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
- Intrallesional 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