Grand Rounds Presentation

SUNY Downstate Department of Ophthalmology

Jason M. Moss, MD

Clinical Assistant Istructor of Ophthalmology

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Patient DS

7/19/12

- CC: 72 year Hispanic F new to clinic for routine checkup. H/o pseudophakia ou x several years prior at OSH. No current ocular complaints.

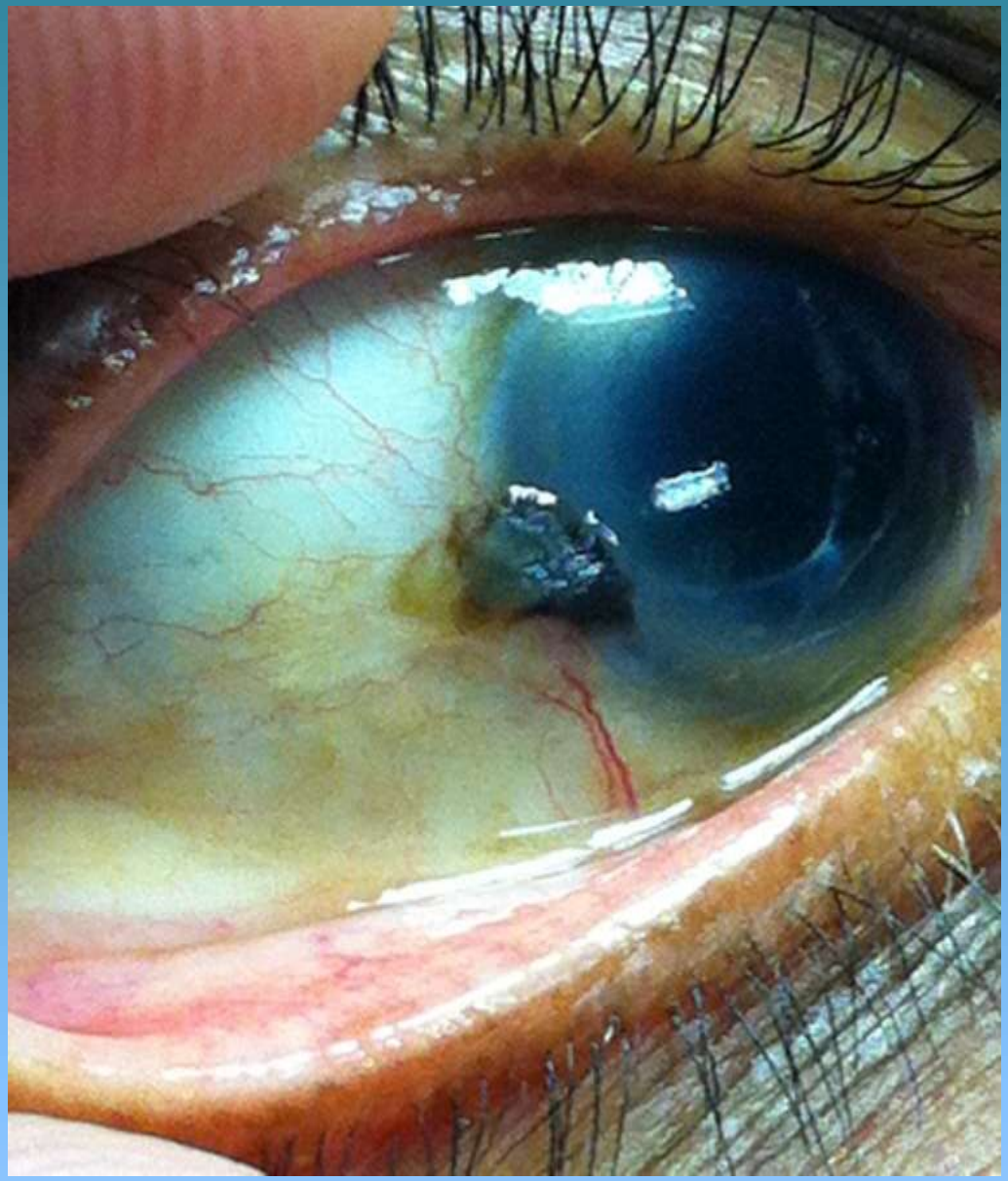
History

- PMHx: DM
- Meds: Metformin
- Allergies: NKDA
- POHx: no h/o trauma OU, CE/PCIOLs OU, + refractive error (readers OU x 30 years)
- Gtts: none
- FOhx: no blindness/glc
- Social Hx: no tob, etoh, id

Exam

- dVAsc
  - OD 20/40
  - OS 20/40
- EOM full ou, no diplopia/pain OU
- CVF FTCF ou
- Pupils R/R ou
- T_{Tp} 12/13 @ 4: 10 PM

Slit Lamp Examination



Patient Care

Slit Lamp Examination

- LLL wnl OU
- C/s elevated pigmented conj lesion 9:00 limbus with dilated vessel inferiorly OD, w/q OS
- K arcus OU
- AC d/q OU
- I/P r/r OU
- Lens PCIOLs OU, well-centered

DFE

- Vitreous –clear OU
- ON – C/d 0.3 s/p OU
- Macula – flat OU
- Vessels – wnl OU
- Periphery – no holes, breaks, or tears OU

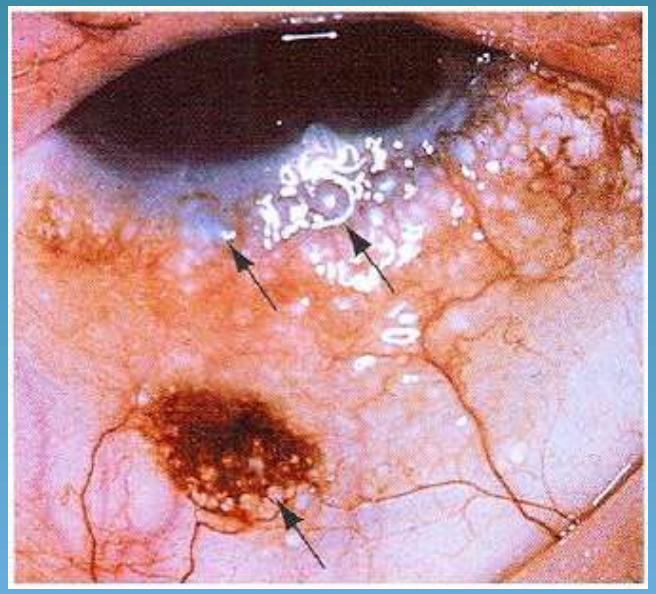
Differential Diagnosis?

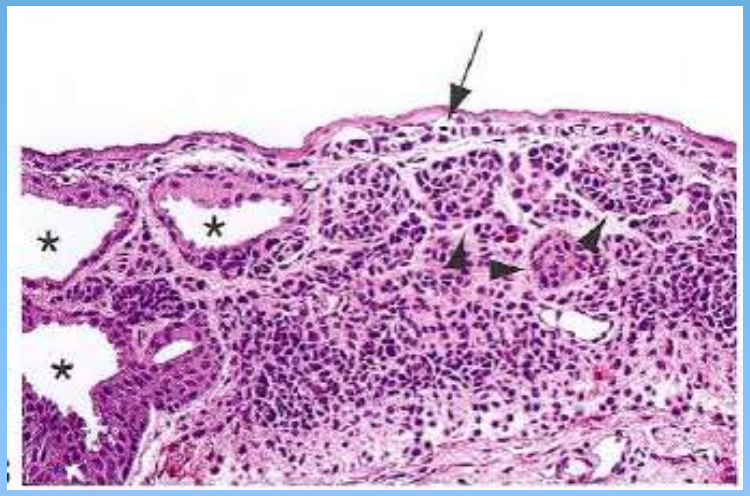
Differential Diagnosis?

- Nevus
- Racial/Benign Acquired Melanosis
- Primary Acquired Melanosis
- Malignant Melanoma
- Secondary Acquired Melanosis
  - Addison's Disease
  - Radiation
  - Pregnancy
  - Epinephrine

Nevus

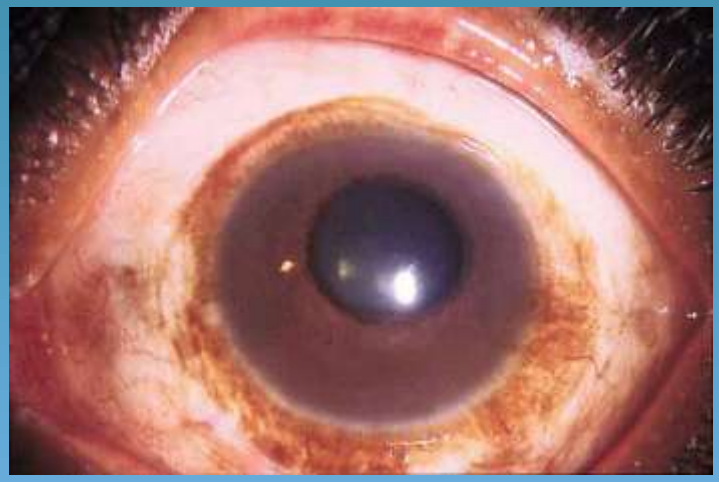
- Typical onset 1-2nd decade
- Discrete, brown or amelanotic cysts
- Very low <1% malignant potential
- Bulbar Conjunctiva/Caruncle
- Junctional, Stromal, Compound
  - Epithelial Inclusion Cysts

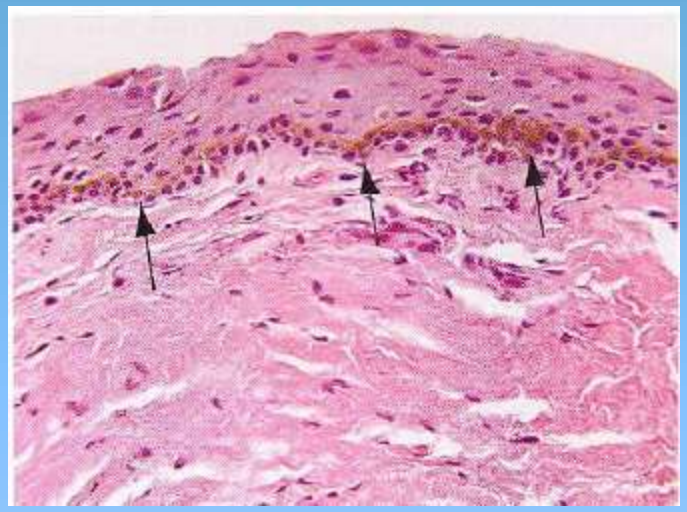




BAM

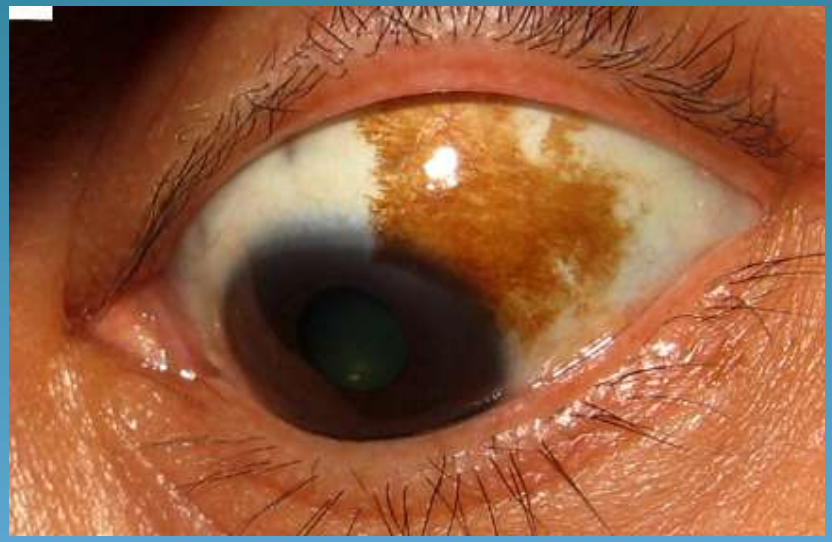
- Bilateral, flat patches of brown pigmentation with irregular margins.
- Usually in individuals with dark skin pigmentation
- Typically involves the bulbar conjunctiva and limbus. The caruncle and palpebral conjunctiva may also be involved.
- *Striate melanokeratosis*
  - Streaks and whorls of melanotic pigmentation extending onto the peripheral cornea. The
- Almost always benign, exceedingly low rate of malignant transformation.
- Histo: lentiginous proliferation of benign-appearing melanocytes along the basal epithelial layer.

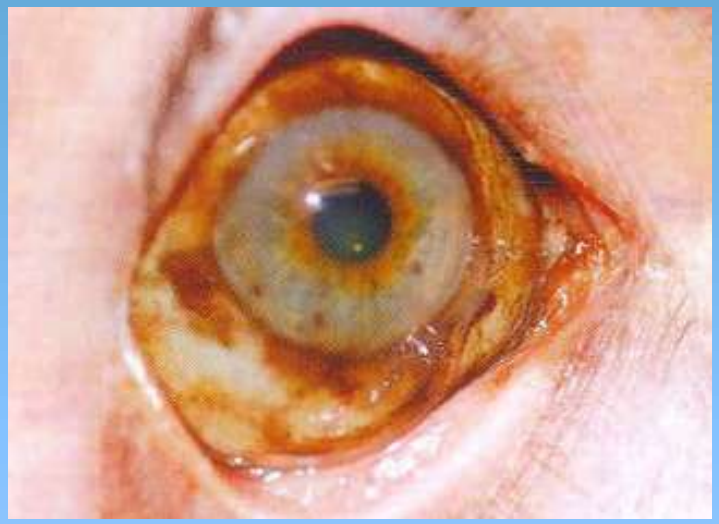




PAM

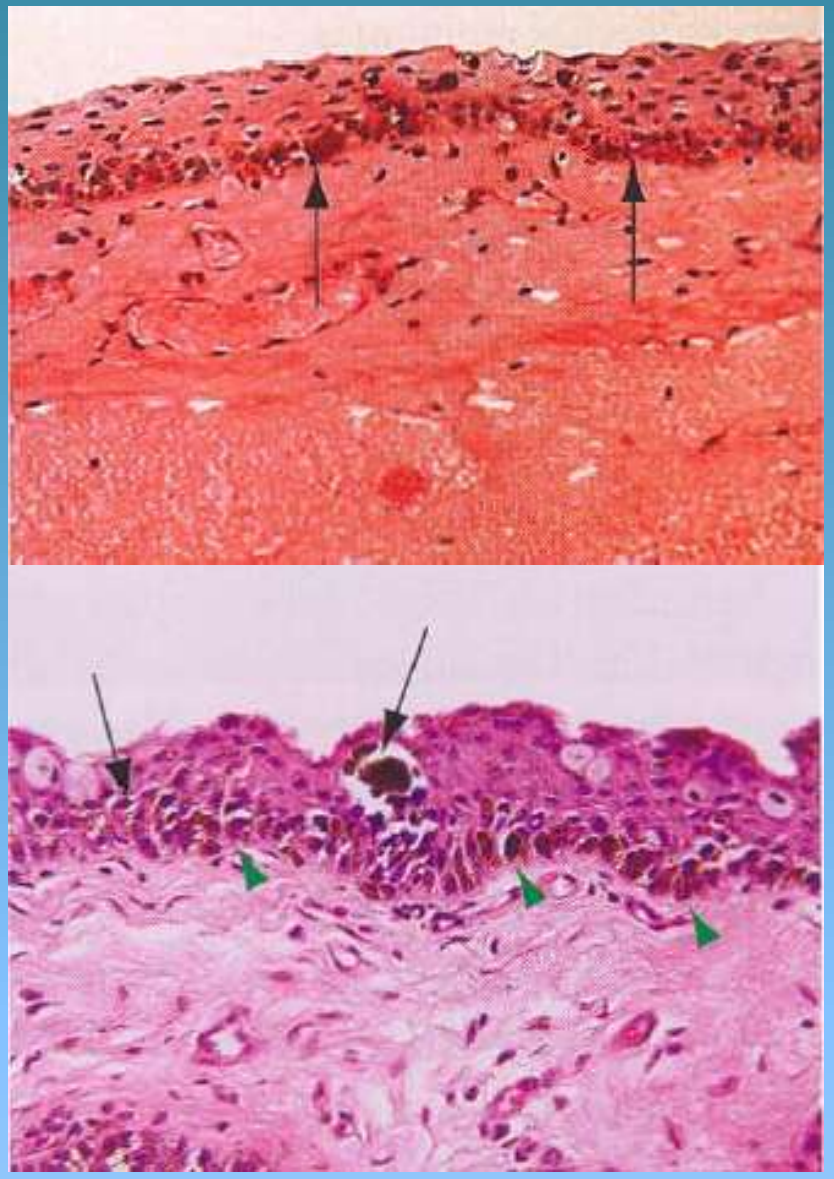
- Most often unilateral in individuals with lighter skin.
- Most commonly presents in middle-aged adults
- May occasionally be amelanotic
- May remain stable or grow slowly over a period of 10 or more years.
- Retrospective data suggest that the number of clock-hours involved may help predict the risk of malignant transformation
  - <1 clock-hour have a low chance of progressing to malignancy, observation recommended
  - ≥3 clock-hours have a greater than 20% chance of malignant transformation, biopsy.
- Involvement of the caruncle, fornix, or palpebral conjunctiva may prompt biopsy, because malignant transformation in these regions would be associated with a worse prognosis.





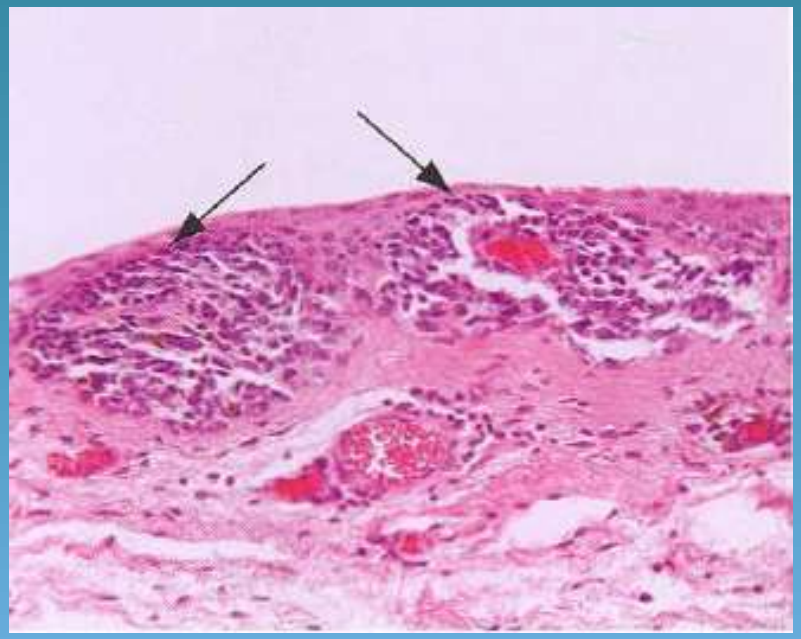
PAM Histology

- PAM without atypia
  - melanocytic proliferation confined to basal layer of the epithelium, no cellular atypia
  - No risk of transformation to melanoma
- PAM with mild/moderate atypia.
  - Most melanocytic proliferation is located in the basal epithelial layer, melanocytes are small, without prominent nucleoli.
  - Some melanocytes are in more superficial epithelium
  - White spaces around many melanocytes ->discohesiveness.
  - Low to moderate risk for transformation to melanoma.



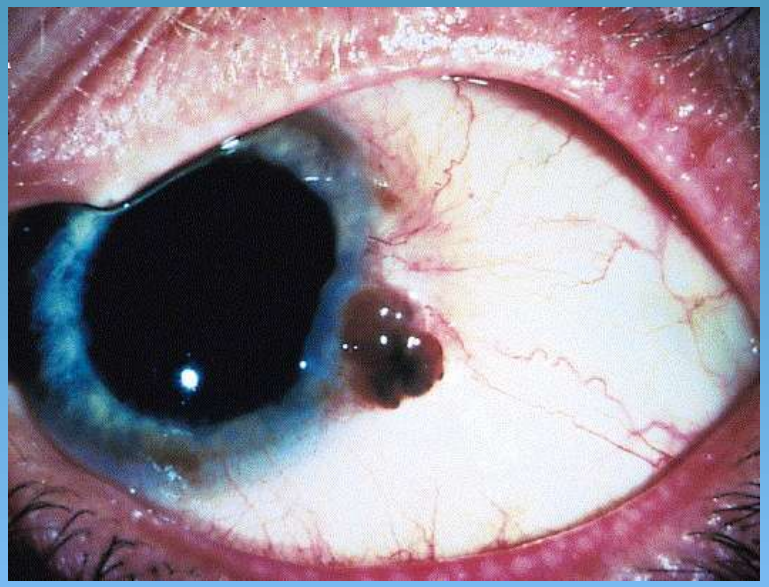
PAM Histology

- PAM with severe atypia
  - Melanocytic proliferation involves most of the epithelial thickness.
  - Epithelioid melanocytes within the epithelium.
  - Significant risk for progression to melanoma.



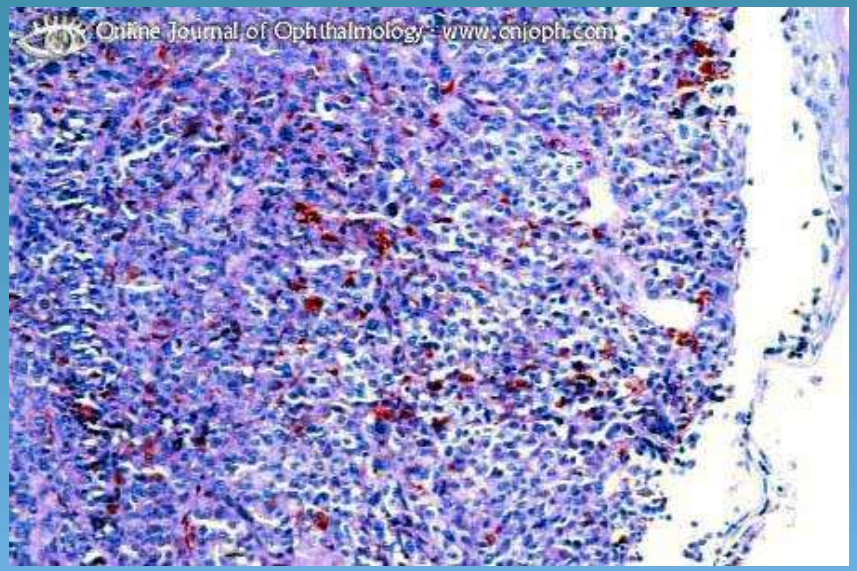
Malignant Melanoma

- Prevalence 1:2 million in Europeans
  - Rare in Black and Asian populations.
- Slightly better prognosis than cutaneous melanoma
  - Overall mortality rate 25%.
- May arise from PAM (70%), nevi (2%), or de novo (10%).
  - Intralymphatic spread increases the risk of metastasis.
  - Mets to regional LN, brain, lungs, liver, and bone.
- Most commonly found in bulbar conjunctiva or at the limbus.
- Degree of pigmentation variable
  - 25% amelanotic



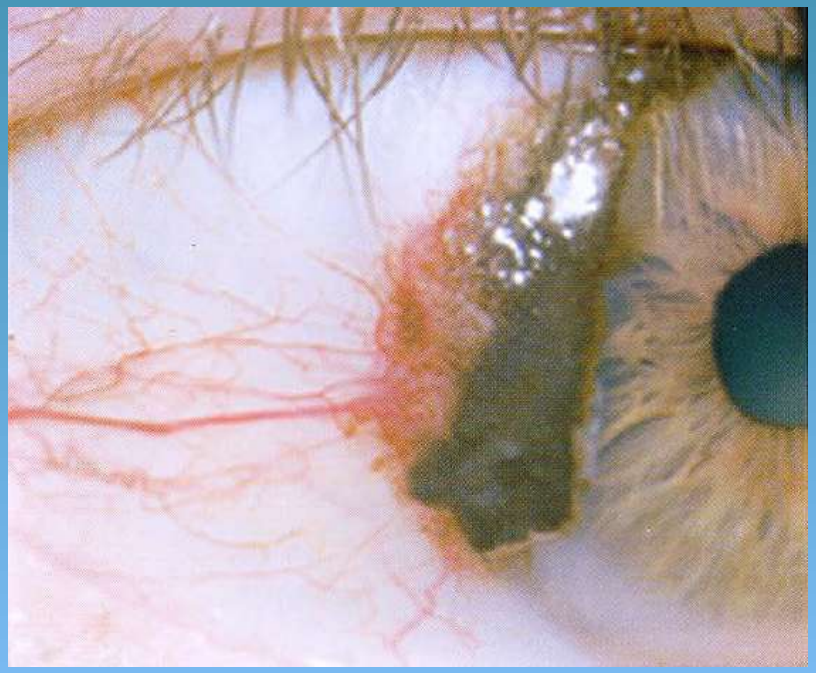
Malignant Melanoma

- Heavy vascularization common
  - Tumors may bleed easily
- Grow in a nodular fashion and can invade the globe or orbit
- Poor prognostic indicators
  - Location
    - Palpebral conjunctiva
    - Caruncle
    - Fornix
  - Invasion into deeper tissues
  - Thickness> 1.8 mm
  - Involvement of eyelid margin
  - Pagetoid or full–thickness intraepithelial spread
  - Lymphatic invasion
  - Mixed cell type.



Malignant Melanoma Management

- Excisionai biopsy should be considered for any suspicious pigmented epibulbar lesions
  - No increased risk of metastasis
- Excision of conjunctiva 4 mm beyond clinically apparent tumor margins
- Excision of a thin lamellar scleral flap beneath the tumor
- Treat remaining sclera with absolute alcohol
- Cryotherapy applied to conjunctival margins
- Primary closure performed when feasible
  - Conjunctival or AMG necessary for large excisions
- Sentinel LN Bx prior to surgical excision may be helpful in establishing prognosis
- Topical MMC after excision and cryotherapy to treat residual disease



Conjunctival melanoma: outcomes based on tumor origin in 382 consecutive cases

Shields CL, Markowitz JS, Belinsky I, Schwartzstein H, George NS, Lally SE, Mashayekhi A, Shields JA

Ophthalmology 2011 **118**: 389-395

Conjunctival Melanomas

Table 1. Conjunctival Melanoma in 382 Consecutive Cases: Demographic Features

Feature	All patients, N = 382 (%)	Origin		
		Primary Acquired Melanosis, n = 284 (%)	*Nevus, n = 26 (%)*	*De Novo, n = 72 (%)*
Mean age (median, range), yrs	59 (62, 10–92)	61 (64, 10–92)	45 (44, 14–73)	56 (59, 15–89)
Gender				
Male	186 (49)	133 (47)	15 (58)	38 (53)
Female	196 (51)	151 (53)	11 (42)	34 (47)
Race				
White	360 (94)	266 (94)	25 (96)	69 (96)
Black	10 (3)	9 (3)	1 (4)	0 (0)
Hispanic	9 (2)	7 (2)	0 (0)	2 (3)
Asian*	3 (1)	2 (1)	0 (0)	1 (1)
Eye				
Right	205 (54)	151 (53)	13 (50)	41 (57)
Left	177 (46)	133 (47)	13 (50)	31 (43)

*Includes East Asian and Indian.

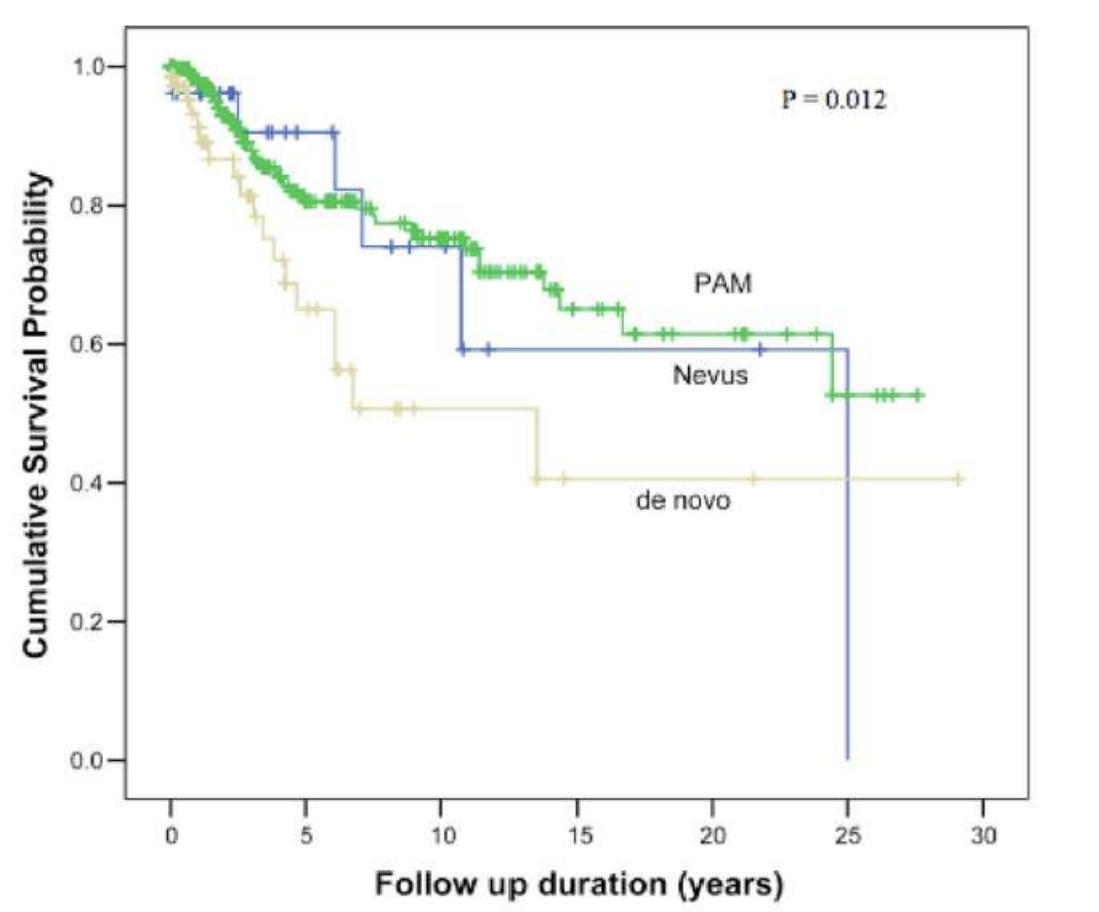
Conjunctival Melanomas

Table 2. Conjunctival Melanoma in 382 Consecutive Cases

Feature	All Patients, N = 382	Origin		
		Primary Acquired Melanosis, n = 284	*Nevus, n = 26*	*De Novo, n = 72*
Mean basal dimension (median, range), mm	11 (9, 0–40)	11 (10, 0–40)	6 (5, 2–24)	10 (9, 0–36)
Quadrant location*				
Superior	128	105	4	19
Inferior	150	115	9	26
Nasal	128	100	6	22
Temporal	101	81	6	14
Anteroposterior location*				
Cornea	140	112	7	21
Limbus	184	145	12	27
Bulbar	228	177	15	36
Fornix	84	70	1	13
Palpebra	64	53	1	10
Plica/caruncle	35	27	2	6
Pigmentation (n = 280 tumors)†				
Pigmented	166 (59)	130 (60)	14 (74)	22 (48)
Nonpigmented	56 (20)	42 (20)	2 (11)	12 (26)
Mixed	58 (21)	43 (20)	3 (16)	12 (26)

The main tumor was assessed for data collection.
*Some melanomas involved more than 1 quadrant or anteroposterior location. Information on location of conjunctival melanoma arising from nevus was not available in 1 patient.
†Tumor pigmentation was visible by photographs or recorded in 280 cases.

Conjunctival Melanomas



Next Step?

Our patient

- Total excisional biopsy performed leaving grossly unaffected underlying sclera and specimen sent for pathologic diagnosis
- Path:
  - Conjunctival melanosis with mild to moderate dysplasia to keratocytes
  - Consistent with pinguecula or ligneous conjunctival changes

Our patient

- Informed of the diagnosis and continues to follow for routine post-operative care

Reflective Practice

- The patient was an example of a potentially deadly pigmented conjunctival lesion requiring biopsy for a histopathologic diagnosis. The lesion was ultimately determined to be benign and the patient was appropriately alleviated of her concerns. The atypical presentation of the pigmented lesion allowed for a thorough review of the differential diagnosis and an appropriate diagnostic algorithm was followed.

Core Competencies

- Patient Care: The case involved thorough patient care and workup for a concerning ophthalmic findings requiring surgical biopsy for diagnosis.
- Medical Knowledge: This presentation allowed us to review the differential diagnosis of pigmented conjunctival lesions with particular attention to malignant melanoma.
- Practice-Based Learning and Improvement: This presentation included a current literature search of conjunctival malignant melanoma and its presentation, treatment options, and prognosis.
- Interpersonal and Communication Skills: The patient was treated with respect throughout this patient encounter and she was given the good news of her favorable pathological findings.
- Professionalism: The underlying condition was identified in the most timely manner possible. An in-depth discussion of planned procedures took place with the patient as a diagnostic plan was formulated.
- Systems-Based Practice: The patient underwent appropriate preoperative photographic documentation of her lesion and the pathology department was appropriately utilized in order to establish a diagnosis.

Thank you

- Dr. D’Amico
- Dr. Zazzali
- Dr. Rizzuti
- RUMC Staff
- Our patient DS