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 Choiroid
- There is benign counter-part 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 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 sparring :
        - 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%).25
    - 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
- ∴ 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"

Morpheaform Basal Cell Carcinoma

- More invasive than nodular BCCa
  ∴ 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

  ≈5-10% 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

  ∴ 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?
  $\Rightarrow$ 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)
  - **A**symmetric shape (cannot “fold” on itself)
  - **B**orders irregular (uncontrolled growth)
  - **C**olor change or multiple colors within lesion
  - **D**iameter >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

Treatmnent

- 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