8.8.3. Neoplastic Disorders of the Conjunctiva and Cornea (III): Benign Pigmented Lesions

Introduction

- **Pigment spot of the sclera**
  - collection of melanocytes associated with
    - intrascleral nerve loop
    - perforating anterior ciliary vessel
- **Melanosis**
  - excessive pigmentation without an elevated mass
    - congenital (whether epithelial or subepithelial)
    - acquired (whether primary or secondary)
- **Conjunctival pigmentation**
  - chronic exposure to
    - epinephrine
    - silver
    - mascara

See Tables 8-2 and 8-3

Congenital epithelial melanosis (freckle/ephelis)

- present at an early age
- flat brown patch
- bulbar conjunctiva near the limbus
- more common in darkly pigmented individuals

Ocular melanocytosis

- 1:2500 individuals
  - more common in the black, hispanic, and asian populations
- usually unilateral
  - 5% bilateral
- pathogenesis
  - focal proliferation of subepithelial melanocytes (blue nevi)
- clinical findings
  - slate gray
  - immobile
  - patches of episcleral pigmentation (congenital melanosis of the episclera)

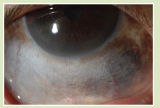

Figure 8-8 (© 2020 American Academy of Ophthalmology)

  - proliferation of dermal melanocytes in the periocular skin
  - dermal melanocytosis (nevus of ota)
    - first and second dermatomes of cranial nerve V
    - 1/2
  - oculodermal melanocytosis
  - increased pigmentation of the iris and choroid
  - secondary glaucoma
    - 10%
  - malignant transformation
    - rare
    - lifetime risk of uveal melanoma in a patient with ocular melanocytosis is about 1/400
    - seems to occur only in patients with fair complexions
  - malignant melanoma
    - skin
    - conjunctiva
    - uvea
    - orbit

Nevus

- arise during childhood and adolescence
- types
  - junctional (pure intraepithelial)
    - rare except in children
    - difficult to distinguish histopathologically from primary acquired melanosis
  - compound
    - most common type
  - subepithelial
    - equivalent of the intradermal nevus
- clinical findings
  - flat or elevated
  - variable pigmentation

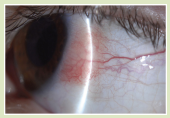

Figure 8-9 (© 2020 American Academy of Ophthalmology)

  - subepithelial nevus often has a cobblestone appearance
  - small epithelial inclusion cysts
    - half of all conjunctival nevi
    - compound or subepithelial nevi
    - secretion of mucin by goblet cells in the inclusion cysts can cause a nevus to enlarge
      - false impression of malignant change
  - when inflamed, an amelanotic, vascularized nevus may resemble
    - angioma
    - chronic conjunctivitis
  - secondary lymphocytic inflammation
  - rapid enlargement at puberty
  - clinical impression of conjunctival melanoma
- management
  - rarely become malignant
  - follow every 6–12 months
    - serial photography
    - lesions that change
  - excisional biopsy should be performed on
    - lesions on the tarsal conjunctiva, the caruncle, or the plica semilunaris or fornix
    - nevi are rare in these locations!

Benign (racial) melanosis

- bilateral
- middle-aged individuals with dark skin
- increasing pigmentation of the conjunctiva
  - most apparent at the limbus
  - less prominent as one approaches the fornix

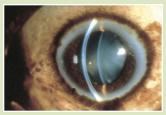

Figure 8-7 (© 2020 American Academy of Ophthalmology)

- **striate melanokeratosis**
  - streaks and whorls of peripheral corneal epithelial pigment

Neurogenic and Smooth-Muscle Tumors

- peripheral nerve sheath tumors
  - types
    - neurofibroma
      - neurofibroma of the conjunctiva or eyelid is almost always a manifestation of neurofibromatosis
    - neurilemoma (schwannoma)
      - from Schwann cells of a peripheral nerve sheath
    - neuroma
      - more common in multiple endocrine neoplasia (MEN)
- leiomyosarcoma
  - very rare limbal lesion
  - potential for orbital invasion