

Approach to Pigmented Cutaneous and Ocular Lesions and Melanoma

Objectives

- Generate a differential diagnosis for pigmented lesions on the eye and skin
- Discuss the key features of common pigmented cutaneous and ocular lesions
- Describe how cutaneous and ocular melanoma can present in pediatric populations
- List the treatment options for ocular and cutaneous melanoma

Disclosure

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Case

You're a fourth-year medical student doing a pediatrics elective. Lucy, who is now 17 years old, presents with a pigmented skin lesion on her right eyelid. It has been present since she was a child but darkened within the past year. As well, there's a brown lesion on her right conjunctiva that has darkened in the same time frame.

She denies any eye discharge, blurry vision, foreign body sensation or pain.

Case

She is otherwise healthy and does not have a history of severe/blistering sunburns. She does not drink alcohol or use recreational drugs.

Physical Exam:

- 4x3mm symmetric, dark brown oval papule with well-demarcated irregular borders on the right nasal lower eyelid
- 1x1mm limbal lesion at 9 o'clock on the right eye that is well-circumscribed

What are these lesions?

Cutaneous Lesions

Congenital Nevi



- Benign
- Present at birth
- Macules or papules
- Pink, brown
- May have hair
- Slowly more prominent and increase with time
- Size correlates with risk of melanoma
 - Small: <1.5cm
 - Medium 1.5-20cm
 - Large 20-40cm
 - Giant >40cm

Cutaneous Lesions

Acquired Nevi



- Appear after 6 months of life
- Symmetric
- Round/oval
- Evenly pigmented
- Regular outline, well demarcated
- Types:
 - Spitz nevus
 - Reed nevus

Cutaneous Lesions

Is it Pediatric Melanoma?



- Differential diagnosis
 - Café au lait macule
 - Vascular lesions
 - Blue nevus
 - Pyogenic granulomas

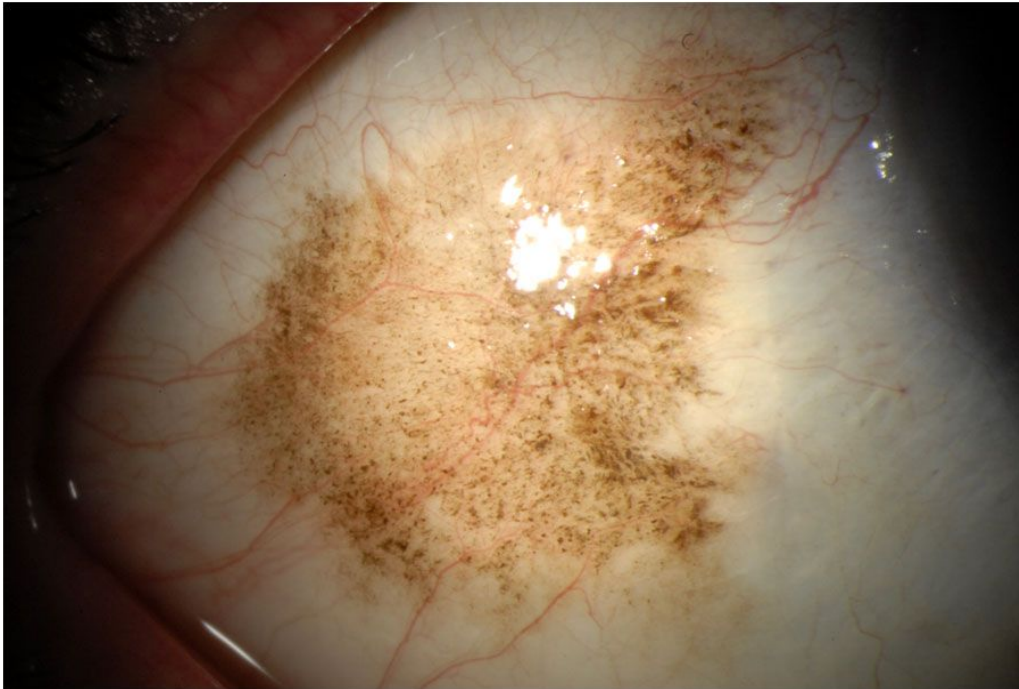
Cutaneous Lesions

Assessing for Suspected Melanoma

- Pediatric ABCD CUP criteria
 - Amelanotic
 - Bleeding, bump
 - Color variation
 - De novo, diameter
 - Colour (pink/red), changing
 - Ulcer, upward thickening
 - Pyogenic granuloma-like

Pigmented Ocular Lesions

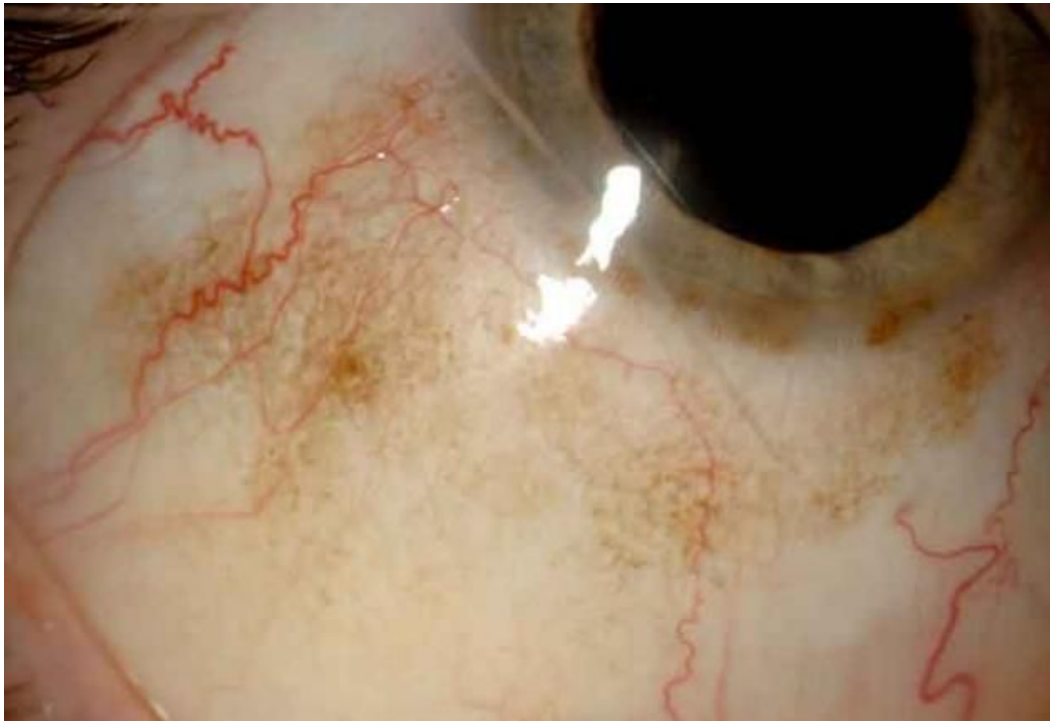
Primary Acquired Melanosis



- Flat
- Unilateral
- Yellow to brown
- Not well circumscribed
- May wax and wane
- **Concerning features:**
 - Increased nodularity
 - Vasculature
 - Thickening

Pigmented Ocular Lesions

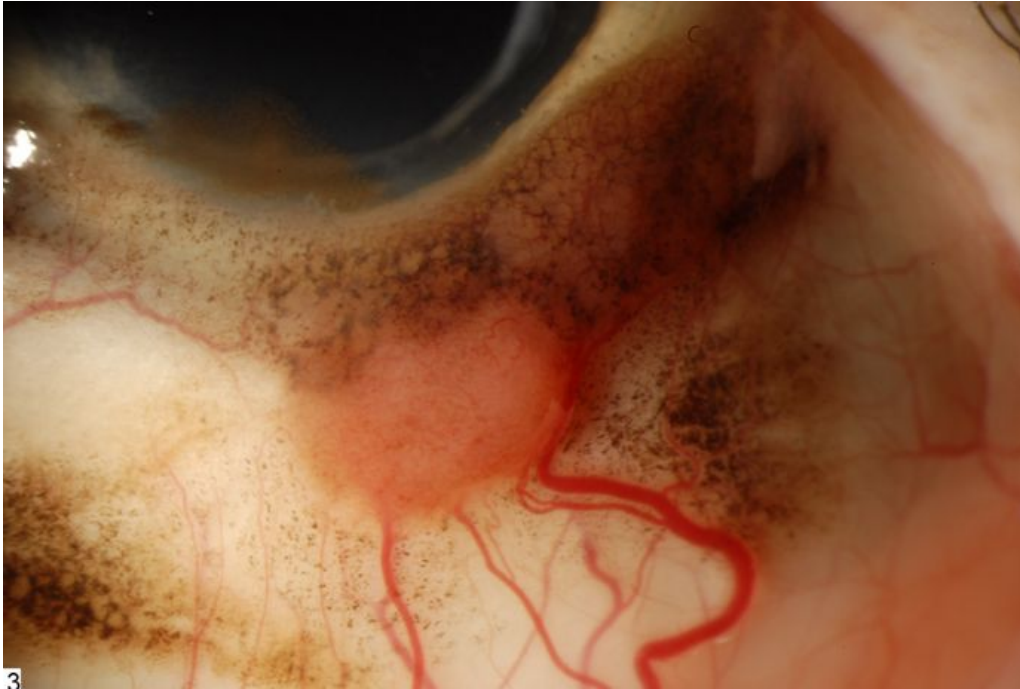
Complexion-Associated Melanosis



- Benign
- Seen in people with darker skin tones
- Usually around the limbus
- Flat
- Noncystic
- Usually bilateral
- Increases with age

Pigmented Ocular Lesions

Conjunctival Melanoma



- Most common at the limbus
- Elevated usually
- Vascular
- Can be de novo, from a nevus or from primary acquired melanosis

Pigmented Ocular Lesions

Lisch Nodules

- Small
- Well-defined
- Usually appear before 5 years old
- Seen in neurofibromatosis-1

Uveal Nevus

- Flat or elevated
- Variable pigment
- Iris freckles
 - Superficial
 - Not elevated or distorted
 - Usually nonprogressive

Uveal Melanomas

- 85% occur within posterior choroid
- Pigmented or nonpigmented
- Iris melanomas
 - Inferior half
 - Vascularized
 - Pigment variable

Pigmented Ocular Lesions

Melanocytoma of the Optic Nerve

- Darkly pigmented
- Within optic nerve

Congenital Hypertrophy of the RPE

- Flat
- Well-circumscribed
- Pigmented
- May have central depigmentation
- May be grouped in bear tracks pattern
- 4 or more:
 - Gardner's Syndrome
 - Adenomatous polyposis of the colon

Treatment for Melanoma

Cutaneous

- Wide local excision to the deep fascia
- Margins depend on depth and location of tumor
- Adjuvant therapy may be used

Ocular

- Conjunctival lesions:
 - Incisional biopsy
 - Excision with dry, no touch technique and wide margins
 - Cryotherapy of edges
 - Sentinel lymph node biopsy
 - Proton beam
 - Plaque brachytherapy
 - Enucleation

Case

- Congenital melanocytic nevus
- Refer to ophthalmology for comprehensive eye exam
- Sun protection counselling

Take Home Points

- Pigmented lesions should be observed carefully and biopsied if there are high risk features or changes
- Pediatric melanoma is rare and can present atypically
- Pediatric ABCD and CUP criteria
- Thorough eye exam with photographs to characterize each lesion and ensure there are no other concerning eye lesions

References

- Balzer BW, Cherepanoff S, Joshua AM, Giblin M, Conway RM, Anazodo AC. Conjunctival Melanoma in Childhood and Adolescence: A Systematic Review. *Ocular oncology and pathology*. 2019;5(6):387-95.
- Bartenstein DW, Kelleher CM, Friedmann AM, Duncan LM, Tsao H, Sober AJ, Hawryluk EB. Contrasting features of childhood and adolescent melanomas. *Pediatric dermatology*. 2018 May;35(3):354-60.
- Cordoro KM, Gupta D, Frieden IJ, McCalmont T, Kashani-Sabet M. Pediatric melanoma: results of a large cohort study and proposal for modified ABCD detection criteria for children. *Journal of the American Academy of Dermatology*. 2013 Jun 1;68(6):913-25.
- Herwig, M. and Wells, J., 2020. Conjunctival Melanocytic Tumors - Eyewiki. [online] Eyewiki.aao.org. Available at: <https://eyewiki.aao.org/Conjunctival_Melanocytic_Tumors>
- Howman-Giles R, Shaw HM, Scolyer RA, Murali R, Wilmott J, McCarthy SW, Uren RF, Thompson JF. Sentinel lymph node biopsy in pediatric and adolescent cutaneous melanoma patients. *Annals of surgical oncology*. 2010 Jan 1;17(1):138-43.
- Navid F, Furman WL, Fleming M, Rao BN, Kovach S, Billups CA, Cain AM, Amonette R, Jenkins JJ, Pappo AS. The feasibility of adjuvant interferon α -2b in children with high-risk melanoma. *Cancer*. 2005 Feb 15;103(4):780-7.
- Paller A., Mancini AJ, Hurwitz S. Hurwitz clinical pediatric dermatology: A textbook of skin disorders of childhood and adolescence. New York: Elsevier/Saunders, 2011. Chapter 9.
- Paller A, Mancini AJ, Hurwitz S. Hurwitz clinical pediatric dermatology: A textbook of skin disorders of childhood and adolescence. 5th ed. New York: Elsevier/Saunders; 2016.
- Paradela S, Fonseca E, Prieto VG. Melanoma in children. *Archives of pathology & laboratory medicine*. 2011 Mar;135(3):307-16.
- Sayyad FFE, Karp CL. Conjunctival Pigmented Lesions: Diagnosis and Management. *AAO EyeNet Magazine*. 2013 Sep; 35-36.
- Shields CL, Shields JA. Tumors of the conjunctiva and cornea. *Survey of ophthalmology*. 2004 Jan 1;49(1):3-24.
- Shields CL, Sioufi K, Alset AE, et al. Clinical Features Differentiating Benign From Malignant Conjunctival Tumors in Children. *JAMA Ophthalmol*. 2017;135(3):215–224.
- Shields CL, Swathi K, Sruthi ABA, et al. Uveal Melanoma in Children and Teenagers. *Saudi J Ophthalmol*. 2013 Sep;27(3):197-201
- Shields JA, Eagle RC Jr, Shields CL, Brown GC, Lally SE. Malignant transformation of congenital hypertrophy of the retinal pigment epithelium. *Ophthalmology*. 2009;116(11):2213-2216. doi:10.1016/j.opthta.2009.04.048
- Silverberg NB, McCuaig CC. Melanoma in childhood: changing our mind-set. *Cutis*. 2013 Nov;92(5):217.
- Wills Eye Hospital. 2020. Conjunctival Primary Acquired Melanosis | Wills Eye Hospital. [online]