Work-up
1. Rule out neoplastic changes—rapid growth or pigmentation and feeder blood vessels, become raised, atypical location ie. palpebral or fornix, extension into cornea

Treatment
1. If suspicious or cosmetic concern, refer for excisional biopsy
2. If no referral, photo-document
3. Monitor 6-12 months or sooner if first presentation

Primary Acquired Melanosis (PAM)
Acquired melanosis—may be benign, precancerous, or cancerous

**General Characteristics**
Later in life, 30-40 years of age (almost always Caucasians), unilateral

15 to 17% of benign melanosis will convert to cancer within 30 years. Carries a higher risk of developing into melanoma

**Signs**
Sudden development of irregular diffuse, flat, grayish-black, brown or tan bulbar pigmentation; may extend onto palpebral conjunctiva, without cysts, any part of conj., lesion can be moved over sclera

Malignant transformation should be suspected when elevation, nodules or increase in vascularity in one of these areas develops

**Treatment**
Monitor closely (photos) - every 3-6 months or refer for biopsy
Dilated fundus exam to R/O choroidal melanoma

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**PAM without atypia**

- Is benign proliferation of normal melanocytes confined to the basal layers of conj.

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**PAM with atypia**

- Is a pre-malignant condition with a 50% chance of malignant transformation within 5 yrs
- Characterized by melanocytes involving all layers of conjunctiva

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**Malignant Melanoma**

**General Characteristics**
Arise spontaneously from a pre-existing nevus (20%) or acquired melanosis (60%), or spontaneously: rare tumor, more like melanomas of the skin than uvea, accounts for 2% of all eye malignancies

Most commonly between the ages of 40 and 60 years in Caucasians.

Common site is limbus, nodular brown or solitary black or grey nodule which is fixed to the episclera, amelanotic tumors are pink and have a characteristic feature of smoothly, finished appearance - dilated feeder vessels

**Signs**
Increased size, change in color, bleeding or ulceration, and feeder blood vessels

**Work-up**
Check for underlying ciliary body melanoma (dilated fundus exam, transillumination, and B-scan ultrasound). Intraocular and orbital extension may occur

**Treatment**
Refer for excision and histological examination—carefully document if refusal of excision; recheck every 6 months after removal

**Prognosis**
70% survival at 10 years
**Metastases**

- Regional lymph nodes, lung, brain & liver

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**Other Conjunctival Pigmentations**

Endogenous—Addison disease (light brown) & Jaundice (yellowish)

Exogenous—long term topical drug administration—epinephrine, mascara, argyrosis

Ciliary staphylomas
Non-Pigmented Tumors

Dermoid cyst – Congenital

General characteristics – Choristoma – a congenital overgrowth of normal tissue in an abnormal location

Composed of tissue not normally found in region

Usually found at lower temporal limbus where it involves cornea, conjunctiva, and sclera

May be associated with eyelid colobomas, preauricular accessory skin tags, and vertebral abnormalities, maldevelopment of jaw (Goldenhar’s syndrome)

Signs

Mass of collagen tissue containing hair, follicles, and glands covered by keratinized epithelium

Smooth, soft, yellowish-white, solid subconjunctival masses at the limbus

They may enlarge, especially at puberty

Treatment

Noted at birth; removal for cosmetic reasons when child is nearing school age

White corneal scar will persist after surgery, underlined cornea or sclera can be quite thin

Larger lesion may require lamellar corneal grafting
Dermatolipomas

**General characteristics**
- Congenital: may occur as part of Goldenhar's syndrome or in isolation

**Symptoms**
- Asymptomatic

**Signs**
- Firm, elevated, movable, yellowish, subconjunctival mass present at outer (superior temporal) canthal angle of eye and is usually obscured in primary position.
- Usually smooth surface but may contain hair follicles, glands, or fatty tissue; fatty tissue is continuous with orbital fat; extend well back into orbit.

**Treatment**
- No treatment unless cosmetic concern; refer to ophthalmic plastic surgeon - but usually avoided because of frequent extension into orbit, involving orbital structures, partial resection

The Epithelial Tumors

**Benign:** Cysts, papilloma, keratoses

**Dysplastic:** CIN, Squamous dysplasia, Carcinoma-in-Situ

**Malignant:** Invasive squamous cell carcinoma, Mucoepidermoid carcinoma

Papillomas

**General Characteristics**
- Occur most commonly at the caruncle, in the fornices, and limbus, maybe cause by a papilloma virus
- Pedunculated (Viral) most commonly in early adulthood
- Sessile is not infectious, usually middle age

**Signs**
- Soft, pink, elevated lesion with a slightly irregular surface, with multiple cores of corkscrew blood vessels, may have rapid growth

**Treatment**
- If suspicious or cosmetic, refer for excision and biopsy

Papilloma

- The tumor may arise from a thin central stalk (pedunculated type) or broad base (sessile type)
- Viral or benign or possibly malignant: difficult to distinguish clinically
- Treatment: viral often left untreated because of frequent recurrence rate and spontaneous resolution. Nonviral (sessile) usually excisional biopsy because may represent precancerous lesion
Carcinoma in Situ – Bowen’s disease – AKA Conjunctival/Corneal Intraepithelial Neoplasia (CCIN), Squamous Dysplasia

General characteristics
Usually in late adult life, fair-skinned people
The lesions are rare, usually unilateral, begin near the limbus, and may evolve into invasive squamous cell carcinoma if not treated early and successfully.
They can spread over the cornea or, less commonly, invade the eye or metastasize.

Signs
The lesions is a leukoplakic or gray-white elevated, gelatinous, highly vascularized, fleshy lesion that usually begins at the limbus with an inflammatory reaction, rose bengal or lissamine green staining of lesion, lesion is superficial to the basement membrane, R/O pterygium

Treatment
Local excision and biopsy often followed by supplemental cryotherapy to the remaining adjacent conjunctiva
Periodic follow-up to detect recurrence

CCIN
- Uncommon benign slowly progressive unilateral lesion
- Histological changes range from mild to severe epithelial dysplasia confined to the basal third of the epithelium to full thickness epithelial involvement (carcinoma in situ)
- Risk factors: UV, human papilloma virus & AIDS

En Plaque CCIN
- A raised, gelatinous or leucoplakic growth with tufts of superficial blood vessels at the limbus within intrapalpebral fissure