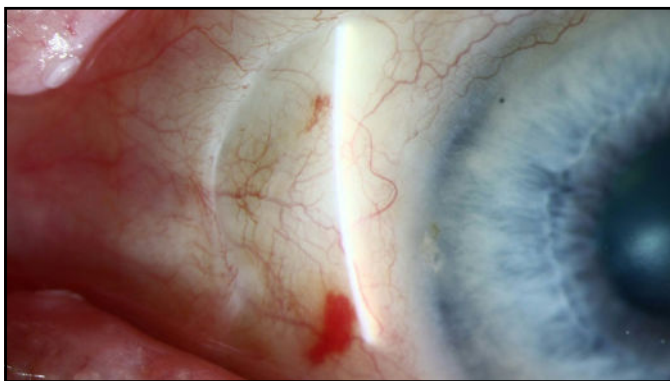


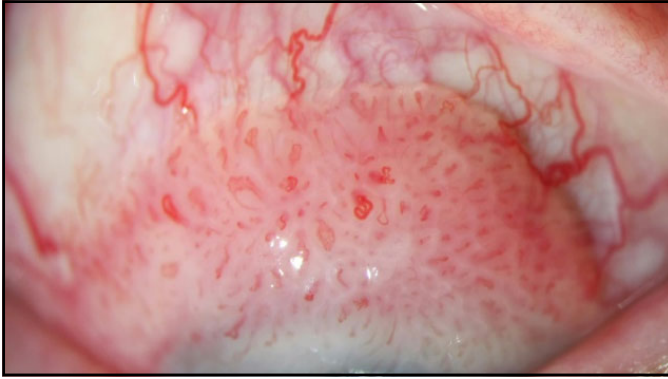
Surgical Technique

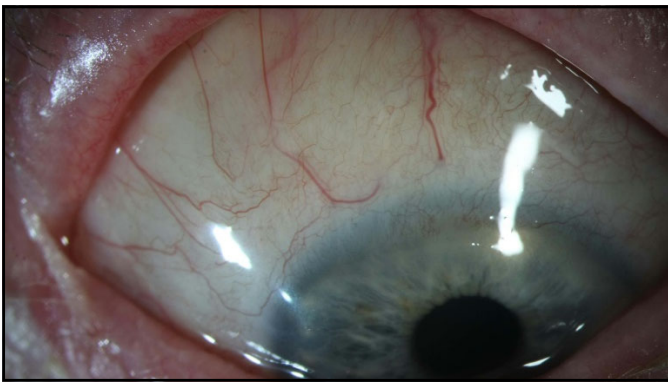
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- ▶ Partial lamellar scleroconjunctivectomy
- ▶ 2-4 mm margins
- ▶ Double freeze thaw cryotherapy
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- ▶ Clean instruments with alcohol if moving to a new location
- ▶ Use different, clean instruments for closure

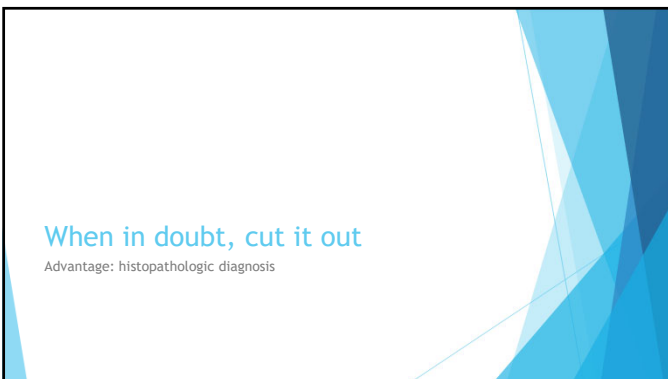


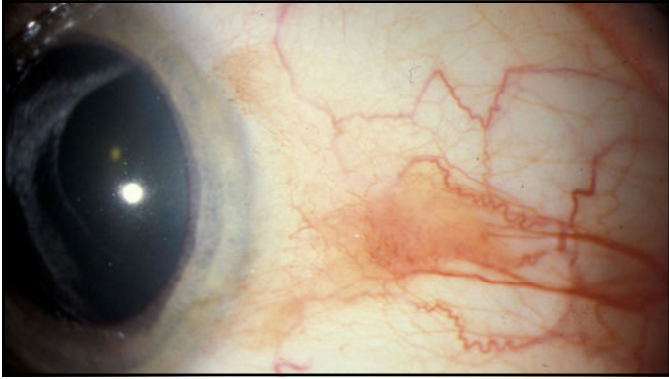


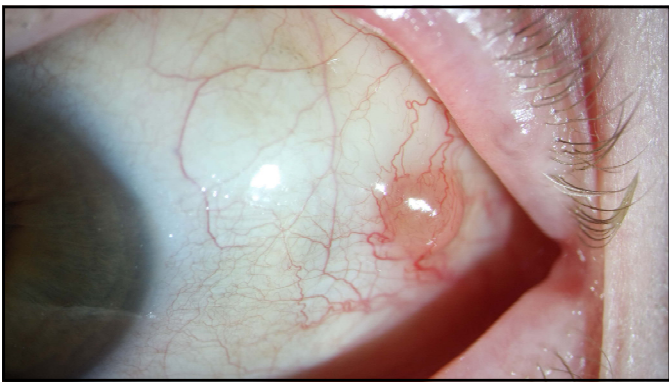












Topical Therapy

- ▶ 5-fluorouracil (1%)
- ▶ Interferon alpha-2b (1 million IU/mL)
- ▶ Mitomycin C (0.02-0.04%)
- ▶ More time, more follow-up required
- ▶ Primary treatment, chemoreduction before surgery, or adjuvant after surgery
- ▶ Insurance coverage can be challenging

5FU

- ▶ Antimetabolite
- ▶ Interrupts DNA replication and cell growth
- ▶ 1% concentration
- ▶ 1 cycle: QID for 1 week, followed by 3 weeks of no medication
- ▶ Repeat until resolution
- ▶ 82-100% response rate
- ▶ 10-14% recurrence

- ▶ Side effects:
 - ▶ Pain
 - ▶ Tearing
 - ▶ Redness
 - ▶ Eyelid edema
 - ▶ Keratopathy
- ▶ Generally well-tolerated
- ▶ Compounded
- ▶ Low cost compared with others in US

IFN

- ▶ Glycoprotein
- ▶ Acts at cell surface
- ▶ 1 million IU/mL
- ▶ Topical and subconjunctival use
 - ▶ 10 MIU for subconj
- ▶ Topical: QID until resolution, then continue for 1-3 additional months
- ▶ Subconj: once per month
- ▶ 81-100% response rate
- ▶ 0-4% recurrence

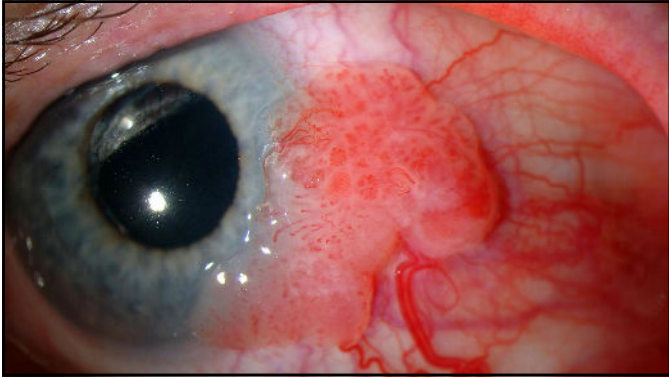
- ▶ Side effects:
 - ▶ Keratoconjunctivitis
 - ▶ Flu-like symptoms with injection
- ▶ Very well-tolerated
- ▶ Compounded
- ▶ Expensive in US, less expensive in other countries
- ▶ Difficult to obtain in US

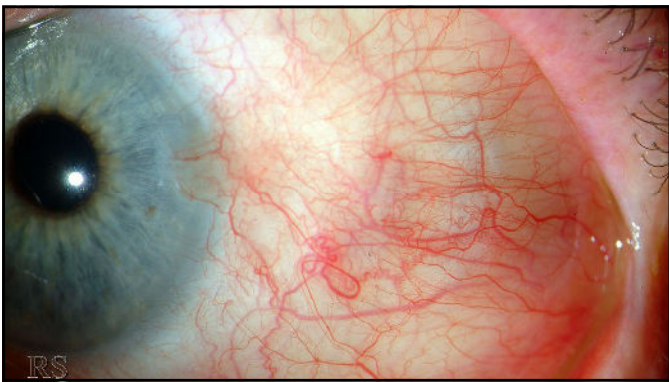


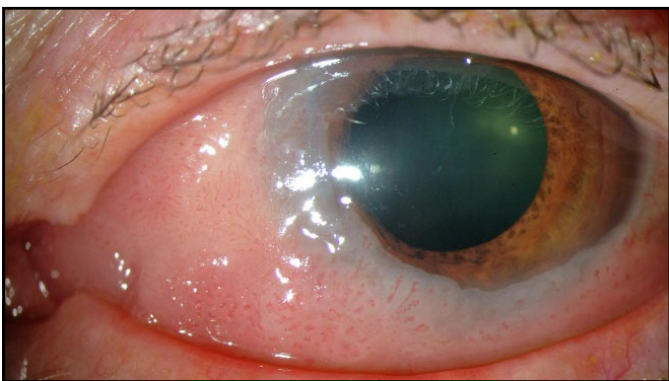
MMC

- ▶ Alkylating agent
- ▶ 0.02-0.04%
- ▶ 1 cycle: QID for 1 week on, 1 week off, 1 week on, 1 week off
 - ▶ Wait AT LEAST 1-2 weeks after surgery for healing
- ▶ Repeat until resolution
- ▶ 76-100% response rate
- ▶ 0-20% recurrence

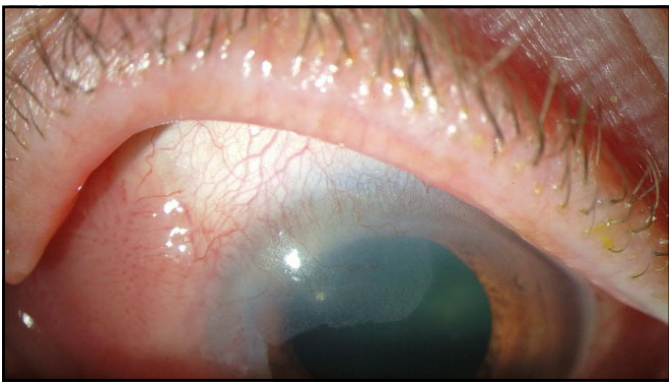
- ▶ Side effects:
 - ▶ Keratoconjunctivitis
 - ▶ Redness, itching, tearing, pain
 - ▶ Corneal erosion, punctate staining
 - ▶ Hyperemia
 - ▶ Punctal stenosis
 - ▶ MUST place punctal plugs
 - ▶ Limbal stem cell deficiency
- ▶ Poorly tolerated compared with others
- ▶ Compounded
- ▶ Moderate cost in US

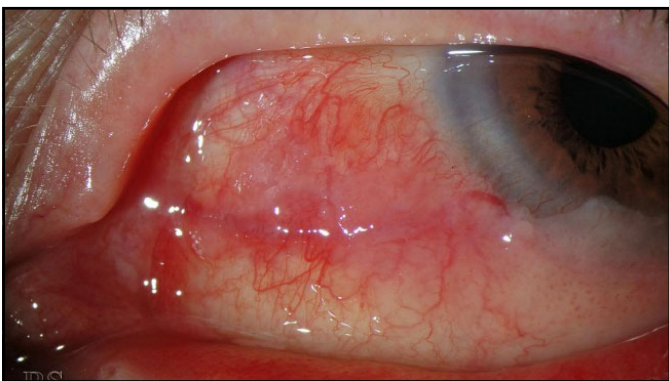


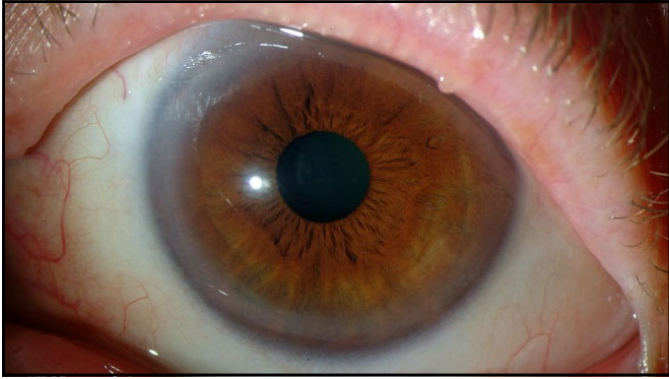




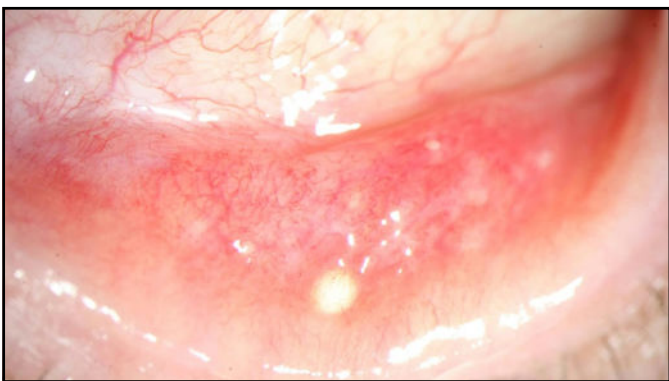













**Conclusions:
Ocular Surface
Tumors**

- ▶ Pigmented or amelanotic
- ▶ Nevus, CAM, PAM, Melanoma, OSSN
- ▶ Image-assisted diagnosis
- ▶ Surgical excision
- ▶ Topical chemotherapy
 - ▶ 5FU, IFN, MMC
- ▶ Combination surgery and chemotherapy
- ▶ Individualized based on extent of involvement and discussion with patient
- ▶ When in doubt, cut it out


Topics: Tumor Tour

- ▶ Ocular surface
- ▶ Iris
- ▶ Posterior segment



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Iris Tumors
The Pigment Spectrum

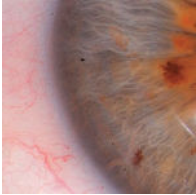
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Iris freckle

- ▶ Most common melanocytic lesion of iris stroma
- ▶ ≥60% of the population
- ▶ Pigmented
- ▶ Flat
- ▶ Does not distort the iris architecture
- ▶ Minimal risk for melanoma



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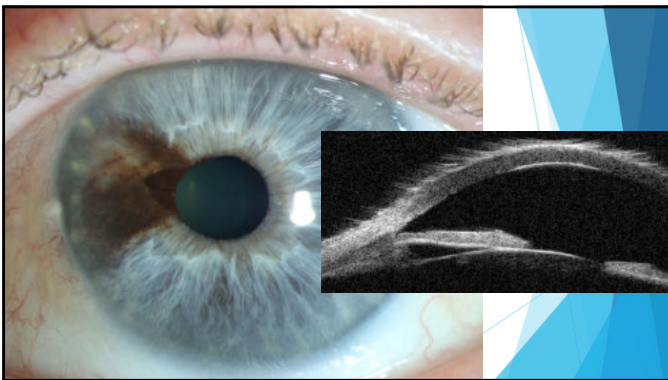
Iris Nevus

- ▶ 5% of the population
- ▶ Iris stroma
- ▶ Pigmented or amelanotic
- ▶ Circumscribed or diffuse
- ▶ Dome-shaped or flat
- ▶ Distorts iris architecture
- ▶ Associated pigment dusting, corectopia, angle involvement, secondary cataract, cyst
- ▶ >80% in inferior iris



Iris Nevus Transformation to Melanoma

- ▶ ~3% risk at 5 years
- ▶ A - age young <40 years
- ▶ B - blood (hyphema or microhyphema)
- ▶ C - inferior clock hour location 4:00-9:00
- ▶ D - diffuse configuration involving entire iris surface
- ▶ E - ectropion uveae
- ▶ F - feathery tumor margins




Sector Iris Nevus

- ▶ Involves section of iris
- ▶ Extends from pupil to angle
- ▶ Typically 3-4 clock hours
- ▶ May be congenital
- ▶ May be variant of melanocytosis



Iris Sector Melanocytosis

- ▶ Mammillations
- ▶ 1 in 400 risk for uveal melanoma with oculodermal melanocytosis
 - ▶ Probably lower for sector melanocytosis



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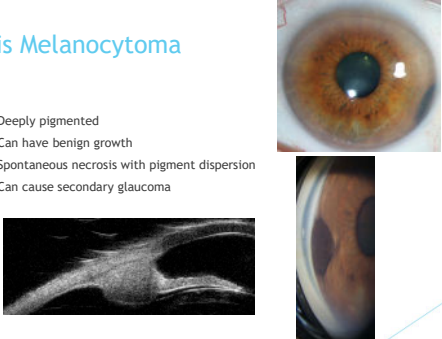
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Iris Melanocytoma

- ▶ Deeply pigmented
- ▶ Can have benign growth
- ▶ Spontaneous necrosis with pigment dispersion
- ▶ Can cause secondary glaucoma



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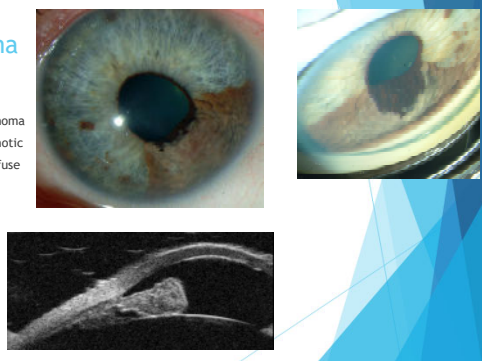
Iris Tumors

The Pigment Spectrum

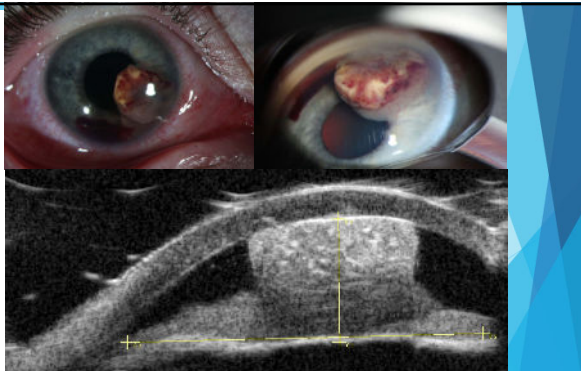
- Freckle
- Nevus
- Melanocytoma
- Malignant melanoma

Iris Melanoma

- ▶ 4% of all uveal melanoma
- ▶ Pigmented or amelanotic
- ▶ Circumscribed or diffuse
- ▶ Dome-shaped or flat
- ▶ Corectopia 45%
- ▶ Ectropion uveae 24%
- ▶ Angle seeding 28%
- ▶ Hyphema 3%
- ▶ EOE 3%



The slide features three images: a clinical photograph of a human eye showing a pigmented iris lesion, a magnified clinical view of a dome-shaped iris melanoma, and a UBM image showing the cross-section of the iris with a melanoma lesion.



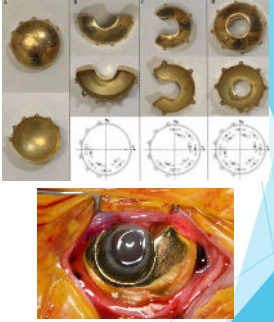
The slide features three images: a clinical photograph of a human eye showing a pigmented iris lesion, a magnified clinical view of a dome-shaped iris melanoma, and a UBM image showing the cross-section of the iris with a melanoma lesion.

Workup

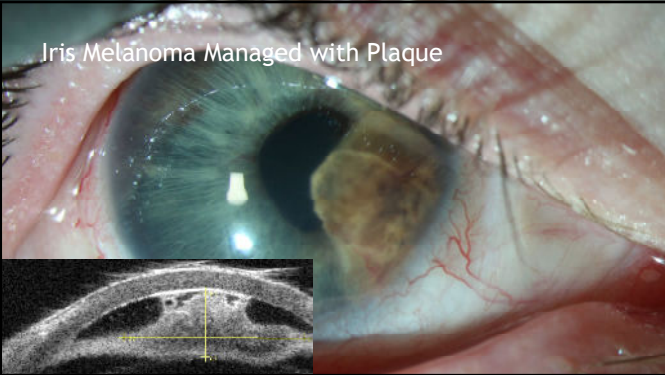
- ▶ Complete ophthalmic examination, including IOP check, gonioscopy, dilated fundusoscopic examination
- ▶ Slit lamp and gonio photography
- ▶ UBM
- ▶ LFTs, lung and liver imaging

Treatment

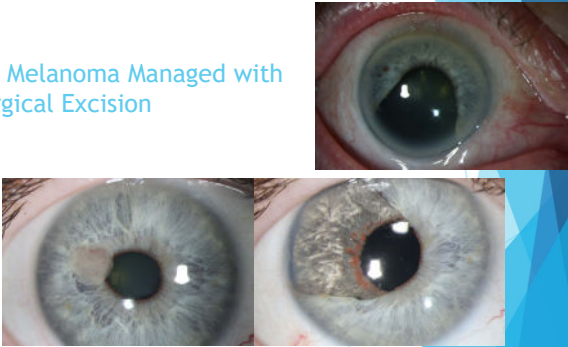
- ▶ Iridocyclectomy
 - ▶ Small
 - ▶ Well-circumscribed
 - ▶ Preferably not in CB
- ▶ Radiation
 - ▶ Most lesions
 - ▶ Plaque
 - ▶ Proton
 - ▶ Gamma
- ▶ Enucleation
 - ▶ Large
 - ▶ Glaucoma
 - ▶ Poor VA potential



Iris Melanoma Managed with Plaque



Iris Melanoma Managed with Surgical Excision



Prognosis

- ▶ Favorable compared with other uveal melanoma
- ▶ <10% metastasis at 10 years
- ▶ Biopsy can be done for cytogenetics

Iris Tumors
Misc

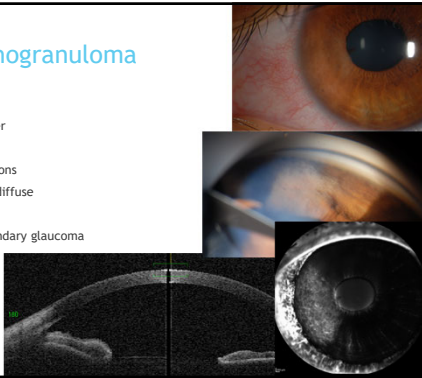
- Histiocytic
- Vascular
- Cystic

Iris Tumors
Misc

- Histiocytic
- Vascular
- Cystic

Iris Juvenile Xanthogranuloma

- ▶ Histiocytic inflammatory disorder
- ▶ Cutaneous pink-yellow lesions
- ▶ Iris may not have cutaneous lesions
- ▶ Pink-brown discrete nodules or diffuse
- ▶ Can simulate uveitis
- ▶ Spontaneous hyphema and secondary glaucoma
- ▶ Fibrillary material by OCT
- ▶ Managed with corticosteroids

The image block contains three photographs. The top right shows a clinical view of an eye with a pinkish-brown lesion on the iris. The middle right shows a close-up of the iris with a similar lesion. The bottom left shows an OCT scan of the iris, highlighting fibrillary material.

Iris Tumors
Misc

- Histiocytic
- Vascular
- Cystic

Iris Tumors
Misc

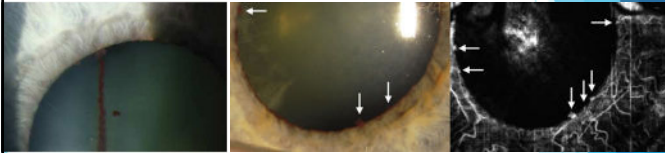
- Histiocytic
- Vascular
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Iris Hemangioma

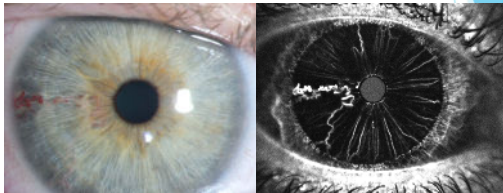
- ▶ Usually sporadic
- ▶ Capillary can regress with time
- ▶ Cavernous can be solitary or microhemangiomatosis
 - ▶ Recurrent hyphema
- ▶ Racemose hemangioma = AV malformation
- ▶ Iris varix appears blue
- ▶ FA can help with diagnosis
- ▶ Laser or excision for recurrent hyphema



Iris Microhemangiomatosis




Iris Racemose Hemangioma



Iris Varix

- ▶ Hypofluorescent on FA due to internal thrombosis
- ▶ Can be excised if recurrent hyphema



Iris Tumors
Misc

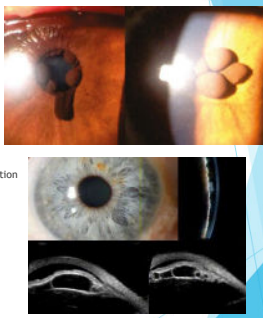
- Histiocytic
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Iris Tumors
Misc

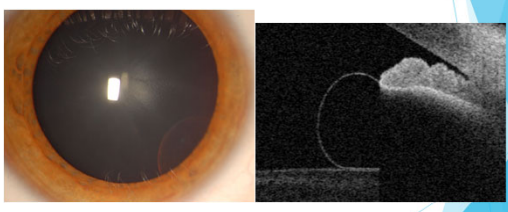
- Histiocytic
- Vascular
- Cystic

Iris IPE Cyst

- ▶ Central (pupillary)
 - ▶ Multiple = autosomal dominant
 - ▶ Wrinkled = iris flocculi
 - ▶ Familial dissecting aortic aneurysm
- ▶ Midzonal (retroiridic)
 - ▶ Elevated mass, sometimes seen better with dilation
 - ▶ Pigmented, blocks light on transillumination
- ▶ Peripheral (iridociliary)
 - ▶ Common
 - ▶ Young women
 - ▶ Can see with dilation sometimes
- ▶ Dislodged
 - ▶ Can float, lodge in angle



Peripheral Iris IPE Cyst

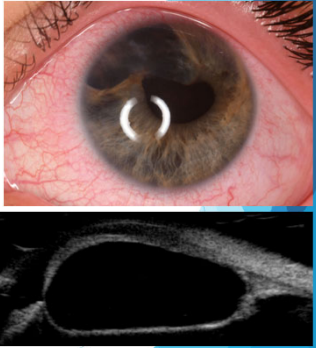


Dislodged Iris IPE Cyst



Iris Stromal Cyst

- ▶ Usually clear (unlike pigmented IPE cyst)
- ▶ Congenital usually found by age 10
- ▶ Acquired can be idiopathic or from trauma/surgery
- ▶ Spontaneous rupture can cause uveitis
- ▶ Needle aspiration to collapse cyst
- ▶ Absolute alcohol to prevent recurrence
- ▶ Complete surgical excision of cyst wall



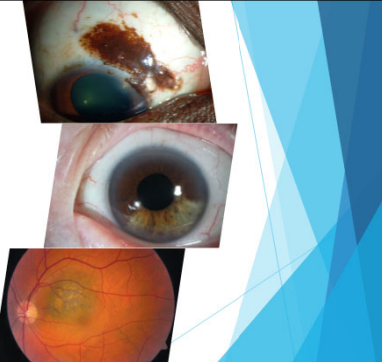
The top image shows a clinical view of a human eye with a white, ring-shaped cyst on the iris. The bottom image is an OCT scan showing a dark, well-defined cystic structure within the iris stroma.

Conclusions: Iris Tumors

- ▶ Freckle, nevus, melanocytoma, melanoma, histiocytic, vascular, cystic
- ▶ Multimodal imaging
- ▶ Surgical excision
- ▶ Radiation
- ▶ Corticosteroids
- ▶ Individualized based on symptoms, location, size

Topics: Tumor Tour

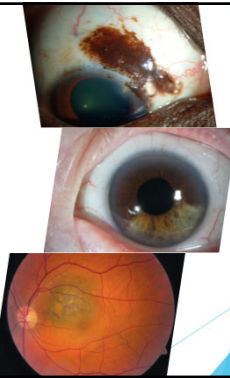
- ▶ Ocular surface
- ▶ Iris
- ▶ Posterior segment



The collage includes: a close-up of a brown pigmented lesion on the ocular surface; a clinical view of a human eye with a large, dark, pigmented mass on the iris; and a fundus photograph showing a large, orange, pigmented lesion in the posterior segment of the eye.

Topics: Tumor Tour

- ▶ Ocular surface
- ▶ Iris
- ▶ Posterior segment



Posterior Segment Tumors

The Pigment Spectrum

- Melanocytosis
- Nevus
- Malignant melanoma


Posterior Segment Tumors

The Pigment Spectrum

- Melanocytosis
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Ocular Melanocytosis

- ▶ Aka Nevus of Ota
- ▶ Sclera, Iris, Choroid - need UBM to r/o CB MM
- ▶ Cutaneous involvement - oculodermal melanocytosis
- ▶ 1 in 400 risk of uveal melanoma
- ▶ Risk for orbital and meningeal melanoma - MRI brain and orbits





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Choroidal and Ciliary Body Nevus

- ▶ Most common benign primary intraocular tumor (except maybe Fuchs adenoma)
- ▶ ~8% of US white population
- ▶ 1 in 8,845 risk of transformation to melanoma
- ▶ Detected after puberty
- ▶ Pigmented or amelanotic
- ▶ Flat or dome
- ▶ Mean age 60, diameter 5.5, thickness <2 mm
- ▶ Associated PED, drusen, RPE trough, CNV



Choroidal Nevus Transformation to Melanoma

Clinical Features

- ▶ Io
- ▶ Find
- ▶ Small
- ▶ Ocular
- ▶ Melanoma
- ▶ Using Helpful
- ▶ Hints
- ▶ Daily
- ▶ Thickness > 2mm
- ▶ Subretinal Fluid
- ▶ Symptoms (flash/floater)
- ▶ Orange pigment
- ▶ Margin < 3 mm from ON
- ▶ Ultrasound Hollow
- ▶ Halo
- ▶ Drusen

Choroidal Nevus Transformation to Melanoma

Multimodal Imaging Features

- ▶ Io
- ▶ Find
- ▶ Small
- ▶ Ocular
- ▶ Melanoma
- ▶ Doing Imaging
- ▶ Thickness > 2mm
- ▶ Subretinal Fluid
- ▶ Symptoms (VA <20/50)
- ▶ Orange pigment
- ▶ Melanoma acoustic hollowness
- ▶ Diameter > 5mm

5-Year Risk for Growth with 1 Factor

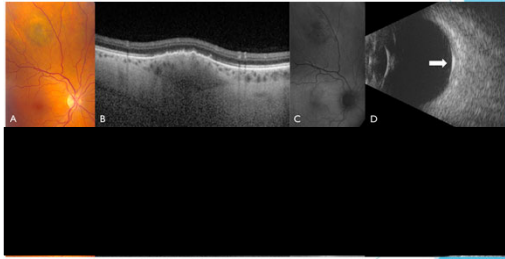
- ▶ **Thickness > 2mm**
- ▶ Subretinal Fluid
- ▶ Symptoms (VA <20/50)
- ▶ **Orange pigment**
- ▶ Melanoma acoustic hollowness
- ▶ Diameter > 5mm
- ▶ 26%, **8-fold**
- ▶ 27%, 3-fold
- ▶ 9%, 2-fold
- ▶ **37%**, 3-fold
- ▶ 23%, 2-fold
- ▶ 12%, NS

Risk Increases with More Factors

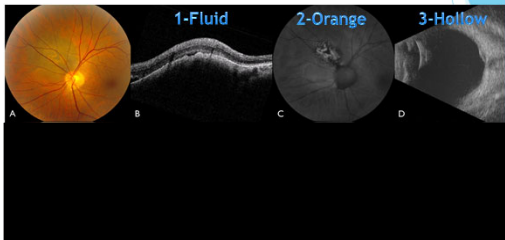
- ▶ 1 Factor: 11%, 3-fold
- ▶ 2 Factors: 22%, 11-fold
- ▶ 3 Factors: 34%, 15-fold
- ▶ 4 Factors: 51%, 15-fold
- ▶ 5 Factors: 55%, 26-fold

← Always Refer

How Many Risk Factors?

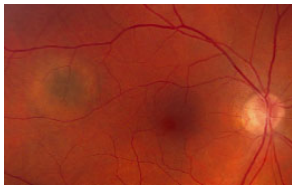


How Many Risk Factors?



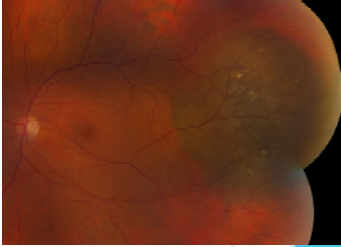
Halo Choroidal Nevus

- ▶ Non-pigmented halo
- ▶ 5% of nevi
- ▶ Associated with cutaneous melanoma history
- ▶ Could be autoimmune phenomenon



Giant Choroidal Nevus

- ▶ LBD >10 mm
- ▶ Features of chronicity
 - ▶ RPE changes
 - ▶ Drusen
- ▶ 18% transform to melanoma



Posterior Segment Tumors

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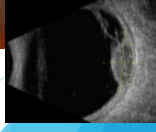
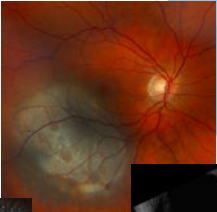
Posterior Segment Tumors

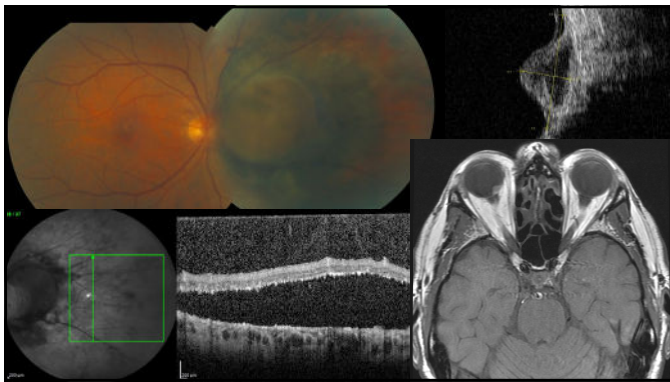
The Pigment Spectrum

- Melanocytosis
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Choroidal and Ciliary Body Melanoma

- ▶ 6 per 1 million
- ▶ Melanotic or amelanotic
- ▶ Dome or mushroom-shaped (broke through Bruch's)
 - ▶ Rarely diffuse
- ▶ CB hidden by iris until large
 - ▶ Sentinel vessel, EOE, lens subluxation, sector cataract
 - ▶ Rarely ring-shaped and diffuse
- ▶ Hollow/low internal reflectivity by ultrasound
- ▶ Double circulation sign by FA/ICG





This composite image illustrates the diagnostic workup for a choroidal melanoma. It includes fundus photographs showing a large, pigmented lesion, an OCT scan showing a dome-shaped elevation of the retina, and an axial MRI scan showing a well-circumscribed, homogeneous enhancing mass in the choroid.

Workup

- ▶ Complete ophthalmic examination
- ▶ Fundus photography
- ▶ OCT
- ▶ US
- ▶ Sometimes MRI brain/orbits
- ▶ LFTs, lung and liver imaging

Treatment

- ▶ Radiation
 - ▶ Most lesions
 - ▶ Plaque
 - ▶ Proton
 - ▶ Gamma
- ▶ Enucleation
 - ▶ Large
 - ▶ Glaucoma
 - ▶ Poor VA potential
- ▶ Less often
 - ▶ TTT - more recurrence
 - ▶ Sclerouvectomy
 - ▶ New nanoparticles

Treatment Side Effects

- ▶ Maculopathy
 - ▶ Anti-VEGF for CME
- ▶ Retinopathy
 - ▶ Anti-VEGF
 - ▶ PRP
 - ▶ PPV for non-clearing VH
- ▶ Papillopathy
 - ▶ Anti-VEGF/corticosteroids attempted
- ▶ Cataract
- ▶ NVG
- ▶ Diplopia

Prognosis

- ▶ Depends on size and cytogenetics
- ▶ 12% for small ≤ 3 mm to 49% for large > 8 mm thick at 10 years
- ▶ FNAB or vitrectomy biopsy for cytogenetics
 - ▶ Class 1A, 1B, 2
 - ▶ TCGA class A, B, C, D
 - ▶ Risks ranging from $< 10\%$ to $> 70\%$ at 5 years
- ▶ Adjuvant therapy
 - ▶ Clinical trials

Posterior Segment Tumors

The Amelanotic

Vascular

Lymphoma

Retinoblastoma

Posterior Segment Tumors

The Amelanotic


Vascular

Lymphoma

Retinoblastoma

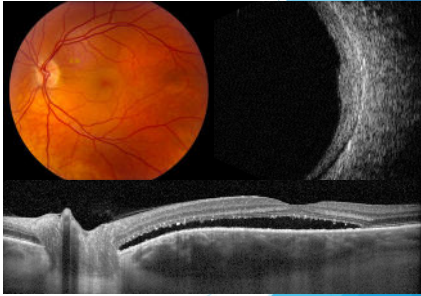
Choroidal Hemangioma

- ▶ Circumscribed type
- ▶ Often unilateral
- ▶ Adults
- ▶ Dome-shaped
- ▶ Red-orange
- ▶ Posterior fundus
- ▶ Dense by US
- ▶ Early hyper with diffuse leakage by FA
- ▶ Early filling with late washout by ICG
- ▶ Not associated with systemic disease



Treatment


- ▶ Observe if asymptomatic
- ▶ Can cause hyperopic shift
- ▶ Associated SRF can reduce VA
- ▶ PDT
- ▶ Radiotherapy if resistant



The image shows a fundus photograph on the left and an OCT scan on the right. The fundus photograph displays a large, orange, well-circumscribed lesion in the choroid. The OCT scan shows a dome-shaped elevation of the choroid with a hyperreflective layer, characteristic of a choroidal hemangioma.

Choroidal Hemangioma


- ▶ Diffuse type
- ▶ Children
- ▶ Associated with Sturge Weber Syndrome
- ▶ Typically ipsilateral to cutaneous port wine mark
- ▶ Tomato catsup fundus
- ▶ Diffuse choroidal thickening
- ▶ Serous retinal detachment
- ▶ Neovascular glaucoma



The image includes a clinical photograph of a child's face showing a port wine stain on the forehead and a fundus photograph showing a diffuse, reddish, 'tomato catsup' appearance of the choroid.

Treatment

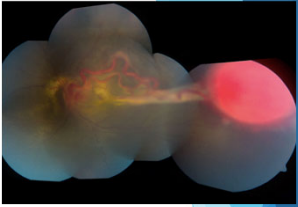
- ▶ Treat amblyopia
- ▶ Oral propranolol
- ▶ External beam radiotherapy
- ▶ Enucleation for blind, painful eye



The image shows a clinical photograph of a child's face with a port wine stain and a fundus photograph showing a large, reddish, lobulated lesion in the choroid.

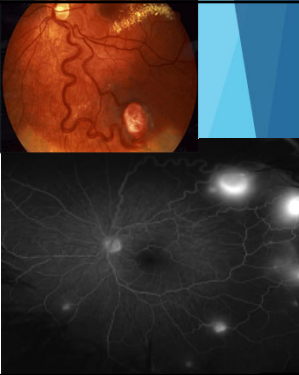
Retinal Hemangioblastoma

- ▶ Solitary without systemic disease
- ▶ Multiple associated with systemic Von Hippel Lindau
 - ▶ Autosomal dominant
 - ▶ Chromosome 3
 - ▶ Inactivation of tumor suppressor gene
- ▶ Three tumors: Retinal/cerebellar hemangioblastoma, pheochromocytoma, renal cell carcinoma
 - ▶ Also pancreatic neuroendocrine tumor, endolymphatic sac tumor, cysts in pancreas, kidney genitourinary tract



Retinal Hemangioblastoma

- ▶ Red-pink tumor in peripheral retina or optic disc
- ▶ Exudation
- ▶ Traction
- ▶ Feeding artery and draining vein
- ▶ Leakage by FA
 - ▶ FA to discover additional smaller, subtle lesions
- ▶ Observe if asymptomatic
- ▶ Laser, cryotherapy, PDT, TTT, radiotherapy
- ▶ Corticosteroids to prevent exudative response
- ▶ Modest benefit of anti-VEGF



Posterior Segment Tumors

The Amelanotic

- Vascular
- Lymphoma
- Retinoblastoma

Posterior Segment Tumors

The Amelanotic


Vascular

Lymphoma

Retinoblastoma

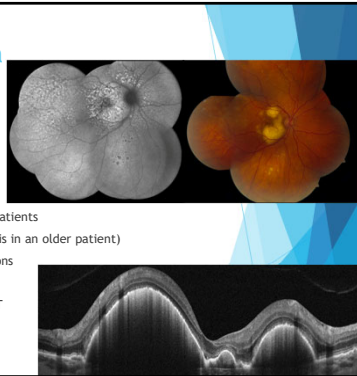
Choroidal Lymphoma

- ▶ Typically non-Hodgkin B-cell
- ▶ Often unilateral
- ▶ Associated conjunctival and orbital involvement
- ▶ Hollow choroid thickening by US
 - ▶ Epibulbar mass
- ▶ Seesick by OCT
- ▶ Look for conjunctival salmon patch to biopsy
- ▶ Systemic lymphoma workup
- ▶ Systemic treatment (rituximab)
- ▶ Or local radiotherapy



Vitreoretinal Lymphoma

- ▶ Typically B cell
- ▶ Bilateral in 90%
- ▶ Associated with CNS lymphoma
 - ▶ Need MRI, lumbar puncture
- ▶ Increased incidence in immunosuppressed patients
- ▶ Vitreous cell (can mimic intermediate uveitis in an older patient)
- ▶ Yellow retinal, subretinal, and sub-RPE lesions
- ▶ Need biopsy for diagnosis (eye or CNS)
- ▶ Intravitreal or systemic chemotherapy, EBRT



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The Amelanotic

Vascular

Lymphoma

Retinoblastoma

Posterior Segment Tumors

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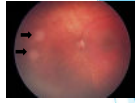
Retinoblastoma

- ▶ Most common intraocular malignancy of childhood
- ▶ Bilateral or unilateral/Somatic or germline
- ▶ Life-threatening if untreated
- ▶ Leukocoria (56%)
- ▶ Strabismus (24%)
- ▶ Decreased vision (7%)



Clinical appearance

- ▶ Small, translucent retinal lesion
- ▶ Large, white calcified mass with feeding artery, draining vein, and retinal detachment
- ▶ Advanced, extraocular orbital mass



Enucleation

- ▶ Unilateral, advanced disease
- ▶ Optic nerve involvement
- ▶ Fewer follow-up visits
- ▶ No chemotherapy or radiation exposure

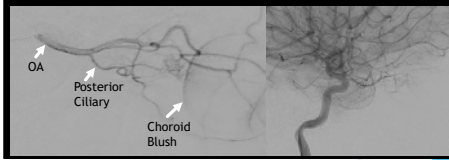


Group E Rb Managed with Enucleation

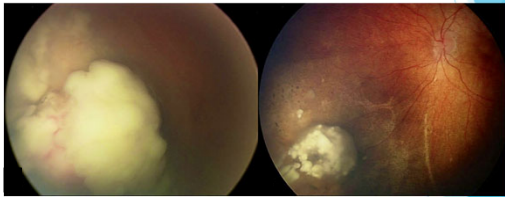


Chemotherapy

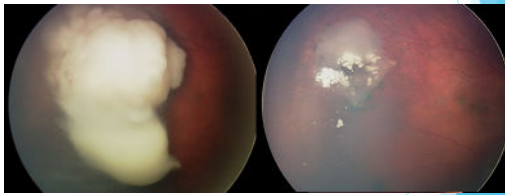
- ▶ Systemic, intravenous chemotherapy
 - ▶ Consolidate: cryotherapy, TTT
- ▶ Targeted intra-arterial chemotherapy
- ▶ Intravitreal chemotherapy



Group D M5 T1



Group D M5, IvitM



Systemic Monitoring

- ▶ Germline at risk for second cancers
- ▶ MUST have a pediatric oncologist
- ▶ Get MRI brain and orbits with and without contrast with fat suppression q6months until age 5
 - ▶ To monitor for pineoblastoma
- ▶ Germline needs to be counseled about in vitro fertilization and preimplantation diagnosis

Conclusions: Posterior Segment Tumors

- ▶ Melanocytosis, nevus, melanoma, vascular, lymphoma, retinoblastoma
- ▶ Multimodal imaging
- ▶ Radiation
- ▶ Chemotherapy
- ▶ Cryotherapy
- ▶ Laser
- ▶ Individualized based on symptoms, location, size
- ▶ Serious systemic implications

End of the Tour

- ▶ Eye tumors can be benign or malignant
- ▶ Treatment and prognosis individualized
- ▶ Can save vision or even save a life!



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