Benign eye lid lesions

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Any Eyelid mass or ulcer could be a benign or a malignant lesion. An accurate diagnosis can be reached based on history and clinical examination. If in doubt surgical biopsy followed by histo pathological evaluation can clinch the diagnosis. In this review article we look at some of the more common eyelid lesions that an ophthalmologist may encounter in a general practice.

BENIGN TUMORS OF EPIDERMIS

Squamous Papilloma

Introduction: It is one of the most common benign eye lid lesion. It is not a specific clino-pathological entity.

Age of Presentation: It is a group of condition that usually presents in middle or elderly age.

Clinical Feature: It can have a varied presentation, ranging from sessile to pedunculated, solitary or multiple, can be pigmented or similar in colour to skin. A rough irregular surface with keratinized crust formation is a common finding.

Prognosis: Their growth pattern is very slow, if rapidly growing, one should suspect other conditions.

Treatment Options: Shave excision is the best option. Other options include ablative carbon dioxide or Argon laser application to the base.

Seboric Keratosis

Introduction: Seborrheic keratosis (Basal cell papilloma, seborrheic warts) are common benign lesions on the face and abdomen. It can also present on the lids of aging individuals.

Clinical Features: They are well circumscribed, waxy, friable and appear stuck on to the skin. Some lesions are covered by an adherent greasy-appearing scale and are raised above the surface of the skin. They can feel soft and greasy.

The shape is round to oval, and multiple lesions may be aligned in the direction of skin folds. The lesion is very superficial and may be pigmented from slight discoloration to deep brown in colour.

Prognosis: They are usually asymptomatic, but can sometimes cause pruritus and irritation.

Treatment Options: Treatment involves surgical excision or laser ablation

Inverted Folicular Keratosis

Introduction: It is a benign cutaneous lesion almost similar in character to seborrheic keratosis. The term Inverted follicular keratosis is a misnomer, as it was thought that

A large pailomatous growth from lateral aspect of lower lid

Pigmented plaque like Folicular Keratosis

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these lesions arose from hair follicle. However recent studies suggest that they are basically an irritated form of Seboric Keratosis.

**Age of Presentation:** It appears in middle age.

**Clinical Presentation:** It can present as nodular, papilomatous or pigmented lesion. The differential point from a Seborric Keratosis, is its rapid growth pattern.

**Treatment:** Treatment consists of surgical removal.

**Keratoacanthoma**

**Introduction:** It is a form of pseudocarcinomatous hyperplasia. It has recently been re-classified as low grade squamous carcinoma.

**Age of Presentation:** It usually occurs in the middle age.

**Clinical Features:** It is similar in clinical look to a nodulo-ulcerative variety of basal cell carcinoma. It has rapid growth pattern. It grows over a period of 2 to 6 months.

**Treatment Options:** Treatment options range from observation, to cryo application, to surgical resection. There have been reports of spontaneous regression.

**Actinic Keratosis**

**Introduction:** Actinic keratosis also called solar keratosis—is associated with chronic sun exposure. Actinic keratoses are significant because they are recognized precursors of squamous cell carcinoma

**Age of Onset:** It is commonly encountered in patients past age 50 who have fair skin.

**Clinical Features:** Clinically, the keratoses usually appear as nondescript, reddish-brown scaly patches in sun-exposed areas of lids 3mm to 10mm in diameter.

**Treatment Options:** Isolated actinic keratoses may be treated with cryotherapy or curettage. However, because these lesions are often precancerous. Excision with biopsy in larger lesions may be advisable.

**BENIGN SEBACEOUS GLAND TUMOURS**

**Sebaceous Gland Hyperplasia and Adenoma**

**Introduction:** The meibomian glands of the tarsal plate, the gland of Zeis of the cilia, and the sebaceous gland of the caruncle are the different types of sebaceous glands of the lid. Tumour arising from these glands can either benign Adenoma, or malignant(Adeno carcinoma)

**Clinical Presentation:** Hyperplasia and adenoma of the sebaceous gland appears almost similar clinically. It appears as a yellowish nodule of the lid with smooth surface. The curuncular variety is slightly more irregular in appearance.

**Age of Presentation:** It can present in any age, but is usually common in young adult hood

**Treatment:** Benign tumours of the sebaceous gland of the lid usually never turn malignant. They can be observed or a simple surgical excision can be planned.

**Systemic Association:** A systemic association with Muir-Torre syndrome has been reported. It is an autosomal dominant condition where patients with cutaneous sebaceous tumours, basal cell carcinoma or keratoacanthoma, have a high incidence of visceral malignancy, special cancer of the colon. Almost 70% of these patients have a positive family history.

**SWEAT GLAND TUMORS**

**Syringoma**

These are benign tumors of the eccrine sweat glands around the lid and adnexae.

**Age of Presentation:** It is common in young females, but can present at any age.

**Clinical Presentation:** They appear as subtle yellowish small nodulated elevations in the periorbital region. They can be single, or appear in clusters.

**Treatment:** As they are benign in nature, a simple observation is enough. If it becomes large or cosmetically ugly, surgical excision is the treatment of choice. Other options include ablative carbondioxide or Argon laser resurfacing.
Prognostic Factors: Good, practically no chances of malignant transformation.

**Eye lid Pleomorphic Adenoma**

Pleomorphic adenoma (also referred to as benign mixed tumor or chondroid syringoma) is among the rarest of the adenomas and adenocarcinomas of the eyelids. Pleomorphic Adenomas (PAs) can arise from sweat glands in the dermis of the eyelid skin. It is a rare cutaneous neoplasm. It may arise from eccrine or apocrine glands.

**Age of Presentation:** It usually affects the elderly.

**Clinical Presentation:** It usually presents as nodulated lump of the upper or lower eye lid margin, which has been slowly increasing in size. Clinical differentiation from other similar nodulated growth is very difficult.

**Treatment:** Surgical excision, followed by reconstruction (Similar to what is done in BCC).

**TRICHOEPITHELIOMA**

**Introduction:** Trichoepithelioma is a benign adnexal neoplasm. The gene involved in the familial form of trichoepithelioma is located on band 9p21. Trichoepitheliomas consisted of nests of basaloid cells. Mitoses are uncommon when compared to basal cell carcinoma.

**Age of Presentation:** Lesions usually start appearing in childhood and gradually increase in number with aging.

**Clinical Presentation:** Slow-growing, single or multiple papules or nodules are typically observed on the lid skin and face. The lesions are rounded, skin-colored, firm papules or nodules that are 2-8 mm in diameter. Lid skin involvement occurs as an associated involvement. The occurrence of multiple trichoepitheliomas is transmitted as an autosomal dominant trait.

An association may exist with other cutaneous tumors (eg, cylindroma or Brooke-Spiegler syndrome, spiradenoma, basal cell carcinoma, ungual fibromas) or dystrophy unguis congenita.

Trichoepithelioma can be part of the Rombo syndrome (ie, vermiculate atrophoderma, milia, hypotrichosis, trichoepithelioma, basal cell carcinoma, peripheral vasodilatation).

**Treatment:** The treatment of the trichoepithelioma lesion is primarily surgical, only for cosmetic purpose. Solitary lesions may be excised. In the case of multiple tumors, this surgical approach is usually not feasible.

Split-thickness skin grafting, dermabrasion, and laser surgery have been tried when huge cluster lesions have to be managed. Recurrence of solitary trichoepithelioma is uncommon. When the multiple lid and lesions are surgically flattened by dermabrasion or laser therapy, they tend to regrow into elevated papules or nodules.

**MELANOCYTIC TUMOUR**

**Melanocytic nevus**

**Introduction:** A Melanocytic Nevus is usually a benign tumour derived from the cutaneous melanocytes.

**Age of Presentation:** Depending on the time of presentation a nevus can either be congenital or acquired. A congenital nevus usually shows up some time after birth, and shows a very slow growth pattern till puberty.

An acquired Melanocytic Nevus starts developing in early adult hood and can occur any where over the eye lid skin.

**Clinical Presentation:** They are commonly found on the skin of the human body. A nervus can also occur on the eyelid skin. These tumors are usually brownish pigmented and have thickness. It can range from being flat non elevated to a large nodular pigmented growth in the eye lid margin, over the eye lid skin or even over the eye brow. The pigmentation can vary from being just few spots of brown to very darkly brown. The eye lid margin lesions usually extend beyond the mucocutaneous junction into the tarsal conjunctiva. In the lid two very characteristic variety have been described.
Kissing nevus or Divided nevus is a rare form of congenital nevus that usually occurs on adjacent parts of the upper and lower eyelids of one eye. Most often, the formation is present from birth, but it may also appear later.

Hair naevus is a dark colored, often hairy patch of skin present in the periocular region present at birth. It can be along the distribution of the lacrimal drainage system. Sometimes it may form a teardrop pigmentation over the periocular region.

PROGNOSTIC FACTORS: A Melanocytic Nevus is usually a benign condition. In very rare situations, if the lesion shows very rapid increase in pigmentation and growth in middle ages, especially in sun exposed areas. A malignant conversion should be suspected.

Treatment: Eyelid nevi are usually benign tumors. They can be photographed and followed for evidence of change or growth prior to consideration of biopsy or treatment. Large eyelid nevi can be a cosmetic problem. It requires complex and challenging rotational flaps to restore and give a better cosmetic outcome.

NEURAL TUMOURS

Neurofibroma

Introduction: Neurofibroma affects the bone, the nervous system, soft tissue, and the skin. Increased concentrations of nerve growth stimulating activity have been linked with the development of neurofibromatosis.

In Neurofibromatosis beside the characteristic skin lesions, other clinical features include skeletal bony abnormalities, mental deficiency, seizures, neurofibromas of the spinal and cranial nerve roots, iris hamartomas, optic nerve gliomas, endocrine disorders, endocrine tissue tumors, other visceral tumors, etc.

The genetic defect is localised to chromosome 17 and is transmitted in an autosomal dominant pattern.

Clinical Presentation: In the eye lid and periorbital region, two forms of presentation are usually noted.

• Plexiform neurofibroma: presents as a diffuse and elongated swelling along in the lateral aspect of the lid and peribital region. It can range from a small bulkiness of the lateral upper lid to a mechanical ‘S’ shaped ptosis due the increased weight of the lid. In severe from a large fold of skin occupying the whole of the lateral side of the face and lid can be found and usually follows the course of facial nerve trunk. These tend to infiltrate into deeper structures like fascia, muscles and bone.

• Neurofibroma nodules: Soft multiple lesions ranging from a small lesion to large nodule or peduncle is seen all over the lids, peribital region and face. Similar lesions are also seen all over the trunk and body. They are usually very slowly growing but it continues to grow lifelong.

Age of Presentation: It starts at or after birth and grows lifelong

Treatment:

• Plexiform neurofibroma: There is no definitive plain of differentiation between the fibromatous growth to healthy tissue. Debulking of the excess tissue is often tried. How much to debulk is a tricky situation to evaluate. Some prefer a pinching technique, where you pinch the excess tissue with your fingers, and make the base, so that the tissue can be aposed after excision. Others prefer to use a template of the other side eye lid and remove the excess tissue. The
difficulty during debulking is because the tissue oozes quite significantly during dissection. Preferably multiple figure of eight sutures should be applied in the deeper plain to help in hemostasis. Then skin is sutured in interrupted layers. Another issue is the relapse or recurrence. This needs to be explained and discussed clearly with the patient.

- **Neurofibroma nodules**: They usually do not require any intervention, as it is not practically possible to remove them all. Only when a lesion becomes cosmetically too large, or is obstructing vision, that specific lesion can be removed.

**Schwannoma**

**Introduction**: Schwannoma also called neurilemmoma, is a benign tumour arising from the Schwan cells of the neural sheath of a peripheral nerve. Isolated eye lid schwannoma is extremely rare.

**Age of Presentation**: Usually occur in adults from age 20 to 70 years.

**Clinical Features**: It presents usually as a solitary smooth very slowly growing nodule over the lid margin or peri orbital area. It is very difficult clinically to differentiate the lesion from other similar benign nodules like chalazion or inclusion cysts. Surgical excision followed by HP evaluation can confirm the pathology. The presence of multiple schwannoma should usually arise a suspicion of associated Neurofibromatosis type 1 or 2. Associated schwannoma have been described in conjunctiva, uvea tract and in the orbit.

**Treatment**: Treatment consists of surgical excision followed by histopathological evaluation. The prognosis is excellent with very rare recurrences.

If it is in the periorbital region, dissection and removal of the nodule usually solves the problem. If the lesion lies over the lid margin, fullthickness lid excision, followed by lid reconstruction, following the principles of reconstruction, gives very good cosmetic outcome.

**VASCULAR TUMOURS**

**Congenital capillary hemangioma**

**Introduction**: It is a benign vascular tumour of the child hood. These lesions are considered hamariforms.

**Age of Presentation**: Usually present congenitaly or arise in early infancy They may appear after birth but usually cease growing by one year of ageand most will shrink by first decade.

**Clinical Presentation**: Multiple Red or purple elevated, subcutaneous soft spongy mass that will Blanch with pressure. The lesions are composed of diluted capillary network, and looks like the surface of a strawberry, so the name “Strawberry marks”. They are usually unilateral and located on the eyelid or brow. Capillary hemangiomas are without pulsation and have no bruit.

A mass effect of the lesion may result in significant ptosis of an involved eyelid. Reduced visual acuity may be noted (either due to amblyopia or uncorrected astigmatism from mass effect of a hemangioma on the cornea).

**Treatment**: Spontaneous involution occurs in most cases, so observation may be appropriate. Usually 40% completely involute by age 4, while 80% completely involute by age 8.

In cases there is occlusion amblyopia or significant astigmatism, prompt treatment should be initiated. The different options are:

- **Intralesional steroid injection** with a mix of long and short acting steroid (i.e. 40mg/ml of triamcinolone and 6mg/ml of betamethasone )
- **Oral prednisone** (1-2mg/kg/day given with the involvement of the child’s pediatrician - treatment is usually for months with slow taper)
- **Surgical excision** (for debulking - lesion is not encapsulated and there is a risk of recurrence.)
- Other treatments have been described (topical steroid, radiation, and a variety of lasers) but they are considered somewhat controversial and not used routinely.

**Associations**:

- In PHACES (Posterior fossa malformations, Hemangiomas, Arterial anomalies, Coarctation of the aorta, Eye abnormalities, Sternal clefting and supraumbilical raphe), especially if the hemangioma involves more than one dermatome.
In Maffucci syndrome the patient has multiple cutaneous and visceral hemangiomas.

In Kasabach-Merritt syndrome there is thrombocytopenic coagulopathy where platelets are rapidly sequestered within the vascular lesion. This is a rare condition with high mortality which requires prompt management by multiple subspecialists.

**Differential Diagnoses:**
- Vascular malformation
- Lymphangioma
- Arteriovenous malformations
- Nevus Flammeus (port-wine stain) (Port-Wine stain will not typically blanch with pressure — a capillary hemangioma will usually blanch)

### Acquired capillary hamangioma

**Introduction:** Acquired capillary haemangioma also called cherry angioma are benign vascular lesions, rarely involving the eye lid.

**Age of Presentation:** It is a common cutaneous lesion of the young adult hood or in middle age.

**Clinical Presentation:** It presents as small nodular reddish structure either at the lid margin or over it. They can become pedunculated with a cherry like surface irregularity. They can bleed on a subtle trauma. These types of localized haemangiomas are usually very compressible.

**Treatment:** If the lesions are small, one can wait and observe. If the lesions increase in size a wedge resection of full thickness lid along with the lesion, followed by lid apposition in layers give excellent result. Radioablation, cryo application or electrodisection are other alternative methods of treatment that have been applied.

**Differential Diagnoses:** The differential diagnosis include Kaposi’s sarcoma, cavernous haemangioma, angiosarcoma and varix.

### Nevus flammeus (port-wine stain)

**Introduction:** A port-wine stain or nevus flammeus is a vascular anomaly consisting of superficial and deep dilated capillaries in the skin lid or one side of the face. They resemble that of port wine.

**Age of Presentation:** Port-wine stains are present at birth and persist throughout life; The area of skin affected grows in proportion to general growth.

**Clinical Presentation:** A reddish to purplish discoloration of the skin typically following the distribution of the Trigeminal nerve. It usually does not cross the midline. Port-wine stains occur most often on the face but can appear anywhere on the body.

Early stains are usually flat and pink in appearance. As the child grows, the color may deepen to a dark red or purplish color. In adulthood, thickening of the lesion or the development of small lumps may occur. It affects both males and females of all ethnic backgrounds.

**Associations:**

- **Sturge–Weber syndrome:** It is one of the phakomatoses and is often associated with port-wine stains of the face, glaucoma, seizures, mental retardation, and ipsilateral leptomeningeal angioma.

Klippel-Trenaunay-Weber syndrome (KTWS) is characterized by a triad of port-wine stain, varicose veins, and bony and soft tissue hypertrophy involving an extremity.

**Treatment:** Different options are tried, ranging from freezing, surgical removal, local radiation, and tattooing. Port-wine stains can also be masked with cosmetics. Lasers have application to the cutaneous capillaries with minimal damage to the overlying skin is one of the recent advances, in the management of this cosmetic blemish.

### Miscellaneous Benign Vascular tumour (LID VARIX and Lymphangioma)

**Introduction:** A varix or lymphangioma consists of abnormally distended vein, artery or lymphatic vessel. Orbital varix and lymphangioma are a common entity but isolated eyelid varix are rare. Orbital and lid varix may be associated with additional venous malformations elsewhere in the body.

**Age of Presentation:** Lymphangioma usually present at birth, which gradually increases. Whereas usually present in adult hood, when the vascular channels become dilated, and lid appers bulky.

**Clinical Presentation:** Clinically the Lid varix and Lymphangioma may present almost similarly. Lymphangioma are usually present as a bluish well circumscribed or lobulated lesion over the lid skin. The lesions often enlarge and increase in size on Valsalva maneuver. Patients with orbital component of varix may present with proptosis or complain of visual disturbance.
On palpation a bag of worm like feeling can sometimes be felt. It is usually soft, is compressible on pressure, again regaining shape on release of pressure over the lesions. A lid varix can rupture on subtle trauma and bleed heavily.

**Differential Diagnosis:** The differential diagnosis includes hemangioma, and lymphangioma.

**Treatment:** They are difficult to treat. Most of the case can be left as it is with assurance. Ligation of small sections of the varix or Lymphangioma, as appropriate, may achieve the desired cosmetic effect and improve patient comfort. Venography should be used to evaluate the extent and supply before surgery.

### CYSTIC LESIONS

- **Epidermal Inclusion Cysts** These are small white-yellow cystic lesions occurring on the lid skin, conjunctiva, face or neck and are very common. They may develop spontaneously or arise following trauma or after surgery along an incision line. They originate from pilosebaceous follicles or invagination of surface epidermis. Some of these cysts occurring amongst the lashes will be difficult to distinguish from a blocked Zeiss gland (sebaceous gland).
- **Sudoriferous Cysts (Retention Cysts, Hidrocystomas)** The lid skin has numerous sweat glands (eccrine glands) and modified sweat glands (apocrine glands) such as the glands of Moll. Blockage of these glands leads to the formation of a translucent (water blister-like) lesion on the lid skin or lid margin amongst the lashes. They may occur as a single lesion or as multiple.
- **Sebaceous cysts** Sebaceous cysts may occur in and around the eyelid area and clinically resemble epidermal inclusion cysts. They are generally found in locations with many hair follicles, particularly the brow area and medial canthus. These cysts can occur secondary to obstruction of the Zeiss gland, meibomian gland or sebaceous glands associated with hair follicles of the lid skin or brow area. Unlike an epidermal inclusion cyst (filled with keratin material), a sebaceous cyst contains epithelial cells, keratin, fats and cholesterol crystals.

**Age of Presentation:** It can present at any age. It can remain static or can grow over time.

**Treatment:** Treatment involves total excision. Otherwise they will recur. Simply stabbing them with a needle is ineffective in allowing their resolution.

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