Conjunctiva
Lecture 3: Cysts and Tumors

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Cysts

- Congenital Cystic lesions:
  - Congenital corneoscleral cyst
  - Cystic form of epibulbar dermoid
- Lymphatic cysts:
  - Lymphangiectasia
  - Lymphangioma
- Retention cysts
- Epithelial Implantation cysts
- Aqueous cysts:
  - Epithelial cysts due to downgrowth of epithelium
- Parasitic cysts
  - Hydatid cyst
  - Cysticercus
  - Filarial cyst
- Pigmented Epithelial cysts: Prolonged topical use of cocaine/epinephrine
Lymphangiectasia

- Appears as irregularly dilated lymphatic channels in bulbar conjunctiva
- May be developmental anomaly
- Can follow trauma or inflammation
- Anomalous communication with venule can lead to spontaneous filling of lymphatic vessels with blood
Lymphangioma

- Proliferations of lymphatic channel elements
- Usually present at birth and enlarge slowly
- Patch of vesicles with edema
- Intralesional hemorrhage – “chocolate cyst”
Subconjunctival cysticercus
Tumors of Conjunctiva:

*Non-pigmented tumours*

I. **Congenital**: dermoid and lipodermoid (choristomas).

II. **Benign**: simple granuloma, papilloma, adenoma, fibroma and angiomas.

III. **Premalignant**: intraepithelial epithelioma (Bowen's disease).

IV. **Malignant**: epithelioma or squamous cell carcinoma, basal cell carcinoma.
Pigmented tumours

I. **Benign**: naevi or congenital moles.

II. **Precancerous melanosis**: superficial spreading melanoma and lentigo maligna (Hutchinson's freckle).

III. **Malignant**: primary melanoma (malignant melanoma).
Dermoid:

Epibulbar Dermoid Tumor
• 1 in 10,000 individuals
• Pathogenesis
  – Displaced embryonic skin tissue
  – Composed of fibrous tissue, hair with sebaceous glands
  – Covered by conjunctival epithelium
• Clinical findings
  – Well-circumscribed, solid, smooth, porcelain white, round to oval elevated lesion embedded in superficial sclera or cornea
  – Most common in inferotemporal limbus
  – Arcus-like deposit of lipid along anterior corneal border
  – Corneal astigmatism – anisometropic amblyopia
Epibulbar Dermoid Tumor Management

- No malignant potential
- Lesion often extends deep into underlying tissues
- Elevated portion may be excised
- Relaxing incision or other corrective measure may be considered
- Lamellar keratoplasty for cosmetic appearance
- Amblyopia treatment
Lipodermoid:

- Found at the limbus or outer canthus.
- Appears as soft, yellowish white, movable subconjunctival mass.
- Consists of fatty tissue and the surrounding dermis-like connective tissue, hence the name lipodermoid.
- Sometimes the epibulbar dermoids or lipodermoids may be associated with accessory auricles and other congenital defects (*Goldenhar's syndrome*).
Conjunctival Inclusion Cyst
Benign Tumors:
Simple Granuloma:

- Consists of an extensive polypoid, cauliflower-like growth of granulation tissue.
- Simple granulomas are common following squint surgery, as foreign body granuloma and following inadequately scraped chalazion.
Papilloma

Pedunculated
– HPV, type 6 or 11
– Fleshy, exophytic growth with fibrovascular core
– Emanates from a stalk with multilobulated appearance with smooth, clear epithelium and small corkscrew vessels
– Inferior fornix, tarsal or bulbar conjunctiva
– May be multiple – more in HIV pts
Papilloma

Sessile

- HPV, type 16 or 18
- More likely dysplastic or carcinomatous
- Limbus
- Flat base with glistening surface and numerous red dots
- Signs of dysplasia
  - Keratinization (leukoplakia)
  - Inflammation
  - Invasion
- Rare variant – Inverted papilloma
Pyogenic granuloma:

Common reactive hemangioma

- Misnamed – not suppurative, no giant cells
- May occur
  - Over chalazion
  - Minor trauma
  - Post op granulation tissue
- Rapidly growing red, pedunculated, smooth lesion
- Bleeds easily and stains with fluorescein dye
Pre-malignant tumours

Bowen's intraepithelial epithelioma (carcinoma in situ):

- Usually occurring at the limbus as a flat, reddish grey, vascularised plaque.
- Histologically, it is confined within the epithelium.
- It should be treated by complete local excision.
Conjunctival Intraepithelial Neoplasia (CIN)

Clinical findings

– 3 clinical variants:
  • Papilliform – sessile papilloma harboring dysplastic cells
  • Gelatinous – result of acanthosis and dysplasia
  • Leukoplakic – hyperkeratosis, parakeratosis, and dyskeratosis

– Mild inflammation and abnormal vascularization

– Classification: Mild, Moderate, Severe (Carcinoma in situ)

– Slow growing tumors

– Potential to spread to other ocular surfaces
Conjunctival Intraepithelial Neoplasia (CIN)

**Management**
- Excisional biopsy with adjunctive cryotherapy
  - Recurrence rates at 10 years
    - Negative surgical margins ~ 33%
    - Positive surgical margins ~ 50%
  - Topical chemotherapeutic agents
    - Interferon, MM-C, 5-FU
    - No long term recurrence studies
Malignant tumors:
Squamous cell carcinoma

Pathogenesis
– Risk factors: UV radiation, viral, genetic
– More common and aggressive in:
  • HIV
  • Xeroderma pigmentosa
Clinical findings SCC:

- Broad based lesion at or near limbus in interpalpebral fissure
- Grow outward with sharp borders
- Can be leukoplakic
- Usually remains superficial rarely penetrating sclera
- Pigmentation in dark-skinned pts
- Engorged conjunctival vessels feeding tumor
- Inflammation
- Locally invasive and can metastasize
Management of SCC:

– Complete local excision
  • 4 mm beyond clinically apparent margins
  • Thin lamellar scleral flap beneath tumor
– Absolute alcohol to remaining underlying sclera
– Adjunctive cryotherapy to margins
– Risk of recurrence related to surgical margins
– Extensive external spread
  • Orbital exenteration and possible radiation therapy
Kaposi Sarcoma

- Malignant neoplasm of vascular endothelium involves skin, mucous membranes and internal organs
- Pathogenesis
  - Infection with HHV-8
  - Occurs in setting of AIDS
- Clinical findings
  - Reddish, highly vascular subconjunctival lesion
- Can be mistaken for subconjunctival hemorrhage
  - Orbital involvement – lid and conjunctival edema
  - Inferior fornix most common
  - Nodular or diffuse
Management
– Treatment may not be curative
– Nodular lesions less responsive to therapy
– Surgical debulking
– Cryotherapy
– Radiotherapy
– Local or systemic chemotherapy
– Intralesional interferon alpha-2a may be effective
Pigmented Tumors:
Nevus

- Nevocellular nevi of conjunctiva – hamartia arising during childhood and adolescence
- Junctional, Compound, Subepithelial
- Flat near limbus, Elevated elsewhere
- Pigmentation variable
- Small epithelial inclusion cysts ~ 50%
- Secretion of mucin in inclusion cysts – enlargement
- Rapid enlargement at puberty
- High prevalence of junctional activity but rarely become malignant
- Excision of suspicious lesions
- Excise nevi on palpebral conjunctiva
Primary Acquired Melanosis

• Preinvasive intraepidermal lesion of sun-exposed skin
• Flat, brown noncystic lesions of conjunctival epithelium
• PAM associated with cellular atypia – progress to melanoma in ~ 46%
• Pathogenesis
  – Abnormal melanocytes proliferate in basal conjunctival epithelium of middle-aged, light-skinned individuals
• Malignant transformation – nodularity, enlargement or increased vascularity
Management of PAM:

– Excisional biopsy

– All palpebral pigmented lesions should be excised

– Lesions that show atypia
  • Adjunctive cryotherapy
  • Mitomycin-C

– Check regional lymph nodes
Melanoma

• Less than 1% of ocular malignancies
• Prevalence:
  ~ 1 per 2 million in population of European ancestry
  – Rare in blacks and Asians
• Better prognosis than cutaneous melanoma
Pathogenesis of Melanoma

– Arise from acquired nevi, PAM, or normal conjunctiva
– Malignant transformation of congenital conjunctival nevus very rare
– Intralymphatic spread increases risk of metastasis
– Underlying ciliary body melanoma can extend through sclera
– Cutaneous melanoma can rarely metastasize to conj
Clinical findings: Melanoma

- Most common on bulbar conj or at limbus
- Variable pigmentation
- Highly vascularized – bleed easily
- Grow in nodular fashion
- Can invade globe or orbit
- Outcome
  - Bulbar melanomas have better prognosis than those on palpebral conj, fornix, or caruncle
  - Metastasis in ~ 26%, Mortality ~ 13% 10 yrs after surgical excision
- Cytologic risk factors for metastasis: large size, multicentricity, epithelioid cell type, lymphatic invasion
- Can metastasize to LN’s brain, and other sites
Melanoma
Management
– Excisional biopsy
– Excision of conjunctiva 4mm beyond clinically apparent margins
– Excision of thin lamellar scleral flap beneath tumor
– Treat remaining sclera with absolute alcohol
– Cryotherapy to conjunctival margins
– Primary closure or conj/amniotic membrane graft
– Topical mitomycin-C – can be used for residual disease
– Orbital exenteration – advanced disease or palliative tx

• Poor prognostic factors
  – Melanomas arising de novo
  – Tumors not involving limbus
  – Residual involvement at surgical margins