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 infertemporal 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)

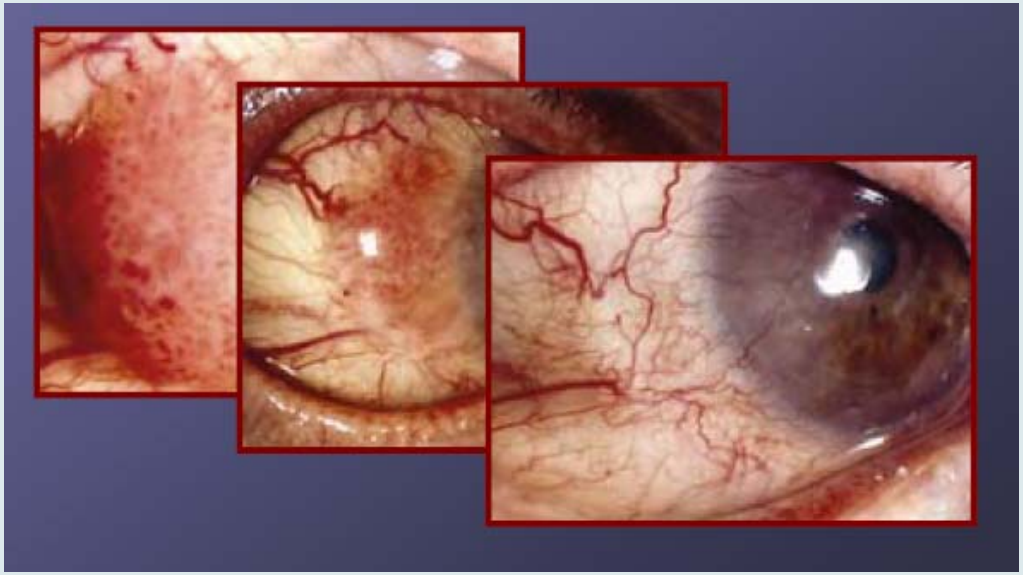
– Slow growing tumors

– 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

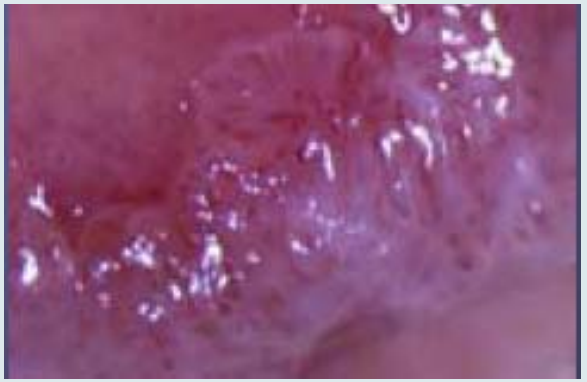
Pathogenesis

– 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 fussure
- 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 membrans 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

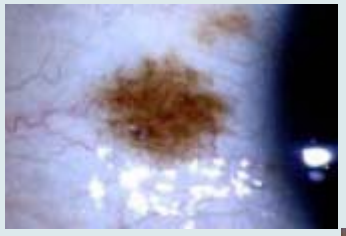
Management

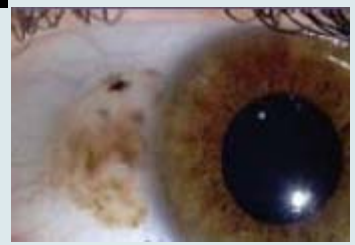
- Treatment may not be curative
- Nodular lesions less responsive to therapy
- Surgical debulking
- Cryotherapy
- Radiotherapy
- Local or systemic chemotherapy
- Intralesional interferon alpha-2a may be effective

Pigmented Tumors:

Nevus

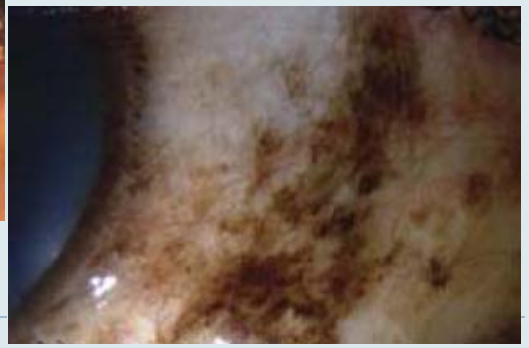
- Nevocellular nevi of conjunctiva – hamartia arising during childhood and adolescence
- Junctional, Compound, Subepithelial
- Flat near limbus, Elevated elsewhere
- Pigmentation variable
- Small epithelial inclusion cysts ~ 50%
- Secretion of mucin in inclusion cysts – enlargement
- Rapid enlargement 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