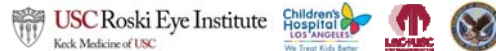


Dispelling Rumors about Tumors

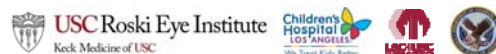
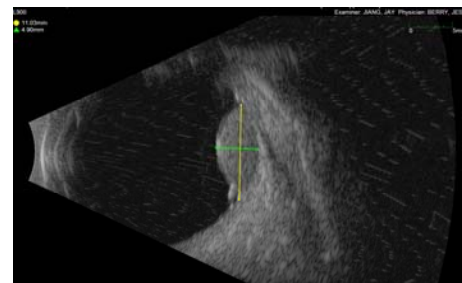
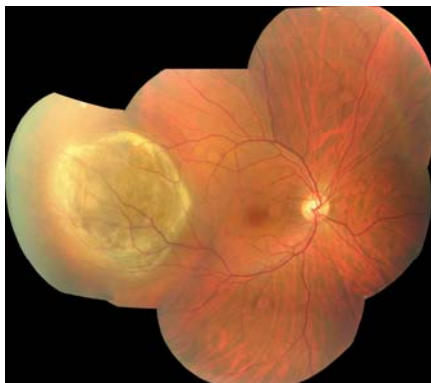
Jesse L. Berry, MD
Arizona Ophthalmology Society
2017

Associate Director, Ocular Oncology Service
Associate Program Director
USC/CHLA, Keck School of Medicine



Case

- 65 year old female presents with flashes



Diagnosis: Choroidal Melanoma

- 5% of all melanomas in the US
- most common primary IO tumor in adults
- 6 cases/million→1500 cases per year in US
- 50-70 years/women=men/Caucasian
- Diagnosis based on fundoscopy + ultrasound



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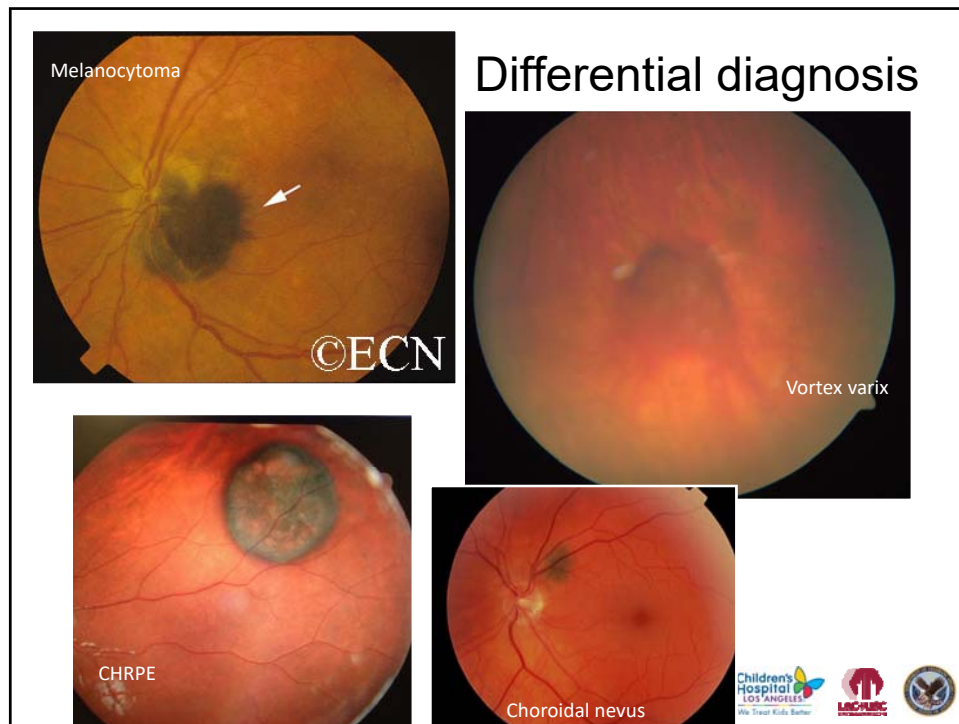


Rumor #1: Everything
that's pigmented is a
melanoma



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Rumor #2: Everything
that's pigmented **and**
elevated must be a
melanoma

Case: Melanoma v. Nevus?



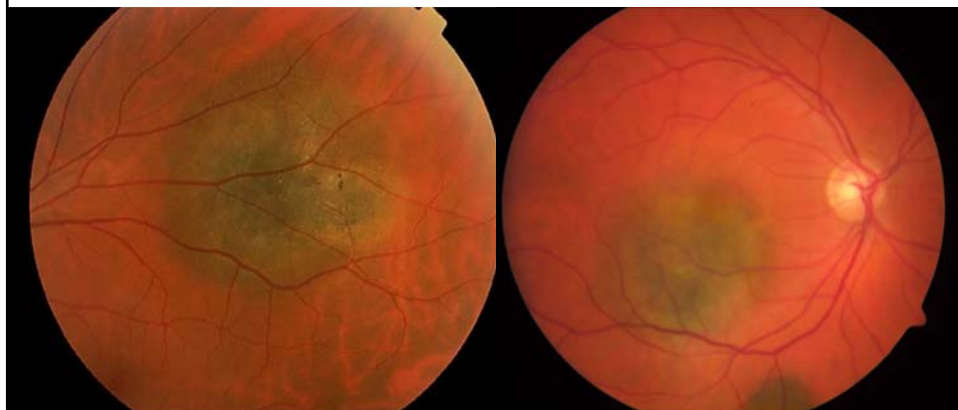
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Melanoma v. Nevus?



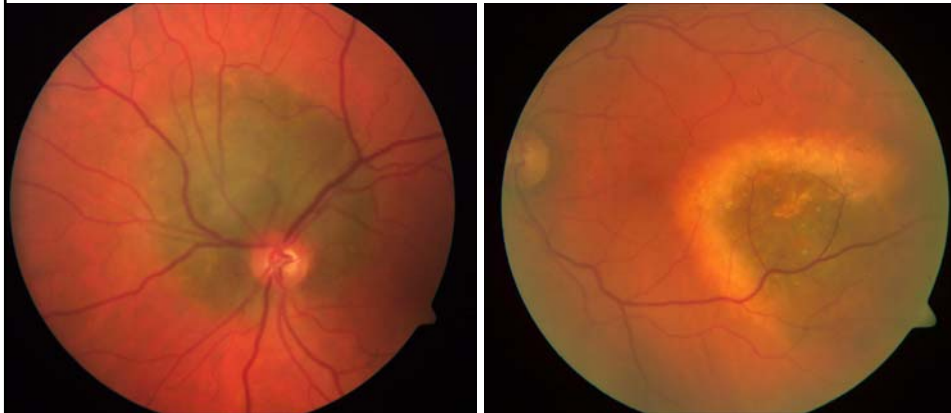
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Melanoma v. Nevus?



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Melanoma v. Nevus?



The only melanoma in the bunch



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Diagnosis: Choroidal Nevus

- Benign tumors
- Collection of bland spindle A melanocytes
- The edges are defined but not sharply demarcated
- Dark brown or grey pigmentation
- Amelanotic not unusual
- High risk features which predispose to growth
- Growth may or may not be a sign of malignancy



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Which nevi grow?

Feature	Feature in Nevi that progress to Melanoma (%)	HR
Thickness > 2mm	19	2
Fluid	27	3
Symptoms	23	2
Orange Pigment	30	3
Margin <3mm to disc	13	2
Ultrasonographic Hollowness	25	3
Halo Absence	7	6

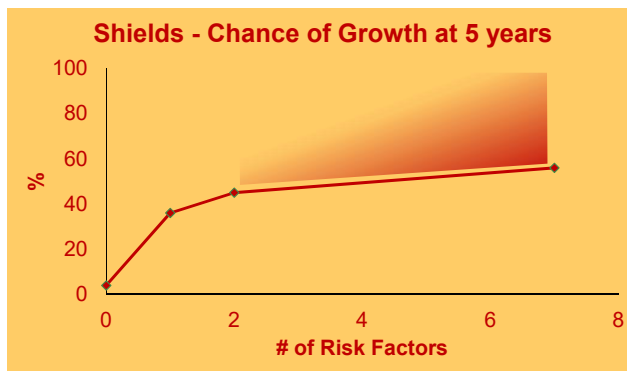
To	Find	Small	Ocular	Melanoma	Using Helpful	Hints
Thickness	Fluid	Symptoms	Orange	Margin to disc	Ultrasound	Halo
Thick	Orange	Fluids	Sometimes	Halo	Hollow	Melanoma Discovery
Thickness	Orange Pigment	Fluid	Symptoms	Halo	U/S Hollowness	Disc distance



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Which nevi grow?



- 27x greater risk ratio for 5 factors vs. 0 factors
- Growth not guarantee of malignancy
- Drusen are a sign of chronicity (favorable)
- Lifetime risk <1%



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Risk Factors

- Shields Combination of clinical factors
- If zero risk factors: 4% chance of growth/5 years
- If one risk factor: 36% chance of growth/5 years
- If 2 risk factors: >45% chance of growth/5 years
- If all risk factors: >56% chance of growth/5 years
- **27x greater risk ratio for 5 factors v 0 factors**
- Growth not guarantee of malignancy
- Drusen are a sign of chronicity (favorable)



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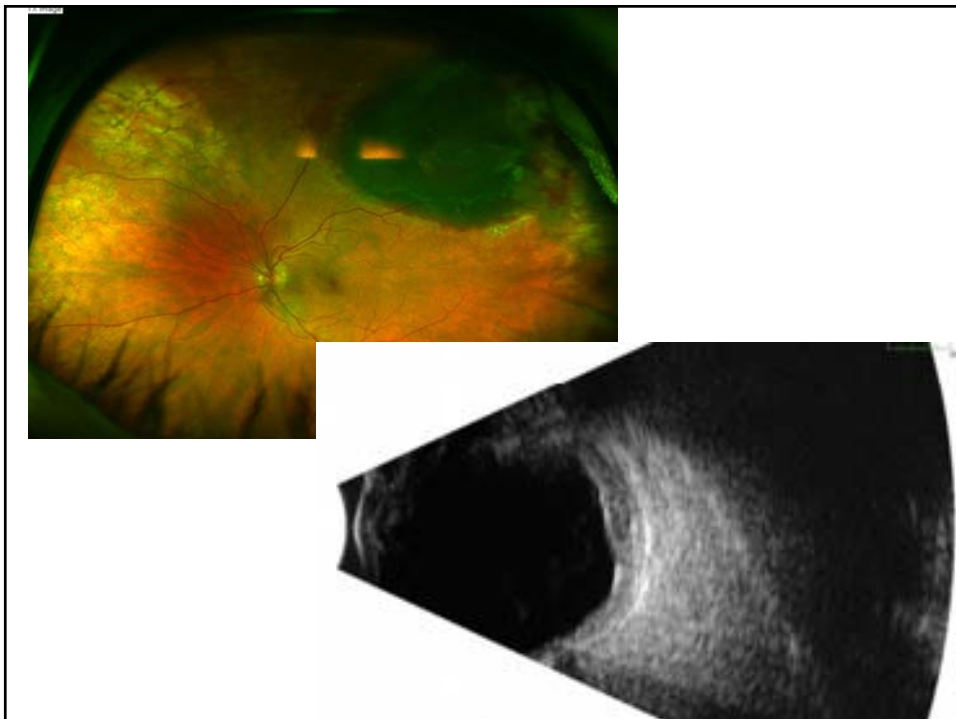
Rumor #3: Everything
that's pigmented **and**
elevated must be a
melanoma or a high
risk choroidal nevus

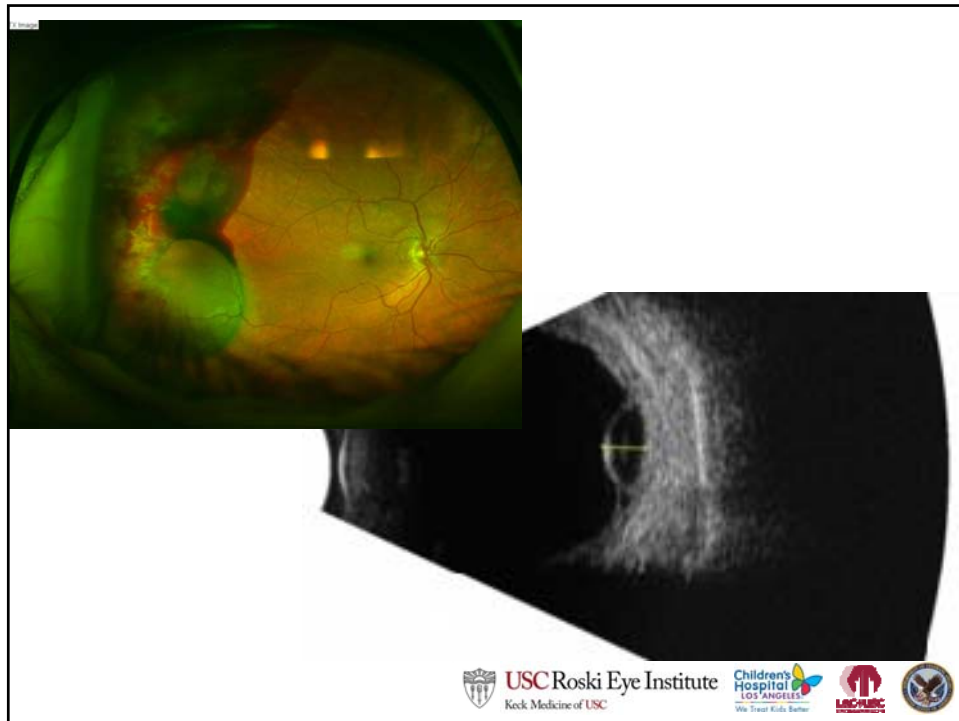


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Diagnosis: PEHCR

(peripheral exudative hemorrhagic chorioretinopathy)

- Large choroidal and subretinal heme
- Elderly, caucasian patients
- Associated with drusen and blood thinner use
- Usually no trauma
- Often temporal
- Lumpy bumpy and cystic spaces on Bscan

Case

- 55 yo F with known breast cancer presents with sudden loss of vision



Diagnosis: Choroidal Metastases

- most common choroidal tumor in adults
- Women=Breast, lung, unknown
- Men=Lung, Unknown, GI
- Lung often precedes the systemic diagnosis
- Breast rarely does
- Poorly circumscribed, amelanotic, associated with subretinal fluid
- Can be bilateral and/or multifocal (20%)

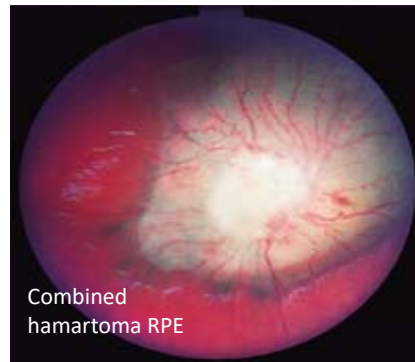
Rumor #4: Everything that's amelanotic must then be a met



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Choroidal hemangioma



Combined
hamartoma RPE

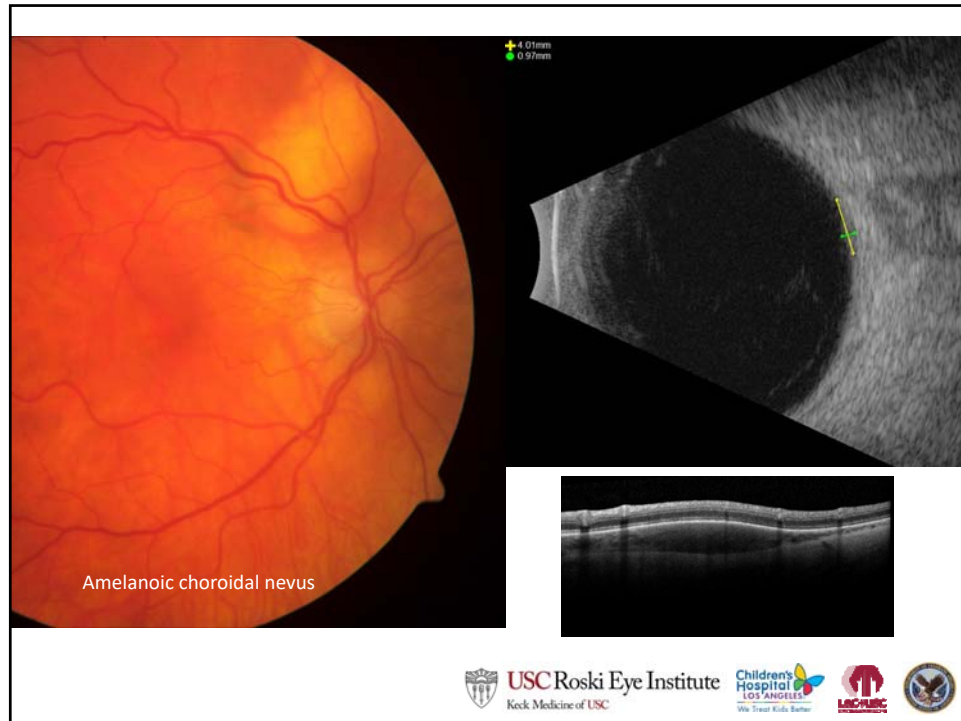


Choroidal osteoma



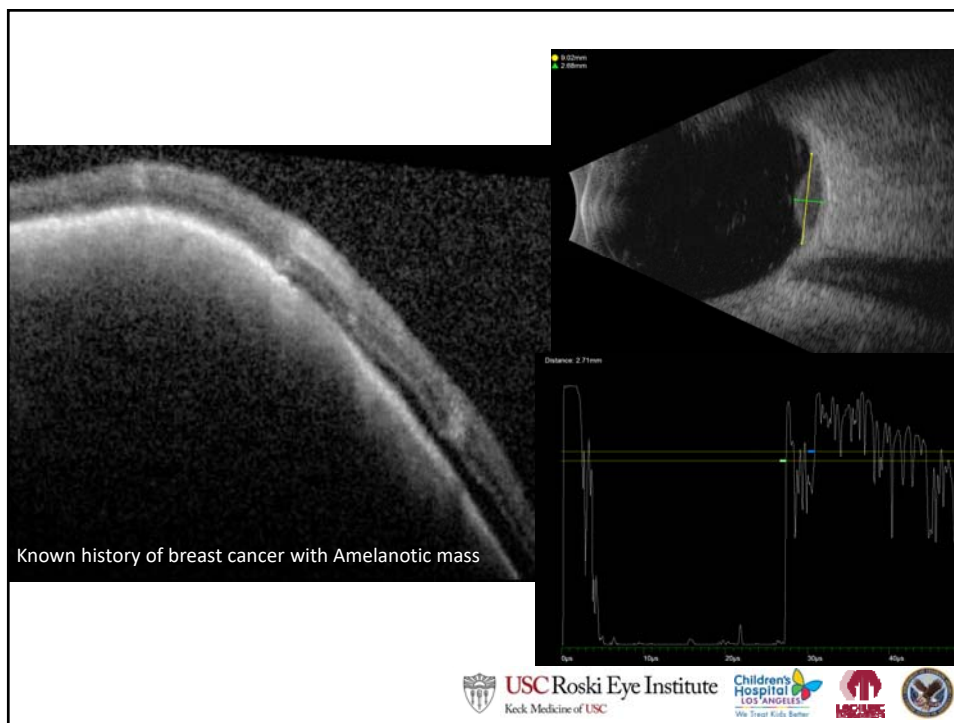
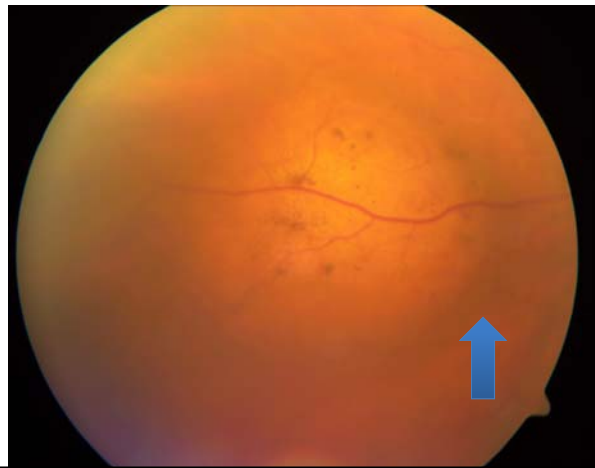
Astrocytic
hamartoma





case

- 65 yo F with known breast cancer presents for routine evaluation

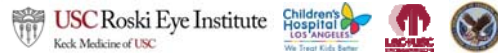


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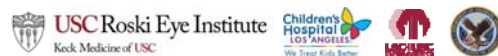


Breast cancer and primary Amelanotic choroidal melanoma

- Occam's razor does not always apply
- Patients can have two primary cancers

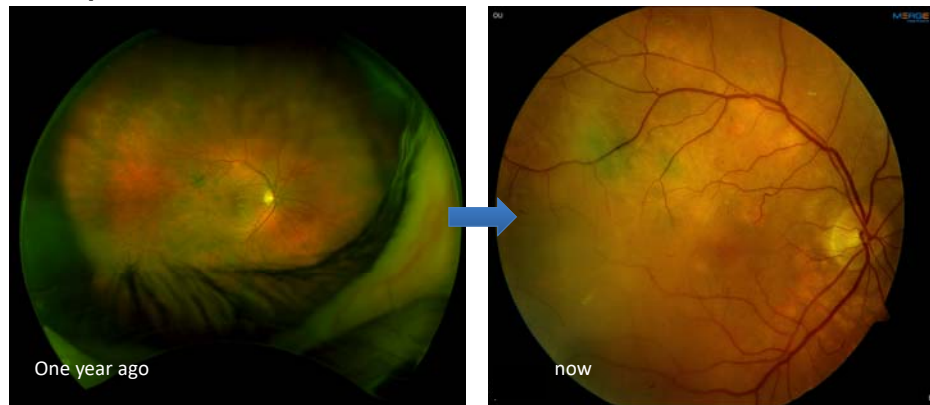


Rumor #5: Rules exist
for a reason
(but they don't always
apply in Ocular
Oncology...)

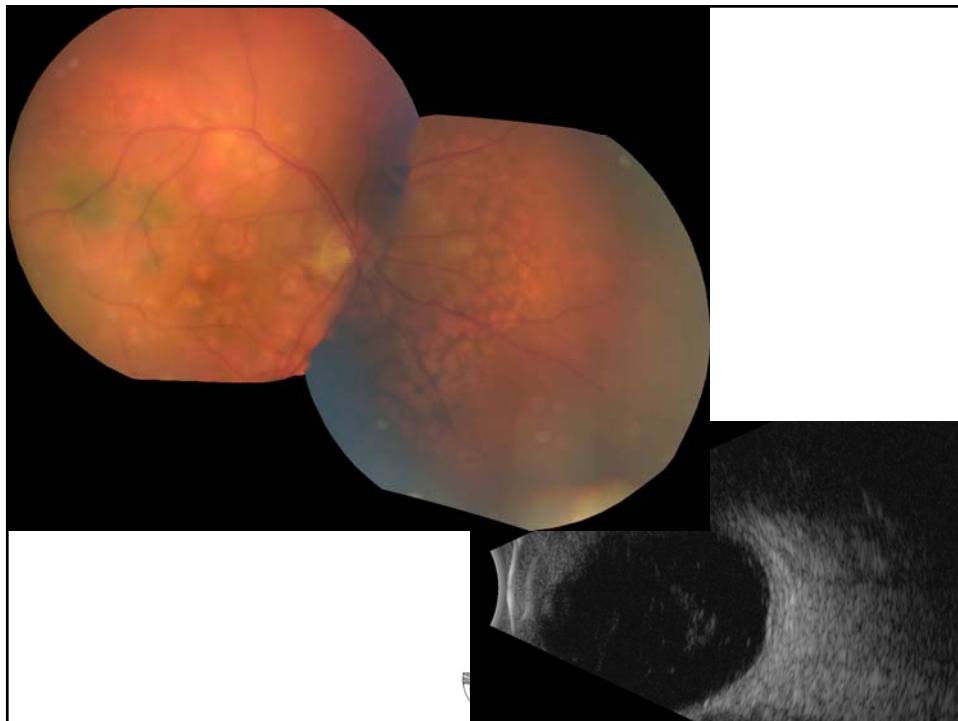


Case

- 65 yo M with a history of a nevus with painless vision loss



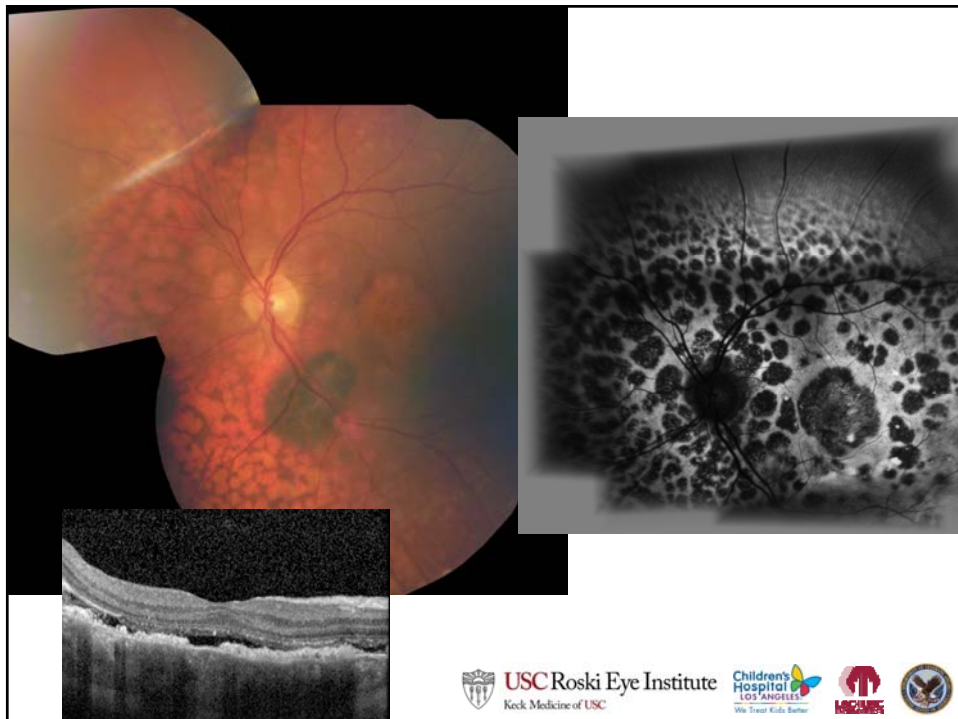
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Rumor #6: Everything that grows is cancer



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Diagnosis: BDUMP

(Bilateral diffuse uveal melanocytic proliferation)

- rare paraneoplastic ocular syndrome
- benign hyperplasia of uveal melanocytes
- The GROWTH is not CANCER
- Painless bilateral vision loss
- Diffuse pigment clumping and orange pigment
- Subretinal fluid is common
- May precede diagnosis of systemic carcinoma by 3-12 months

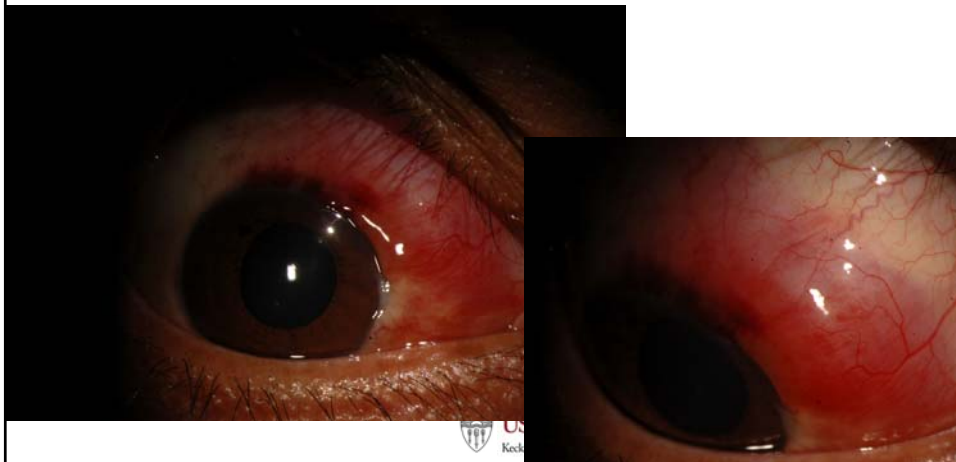


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Case

- A 45 year old male presents with a red, painless eye



Diagnosis: Ocular Adnexal Lymphoma

- Low grade Non-Hodgkins B-cell Lymphoma
 - 80% Extranodal marginal Zone lymphoma/Mucosa associated lymphoma
- Often affects the orbit, lacrimal gland, lids and conjunctiva
- Associated with systemic disease in 30%
- Conjunctival involvement is most associated with systemic disease
- Presents as a thick, velvety salmon patch



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Do not confuse with OSSN!



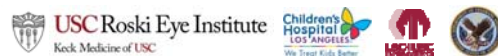
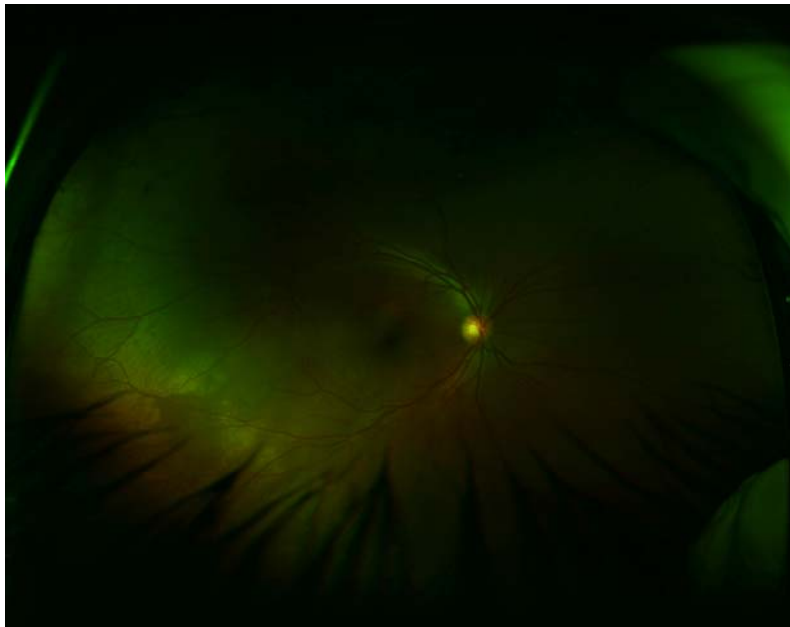
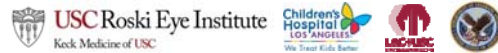
No touch
with cryo



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Rumor #7: Ocular adnexal lymphoma is always an external disease





Diagnosis: Uveal Lymphoma

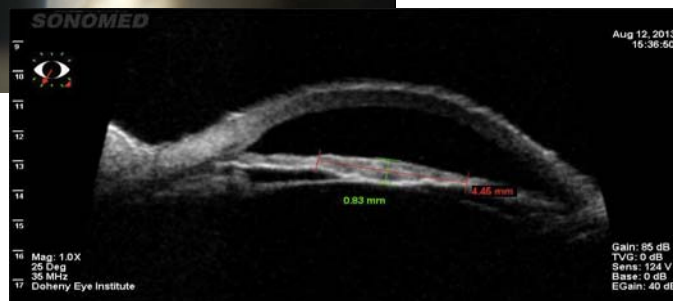
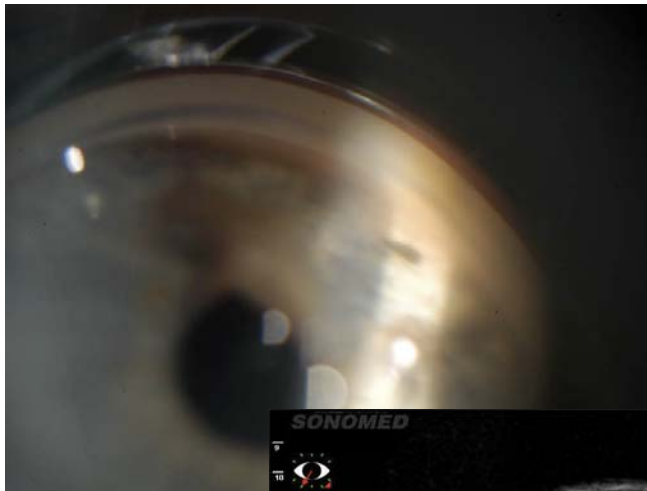
- Low-grade Non-Hodgkins B-cell Lymphoma (MALT)
- Often affects the choroid, iris and/or ciliary body
- Prolonged indolent course often misdiagnosed as birdshot, white dots syndrome or VKH
- Key finding: yellow-white choroidal infiltrates with associated crescentic choroidal thickening and hypofluorescence of ICG
- It is NOT vitreoretinal lymphoma (worse prognosis by far)
- 60% of patients with uveal lymphoma have OAL overlap
- 50% are bilateral
- 30% have systemic involvement
- Don't fall for the rumor - Dilate patients with a salmon patch

Case

- 56 yo M presented for evaluation of a 'spot', recently started timolol in the right eye only



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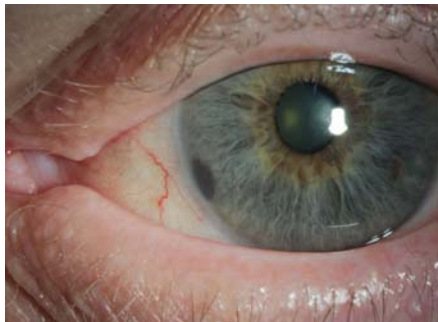


Diagnosis: Iris Nevus

- Common iris tumor
- Concern for melanoma with thickness >1 mm (average is 2mm), distortion of iris stroma, correctopia, ectropion uveae, feather borders, angle involvement
- Risk for malignant conversion ~8%
- Risk for metastatic disease is low ~3%
- Other high risk features: ABCDE
 - young **A**ge, **B**lood (hyphema), inferior **C**lock hour, **D**iffuse, **E**ctropion uveae
 - High pressure also a risk factor



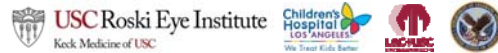
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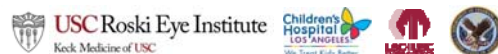


Rumor #8: Iris nevi are no big deal



Case continues

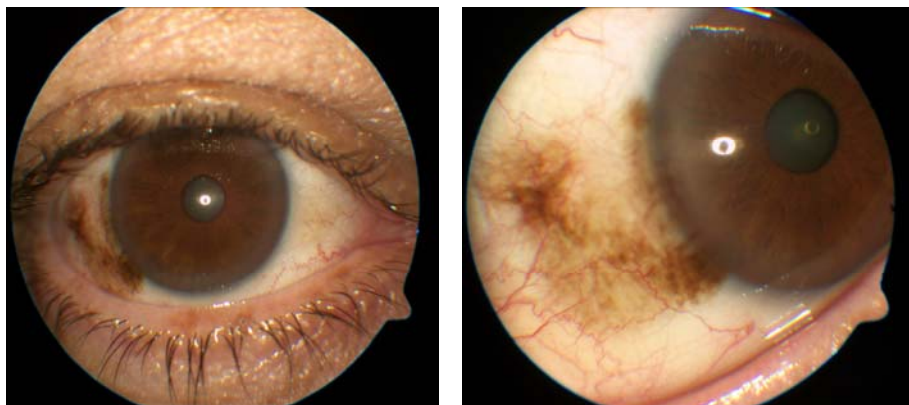
- Treated for recalcitrant unilateral glaucoma





case

- 58 yo Male from Egypt is referred for evaluation of a conjunctival nevus



Diagnosis: Primary Acquired Melanosis

- Painless, flat brown spot
- Often misdiagnosed as freckle or nevus
- Benign
- PAM with atypia – precancerous lesion with ~15% risk of progression to conjunctival melanoma
- Conjunctival melanoma ~50% mortality at 3 years, worse with >2mm, ulceration, caruncular involvement



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Rumor #9: Conjunctival
'nevi'
are no big deal either

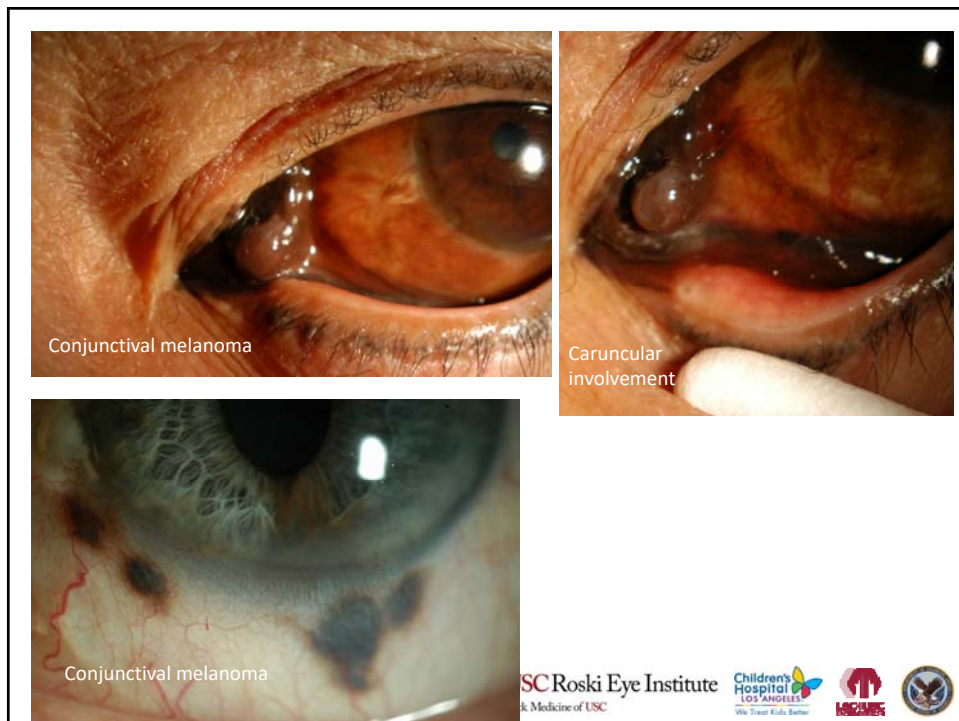
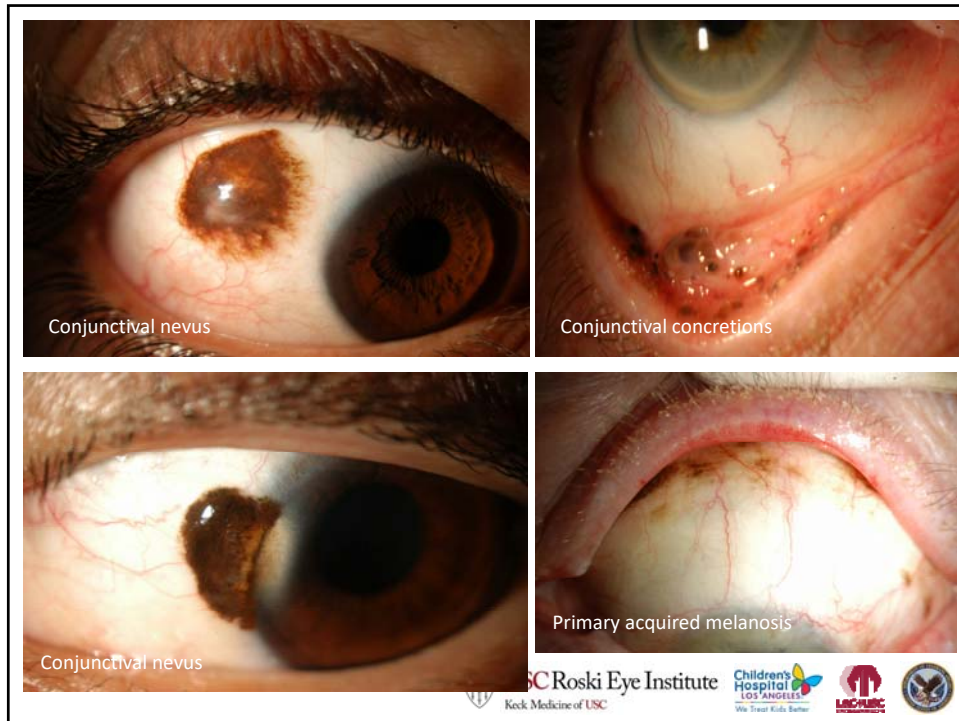


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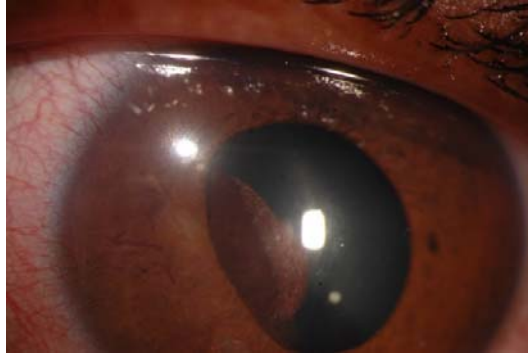
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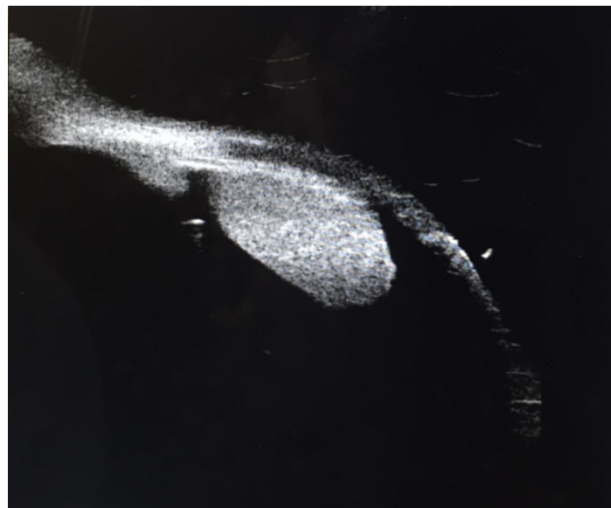


case

- 28 yo Hispanic Female presents for evaluation of decrease vision x 6 months, worse after becoming pregnant



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Diagnosis: Pigmented IridoCiliary Body Mass

- Often a late diagnosis
- May cause sectoral cataract
- Look for a sentinel vessel (important clue!)
- Considered a worse prognostic feature for uveal melanoma because it is detected later



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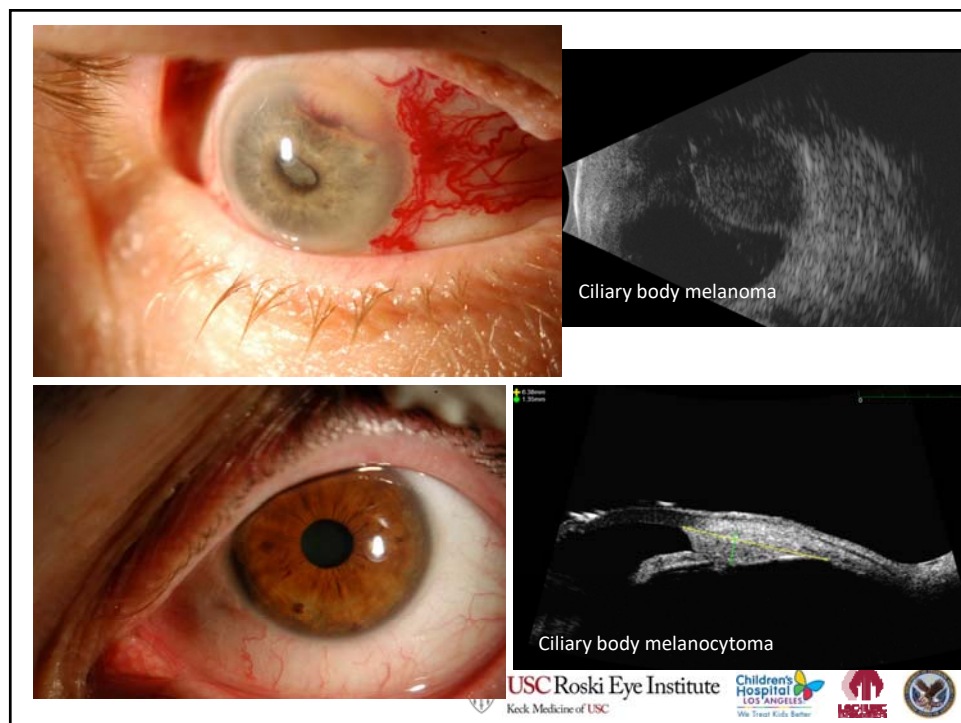
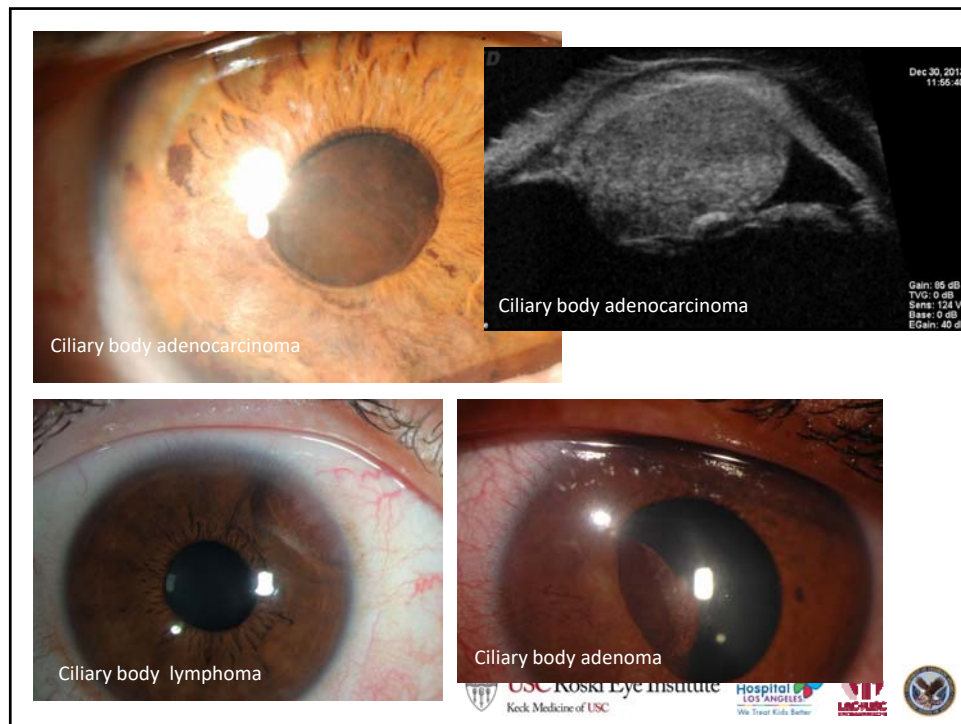


Rumor #10: Ciliary body tumors are bad, bad, bad

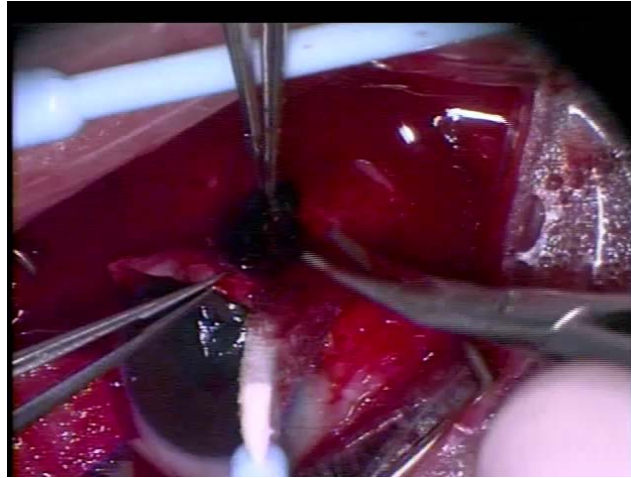


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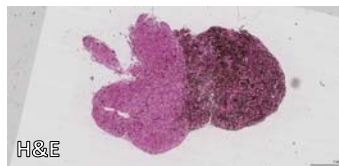
Case continues: Biopsy...



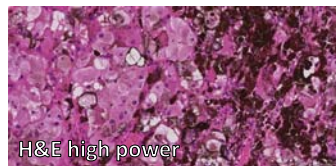
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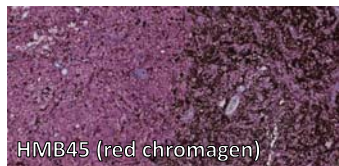
Pathology: adenoma



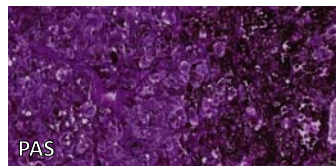
H&E



H&E high power



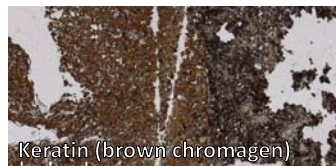
HMB45 (red chromagen)



PAS



MelanA (red chromagen)



Keratin (brown chromagen)



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I love rumors. I
always find out
amazing
things
about
myself
i never knew.



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Thank you!



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