Anatomic Divisions

- Ocular Surface
- Intraocular
- Orbital
- Lacrimal
- Eyelid
Ocular Surface

- Melanocytic
- Squamous Neoplasia
- Lymphoid
Melanocytic

- Nevi
- PAM (Primary Acquired Melanosis)
- Ocular Melanocytosis
- Melanoma
  - In-situ
  - Invasive
Melanocytic

- Nevi
  - Typically, unilateral, pigmented, may be cystic
  - Generally intrapalpebral in location
  - NO FEEDER vessel
  - ? Rare malignant transformation
PAM

- Generally unilateral, flat, noncystitic

- Often stippled pigmentation away from the main pigmentation

- Involves conjunctiva only, not sclera — unlike Ocular melanocytosis

- Therefore, overlying pigmented conj may be moved over the non pigmented sclera
Ocular Melanocytosis

• Unilateral pigmented and congenital
• Located in deep episclera, sclera, and uveal tract
• Form of a blue nevus
• Therefore no movement with Q-tip
Melanoma

- Raised, thickened, usually pigmented 80%
- Feeder Vessel
- Can occur anywhere on conjunctiva, even caruncle
  - Usually on bulbar conjunctiva
- May metastasize to preauricular or cervical lymph nodes
Squamous cell Ca of Conjunctiva

- End-stage of a spectrum of disease referred to as ocular surface squamous neoplasia (OSSN)
- Main risk factors for both are exposure to solar ultraviolet radiation outdoors, HIV/AIDS, human papilloma virus
Squamous cell Ca of Conjunctiva

• The tumor tends to be found between the eyelids (interpalpebral space), and at the limbus (border of the white sclera and clear cornea).

• This tumor can extend onto the cornea, around the limbus

• Rarely invades orbit
Squamous cell Ca of Conjunctiva

• Diagnosis

• The diagnosis of squamous conjunctival neoplasia is typically made by biopsy.

• Like most squamous epithelial tumors, invasion beneath the epithelium into the substantia propria defines these lesions as carcinoma.

• When the tumor is contained within the epithelium it does not have access to the lymphatic system (metastatic potential)
Squamous cell Ca of Conjunctiva

• Treatment

  • Surgical excision (alone) has been associated with high rates of recurrence. This is because the tumor’s edges and deep margins are often difficult to determine.

  • Local Ancillary treatments include:

    • Local superficial Cryo of the tumor bed (sclera and adjacent conjunctiva)

    • radiation therapy to decrease tumor recurrence.

    • Most recently, topical chemotherapy, or “chemotherapy eye drops”—usually mitomycin C— have been found effective.
Conjunctival lymphoid lesions

- Conjunctival Reactive lymphoid hyperplasia
  - Lymphoid proliferations, which are either polyclonal or occasionally oligoclonal
- Conjunctival Lymphoma
  - Monoclonal lymphoid proliferations with evidence of cytological atypia and malignant behavior
Conjunctival Lymphoid proliferations

- Classically a salmon colored patch of elevated conjunctiva
- Virtually impossible to distinguish on exam alone
  - Biopsy and Pathology critical
Conjunctival Lymphoid proliferations

- Lymphoma is generally an extra nodal B-cell lymphoma
  - Tx consist of referral to oncologist for systemic work-up
  - Isolated disease may be treated with low dose external beam radiation
Intraocular malignant lesion

- Most common
  - Adults
    - Uveal melanoma
      - Iris, ciliary body, choroid
  - Children
    - Retinoblastoma
Uveal melanoma

- Arise from the melanocytes in the uvea
- May be divided into anterior and posterior
  - Anterior - Iris
  - Posterior - Ciliary body and Chorioid
- There is benign counterpart to each which is perfectly harmless except in the event of malignant transformation
  - Surveillance important
Iris Melanoma

• @5% of Uveal melanoma

• Noted on routine exam

  • May be circumscribed mass or diffuse iris mass

  • Other signs— heterochromia, chronic uveitis, glaucoma, spontaneous hyphema

• Show characteristic low-medium internal reflectivity on U/S

• Less likely to metastasize
**Posterior melanoma**

- Presenting symptoms:
  - blurred vision - most common
  - Asymptomatic - second most common
  - Other symptoms at presentation include photopsia, floaters, visual field loss
Posterior melanoma

- Important to distinguish from benign nevi
- Signs that favor melanoma include:
  - thickness greater than 2 mm, subretinal fluid, symptoms, orange pigment, tumor margin less than 3 mm to disk, ultrasonographic hollowness-low internal reflectivity,
Posterior melanoma

• Treatment

  • Enucleation generally reserved for a large tumor load and with vision loss

• Globe sparing:

  • Plaque brachytherapy, external beam radiation, trans pupillary thermal therapy, Photodynamic laser photocoagulation
Posterior melanoma

• Prognosis

• Despite excellent rates of local disease control with surgery or radiotherapy, up to 50% of patients will ultimately develop metastatic disease, with the most common initial sites being the liver (60.5%), the lung (24.4%), skin/soft tissue (10.9%), and bone (8.4%).

• The overall rate of survival from initial diagnosis is 69% at 5 years, 55% at 15 years, and 51% at 25 years.

• Predictors of metastasis: base diameter, thickness, ciliary body involvement, and distance of extraocular extension.

• Median overall survival is approximately 13.4 months, with only 8% surviving 2 years.
Retinoblastoma

- Rare tumor of retina generally diagnosed before age three
  - 250-300 cases annually in the United States
- Two forms/groups:
  - Heritable - transmitted genetic defect
  - Non-heritable - Spontaneous
Retinoblastoma

• Presentation:
  • White reflex/ Cat Eye
  • Exotropia on esotropia
  • Pain ——> increase pressure
  • Poor vision
Retinoblastoma

- Treatment: Very Complex and individualized
  - Can include surgery, chemotherapy, focal therapy and radiation therapy
- Eye(s) may be removed in children with advanced retinoblastoma.
- In patients with only one affected eye: When that one eye is removed, more than 90 percent of those patients do not need any more treatment.
Retinoblastoma

• If tumor has spread into the tissues surrounding the eye or the eye socket, the patient is treated with chemotherapy after surgery.

• Chemotherapy alone cannot cure retinoblastoma, so patients often receive “focal therapy.”

• Focal therapy — is treatment with laser therapy or freezing treatments (cryotherapy) while under anesthesia

• if lesions are small enuf, then focal therapy may sufficient
Rhabdomyosarcoma

- Most common primary malignancy of orbit in children
  - @35 Cases/year
  - Initially was believed to originate directly from striated muscle
  - Now believed to originate from primitive pleuripotential mesenchymal cells that possess the ability to differentiate into striated muscle
Rhabdomyosarcoma

- Most cases of orbital RMS present initially to ophthalmologists as a space occupying lesion.
- Disease of young children, mean age of presentation 5-7.
- Patients with orbital RMS usually present with proptosis developing rapidly over weeks (80–100%), or globe displacement (80%) which is usually downward and outward because two-thirds of these tumors are supero-nasal.
Rhabdomyosarcoma

- Treatment of orbital RMS typically includes a combination of surgery, irradiation, and chemotherapy

- Specifics depend on classification as defined by Intergroup Rhabdomyosarcoma Studies (IRSG
Lacrimal Tumors

- Lacrimal Gland (orbital)
  - Lymphoproliferative disease
    - Range from reactive benign hyperplasia to malignant lymphoma
  - Pleomorphic Adenoma
- Lacrimal Sac
  - Inverted papilloma
  - Squamous cell
Lymphoproliferative tumors

• Presentation:
  • Depends on size and location
  • Some asymptomatic
  • Pain, proptosis, diplopia, Ptosis, eyelid swelling
Lymphoproliferative tumors

- Clinical exam and orbital imaging is not enough to make the diagnosis with certainty

- Diagnosis is made based on a combination of histopathologic, immunophenotypic, and molecular genetic studies

  - Therefore, open biopsy needed

- 50% will have systemic disease at time of DX

  - Oncologic workup needed
Lymphoproliferative tumors

- Treatment
  - If localized: Radiation
  - If disseminated
    - Standard lymphoma chemotherapy
Pleomorphic adenoma

- Most common epithelial tumor of the lacrimal gland
- Presents as gradual painless proptosis
- Dx can general be made based on the clinical history and Radiographic finding
Pleomorphic adenoma

- Treatment
  - Biopsy in questionable cases
  - Complete excision via a lateral orbitotomy
Inverted Papilloma

- Inverted papilloma is a rare tumor comprising 0.5-4% of all sinonasal neoplasms.

- Benign sinonasal tumor with potential for malignant transformation
Inverted Papilloma

- Presentation
  - epiphora or dacryocystitis
  - Often have a mass below the medial canthal tendon
  - Irrigation may cause bloody reflux
  - Rarely diplopia from tumor extension
  - Found at time of DCR
Inverted Papilloma

- Treatment is directed at complete excision
  - Has a high recurrence rate (up to 70%)
  - Potential for malignant transformation —> 5-10%
Squamous cell carcinoma

- @300 cases per year

- May present with mass under medial cantonal tendon, epiphora, blood on irrigation of NLD

- Usually associated w HPV type 16 or 18, sometime 6 and 11
Squamous cell carcinoma

- Definitive diagnosis dependent on Biopsy
- ENT consult to evaluate Nasal cavity
- Wise to treat ocular surface w mitomycin C to take care of potential service
- Systemic workup to look for metastasis
Two Common, Two Deadly

• **Common**
  – Basal cell carcinoma
  – Squamous cell carcinoma

• **Deadly**
  – Sebaceous cell carcinoma
  – Malignant Melanoma
Basal Cell Carcinoma

- Pearly borders with telangiectasia
- Central ulceration is common
- Originates from basal layer of skin
- \[ \therefore \] no extra keratinization
- Indurated, not painful, irregular
- Loss of lid architecture when lid margin is involved; lashes often lost
Basal Cell Carcinoma

- Low risk for metastasis
- May/do invade locally
- Mortality ≈3%
- Morbidity & mortality most common with:
  - medial canthal lesions
  - h/o treatment with radiation
  - clinically neglected tumors
Types of Basal Cell Carcinoma

- Nodular – most common
- Morpheaform or sclerosing
- Superficial – least common on eyelids
Nodular Basal Cell Carcinoma

- Firm, raised, pearly nodule
- Telangiectasia common
- Central ulceration may be present
  “rodent ulcer”
Morpheiform Basal Cell Carcinoma

- More invasive than nodular BCCa
  \[ \therefore \text{ worse prognosis} \]
- Involvement usually underestimated on examination (margins difficult to discern)
- Firm lesions
- Ulceration common
Superficial BCCa

- Relatively rare on the face
- Slightly elevated, erythematous, scaling patches
Squamous Cell Carcinoma

• Much less common than BCCa
  \( \approx 5\text{-}10\% \) of lid malignancies
  BCCa to SCCa ratio 40:1

• Arises in sun-damaged skin (de novo or from actinic keratosis)

• Can appear as nodule or an indurated plaque
Squamous Cell Carcinoma

- Hyperkeratosis
  if the scaly skin falls off easily, strongly suggests SCCa or its precursor, actinic keratosis
- ± ulceration, telangiectasia, & pearly borders
Spread of SCCa

- Direct extension by narrow cellular strands
- Perineural invasion
- Lymphatic spread
- Hematogenous
Sebaceous Cell Carcinoma

• Rare (1 to 5% of eyelid cancers)
• Arises within sebaceous glands of the skin
  \[\therefore \text{adnexal malignancy}\]
• BCCa, SCCa, and sebaceous cell carcinoma comprise >95% of eyelid malignancies

(if you have to guess what a malignancy is, guess BCCa)
Sebaceous Cell Carcinoma

• Sebaceous glands in the periocular region:
  – meibomian glands in tarsus
  – glands of Zeis associated with lash follicles
  – in the periocular skin
  – in the caruncle
  – associated with eyebrow follicles

⇒ sebaceous cell carcinoma is more common in the periocular area than anywhere else in the body (because there are so many sebaceous glands in this region)
Sebaceous Cell Carcinoma

- No characteristic appearance → "masquerader"
- May present as unilateral blepharoconjunctivitis or a chronic/recurrent chalazion
- Subtle thickening of lid & lid margin
- Yellowish material within any suspicious lesion should suggest sebaceous cell carcinoma
- More common on upper lid → more meibomian glands in the superior tarsus
Sebaceous Cell Carcinoma

Two unusual growth patterns make complete excision difficult

1. pagetoid spread—may spread superficially over large areas; margins not clinically visible

2. Multifocal, noncontiguous tumor origins with “skip” areas between
Sebaceous Cell Carcinoma

• Preoperative map biopsies of the conjunctiva are done to assess peripheral pagetoid spread

• Generous margins of full-thickness lid

• Frozen sections can be unreliable

  Treatment may require 2 stages– excision followed by reconstruction
Sebaceous Cell Carcinoma

• Why spend so much time on a rare tumor?
  ⇒ it can be deadly

• Regional lymph node metastasis possible

• The longer the duration of symptoms before treatment, the poorer the prognosis for survival
Melanoma

• Eyelid melanomas are rare (<1%)
• Clinical features
  – recent onset
  – change in existing lesion (uncontrolled growth)
  – Asymmetric shape (cannot “fold” on itself)
  – Borders irregular (uncontrolled growth)
  – Color change or multiple colors within lesion
  – Diameter >6mm in diameter (large lesion)
Melanoma

- May metastasize
- Risk of metastasis increases with increasing depth of lesion
  - Therefore never do shave biopsy if melanoma suspected
- Lymph node mapping (sentinel node biopsy) is new popular modality in surgical therapy
Treatment

- Medical

- Surgical
Eyelid Reconstruction - Goals

• Only after the lesion is completely excised is reconstruction considered
• Globe protection
  – Adequate mucous membrane
  – Static position
  – Closure
• Lacrimal function
• Cosmesis
Eyelid Reconstruction - Principles

• Reconstitute BOTH eyelid lamella
  – Anterior lamella: skin/orbicularis oculi
  – Posterior lamella: conjunctiva/ retractor band/ tarsus

• Reconstitute canthal attachments

• Reconstitute lacrimal drainage system